Table 10: *H. sapiens* Recon 1 network confidence scores and citations. Alphabetized list of reactions and their corresponding confidence scores, literature citations, and curator notes. Confidence scores (ranging from 0 to 3) are defined in the text.

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
24,25VITD2Hm	3	Labuda M, Lemieux N, Tihy F, Prinster C, Glorieux FH.	Human 25-hydroxyvitamin D 24-hydroxylase cytochrome P450 subunit maps to a different chromosomal location than that of pseudovitamin D-deficient rickets.	J Bone Miner Res	1993	8266831	This reaction takes place in kidney based on Vitamins, G.F.M. Ball,2004, Blackwell publishing, 1st ed (book) pg.196 1-4 ng/ml blood is produced if neither ca2+ nor pi i needed (regulated by these compounds concentration) IT
24,25VITD2Hm	3	Kusudo T, Sakaki T, Abe D, Fujishima T, Kittaka A, Takayama H, Hatakeyama S, Ohta M, Inouye K.	Metabolism of A-ring diastereomers of 1alpha,25-dihydroxyvitamin D3 by CYP24A1.	Biochem Biophys Res Commun	2004	15358094	This reaction takes place in kidney based on Vitamins, G.F.M. Ball,2004, Blackwell publishing, 1st ed (book) pg.196 1-4 ng/ml blood is produced if neither ca2+ nor pi i needed (regulated by these compounds concentration) IT
25VITD3Hm	3	St-Arnaud R, Messerlian S, Moir JM, Omdahl JL, Glorieux FH.	The 25-hydroxyvitamin D 1-alpha-hydroxylase gene maps to the pseudovitamin D-deficiency rickets (PDDR) disease locus.	J Bone Miner Res	1997	9333115	This reaction takes place in kidney based on Vitamins, G.F.M. Ball,2004, Blackwell publishing, 1st ed (book) pg.196 this form is the biological acitve hormon (25-70pg/ml blood) IT is produced if ca2+ or pi i needed (regulated by these compounds concentration)
25VITD3Hm	3	Yokomura K, Suda T, Sasaki S, Inui N, Chida K, Nakamura H.	Increased expression of the 25-hydroxyvitamin D(3)-1alpha-hydroxylase gene in alveolar macrophages of patients with lung cancer.	J Clin Endocrinol Metab	2003	14671156	This reaction takes place in kidney based on Vitamins, G.F.M. Ball,2004, Blackwell publishing, 1st ed (book) pg.196 this form is the biological acitve hormon (25-70pg/ml blood) IT is produced if ca2+ or pi i needed (regulated by these compounds concentration)
2AMACHYD	3	Gavaret JM, Cahnmann HJ, Nunez J	The fate of the "lost side chain" during thyroid hormonogenesis	J Biol Chem	1979	500639	The reference indicates this is a spontaneous reaction. It is needed to get rid of 2amac that is a byproduct of thyroid hormone synthesis. added by MM- PMID 14596599: Formation of pyruvate by SDH is a two-step reaction in which the hydroxyl group of serine is cleaved to produce aminoacrylate, and then the aminoacrylate is deaminated by nonenzymatic hydrolysis to produce pyruvate.
2AMACSULT	2	Cooper AJ	Biochemistry of sulfur-containing amino acids.		1983	6351723	-L-cysteate can be synthesized from 2-aminoacrylate in rat (PMID: 6351723) -this is a lumped rxn w/ an inferred reduction rxn that occurs in conjunction w/ the sulfotransferase step; this reduction step has to occur in order for the transformation (2amac-->Lcyst) to be valid -used NADPH as electron donor since it is a common cofactor involved in biosynthetic steps utilizing oxygen (see Neema for further explanations) MM
2HBO	3	Komoda T, Sakagishi Y, Mizushima H	Determination of isoenzyme contents of lactic dehydrogenase activity and 2-hydroxybutyric dehydrogenase activity in lactic dehydrogenase preparations	Clin Chim Acta	1976	10108	- another function of L-lactate dehydrogenase [Naghizadeh, Clin Chim Acta 1977], [Komoda, Clin Chim Acta 1976]
2HBO	3	Naghizadeh F	Oxidation of alpha-hydroxybutyrate by human serum	Clin Chim Acta	1977	21765	- another function of L-lactate dehydrogenase [Naghizadeh, Clin Chim Acta 1977], [Komoda, Clin Chim Acta 1976]
2OXOADOXm	2	Cox, RP	Errors of lysine metabolism	The Metabolic and Molecular Bases of Inherited Disease, 8th ed	2001		0
2OXOADPTm	3	Fiermonte G, Dolce V, Palmieri L, Ventura M, Runswick MJ, Palmieri F, Walker JE	Identification of the human mitochondrial oxodicarboxylate carrier. Bacterial expression, reconstitution, functional characterization, tissue distribution, and chromosomal location	J Biol Chem	2001	11083877	reference says other transport reactions done by this gene as well
34DHOXPEGOX	3	Mardh G, Dingley AL, Auld DS, Vallee BL	Human class II (pi) alcohol dehydrogenase has a redox-specific function in norepinephrine metabolism	Proc Natl Acad Sci U S A	1986	3466164	Citation in abstract indicates that ADH does this reaction.
34DHOXPEGt	2	Kurz T, Richardt G, Hagl S, Seyfarth M, Schomig A	Two different mechanisms of noradrenaline release during normoxia and simulated ischemia in human cardiac tissue	J Mol Cell Cardiol	1995	7473774	Citation indicates that this compound is found in the blood. Mechanism and gene unknown.
34DHPLACOX(NADP)	3	Ambroziak W, Pietruszko R	Human aldehyde dehydrogenase. Activity with aldehyde metabolites of monoamines, diamines, and polyamines	J Biol Chem	1991	2071588	First citation has catalytic constants. Only Aldh3a1 is known to use NADP, but the other two have not yet been determined.
34DHPLACOX(NADP)	3	Keung WM	Biogenic aldehyde(s) derived from the action of monoamine oxidase may mediate the antidipsotropic effect of daidzin	Chem Biol Interact	2001	11306106	First citation has catalytic constants. Only Aldh3a1 is known to use NADP, but the other two have not yet been determined.
34DHXMANDACOX(NADP)	3	Kawamura M, Eisenhofer G, Kopin IJ, Kador PF, Lee YS, Tsai JY, Fujisawa S, Lizak MJ, Sinz A, Sato S	Aldose reductase, a key enzyme in the oxidative deamination of norepinephrine in rats	Biochem Pharmacol	1999	10424772	First citation indicates that aldh1a3 does not use NADP. Aldh3b1, 3b2 are yet to be determined according to citation and left with both forms of the reaction. Second citation is based on evidence in rats. This reaction is left reversible because the loop was eliminated by making the NAD dependent reaction irreversible.

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
3DPHBH1	2	Szkopinska A.	Ubiquinone. Biosynthesis of quinone ring and its isoprenoid side chain. Intracellular localization.	Acta Biochim Pol	2000	11051212	IT the enzyme responsible for this reaction has not been identified in any organism. Szekopinspka (2000) describes reaction as hydroxylation, Stryer (Biochemistry) notes that NADPH is a required secondary metabolite for the hydroxylation reaction of sterols. Based on these information the reaction was created (plus for NADH since we cannot exclude the action of NADH instead of NADPH)
3HAO	3	Malherbe P, Kohler C, Da Prada M, Lang G, Kiefer V, Schwarcz R, Lahm HW, Cesura AM	Molecular cloning and functional expression of human 3-hydroxyanthranilic-acid dioxygenase	J Biol Chem	1994	7514594	Reaction and gene detailed in citation.
3HKYNAKGAT	2	BONNER DM, JAKOBY WB	Kynurenine transaminase from neurospora	J Biol Chem	1956	13357462	The citation states that the same enzyme that takes Lkynr as a substrate in neurospora also takes this as a substrate. Physiological data was chosen for this reaction, but that characterization may not be entirely accurate.
3HPCOAHYD	3	Hawes JW, Jaskiewicz J, Shimomura Y, Huang B, Bunting J, Harper ET, Harris RA.	Primary structure and tissue-specific expression of human beta-hydroxyisobutyryl-coenzyme A hydrolase.	J Biol Chem	1996	8824301	Citation has details. It is unclear why it is included in this pathway.
3MLDAt	2	Johansson AC, Lonnqvist B, Granerus G	The relationship between body size and the urinary excretion of the main histamine metabolite tele-methylimidazoleacetic acid in man	Inflamm Res	2001	11411609	SAB Added because this compound is known to be in the urine. Mechanism and gene unknown.
3MOPt2im	3	Hutson SM, Hall TR.	Identification of the mitochondrial branched chain aminotransferase as a branched chain alpha-keto acid transport protein.		1993	8428987	PMID 8428987: transport of branched chain alpha-keto acid via proton symport shown in rat mitochondria; possibly a bifunctional activity of mitochondrial branched chain aminotransferase MM
3MOXTYROX	3	Jonsson EG, Norton N, Gustavsson JP, Oreland L, Owen MJ, Sedvall GC	A promoter polymorphism in the monoamine oxidase A gene and its relationships to monoamine metabolite concentrations in CSF of healthy volunteers	J Psychiatr Res	2000	10867119	MAOA seems to participate a bit more than MAOB in this reaction
3NTD7l	3	R.L. Pisoni	Lysosomal Nucleic Acid and Phosphate Metabolism and Related Metabolic Reactions, Chapter 9		1996		nucleotide monophosphates come from DNA and RNA degradation in lysosome (by acid exonucleoase and acid ribonuclease). Will be dead-end in model as well as their efflux transport systems. IT Their is also a acid nucleotidase which carries out these reactions but i could not find the gene for this lysososmal enzyme
4ABUTtm	2	Medina-Kauwe LK, Tobin AJ, De Meirleir L, Jaeken J, Jakobs C, Nyhan WL, Gibson KM.	4-Aminobutyrate aminotransferase (GABA-transaminase) deficiency.		1999	10407778	- needed in the degradation of 4abut (GABA) in the mitochondria -allows 4abut (which resides/is synthesized in cytosol) degradation to succ, which occurs in the mitochondria; transport mechanism not found/specified for humans so this rxn is used for the time being, although there is a possible 4abut/glu L antiport mechanism based on evidence in rat mitochondria (PMID:10407778) MM
4HBZCOAFm	2	Meganathan R.	Ubiquinone biosynthesis in microorganisms.	FEMS Microbiol Lett	2001	11583838	IT I assumed that reaction takes place in mito as whole ubiquinone biosynthesis.- first reaction from tyr to 34hpp has also been assigned to the mito Booth et al proposed this mechanism from tyrosine to 4hbz. Loescher + Heide found this mechanism in higher plant cells and these results make it highly probable that this mechanism also occurs in mammalian cells.
4HBZCOAFm	2	Loscher R, Heide L.	Biosynthesis of p-Hydroxybenzoate from p-Coumarate and p-Coumaroyl-Coenzyme A in Cell-Free Extracts of Lithospermum erythrorhizon Cell Cultures.	Plant Physiol	1994	12232327	IT I assumed that reaction takes place in mito as whole ubiquinone biosynthesis.- first reaction from tyr to 34hpp has also been assigned to the mito Booth et al proposed this mechanism from tyrosine to 4hbz. Loescher + Heide found this mechanism in higher plant cells and these results make it highly probable that this mechanism also occurs in mammalian cells.
4HBZCOAFm	2	Booth, A. N., Masri, M. S., Robbins, D. J., Emerson, O. H., Jones, F. T. & DeEds, F.	Urinary Phenolic Acid Metabolites of Tyrosine	J Biol Chem	1960		IT I assumed that reaction takes place in mito as whole ubiquinone biosynthesis.- first reaction from tyr to 34hpp has also been assigned to the mito Booth et al proposed this mechanism from tyrosine to 4hbz. Loescher + Heide found this mechanism in higher plant cells and these results make it highly probable that this mechanism also occurs in mammalian cells.

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
4HGLSDm	1	Knight J, Holmes RP	Mitochondrial hydroxyproline metabolism: implications for primary hyperoxaluria	Am J Nephrol	2005	15849464	included based on KEGG indicating that the enzyme does this substrate too, but could not track evidence back to anything certain enzyme may use other substrates as well (other 1-pyrrolines) this reaction is written wrong in KEGG (NAD and NADH on wrong side of eqn) - isolated mouse liver mitochondria produced glyoxylate from hydroxyproline [Knight 2005]
4HOXPACDOX(NADP)	2	Shimamura M, Kamada S, Hayashi T, Naruse H, Iida Y.	Sensitive determination of tyrosine metabolites, p-hydroxyphenylacetic acid, 4-hydroxy-3-methoxyphenyl-acetic acid and 4-hydroxy-3-methoxymandelic acid, by gas chromatography-negative-ion chemical-ionization mass spectrometry.	J Chromatogr	1986	3753983	Physiological data based on citation. This metabolite comes from tyrosine.
5HLTDL	3	Sumi-Ichinose C, Ichinose H, Takahashi E, Hori T, Nagatsu T	Molecular cloning of genomic DNA and chromosomal assignment of the gene for human aromatic L-amino acid decarboxylase, the enzyme for catecholamine and serotonin biosynthesis	Biochemistry	1992	1540578	Gene and enzyme characterized.
5HOXINOXDA	3	Geha RM, Rebrin I, Chen K, Shih JC	Substrate and inhibitor specificities for human monoamine oxidase A and B are influenced by a single amino acid	J Biol Chem	2001	11134050	Fourth citation gives some information about substrate specificities for each enzyme.
5HTRPVESSEC	3	Eiden LE, Schafer MK, Weihe E, Schutz B	The vesicular amine transporter family (SLC18): amine/proton antiporters required for vesicular accumulation and regulated exocytotic secretion of monoamines and acetylcholine	Pflugers Arch	2004	12827358	From PMID 12827358: VATs accumulate singly positively-charged amines into the relatively proton-impermeable acidic secretory vesicles at the expense of proton antiport through the transporter protein (protons are first accumulated in secretory vesicles via a vacuolar ATPase not physically associated with the transporter) with a two proton:one amine stoichiometry, and to a final substrate concentration of up to 500 mM, exceeding that found in the cytosol by 100-fold (ACh) to 10,000-fold (biogenic amines)[29]. From PMID 15383652: In addition to this intrinsic dependency on the transmembrane electrochemical gradient, the transport rate can also be modulated by alterations in the rate of ATP hydrolysis and its coupling to H+ translocation. A recent detailed analysis of current-voltage relationships in the absence and presence of several ions, ATP or ADP and imposing different pH gradients, described different coupling ratios for vacuolar H+-ATPases from yeast. In the presence of large pH gradients (4 pH units), the approximate ratio was 2 H+/ATP and increased to more tha 3 H: 1 ATP averaged to arrive at 3 substrate: 2 ATP transport vesicle not accounted for--this is a net transport reaction
A_MANASEly	0	Liao YF, Lal A, Moremen KW.	Cloning, expression, purification, and characterization of the human broad specificity lysosomal acid alpha-mannosidase	J Biol Chem	1996	8910458	MAN2B1 encodes an enzyme that hydrolyzes terminal, non-reducing alpha-D-mannose residues in alpha-D-mannosides. Its activity is necessary for the catabolism of N-linked carbohydrates released during glycoprotein turnover and it is member of family 38 of glycosyl hydrolases. Defects in this gene have been shown to be the cause of lysosomal alpha-mannosidosis (AM), a lysosomal storage disease characterized by the accumulation of unbranched oligosaccharide chains. [RefSeq] Man2b1p ubiquitously expressed [Liao et al, J Biol Chem 1996]
A4GALTg	3	Steffensen R, Carlier K, Wiels J, Levery SB, Stroud M, Cedergren B, Nilsson Sojka B, Bennett EP, Jersild C, Clausen H.	Cloning and expression of the histo-blood group Pk UDP-galactose: Ga1beta-4G1cbeta1-cer alpha1, 4-galactosyltransferase. Molecular genetic basis of the p phenotype.	J Biol Chem	2000	10747952	Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot also cytoplasmic domain - see ref Kojima strongly expressed in heart, kidney, spleen, and placenta and weakly in colon, small intestine, and brain NJ
A4GALTg	3	Kojima Y, Fukumoto S, Furukawa K, Okajima T, Wiels J, Yokoyama K, Suzuki Y, Urano T, Ohta M, Furukawa K.	Molecular cloning of globotriaosylceramide/CD77 synthase, a glycosyltransferase that initiates the synthesis of globo series glycosphingolipids.	J Biol Chem	2000	10748143	Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot also cytoplasmic domain - see ref Kojima strongly expressed in heart, kidney, spleen, and placenta and weakly in colon, small intestine, and brain NJ
A4GNT2g	0	Nakayama J, Yeh JC, Misra AK, Ito S, Katsuyama T, Fukuda M.	Expression cloning of a human alpha1, 4-N-acetylglucosaminyltransferase that forms GlcNAcalpha1-->4Galbeta-->R, a glycan specifically expressed in the gastric gland mucous cell-type mucin	Proc Natl Acad Sci U S A	1999	10430883	A4gnt attaches GlcNAc to Core 2 glycans, also acts less efficiently on Core 1 glycans; characteristic for gastric glan mucous cell-type mucins; found primarily in the stomach and pancreas [Nakayama et al, PNAS 1999]
AACTOOR	3	Deng Y, Yu PH.	Assessment of the deamination of aminoacetone, an endogenous substrate for semicarbazide-sensitive amine oxidase.		1999	10328770	this rxn is catalyzed only by SSAO (AOC3) (see PMID: 10328770) MM
AACTOOR	3	Dalfo E, Hernandez M, Lizcano JM, Tipton KF, Unzeta M.	Activation of human lung semicarbazide sensitive amine oxidase by a low molecular weight component present in human plasma.		2003	12878330	this rxn is catalyzed only by SSAO (AOC3) (see PMID: 10328770) MM
AATAi	3	Goh,D.L.M., Patel,A., Thomas,G.H., Salomons,G.S., Schor,D.S.M., Jakobs,C., Geraghty,M.T.,	Characterization of the human gene encoding alpha-aminoadipate aminotransferase (AADAT).		2002	12126930	cytosolic according to Reactome also needs to be cytosolic since 2-oxoadipate is produced in cytosol and subsequently transported to mitochondria (PMID: 11083877)

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
ABO1g	3	Yamamoto F.-I., Hakomori S.-I.;	Sugar-nucleotide donor specificity of histo-blood group A and B transferases is based on amino acid substitutions	J Biol Chem	1990	2121736	localization: golgi - Type II membrane protein. Membrane-bound form in trans cisternae of Golgi. Soluble form in body fluids. biochemistry and review of blood group Ag synthesis: PMID: 11421346. structure: PMID: 12198488 characterization: PMID 2121736 This protein is the basis of the ABO blood group system. The histo-blood group ABO involves three carbohydrate antigens: A, B, and H. A, B, and AB individuals express a glycosyltransferase activity that converts the H antigen to the A antigen (by addition of UDP-GalNac) or to the B antigen (by addition of UDP-Gal), whereas O individuals lack such activity. NJ
ABO1g	3	Lloyd KO.	The chemistry and immunochemistry of blood group A, B, H, and Lewis antigens: past, present and future.	Glycoconj J	2000	11421346	localization: golgi - Type II membrane protein. Membrane-bound form in trans cisternae of Golgi. Soluble form in body fluids. biochemistry and review of blood group Ag synthesis: PMID: 11421346. structure: PMID: 12198488 characterization: PMID 2121736 This protein is the basis of the ABO blood group system. The histo-blood group ABO involves three carbohydrate antigens: A, B, and H. A, B, and AB individuals express a glycosyltransferase activity that converts the H antigen to the A antigen (by addition of UDP-GalNac) or to the B antigen (by addition of UDP-Gal), whereas O individuals lack such activity. NJ
ABO1g	3	Patenaude S.I., Seto N.O.L., Borisova S.N., Szpacenko A., Marcus S.L., Palcic M.M., Evans S.V.;	The structural basis for specificity in human ABO(H) blood group biosynthesis	Nat Struct Biol	2002	12198488	localization: golgi - Type II membrane protein. Membrane-bound form in trans cisternae of Golgi. Soluble form in body fluids. biochemistry and review of blood group Ag synthesis: PMID: 11421346. structure: PMID: 12198488 characterization: PMID 2121736 This protein is the basis of the ABO blood group system. The histo-blood group ABO involves three carbohydrate antigens: A, B, and H. A, B, and AB individuals express a glycosyltransferase activity that converts the H antigen to the A antigen (by addition of UDP-GalNac) or to the B antigen (by addition of UDP-Gal), whereas O individuals lack such activity. NJ
ABTArm	3	Osei,Y.D. , Churchich,J.E.	Screening and sequence determination of a cDNA encoding the human brain 4-aminobutyrate aminotransferase.		1995	7721088	Entrez Gene - 4-aminobutyrate aminotransferase (ABAT) is responsible for catabolism of gamma-aminobutyric acid (GABA), an important, mostly inhibitory neurotransmitter in the central nervous system, into succinic semialdehyde. The active enzyme is a homodimer of 50-kD subunits complexed to pyridoxal-5-phosphate. The protein sequence is over 95% similar to the pig protein. GABA is estimated to be present in nearly one-third of human synapses. ABAT in liver and brain is controlled by 2 codominant alleles with a frequency in a Caucasian population of 0.56 and 0.44. The ABAT deficiency phenotype includes psychomotor retardation, hypotonia, hyperreflexia, lethargy, refractory seizures, and EEG abnormalities. Two alternatively spliced transcript variants encoding the same protein isoform have been found for this gene. MM
ABTD	2	Touster O, Shaw DR	Biochemistry of the acyclic polyols	Physiol Rev	1962	13922173	- authors infer L-arabitol dehydrogenase exists in humans based on observations of patient with presumed deficiency (L-arabinose in diet led to high excretion of L-arabitol and arabinoic acid (-lactone)) [Onkenhout, Mol Genet Metab 2002] - accumulated L-xylulose in pentosuric individuals probably is reduced in part to L-arabitol by the relatively nonspecific DPN-linked polyol dehydrogenase; urinary L-arabitol is in fact labeled when a pentosuric is given D-glucuronolactone-C13 (see refs in [Touster 1962]) - utilization of D-arabitol-C14 and L-arabitol-C14 in rat has been studied; D-isomer led to extremely low labeling of liver glycogen, but the L-isomer caused appreciable incorporation of the isotope (see refs in [Touster 1962])

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
ABUTD	3	Lin SW, Chen JC, Hsu LC, Hsieh CL, Yoshida A.	Human gamma-aminobutyraldehyde dehydrogenase (ALDH9): cDNA sequence, genomic organization, polymorphism, chromosomal localization, and tissue expression.		1996	8786138	gene and reaction characterized
ACACT1	3	Olivier LM, Krisans SK	Peroxisomal protein targeting and identification of peroxisomal targeting signals in cholesterol biosynthetic enzymes	Biochimica et Biophysica Acta	2000		ACAT1 - mitochondrial: involved in beta ox ACAT2 - cytoplasmic - involved in HMG-CoA synthesis for subsequent xol biosynthesis also peroxisomal and mitochondrial subtypes - see liter refs uniprot + lit NJ
ACACt2m	3	Halestrap AP.	Pyruvate and ketone-body transport across the mitochondrial membrane. Exchange properties, pH-dependence and mechanism of the carrier.	Biochem J	1978	28726	- Added by RS/TV - No genes found. "acac can also travel by diffusion" Halestrap AP Biochem J. 1978 Jun 15;172(3):377-87. Pyruvate and ketone-body transport across the mitochondrial membrane. Exchange properties, pH-dependence and mechanism of the carrier - Possible mechanism for acetoacetate transport via proton symport. Pande SV, Parvin R J Biol Chem. 1978 Mar 10;253(5):1565-73. Pyruvate and acetoacetate transport in mitochondria. A reappraisal
ACACt2m	3	Pande SV, Parvin R.	Pyruvate and acetoacetate transport in mitochondria. A reappraisal.	J Biol Chem	1975	627555	- Added by RS/TV - No genes found. "acac can also travel by diffusion" Halestrap AP Biochem J. 1978 Jun 15;172(3):377-87. Pyruvate and ketone-body transport across the mitochondrial membrane. Exchange properties, pH-dependence and mechanism of the carrier - Possible mechanism for acetoacetate transport via proton symport. Pande SV, Parvin R J Biol Chem. 1978 Mar 10;253(5):1565-73. Pyruvate and acetoacetate transport in mitochondria. A reappraisal
ACACT8p	3	Bout A, Franse MM, Collins J, Blonden L, Tager JM, Benne R.	Characterization of the gene encoding human peroxisomal 3-oxoacyl-CoA thiolase (ACAA). No large DNA rearrangement in a thiolase-deficient patient.	Biochim Biophys Acta	1991	1679347	Acetyl-Coenzyme A acyltransferase (ACAA1) is an enzyme operative in the beta-oxidation system of the peroxisomes. Deficiency of this enzyme leads to pseudo-Zellweger syndrome. NJ
ACALDtm	2	Lieber CS	Ethanol metabolism, cirrhosis and alcoholism	Clin Chim Acta	1997	9028626	- in hepatocytes, acetaldehyde is transported from cytosol to mitochondria for oxidation to acetate (see [Lieber 1997])
ACCOACm	3	Abu-Elheiga L, Brinkley WR, Zhong L, Chirala SS, Woldegiorgis G, Wakil SJ.	The subcellular localization of acetyl-CoA carboxylase 2.	Proc Natl Acad Sci U S A	2000	10677481	- Added by RS/TV NOTE: This reaction was under the accoa subsystem in Thuy's model; however, since all the reactions in this subsystem are scattered throughout various pathways in this model, I haven't assigned a subsystem. - ACC catalyzes the carboxylation of acetyl-CoA to form malonyl-CoA. - There are two isozymes enocded by separate genes: 1) Acaca (ACC1) are highly expressed in lipogenic tissues, such as liver and adipose. 7 transcriptional variants have been found for this protein. 2) Acacb(ACC2) is also expressed in the liver, but is the predominant form of carboxylase in heart and skeletal muscle. Both isozymes are shown to occur in the mitochondria. All this according to Abu-Elheiga L, Brinkley WR, Zhong L, Chirala SS, Woldegiorgis G, Wakil SJ.Proc Natl Acad Sci U S A. 2000 Feb 15;97(4):1444-9.
ACCOAgt	3	Hirabayashi Y, Kanamori A, Nomura KH, Nomura K.	The acetyl-CoA transporter family SLC33.	Pflugers Arch	2004	12739170	AcCoa transport into Golgi/ER where o-acetylation can occur. Varying evidence for cytoplasmic vs Golgi localization of rxn. Hirabayashi paper describing cloning (PMID: 9096318) supports cytoplasmic localization, but Hirabayashi paper reviewing SLC33A1 (PMID: 12739170) describes rxn occuring in Golgi lumen. Uniprot: The encoded protein is required for the formation of O acetylated (Ac) gangliosides. It is predicted to contain 6 to 10 transmembrane domains, and a leucine zipper motif in transmembrane domain III. Studies indicate that the protein is localized to the cytoplasm. NJ

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
ACGALtly	2	Jonas AJ, Speller RJ, Conrad PB, Dubinsky WP	Transport of N-acetyl-D-glucosamine and N-acetyl-D-galactosamine by rat liver lysosomes	J Biol Chem	1989	2784441	-N-acetyl-glucosamine and N-acetyl-galactosamine are transported out of rat liver lysosome via carrier-mediated transport; biochemical characterization described in [Jonas 1989] - transport was not dependent upon NaCl, KCl, MgCl2, or ATP/MgCl2 and was unaffected by 5 mM dithiothreitol or variation of buffer pH between 6.0 and 8.0 [Jonas 1989] - studies with cultured human fibroblasts have shown that free N-acetyl-glucosamine and N-acetyl-galactosamine are released from lysosomes and recycled by the cell [Rome 1986]
ACGALtly	2	Rome LH, Hill DF	Lysosomal degradation of glycoproteins and glycosaminoglycans. Efflux and recycling of sulphate and N-acetylhexosamines	Biochem J	1986	3753439	-N-acetyl-glucosamine and N-acetyl-galactosamine are transported out of rat liver lysosome via carrier-mediated transport; biochemical characterization described in [Jonas 1989] - transport was not dependent upon NaCl, KCl, MgCl2, or ATP/MgCl2 and was unaffected by 5 mM dithiothreitol or variation of buffer pH between 6.0 and 8.0 [Jonas 1989] - studies with cultured human fibroblasts have shown that free N-acetyl-glucosamine and N-acetyl-galactosamine are released from lysosomes and recycled by the cell [Rome 1986]
ACGAM2E	3	Luchansky SJ, Yarema KJ, Takahashi S, Bertozzi CR	GlcNAc 2-epimerase can serve a catabolic role in sialic acid metabolism	J Biol Chem	2003	12499362	- reversible [Luchansky, J Biol Chem 2003]
ACGAMK	3	Hinderlich S, Berger M, Schwarzkopf M, Effertz K, Reutter W	Molecular cloning and characterization of murine and human N-acetylglucosamine kinase	Eur J Biochem	2000	10824116	- gene has been cloned and characterized [Hinderlich, Eur J Biochem 2000] - reaction shown as irreversible in Varki p. 74 - kinase can use either GlcNAc or ManNac [Varki, p. 77]
ACGAMPM	3	Mio T, Yamada-Okabe T, Arisawa M, Yamada-Okabe H	Functional cloning and mutational analysis of the human cDNA for phosphoacetylglucosamine mutase: identification of the amino acid residues essential for the catalysis	Biochim Biophys Acta	2000	11004509	- shown as reversible in Devlin p. 672, Varki p. 74, Orten p. 246 - Found in many tissues except lung. Relatively high expression in pancreas, heart, liver, and placenta, and relatively low expression in brain, skeletal muscle and kidney [UniProt]
ACGPID	3	Watanabe R, Ohishi K, Maeda Y, Nakamura N, Kinoshita T	Mammalian PIG-L and its yeast homologue Gpi12p are N-acetylglucosaminylphosphatidylinositol de-N-acetylases essential in glycosylphosphatidylinositol biosynthesis	Biochem J	1999	10085243	- catalyzes de-N-acetylation of GlcNAc-PI [RefSeq], [UniProt] - gene was identified, 77% similarity to rat homolog [Watanabe, Biochem J 1999] - protein was characterized, localization analyzed Pottekat, J Biol Chem 2004]
ACGPID	3	Pottekat A, Menon AK	Subcellular localization and targeting of N-acetylglucosaminyl phosphatidylinositol de-N-acetylase, the second enzyme in the glycosylphosphatidylinositol biosynthetic pathway	J Biol Chem	2004	14742432	- catalyzes de-N-acetylation of GlcNAc-PI [RefSeq], [UniProt] - gene was identified, 77% similarity to rat homolog [Watanabe, Biochem J 1999] - protein was characterized, localization analyzed Pottekat, J Biol Chem 2004]
ACGSm	3	Caldovic L, Morizono H, Gracia Panglao M, Gallegos R, Yu X, Shi D, Malamy MH, Allewell NM, Tuchman M	Cloning and expression of the human N-acetylglutamate synthase gene	Biochem Biophys Res Commun	2002	12459178	Gene characterized and sequence encodes functional enzyme in E. coli system. Biochemical evidence noted, but is perhaps questionable.
ACHEe	3	Shafferman A, Kronman C, Flashner Y, Leitner M, Grosfeld H, Ordentlich A, Gozes Y, Cohen S, Ariel N, Barak D, et al.	Mutagenesis of human acetylcholinesterase. Identification of residues involved in catalytic activity and in polypeptide folding.	J Biol Chem	1992		extraceullular - from phys data - no refs found for other compartments - f/u in future Acetylcholinesterase hydrolyzes the neurotransmitter, acetylcholine at neuromuscular junctions and brain cholinergic synapses, and thus terminates signal transmission. It is also found on the red blood cell membranes, where it constitutes the Yt blood group antigen. Acetylcholinesterase exists in multiple molecular forms which possess similar catalytic properties, but differ in their oligomeric assembly and mode of cell attachment to the cell surface. It is encoded by the single ACHE gene, and the structural diversity in the gene products arises from alternative mRNA splicing, and post-translational associations of catalytic and structural subunits. The major form of acetylcholinesterase found in brain, muscle and other tissues is the hydrophilic species, which forms disulfide-linked oligomers with collagenous, or lipid-containing structural subunits. The other, alternatively spliced form, expressed primarily in the erythroid tissues, differs at the C-terminal end, and contains a cleavable hydrophobic peptide with a GPI-anchor site. It associ NJ

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ACHEe	3	Soreq H, Seidman S.	Acetylcholinesterase--new roles for an old actor.	Nat Rev Neurosci	2001	11283752	extraceullular - from phys data - no refs found for other compartments - f/u in future Acetylcholinesterase hydrolyzes the neurotransmitter, acetylcholine at neuromuscular junctions and brain cholinergic synapses, and thus terminates signal transmission. It is also found on the red blood cell membranes, where it constitutes the Yt blood group antigen. Acetylcholinesterase exists in multiple molecular forms which possess similar catalytic properties, but differ in their oligomeric assembly and mode of cell attachment to the cell surface. It is encoded by the single ACHE gene, and the structural diversity in the gene products arises from alternative mRNA splicing, and post-translational associations of catalytic and structural subunits. The major form of acetylcholinesterase found in brain, muscle and other tissues is the hydrophilic species, which forms disulfide-linked oligomers with collagenous, or lipid-containing structural subunits. The other, alternatively spliced form, expressed primarily in the erythroid tissues, differs at the C-terminal end, and contains a cleavable hydrophobic peptide with a GPI-anchor site. It associ NJ
ACITL	0	Elshourbagy NA, Near JC, Kmetz PJ, Sathe GM, Southan C, Strickler JE, Gross M, Young JF, Wells TN, Groot PH.	Rat ATP citrate-lyase. Molecular cloning and sequence analysis of a full-length cDNA and mRNA abundance as a function of diet, organ, and age	J Biol Chem	1990	2295639	- responsible for cytosolic accoa production in many tissues [RefSeq] - cytoplasmic [UniProt] - Additional info added by RS/TV: Myriame Poirier1, Geneviève Vincent1, et al. Probing the link between citrate and malonyl-CoA in perfused rat hearts. Am J Physiol Heart Circ Physiol. 2002 Oct;283(4):H1379-86. High activity in liver, lower in heart Cytosolic according to Entrez Gene Database Catalytic Activity: Catalyzes the formation of acetyl-CoA and oxaloacetate from citrate and CoA with a concomitant hydrolysis of ATP to ADP and phosphate Two transcript variants according to Entrez Gene Database. Tissue Specificity: Although expressed in a wide variety of tissues, it is found to be in greater concentrations in the liver, kidney, lung, and brain. All this according to Elshourbagy NA, etc; J Biol Chem. 1990 Jan 25;265(3):1430-5; Rat ATP citrate-lyase. Molecular cloning and sequence analysis of a full-length cDNA and mRNA abundance as a function of diet, organ, and age. Awan MM, Saggerson ED.; Biochem J. 1993 Oct 1;295 (Pt 1):61-6; Malonyl-CoA metabolism in cardiac myocytes and its Pierce MW, Palmer JL, Keutmann HT, Avruch J; J Biol Chem. ATP-citrate lyase. Structure of a tryptic peptide containing the
ACN13ACNGALGBSIDEtg	1	Sandhoff K, Klein A.	Intracellular trafficking of glycosphingolipids: role of sphingolipid activator proteins in the topology of endocytosis and lysosomal digestion.	FEBS Lett	1994	8206147	Since specific mechanism is unknown (caveoli vs vesicular vs ...) and energy dependence/requirement is not known either, evidence is left as modeling only, however intracellular trafficking and transport is known to occur for sphingolipids, additionally some forms are transferred to the outer plasma membrane, so a mechanism for EC transport is also present. See Varki glycolipids text (notably glycosphingolipids chapter - p121) and various literature reviews (PMID: 8206147). NJ
ACNAM9PL	3	Lawrence SM, Huddleston KA, Pitts LR, Nguyen N, Lee YC, Vann WF, Coleman TA, Betenbaugh MJ	Cloning and expression of the human N-acetylneuraminic acid phosphate synthase gene with 2-keto-3-deoxy-D-glycero- D-galacto-nononic acid biosynthetic ability	J Biol Chem	2000	10749855	- shown as irreversible in Devlin p. 672 & 677, Orten p.245, Varki p. 74 - NeuAcP synthase uses ManNAc6P and man6p as substrates, but exhibits much higher activity towards ManNAc6P [Lawrence, J Biol Chem 2000] - unlike the E. coli homolog, the human enzyme only uses phosphorylate substrates [Lawrence, J Biol Chem 2000] - cytosolic [Varki, p.78]
ACOAD10m	3	Andresen,B.S., Christensen,E., Corydon,T.J., Bross,P., etc.	Isolated 2-methylbutyrylglycinuria caused by short/branched-chain acyl-CoA dehydrogenase deficiency....		2000	11013134	0
ACOAD8m	3	Tiffany,K.A., Roberts,D.L., Wang,M., Paschke,R., Mohsen,A.W., Vockley,J., Kim,J.-J.P.,	Structure of human isovaleryl-CoA dehydrogenase at 2.6-A resolution: structural basis for substrate specificity. .		1997	9214289	mitochondrial, FAD used as cofactor - see references MM
ACOAD8m	3	Vockley J, Nagao M, Parimoo B, Tanaka K.	The variant human isovaleryl-CoA dehydrogenase gene responsible...		1992		mitochondrial, FAD used as cofactor - see references MM

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ACOAO7p	3	Chu R, Varanasi U, Chu S, Lin Y, Usuda N, Rao MS, Reddy JK.	Overexpression and characterization of the human peroxisomal acyl-CoA oxidase in insect cells.	J Biol Chem	1995	7876265	peroxisomal - uniprot seq + kinetic data in chu ref Defects in ACOX1 are the cause of pseudoneonatal adrenoleukodystrophy [MIM:264470]. It is a disease biochemically characterized by an accumulation of very long chain fatty acids. NJ
ACOAO7p	3	Varanasi U, Chu R, Chu S, Espinosa R, LeBeau MM, Reddy JK.	Isolation of the human peroxisomal acyl-CoA oxidase gene: organization, promoter analysis, and chromosomal localization.	Proc Natl Acad Sci	1994	8159712	peroxisomal - uniprot seq + kinetic data in chu ref Defects in ACOX1 are the cause of pseudoneonatal adrenoleukodystrophy [MIM:264470]. It is a disease biochemically characterized by an accumulation of very long chain fatty acids. NJ
ACODA	1	Jones WM, Scaloni A, Bossa F, Popowicz AM, Schneewind O, Manning JM	Genetic relationship between acylpeptide hydrolase and acylase, two hydrolytic enzymes with similar binding but different catalytic specificities	Proc Natl Acad Sci U S A	1991	2006156	based on KEGG map and broad enzyme specificity
ACOX2x	3	Baumgart E, Vanhooren JCT, Fransen M, Marynen P, Puype M, Vandekerckhove J, Leunissen JAM, Fahimi HD, Mannaerts GP, Veldhoven PP	Molecular characterization of the human peroxisomal branched-chain acyl-CoA oxidase	Proc Natl Acad Sci	1996		peroxisome - literature The product of this gene belongs to the acyl-CoA oxidases family. It encodes the branched-chain acyl-CoA oxidase which is involved in the degradation of long branched fatty acids and bile acid intermediates in peroxisomes. Deficiency of this enzyme results in the accumulation of branched fatty acids and bile acid intermediates, and may lead to Zellweger syndrome, severe mental retardation and death in children. NJ
ACRNtm	3	Pande SV, Parvin R.	Characterization of carnitine acylcarnitine translocase system of heart mitochondria.	J Biol Chem	1976	977593	- Added by RS/TV Pande SV, Parvin R. J Biol Chem. 1976 Nov 10;251(21):6683-91 Characterization of carnitine acylcarnitine translocase system of heart mitochondria Mitochondrial according to Entrez gene database. Reaction description: Slc25a20.1-m catalyses the electroneutral exchange of cytosolic acylcarnitine for mitochondrial carnitine. Thus preferred substrates include carnitine and acylcarnitine in exchange for carnitine or acylcarnitines. Tissue localization: Slc25a20.1-m is expressed in the heart, skeletal muscle, liver, lung, kidney, brain, pancreas, and placenta. All of this according to Table 1 in Palmieri, F. The mitochondrial transporter family (SLC25): physiological and pathological implications. Pflugers Arch. 2004 Feb. (PMID: 14598172)
ACS	3	Luong A, Hannah VC, Brown MS, Goldstein JL	Molecular characterization of human acetyl-CoA synthetase, an enzyme regulated by sterol regulatory element-binding proteins	J Biol Chem	2000	10843999	55902: - catalyzes irreversible reaction between acetate and ATP to acetyl-CoA [RefSeq], [Luong, J Biol Chem 2000] - cytosolic [LocusLink], [UniProt], [Luong, J Bil Chem 2000] 65985: - annotated as 6.2.1.1 [HInv-Db] - cytosolic [Ohgami et al,Biochem Pharmacol. 2003 Mar 15;65(6):989-94]
ACS	3	Ohgami M, Takahashi N, Yamasaki M, Fukui T	Expression of acetoacetyl-CoA synthetase, a novel cytosolic ketone body-utilizing enzyme, in human brain	Biochem Pharmacol	2003	12623130	55902: - catalyzes irreversible reaction between acetate and ATP to acetyl-CoA [RefSeq], [Luong, J Biol Chem 2000] - cytosolic [LocusLink], [UniProt], [Luong, J Bil Chem 2000] 65985: - annotated as 6.2.1.1 [HInv-Db] - cytosolic [Ohgami et al,Biochem Pharmacol. 2003 Mar 15;65(6):989-94]

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ACSm	2	Scholte HR, Wit-Peeters EM, Bakker JC.	The intracellular and intramitochondrial distribution of short-chain acyl-CoA synthetases in guinea-pig heart.	Biochim Biophys Acta	1971	4326157	- catalyzes irreversible reaction between acetate and ATP to acetyl-CoA [RefSeq] - mitochondrial matrix [UniProt] - primarily a cardiac enzyme [RefSeq] - enzyme has been characterized in mouse and cow, assumed to have a similar function in humans [Fujino, J Biol Chem 2001] - Additional information by RS/TV: 1) localized in the mitochondria 2) Loosely bound to the inner side of the inner membrane 3) Found to be localized in the gut, liver, and heart. 4) In rat-liver it is found to be mainly localized on the outer membrane All this according to H. R. Scholte, E. M. Wit-Peeters and J. C. Bakker The intraçellular and intramitochondrial distribution of short-chain acyl-CoA synthetases in guinea-pig heart
ACSm	2	Fujino T, Kondo J, Ishikawa M, Morikawa K, Yamamoto TT	Acetyl-CoA synthetase 2, a mitochondrial matrix enzyme involved in the oxidation of acetate	J Biol Chem	2001	11150295	- catalyzes irreversible reaction between acetate and ATP to acetyl-CoA [RefSeq] - mitochondrial matrix [UniProt] - primarily a cardiac enzyme [RefSeq] - enzyme has been characterized in mouse and cow, assumed to have a similar function in humans [Fujino, J Biol Chem 2001] - Additional information by RS/TV: 1) localized in the mitochondria 2) Loosely bound to the inner side of the inner membrane 3) Found to be localized in the gut, liver, and heart. 4) In rat-liver it is found to be mainly localized on the outer membrane All this according to H. R. Scholte, E. M. Wit-Peeters and J. C. Bakker The intraçellular and intramitochondrial distribution of short-chain acyl-CoA synthetases in guinea-pig heart
ACSRTNMT	3	Donohue SJ, Roseboom PH, Illnerova H, Weller JL, Klein DC	Human hydroxyindole-O-methyltransferase: presence of LINE-1 fragment in a cDNA clone and pineal mRNA	DNA Cell Biol	1993	8397829	Catalyzes the last step in melatonin synthesis.
ACt2m	2	Casal M, Paiva S, Andrade RP, Gancedo C, Leao C.	The lactate-proton symport of Saccharomyces cerevisiae is encoded by JEN1.	J Bacteriol	1999	10198029	- Added by RS/TV - No genes found. - However, there is strong physiological results indicating the existence of different monocarboxylate permeases in S. cerevisiae, four open reading frames (ORFs) with important similarities to mammalian monocarboxylate permeases were found in the genome of S. cerevisiae. (Casal M, Paiva S, Andrade RP, Gancedo C, Leao C.J Bacteriol. 1999 Apr;181(8):2620-3.) - [Wolfe 2005] suggest that acetate can freely diffuse across subcellular membranes since it is lipophilic
ACt2m	2	Wolfe AJ.	The acetate switch	Microbiol Mol Biol Rev	2005	15755952	- Added by RS/TV - No genes found. - However, there is strong physiological results indicating the existence of different monocarboxylate permeases in S. cerevisiae, four open reading frames (ORFs) with important similarities to mammalian monocarboxylate permeases were found in the genome of S. cerevisiae. (Casal M, Paiva S, Andrade RP, Gancedo C, Leao C.J Bacteriol. 1999 Apr;181(8):2620-3.) - [Wolfe 2005] suggest that acetate can freely diffuse across subcellular membranes since it is lipophilic
ACt2r	3	Waniewski RA, Martin DL.	Preferential utilization of acetate by astrocytes is attributable to transport.	J Neurosci	1998	9651205	- acetate generated from ethanol in hepatocytes can be excreted into the blood [Smith 2004] or be taken up by other cells such as astrocytes - acetate uptake by astrocytes has many of the properties of the proton-coupled, monocarboxylate transport process found in erythrocytes, heart, kidney, and skeletal muscle cells [Waniewski 1998]
ACt2r	3	Colleen Smith, Allan Marks, Michael Lieberman	Marks' Basic Medical Biochemistry		2004		- acetate generated from ethanol in hepatocytes can be excreted into the blood [Smith 2004] or be taken up by other cells such as astrocytes - acetate uptake by astrocytes has many of the properties of the proton-coupled, monocarboxylate transport process found in erythrocytes, heart, kidney, and skeletal muscle cells [Waniewski 1998]
ADA	3	Iwaki-Egawa S, Watanabe Y.	Characterization and purification of adenosine deaminase 1 from human and chicken liver.	Comp Biochem Physiol B Biochem Mol Biol	2002	12381379	some paper mentioned a ADA2, however, I could not find a gene for it --> keep in mind IT
ADA	3	Gonzalez-Gronow M, Hershfield MS, Arredondo-Vega FX, Pizzo SV.	Cell surface adenosine deaminase binds and stimulates plasminogen activation on 1-LN human prostate cancer cells.	J Biol Chem	2004	15016824	some paper mentioned a ADA2, however, I could not find a gene for it --> keep in mind IT

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ADK1	3	Zuffardi O, Caiulo A, Maraschio P, Tupler R, Bianchi E, Amisano P, Beluffi G, Moratti R, Liguri G.	Regional assignment of the loci for adenylate kinase to 9q32 and for alpha 1-acid glycoprotein to 9q31-q32. A locus for Goltz syndrome in region 9q32-qter?	Hum Genet	1989	2541064	IT
ADK1	3	Matsuura S, Igarashi M, Tanizawa Y, Yamada M, Kishi F, Kajii T, Fujii H, Miwa S, Sakurai M, Nakazawa A.	Human adenylate kinase deficiency associated with hemolytic anemia. A single base substitution affecting solubility and catalytic activity of the cytosolic adenylate kinase.	J Biol Chem	1989	2542324	IT
ADK1	3	Miwa S, Fujii H, Tani K, Takahashi K, Takizawa T, Igarashi T.	Red cell adenylate kinase deficiency associated with hereditary nonspherocytic hemolytic anemia: clinical and biochemical studies.	Am J Hematol	1983	6305188	IT
ADK1	3	Van Rompay AR, Johansson M, Karlsson A.	Identification of a novel human adenylate kinase. cDNA cloning, expression analysis, chromosome localization and characterization of the recombinant protein.	Eur J Biochem	1999	10215863	IT
ADK1m	3	Bruns GA, Regina VM.	Adenylate kinase 2, a mitochondrial enzyme.	Biochem Genet	1977	195572	IT 204:mRNAs:strongly expressed in liver, heart, skeletal muscle and pancreas, and moderately in kidney, placenta and brain, and weakly in lung AK2 protein was present in large amounts in liver, heart, kidney, and in a small amount in lung, and undetectable in brain and skeletal muscle
ADK1m	3	Lee Y, Kim JW, Lee IA, Kang HB, Choe YK, Lee HG, Lim JS, Kim HJ, Park C, Choe IS.	Cloning and characterization of cDNA for human adenylate kinase 2A.	Biochem Mol Biol Int	1996	8843353	IT 204:mRNAs:strongly expressed in liver, heart, skeletal muscle and pancreas, and moderately in kidney, placenta and brain, and weakly in lung AK2 protein was present in large amounts in liver, heart, kidney, and in a small amount in lung, and undetectable in brain and skeletal muscle
ADK1m	3	Noma T, Fujisawa K, Yamashiro Y, Shinohara M, Nakazawa A, Gondo T, Ishihara T, Yoshinobu K.	Structure and expression of human mitochondrial adenylate kinase targeted to the mitochondrial matrix.	Biochem J	2001	11485571	IT 204:mRNAs:strongly expressed in liver, heart, skeletal muscle and pancreas, and moderately in kidney, placenta and brain, and weakly in lung AK2 protein was present in large amounts in liver, heart, kidney, and in a small amount in lung, and undetectable in brain and skeletal muscle
ADK3	3	Noma T, Song S, Yoon YS, Tanaka S, Nakazawa A.	cDNA cloning and tissue-specific expression of the gene encoding human adenylate kinase isozyme 2.	Biochim Biophys Acta	1998	9434148	IT
ADMDC	3	Pajunen A, Crozat A, Janne OA, Ihalainen R, Laitinen PH, Stanley B, Madhubala R, Pegg AE	Structure and regulation of mammalian S-adenosylmethionine decarboxylase	J Biol Chem	1988	2460457	Enzyme and reaction characterized
ADNCYC	3	Stengel D, Parma J, Gannage MH, Roeckel N, Mattei MG, Barouki R, Hanoune J.	Different chromosomal localization of two adenylyl cyclase genes expressed in human brain.	Hum Genet	1992	1427768	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNCYC	3	Hellevuo K, Berry R, Sikela JM, Tabakoff B.	Localization of the gene for a novel human adenylyl cyclase (ADCY7) to chromosome 16.	Hum Genet	1995	7860067	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells

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ADNCYC	3	Gaudin C, Homcy CJ, Ishikawa Y.	Mammalian adenylyl cyclase family members are randomly located on different chromosomes.	Hum Genet	1994	7959689	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNCYC	3	Defer N, Marinx O, Stengel D, Danisova A, Iourgenko V, Matsuoka I, Caput D, Hanoune J.	Molecular cloning of the human type VIII adenylyl cyclase.	FEBS Lett	1994	8076676	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNCYC	3	Raimundo S, Giray J, Volff JN, Schwab M, Altenbuchner J, Ratge D, Wisser H.	Cloning and sequence of partial cDNAs encoding the human type V and VI adenylyl cyclases and subsequent RNA-quantification in various tissues.	Clin Chim Acta	1999	10481931	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNCYC	3	Patrizio M, Colucci M, Levi G.	Human immunodeficiency virus type 1 Tat protein decreases cyclic AMP synthesis in rat microglia cultures.	J Neurochem	2001	11299302	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells

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ADNCYC	3	Jaiswal BS, Conti M.	Identification and functional analysis of splice variants of the germ cell soluble adenylyl cyclase.	J Biol Chem	2001	11423534	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNCYC	3	Toyota T, Hattori E, Meerabux J, Yamada K, Saito K, Shibuya H, Nankai M, Yoshikawa T.	Molecular analysis, mutation screening, and association study of adenylate cyclase type 9 gene (ADCY9) in mood disorders.	Am J Med Genet	2002	11840511	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNCYC	3	Ludwig MG, Seuwen K.	Characterization of the human adenylyl cyclase gene family: cDNA, gene structure, and tissue distribution of the nine isoforms.	J Recept Signal Transduct Res	2002	12503609	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNCYC	3	Litvin TN, Kamenetsky M, Zarifyan A, Buck J, Levin LR.	Kinetic properties of "soluble" adenylyl cyclase. Synergism between calcium and bicarbonate.	J Biol Chem	2003	12609998	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells

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ADNCYC	3	Katsel PL, Tagliente TM, Schwarz TE, Craddock-Royal BD, Patel ND, Maayani S.	Molecular and biochemical evidence for the presence of type III adenylyl cyclase in human platelets.	Platelets	2003	12623444	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNCYC	3	Cumbay MG, Watts VJ.	Novel regulatory properties of human type 9 adenylate cyclase.	J Pharmacol Exp Ther	2004	14996950	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNCYC	3	Ding Q, Gros R, Chorazyczewski J, Ferguson SS, Feldman RD.	Isoform-specific regulation of adenylyl cyclase function by disruption of membrane trafficking.	Mol Pharmacol	2005	15547246	membrane protein but Ding et al. 2005 report 2 cytoplasmatic domain of ADNCYC protein (in their introduction) IT 107(1): brain 108(2): brain (also olfactory cilia - locuslink) 109(3): paletes, thyroid, germ cells (also olfactory cilia - locuslink) 196883(4): thyroid (also olfactory cilia - locuslink) 111(5): colon, heart, liver, lung (mRNA 60 more abundant in heart than 112) Raimundo et al. 1999 112(6): colon, heart, liver, lung and MNL - Raimundo et al. 1999 113(7): paletes 114(8):brain, May be involved in learning, in memory and in drug dependence.(Genecards) 115(9): brain, neuronal signaling all bind 2 Mg2+ per subunit some isoforms are sensitive to calmodulin/calcium other ones are not 55811(SAC): soluble ADNCYC - is regulated by bicarbonate instead of protein G; broadly present in human tissue, also germ cells
ADNK1	3	McNally T, Helfrich RJ, Cowart M, Dorwin SA, Meuth JL, Idler KB, Klute KA, Simmer RL, Kowaluk EA, Halbert DN.	Cloning and expression of the adenosine kinase gene from rat and human tissues.	Biochem Biophys Res Commun	1997	9070863	monomer needs Mg2+ IT
ADPRDP	3	Gasmi L, Cartwright JL, McLennan AG.	Cloning, expression and characterization of YSA1H, a human adenosine 5'-diphosphosugar pyrophosphatase possessing a MutT motif.	Biochem J	1999	10567213	no information for compartiment. Activity is relatively the same for Mannose, Glucose and ribose. Enzyme cannot act on ribo - or deoxyribonucleoside-tri-phosphates. Needs Mg2+. Is widely expressed, but most abundant in liver (Genecards) ADPglucose is an important precursor bacterial glycogen and plant starch synthesis, whereas ADP-mannose has no known physiological function, although the commercially available synthetic compound can replace ADP-heptoses in bacterial outer-membrane lipopolysaccharide synthesis in vitro . ADP-ribose might therefore also be the most important substrate for human in vivo (from discussion of Gasmi et al, 1999) IT

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
ADPRDP	3	Yang H, Slupska MM, Wei YF, Tai JH, Luther WM, Xia YR, Shih DM, Chiang JH, Baikalov C, Fitz-Gibbon S, Phan IT, Conrad A, Miller JH.	Cloning and characterization of a new member of the Nudix hydrolases from human and mouse.	J Biol Chem	2000	10722730	no information for compartiment. Activity is relatively the same for Mannose, Glucose and ribose. Enzyme cannot act on ribo - or deoxyribonucleoside-tri-phosphates. Needs Mg2+. Is widely expressed, but most abundant in liver (Genecards) ADPglucose is an important precursor bacterial glycogen and plant starch synthesis, whereas ADP-mannose has no known physiological function, although the commercially available synthetic compound can replace ADP-heptoses in bacterial outer-membrane lipopolysaccharide synthesis in vitro . ADP-ribose might therefore also be the most important substrate for human in vivo (from discussion of Gasmi et al, 1999) IT
ADPRDP	3	Ishibashi T, Hayakawa H, Sekiguchi M.	A novel mechanism for preventing mutations caused by oxidation of guanine nucleotides.	EMBO Rep	2003	12717453	no information for compartiment. Activity is relatively the same for Mannose, Glucose and ribose. Enzyme cannot act on ribo - or deoxyribonucleoside-tri-phosphates. Needs Mg2+. Is widely expressed, but most abundant in liver (Genecards) ADPglucose is an important precursor bacterial glycogen and plant starch synthesis, whereas ADP-mannose has no known physiological function, although the commercially available synthetic compound can replace ADP-heptoses in bacterial outer-membrane lipopolysaccharide synthesis in vitro . ADP-ribose might therefore also be the most important substrate for human in vivo (from discussion of Gasmi et al, 1999) IT
ADPRDPm	3	Perraud AL, Shen B, Dunn CA, Rippe K, Smith MK, Bessman MJ, Stoddard BL, Scharenberg AM.	NUDT9, a member of the Nudix hydrolase family, is an evolutionarily conserved mitochondrial ADP-ribose pyrophosphatase.	J Biol Chem	2003	12427752	IT location in mito based on signal-sequence
ADPRDPm	3	Shen BW, Perraud AL, Scharenberg A, Stoddard BL.	The crystal structure and mutational analysis of human NUDT9.	J Mol Biol	2003	12948489	IT location in mito based on signal-sequence
ADPRDPm	3	Kuhn FJ, Luckhoff A.	Sites of the NUDT9-H domain critical for ADP-ribose activation of the cation channel TRPM2.	J Biol Chem	2005	15347676	IT location in mito based on signal-sequence
ADPT	3	Crespillo J, Llorente P, Argomaniz L, Montero C.	APRT from erythrocytes of HGPRT deficient patients: kinetic, regulatory and thermostability properties.	Mol Cell Biochem	2003	14674717	no infos for compartiment IT purine salvage
ADPT	3	Silva M, Silva CH, Iulek J, Thiemann OH.	Three-dimensional structure of human adenine phosphoribosyltransferase and its relation to DHA-urolithiasis.	Biochemistry	2004	15196008	no infos for compartiment IT purine salvage
ADSL1	3	Stone RL, Aimi J, Barshop BA, Jaeken J, Van den Berghe G, Zalkin H, Dixon JE.	A mutation in adenylosuccinate lyase associated with mental retardation and autistic features.	Nat Genet	1992	1302001	IT no info for compartiment
ADSL1	3	Barton JW, Hart IM, Patterson D.	Mapping of a locus correcting lack of phosphoribosylaminoimidazole carboxylase activity in Chinese hamster ovary cell Ade-D mutants to human chromosome 4.	Genomics	1991	2004782	IT no info for compartiment
AG13T4g	3	Sasaki K, Kurata-Miura K, Ujita M, Angata K, Nakagawa S, Sekine S, Nishi T, Fukuda M	Expression cloning of cDNA encoding a human beta-1,3-N-acetylglucosaminyltransferase that is essential for poly-N-acetyllactosamine synthesis	Proc Natl Acad Sci U S A	1997	9405606	- N-glycans: most efficiently synthesized by beta4Gal-TI and iGnT; iGnT acts less efficiently on acceptors w/ large number of repeats, beta4Gal-TI exhibits no significant change - O-glycans: extension of core 4 branches synthesized most efficiently by iGnT and beta4Gal-TI; core 2 branch synthesis requires iGnT and beta4Gal-TIV - extension of core 4 branches less efficient than N-glycans, core 2 from [Shiraishi, J Biol Chem 2001]: - 10331, 10678, 79369 identified and characterized - 10331, 10678, 79369 involved in the initiation and elongation of poly-N-acetyllactosamine synthesis - sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] 10331: - Golgi [UniProt] - expressed in colon, jejunum, stomach, esophagus, placenta, and trachea [Shiraishi, J Biol Chem 2001] 10678: - Golgi [UniProt] - ubiquitously expressed [Shiraishi, J Biol Chem 2001] 79369: - Golgi [UniProt] - expressed in brain [Shiraishi, J Biol Chem 2001] 93010: - mouse gene was identified and characterized; human ortholog identified by BLAST, human and mouse genes have 87% ident 11041: - essential for NAcLac chain biosynthesis [RefSeq] - Golgi [UniProt] - gene was isolated and expressed [Sasaki, PNAS 1997]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
AG13T4g	3	Ujita M, Misra AK, McAuliffe J, Hindsgaul O, Fukuda M	Poly-N-acetyllactosamine extension in N-glycans and core 2- and core 4-branched O-glycans is differentially controlled by i-extension enzyme and different members of the beta 1,4-galactosyltransferase gene family	J Biol Chem	2000	10747980	- N-glycans: most efficiently synthesized by beta4Gal-TI and iGnT; iGnT acts less efficiently on acceptors w/ large number of repeats, beta4Gal-TI exhibits no significant change - O-glycans: extension of core 4 branches synthesized most efficiently by iGnT and beta4Gal-TI; core 2 branch synthesis requires iGnT and beta4Gal-TIV - extension of core 4 branches less efficient than N-glycans, core 2 from [Shiraishi, J Biol Chem 2001]: - 10331, 10678, 79369 identified and characterized - 10331, 10678, 79369 involved in the initiation and elongation of poly-N-acetyllactosamine synthesis - sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] 10331: - Golgi [UniProt] - expressed in colon, jejunum, stomach, esophagus, placenta, and trachea [Shiraishi, J Biol Chem 2001] 10678: - Golgi [UniProt] - ubiquitously expressed [Shiraishi, J Biol Chem 2001] 79369: - Golgi [UniProt] - expressed in brain [Shiraishi, J Biol Chem 2001] 93010: - mouse gene was identified and characterized; human ortholog identified by BLAST, human and mouse genes have 87% ident 11041: - essential for NAcLac chain biosynthesis [RefSeq] - Golgi [UniProt] - gene was isolated and expressed [Sasaki, PNAS 1997]
AG13T4g	3	Shiraishi N, Natsume A, Togayachi A, Endo T, Akashima T, Yamada Y, Imai N, Nakagawa S, Koizumi S, Sekine S, Narimatsu H, Sasaki K	Identification and characterization of three novel beta 1,3-N-acetylglucosaminyltransferases structurally related to the beta 1,3-galactosyltransferase family	J Biol Chem	2001	11042166	- N-glycans: most efficiently synthesized by beta4Gal-TI and iGnT; iGnT acts less efficiently on acceptors w/ large number of repeats, beta4Gal-TI exhibits no significant change - O-glycans: extension of core 4 branches synthesized most efficiently by iGnT and beta4Gal-TI; core 2 branch synthesis requires iGnT and beta4Gal-TIV - extension of core 4 branches less efficient than N-glycans, core 2 from [Shiraishi, J Biol Chem 2001]: - 10331, 10678, 79369 identified and characterized - 10331, 10678, 79369 involved in the initiation and elongation of poly-N-acetyllactosamine synthesis - sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] 10331: - Golgi [UniProt] - expressed in colon, jejunum, stomach, esophagus, placenta, and trachea [Shiraishi, J Biol Chem 2001] 10678: - Golgi [UniProt] - ubiquitously expressed [Shiraishi, J Biol Chem 2001] 79369: - Golgi [UniProt] - expressed in brain [Shiraishi, J Biol Chem 2001] 93010: - mouse gene was identified and characterized; human ortholog identified by BLAST, human and mouse genes have 87% ident 11041: - essential for NAcLac chain biosynthesis [RefSeq] - Golgi [UniProt] - gene was isolated and expressed [Sasaki, PNAS 1997]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
AG13T4g	3	Kataoka K, Huh NH	A novel beta1,3-N-acetylglucosaminyltransferase involved in invasion of cancer cells as assayed in vitro	Biochem Biophys Res Commun	2002	12061784	- N-glycans: most efficiently synthesized by beta4Gal-TI and iGnT; iGnT acts less efficiently on acceptors w/ large number of repeats, beta4Gal-TI exhibits no significant change - O-glycans: extension of core 4 branches synthesized most efficiently by iGnT and beta4Gal-TI; core 2 branch synthesis requires iGnT and beta4Gal-TIV - extension of core 4 branches less efficient than N-glycans, core 2 from [Shiraishi, J Biol Chem 2001]: - 10331, 10678, 79369 identified and characterized - 10331, 10678, 79369 involved in the initiation and elongation of poly-N-acetyllactosamine synthesis - sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] 10331: - Golgi [UniProt] - expressed in colon, jejunum, stomach, esophagus, placenta, and trachea [Shiraishi, J Biol Chem 2001] 10678: - Golgi [UniProt] - ubiquitously expressed [Shiraishi, J Biol Chem 2001] 79369: - Golgi [UniProt] - expressed in brain [Shiraishi, J Biol Chem 2001] 93010: - mouse gene was identified and characterized; human ortholog identified by BLAST, human and mouse genes have 87% ident 11041: - essential for NAcLac chain biosynthesis [RefSeq] - Golgi [UniProt] - gene was isolated and expressed [Sasaki, PNAS 1997]
AG13T4g	3	Zheng H, Li Y, Ji C, Li J, Zhang J, Yin G, Xu J, Ye X, Wu M, Zou X, Gu S, Xie Y, Mao Y	Characterization of a cDNA encoding a protein with limited similarity to beta1, 3-N-acetylglucosaminyltransferase	Mol Biol Rep	2004	15560372	- N-glycans: most efficiently synthesized by beta4Gal-TI and iGnT; iGnT acts less efficiently on acceptors w/ large number of repeats, beta4Gal-TI exhibits no significant change - O-glycans: extension of core 4 branches synthesized most efficiently by iGnT and beta4Gal-TI; core 2 branch synthesis requires iGnT and beta4Gal-TIV - extension of core 4 branches less efficient than N-glycans, core 2 from [Shiraishi, J Biol Chem 2001]: - 10331, 10678, 79369 identified and characterized - 10331, 10678, 79369 involved in the initiation and elongation of poly-N-acetyllactosamine synthesis - sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] 10331: - Golgi [UniProt] - expressed in colon, jejunum, stomach, esophagus, placenta, and trachea [Shiraishi, J Biol Chem 2001] 10678: - Golgi [UniProt] - ubiquitously expressed [Shiraishi, J Biol Chem 2001] 79369: - Golgi [UniProt] - expressed in brain [Shiraishi, J Biol Chem 2001] 93010: - mouse gene was identified and characterized; human ortholog identified by BLAST, human and mouse genes have 87% ident 11041: - essential for NAcLac chain biosynthesis [RefSeq] - Golgi [UniProt] - gene was isolated and expressed [Sasaki, PNAS 1997]

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AG13T4g	3	Ishida H, Togayachi A, Sakai T, Iwai T, Hiruma T, Sato T, Okubo R, Inaba N, Kudo T, Gotoh M, Shoda J, Tanaka N, Narimatsu H	A novel beta1,3-N-acetylglucosaminyltransferase (beta3Gn-T8), which synthesizes poly-N-acetyllactosamine, is dramatically upregulated in colon cancer	FEBS Lett	2005	15620693	- N-glycans: most efficiently synthesized by beta4Gal-TI and iGnT; iGnT acts less efficiently on acceptors w/ large number of repeats, beta4Gal-TI exhibits no significant change - O-glycans: extension of core 4 branches synthesized most efficiently by iGnT and beta4Gal-TI; core 2 branch synthesis requires iGnT and beta4Gal-TIV - extension of core 4 branches less efficient than N-glycans, core 2 from [Shiraishi, J Biol Chem 2001]: - 10331, 10678, 79369 identified and characterized - 10331, 10678, 79369 involved in the initiation and elongation of poly-N-acetyllactosamine synthesis - sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] 10331: - Golgi [UniProt] - expressed in colon, jejunum, stomach, esophagus, placenta, and trachea [Shiraishi, J Biol Chem 2001] 10678: - Golgi [UniProt] - ubiquitously expressed [Shiraishi, J Biol Chem 2001] 79369: - Golgi [UniProt] - expressed in brain [Shiraishi, J Biol Chem 2001] 93010: - mouse gene was identified and characterized; human ortholog identified by BLAST, human and mouse genes have 87% ident 11041: - essential for NAcLac chain biosynthesis [RefSeq] - Golgi [UniProt] - gene was isolated and expressed [Sasaki, PNAS 1997]
AGLPR	0	Brites P, Waterham HR, Wander RJA	Functions and biosynthesis of plasmalogens in health and disease	Biochimica et Biophysica Acta	2004		cytosolic - integral membrane protein on outer membrane of peroxisomes and ER - uniport and refs. Gene for this reaction has not yet been identified, however it is known to exist. Additionally, the transmembrane reaction takes in substrate from peroxisome/ER and produces product on cytosolic side of organelle - AGPex added to accomadate this. NJ
AGLPT	0	Nagan N, Zoeller RA	Plasmalogens: biosynthesis and functions	Progress in Lipid Research	2001		cytosol - see refs no GPR - documented biochemical activity, but no ORF associated w/ it at this time NJ
AGMTm	3	Mistry SK, Burwell TJ, Chambers RM, Rudolph-Owen L, Spaltmann F, Cook WJ, Morris SM Jr.	Cloning of human agmatinase. An alternate path for polyamine synthesis induced in liver by hepatitis B virus.	Am J Physiol Gastrointest Liver Physiol	2002	11804860	cellular location not specified in papers 4th citation indicates bidirectionality of pathway
AGMTm	3	Iyer RK, Kim HK, Tsoa RW, Grody WW, Cederbaum SD.	Cloning and characterization of human agmatinase.	Mol Genet Metab	2002	11914032	cellular location not specified in papers 4th citation indicates bidirectionality of pathway
AGMTm	3	Morris SM Jr.	Vertebrate agmatinases: what role do they play in agmatine catabolism?	Ann N Y Acad Sci	2003	15028567	cellular location not specified in papers 4th citation indicates bidirectionality of pathway
AGMTm	3	Morris SM Jr	Enzymes of arginine metabolism	J Nutr	2004	15465778	cellular location not specified in papers 4th citation indicates bidirectionality of pathway
AGPSx	3	deVet ECJM, vandenBosch H	Alkyl-dihydroxyacetonephosphate synthase	Cell Biochemsitry and Biophysics	2000		peroxisomal inner membrane - uniprot + refs NJ
AGPSx	3	Hajra AK	Glycerolipid biosynthesis in peroxisomes	Prog Lipid Res	1995		peroxisomal inner membrane - uniprot + refs NJ
AGTix	3	Purdue PE, Takada Y, Danpure CJ.	Identification of mutations associated with peroxisome-to-mitochondrion mistargeting of alanine/glyoxylate aminotransferase in primary hyperoxaluria type 1.		1990	1703535	Entrez Gene - The human AGXT protein product is normally localized in the peroxisomes of liver where it is involved in glyoxylate detoxification. Defects in the AGXT gene, some of which alter subcellular targetting, are the cause of Oxalosis I. MM - primarily found in the peroxisome; has a high affinity for glyoxylate; removes >99% of glyoxylate, permitting only a small fraction to be oxidize to oxalate [Poore 1998]
AHC	3	Coulter-Karis DE, Hershfield MS.	Sequence of full length cDNA for human S-adenosylhomocysteine hydrolase.		1989	2596825	Entrez Gene: S-adenosylhomocysteine hydrolase catalyzes the reversible hydrolysis of S-adenosylhomocysteine (AdoHcy) to adenosine (Ado) and L-homocysteine (Hcy). Thus, it regulates the intracellular S-adenosylhomocysteine (SAH) concentration thought to be important for transmethylation reactions. Deficiency in this protein is one of the different causes of hypermethioninemia. S-adenosylhomocysteine hydrolase belongs to the adenosylhomocysteinase family.

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AHC	3	Yang X, Hu Y, Yin DH, Turner MA, Wang M, Borchardt RT, Howell PL, Kuczera K, Schowen RL.	Catalytic strategy of S-adenosyl-L-homocysteine hydrolase: transition-state stabilization and the avoidance of abortive reactions.		2003	12590576	Entrez Gene: S-adenosylhomocysteine hydrolase catalyzes the reversible hydrolysis of S-adenosylhomocysteine (AdoHcy) to adenosine (Ado) and L-homocysteine (Hcy). Thus, it regulates the intracellular S-adenosylhomocysteine (SAH) concentration thought to be important for transmethylation reactions. Deficiency in this protein is one of the different causes of hypermethioninemia. S-adenosylhomocysteine hydrolase belongs to the adenosylhomocysteinase family.
AHEXASE2ly	3	Liu B, Ahmad W, Aronson NN Jr	Structure of the human gene for lysosomal di-N-acetylchitobiase	Glycobiology	1999	10336991	3073, 3074: -The subunits encoded by the genes HEXA and HEXB are synthesized as precursor proteins; processing and subunit assembly in the endoplasmatic reticulum yields three isoforms; beta-hexosaminidase A (alpha, beta), beta-hexosaminidase B (beta, beta) and beta-hexosaminidase S (alpha, alpha). The proteins are targeted to the lysosomes, where final processing produces the mature enzymes. [Maier et al, J Mol Biol. 328(3):669-81 (2003)] - lysosomal [Liu, Glycobiology 1999] - occurs in degradation of Asn-linked glycoproteins [Liu, Glycobiology 1999] - the active site of the beta -subunit hydrolyzes uncharged substrates, whereas the alpha -subunit, in addition, cleaves negatively charged substrate [Hepbildikler 2002] - Hexb -/- mice (expressing only HexS) showed no increased accumulation of glycosaminoglycans, indicating that Hex S involved in their catabolism [Hepbildikler 2002]
AHEXASE2ly	3	Hepbildikler ST, Sandhoff R, Kolzer M, Proia RL, Sandhoff K	Physiological substrates for human lysosomal beta -hexosaminidase S.	J Biol Chem	2002	11707436	3073, 3074: -The subunits encoded by the genes HEXA and HEXB are synthesized as precursor proteins; processing and subunit assembly in the endoplasmatic reticulum yields three isoforms; beta-hexosaminidase A (alpha, beta), beta-hexosaminidase B (beta, beta) and beta-hexosaminidase S (alpha, alpha). The proteins are targeted to the lysosomes, where final processing produces the mature enzymes. [Maier et al, J Mol Biol. 328(3):669-81 (2003)] - lysosomal [Liu, Glycobiology 1999] - occurs in degradation of Asn-linked glycoproteins [Liu, Glycobiology 1999] - the active site of the beta -subunit hydrolyzes uncharged substrates, whereas the alpha -subunit, in addition, cleaves negatively charged substrate [Hepbildikler 2002] - Hexb -/- mice (expressing only HexS) showed no increased accumulation of glycosaminoglycans, indicating that Hex S involved in their catabolism [Hepbildikler 2002]
AHEXASE2ly	3	Maier T, Strater N, Schuette CG, Klingenstein R, Sandhoff K, Saenger W.	The X-ray crystal structure of human beta-hexosaminidase B provides new insights into Sandhoff disease	J Mol Biol	2003	12706724	3073, 3074: -The subunits encoded by the genes HEXA and HEXB are synthesized as precursor proteins; processing and subunit assembly in the endoplasmatic reticulum yields three isoforms; beta-hexosaminidase A (alpha, beta), beta-hexosaminidase B (beta, beta) and beta-hexosaminidase S (alpha, alpha). The proteins are targeted to the lysosomes, where final processing produces the mature enzymes. [Maier et al, J Mol Biol. 328(3):669-81 (2003)] - lysosomal [Liu, Glycobiology 1999] - occurs in degradation of Asn-linked glycoproteins [Liu, Glycobiology 1999] - the active site of the beta -subunit hydrolyzes uncharged substrates, whereas the alpha -subunit, in addition, cleaves negatively charged substrate [Hepbildikler 2002] - Hexb -/- mice (expressing only HexS) showed no increased accumulation of glycosaminoglycans, indicating that Hex S involved in their catabolism [Hepbildikler 2002]
AICART	3	Sugita T, Aya H, Ueno M, Ishizuka T, Kawashima K.	Characterization of molecularly cloned human 5-aminoimidazole-4-carboxamide ribonucleotide transformylase.	J Biochem (Tokyo)	1997	9378707	no infos for compartiment IT
AICART	3	Bulock KG, Beardsley GP, Anderson KS.	The kinetic mechanism of the human bifunctional enzyme ATIC (5-amino-4-imidazolecarboxamide ribonucleotide transformylase/inosine 5'-monophosphate cyclohydrolase). A surprising lack of substrate channeling.	J Biol Chem	2002	11948179	no infos for compartiment IT
AICART	3	Cheong CG, Wolan DW, Greasley SE, Horton PA, Beardsley GP, Wilson IA.	Crystal structures of human bifunctional enzyme aminoimidazole-4-carboxamide ribonucleotide transformylase/IMP cyclohydrolase in complex with potent sulfonyl-containing antifolates.	J Biol Chem	2004	14966129	no infos for compartiment IT
AIRCr	3	Minet M, Lacroute F.	Cloning and sequencing of a human cDNA coding for a multifunctional polypeptide of the purine pathway by complementation of the ade2-101 mutant in Saccharomyces cerevisiae.	Curr Genet	1990	2253271	IT (no info for compartiment)

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
AIRCr	3	Brayton KA, Chen Z, Zhou G, Nagy PL, Gavalas A, Trent JM, Deaven LL, Dixon JE, Zalkin H.	Two genes for de novo purine nucleotide synthesis on human chromosome 4 are closely linked and divergently transcribed.	J Biol Chem	1994	8106516	IT (no info for compartiment)
AKR1C1	3	Stolz A, Hammond L, Lou H, Takikawa H, Ronk M, Shively JE.	cDNA cloning and expression of the human hepatic bile acid-binding protein. A member of the monomeric reductase gene family.	J Biol Chem	1993	8486699	localization: cytosol (uniprot) specificity: Expressed in all tissues tested including liver, prostate, testis, adrenal gland, brain, uterus, mammary gland and keratinocytes. Highest levels found in liver, mammary gland and brain. This gene encodes a member of the aldo/keto reductase superfamily, which consists of more than 40 known enzymes and proteins. These enzymes catalyze the conversion of aldehydes and ketones to their corresponding alcohols by utilizing NADH and/or NADPH as cofactors. The enzymes display overlapping but distinct substrate specificity. This enzyme catalyzes the reaction of progesterone to the inactive form 20-alpha-hydroxy-progesterone. This gene shares high sequence identity with three other gene members and is clustered with those three genes at chromosome 10p15-p14. May assist in the rapid intracellular transport of bile acids from the sinusoidal to the canalicular pole of the cell. Have a role in monitoring the intrahepatic bile acid concentration. NJ
ALASm	2	Bishop DF.	Two different genes encode delta-aminolevulinate synthase in humans: nucleotide sequences of cDNAs for the housekeeping and erythroid genes.		1990	2263504	TV/RS
ALAt2r	2	Sagne C, Agulhon C, Ravassard P, Darmon M, Hamon M, El Mestikawy S, Gasnier B, Giros B	Identification and characterization of a lysosomal transporter for small neutral amino acids	Proc Natl Acad Sci U S A	2001	11390972	The transporter here is thought to principally transport small neutral amino acids and related metabolites from the lysosome to the cytosol. Overexpression experiments have clearly shown that transport from the extracellular media into the cell cytoplasm occurs, but is is not clear if this is an experimental artifact. The evidence is thus physiological for extracellular transport and biochemical for lysosomal transport.
ALAt2r	2	Boll M, Foltz M, Rubio-Aliaga I, Kottra G, Daniel H	Functional characterization of two novel mammalian electrogenic proton-dependent amino acid cotransporters	J Biol Chem	2002	11959859	The transporter here is thought to principally transport small neutral amino acids and related metabolites from the lysosome to the cytosol. Overexpression experiments have clearly shown that transport from the extracellular media into the cell cytoplasm occurs, but is is not clear if this is an experimental artifact. The evidence is thus physiological for extracellular transport and biochemical for lysosomal transport.
ALAt4	3	Albritton LM, Bowcock AM, Eddy RL, Morton CC, Tseng L, Farrer LA, Cavalli-Sforza LL, Shows TB, Cunningham JM	The human cationic amino acid transporter (ATRC1): physical and genetic mapping to 13q12-q14	Genomics	1992	1348489	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly

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ALAt4	3	Furesz TC, Moe AJ, Smith CH	Lysine uptake by human placental microvillous membrane: comparison of system y+ with basal membrane	Am J Physiol	1995	7534987	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly
ALAt4	3	Hoshide R, Ikeda Y, Karashima S, Matsuura T, Komaki S, Kishino T, Niikawa N, Endo F, Matsuda I	Molecular cloning, tissue distribution, and chromosomal localization of human cationic amino acid transporter 2 (HCAT2)	Genomics	1996	8954799	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly
ALAt4	3	Sugawara M, Nakanishi T, Fei YJ, Huang W, Ganapathy ME, Leibach FH, Ganapathy V	Cloning of an amino acid transporter with functional characteristics and tissue expression pattern identical to that of system A	J Biol Chem	2000	10747860	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly

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ALAt4	3	Wang H, Huang W, Sugawara M, Devoe LD, Leibach FH, Prasad PD, Ganapathy V	Cloning and functional expression of ATA1, a subtype of amino acid transporter A, from human placenta	Biochem Biophys Res Commun	2000	10891391	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly
ALAt4	3	Hatanaka T, Huang W, Wang H, Sugawara M, Prasad PD, Leibach FH, Ganapathy V	Primary structure, functional characteristics and tissue expression pattern of human ATA2, a subtype of amino acid transport system A	Biochim Biophys Acta	2000	10930503	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly
ALAt4	3	Gu S, Roderick HL, Camacho P, Jiang JX	Characterization of an N-system amino acid transporter expressed in retina and its involvement in glutamine transport	J Biol Chem	2001	11325958	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly

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ALAt4	3	Hatanaka T, Huang W, Ling R, Prasad PD, Sugawara M, Leibach FH, Ganapathy V	Evidence for the transport of neutral as well as cationic amino acids by ATA3, a novel and liver-specific subtype of amino acid transport system A	Biochim Biophys Acta	2001	11342143	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly
ALAt4	3	Gu S, Adan-Rice D, Leach RJ, Jiang JX	A novel human amino acid transporter, hNAT3: cDNA cloning, chromosomal mapping, genomic structure, expression, and functional characterization	Genomics	2001	11414754	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly
ALAt4	3	Vekony N, Wolf S, Boissel JP, Gnauert K, Closs EI	Human cationic amino acid transporter hCAT-3 is preferentially expressed in peripheral tissues	Biochemistry	2001	11591158	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly

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ALAt4	3	Cariappa R, Heath-Monnig E, Furesz TC, Kamath SG, Smith CH	Stable polarized expression of hCAT-1 in an epithelial cell line	J Membr Biol	2002	11891586	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly
ALAt4	3	Wolf S, Janzen A, Vekony N, Martine U, Strand D, Closs EI	Expression of solute carrier 7A4 (SLC7A4) in the plasma membrane is not sufficient to mediate amino acid transport activity	Biochem J	2002	12049641	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly
ALAt4	3	Verrey F, Closs EI, Wagner CA, Palacin M, Endou H, Kanai Y	CATs and HATs: the SLC7 family of amino acid transporters	Pflugers Arch	2004	14770310	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] When expressed in mammalian cells, hATA2 mediates Na+-dependent transport of alpha-(methylamino)isobutyric acid, a specific model substrate for system A. The transporter is specific for neutral amino acids. It is pH-sensitive and Li+-intolerant. The Na+:amino acid stoichiometry is 1:1. (PMID 10930503) Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, ly

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ALATA_L	3	Yang RZ, Blaileanu G, Hansen BC, Shuldiner AR, Gong DW.	cDNA cloning, genomic structure, chromosomal mapping, and functional expression of a novel human alanine aminotransferase.	Genomics	2002	11863375	- Some info added by RS/TV - alanine aminotransferase catalyzes the reversible transamination between alanine and 2-oxoglutarate to form pyruvate and glutamate. - two isozymes are known to occur GPT and GPT2 - GPT2 is highly expressed in muscle, fat, and kidney. - GPT mainly expressed in kidney, liver, and heart. All this according to Yang RZ, Blaileanu G, Hansen BC, Shuldiner AR, Gong DW. Genomics. 2002 Mar;79(3):445-50.
ALCD1	3	Dawidek-Pietryka K, Szczepaniak S, Dudka J, Mazur M	In vitro studies of human alcohol dehydrogenase inhibition in the process of methanol and ethylene glycol oxidation	Arch Toxicol	1998	9806434	- Methanol is oxidized by alcohol dehydrogenase in the presence of NAD to formaldehyde [Abramson 2000] -Ethanol is preferentially metabolized by alcohol dehydrogenase and therefore can be effective in treatment of methanol poisoning [Dawidek-Pietryka 1998] - alcohol dehydrogenase is a predominant enzyme for methanol and ethylene glycol ox- idation (see [Ziegler 1988] ref in [Dawidek-Pietryka 1998] - ethanol metabolism by alcohol dehydrogenases occurs in the cytosol [Salway] - Class I isozymes are homo- and heterodimers (aa, bb, cc, ab, ac, bc) [Ramchandani et al, Pathol Biol 2001] Also from Ramchandani paper: - 124 (formerly ADH1 alpha subunit, now ADH1A) has low ethanol catalytic efficiency - 125 (formerly ADH2 beta subunit, now ADH1B) has high ethanol catalytic efficiency -126 (formerly ADH3 gamma subunit, now ADH1C) has high ethanol catalytic efficiency -127 (formerly ADH4 pi subunit, now ADH2) has low ethanol catalytic efficiency and is only expressed in the liver -128 (formerly ADH chi subunit, now ADH3) has very low ethanol catalytic efficiency and is ubiquitously expressed -131 (formerly ADH7 sigma subunit, now ADH4) has high etha - 130 (formerly ADH6, now ADH) ethanol catalytic activity no 137872: -only expressed in the liver [Deng et al, DNA Seq 2002]
ALCD1	3	Abramson S, Singh AK.	Treatment of the alcohol intoxications: ethylene glycol, methanol and isopropanol.	Curr Opin Nephrol Hypertens	2000	11128434	- Methanol is oxidized by alcohol dehydrogenase in the presence of NAD to formaldehyde [Abramson 2000] -Ethanol is preferentially metabolized by alcohol dehydrogenase and therefore can be effective in treatment of methanol poisoning [Dawidek-Pietryka 1998] - alcohol dehydrogenase is a predominant enzyme for methanol and ethylene glycol ox- idation (see [Ziegler 1988] ref in [Dawidek-Pietryka 1998] - ethanol metabolism by alcohol dehydrogenases occurs in the cytosol [Salway] - Class I isozymes are homo- and heterodimers (aa, bb, cc, ab, ac, bc) [Ramchandani et al, Pathol Biol 2001] Also from Ramchandani paper: - 124 (formerly ADH1 alpha subunit, now ADH1A) has low ethanol catalytic efficiency - 125 (formerly ADH2 beta subunit, now ADH1B) has high ethanol catalytic efficiency -126 (formerly ADH3 gamma subunit, now ADH1C) has high ethanol catalytic efficiency -127 (formerly ADH4 pi subunit, now ADH2) has low ethanol catalytic efficiency and is only expressed in the liver -128 (formerly ADH chi subunit, now ADH3) has very low ethanol catalytic efficiency and is ubiquitously expressed -131 (formerly ADH7 sigma subunit, now ADH4) has high etha - 130 (formerly ADH6, now ADH) ethanol catalytic activity no 137872: -only expressed in the liver [Deng et al, DNA Seq 2002]

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ALCD1	3	Ramchandani VA, Bosron WF, Li TK	Research advances in ethanol metabolism	Pathol Biol (Paris)	2001	11762128	- Methanol is oxidized by alcohol dehydrogenase in the presence of NAD to formaldehyde [Abramson 2000] -Ethanol is preferentially metabolized by alcohol dehydrogenase and therefore can be effective in treatment of methanol poisoning [Dawidek-Pietryka 1998] - alcohol dehydrogenase is a predominant enzyme for methanol and ethylene glycol ox- idation (see [Ziegler 1988] ref in [Dawidek-Pietryka 1998] - ethanol metabolism by alcohol dehydrogenases occurs in the cytosol [Salway] - Class I isozymes are homo- and heterodimers (aa, bb, cc, ab, ac, bc) [Ramchandani et al, Pathol Biol 2001] Also from Ramchandani paper: - 124 (formerly ADH1 alpha subunit, now ADH1A) has low ethanol catalytic efficiency - 125 (formerly ADH2 beta subunit, now ADH1B) has high ethanol catalytic efficiency -126 (formerly ADH3 gamma subunit, now ADH1C) has high ethanol catalytic efficiency -127 (formerly ADH4 pi subunit, now ADH2) has low ethanol catalytic efficiency and is only expressed in the liver -128 (formerly ADH chi subunit, now ADH3) has very low ethanol catalytic efficiency and is ubiquitously expressed -131 (formerly ADH7 sigma subunit, now ADH4) has high etha - 130 (formerly ADH6, now ADH) ethanol catalytic activity no 137872: -only expressed in the liver [Deng et al, DNA Seq 2002]
ALCD1	3	Deng Y, Wang Z, Gu S, Ji C, Ying K, Xie Y, Mao Y	Cloning and characterization of a novel human alcohol dehydrogenase gene (ADHFe1)	DNA Seq	2002	12592711	- Methanol is oxidized by alcohol dehydrogenase in the presence of NAD to formaldehyde [Abramson 2000] -Ethanol is preferentially metabolized by alcohol dehydrogenase and therefore can be effective in treatment of methanol poisoning [Dawidek-Pietryka 1998] - alcohol dehydrogenase is a predominant enzyme for methanol and ethylene glycol ox- idation (see [Ziegler 1988] ref in [Dawidek-Pietryka 1998] - ethanol metabolism by alcohol dehydrogenases occurs in the cytosol [Salway] - Class I isozymes are homo- and heterodimers (aa, bb, cc, ab, ac, bc) [Ramchandani et al, Pathol Biol 2001] Also from Ramchandani paper: - 124 (formerly ADH1 alpha subunit, now ADH1A) has low ethanol catalytic efficiency - 125 (formerly ADH2 beta subunit, now ADH1B) has high ethanol catalytic efficiency -126 (formerly ADH3 gamma subunit, now ADH1C) has high ethanol catalytic efficiency -127 (formerly ADH4 pi subunit, now ADH2) has low ethanol catalytic efficiency and is only expressed in the liver -128 (formerly ADH chi subunit, now ADH3) has very low ethanol catalytic efficiency and is ubiquitously expressed -131 (formerly ADH7 sigma subunit, now ADH4) has high etha - 130 (formerly ADH6, now ADH) ethanol catalytic activity no 137872: -only expressed in the liver [Deng et al, DNA Seq 2002]
ALDD21	2	De Laurenzi V, Rogers GR, Hamrock DJ, Marekov LN, Steinert PM, Compton JG, Markova N, Rizzo WB.	Sjogren-Larsson syndrome is caused by mutations in the fatty aldehyde dehydrogenase gene.	Nat Genet	1996	8528251	Role in fatty acid metabolism - see PMID 11591435. Aldehyde dehydrogenase isozymes are thought to play a major role in the detoxification of aldehydes generated by alcohol metabolism and lipid peroxidation. This gene product catalyzes the oxidation of long-chain aliphatic aldehydes to fatty acid. Mutations in the gene cause Sjogren-Larsson syndrome. NJ
ALDD21	2	Verhoeven NM, Jakobs C.	Human metabolism of phytanic acid and pristanic acid.	Prog Lipid Res	2001	11591435	Role in fatty acid metabolism - see PMID 11591435. Aldehyde dehydrogenase isozymes are thought to play a major role in the detoxification of aldehydes generated by alcohol metabolism and lipid peroxidation. This gene product catalyzes the oxidation of long-chain aliphatic aldehydes to fatty acid. Mutations in the gene cause Sjogren-Larsson syndrome. NJ

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ALDD2x	0	Sladek NE	Human aldehyde dehydrogenases: potential pathological, pharmacological, and toxicological impact	J Biochem Mol Toxicol	2003	12616643	-cytosolic [Sladek, J Biocehm Molecular Toxicology, 2003], [RefSeq] - acetaldehdyde is known substrate; NAD is cofactor [Sladek, J Biocehm Molecular Toxicology, 2003] 8854: -cytosolic [Sladek, J Biocehm Molecular Toxicology, 2003] - doesn't metabolize acetaldehyde very efficiently; NAD, NADP are cofactors [Sladek, J Biocehm Molecular Toxicology, 2003] 220: -cytosolic [Sladek, J Biocehm Molecular Toxicology, 2003], [UniProt] - NAD is cofactor [Sladek, J Biocehm Molecular Toxicology, 2003] 217, 219: - mitochondrial [Sladek, J Biocehm Molecular Toxicology, 2003], [UniProt] - acetaldehyde is preferred substrate, NAD is cofactor [Sladek, J Biocehm Molecular Toxicology, 2003] 218: -cytosolic [Sladek, J Biocehm Molecular Toxicology, 2003] - NAD, NADP are cofactors [Sladek, J Biocehm Molecular Toxicology, 2003] 224: - integral to ER membrane (GO) - cytoplasmic surface of ER mem [UniProt] - NAD, NADP are cofactors [Sladek, J Biocehm Molecular Toxicology, 2003] 221: - inferred to be associated with ER; assume its localization is similar to 224
ALKP	3	Knoll BJ, Rothblum KN, Longley M.	Nucleotide sequence of the human placental alkaline phosphatase gene. Evolution of the 5' flanking region by deletion/substitution.	J Biol Chem	1988	3042787	cytosolic - uniprot may have activity of fatty acid phosphate esters (?) There are at least four distinct but related alkaline phosphatases: intestinal, placental, placental-like, and liver/bone/kidney (tissue non-specific). The first three are located together on chromosome 2 while the tissue non-specific form is located on chromosome 1. The product of this gene is a membrane bound glycosylated enzyme that is not expressed in any particular tissue and is, therefore, referred to as the tissue-nonspecific form of the enzyme. The exact physiological function of the alkaline phosphatases is not known. A proposed function of this form of the enzyme is matrix mineralization, however, mice that lack a functional form of this enzyme show normal skeletal development. This enzyme has been linked directly to a disorder known as hypophosphatasia, a disorder that is characterized by hypercalcemia and includes skeletal defects. The character of this disorder can vary, however, depending on the specific mutation since this determines age of onset and severity of symptoms. The coding sequence for this form of alkaline phosphatase is un NJ
ALOX12	3	Izumi T, Hoshiko S, Radmark O, Samuelsson B.	Cloning of the cDNA for human 12-lipoxygenase.	Proc Natl Acad Sci	1990	2217179	cytoplasmic - uniprot + refs Leukotrienes biosynthesis gene identification - Izumi and Funk refs NJ
ALOX12	3	Funk CD, Furci L, FitzGerald GA.	Molecular cloning, primary structure, and expression of the human platelet/erythroleukemia cell 12-lipoxygenase.	Proc Natl Acad Sci	1990	2377602	cytoplasmic - uniprot + refs Leukotrienes biosynthesis gene identification - Izumi and Funk refs NJ
ALOX12R	3	Boeglin WE, Kim RB, Brash AR.	A 12R-lipoxygenase in human skin: mechanistic evidence, molecular cloning, and expression.	Proc Natl Acad Sci	1998	9618483	cyt - uniprot Expressed in B-cells, hair follicles, foreskin keratinocytes and adult skin. Also expressed in psoriatic tissue. Converts arachidonic acid to 12R- hydroperoxyeicosatetraenoic acid (12R-HPETE). NJ
ALOX15	3	Tang S, Bhatia B, Maldonado CJ, Yang P, Newman RA, Liu J, Chandra D, Traag J, Klein RD, Fischer SM, Chopra D, Shen J, Zhau HE, Chung LW, Tang DG.	Evidence that arachidonate 15-lipoxygenase 2 is a negative cell cycle regulator in normal prostate epithelial cells.	J Biol Chem	2002	11839751	cytoplasmic - uniprot + refs Leukotrienes biosynthesis gene identification - Tang ref NJ
ALOX5	3	Matsumoto T, Funk CD, Radmark O, Hoog JO, Jornvall H, Samuelsson B.	Molecular cloning and amino acid sequence of human 5-lipoxygenase.	Proc Natl Acad Sci	1988	2829172	cytoplasmic - uniprot + refs Leukotrienes biosynthesis seq see Matsuomoto ref NJ

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ALOX52	3	Leff AR.	Regulation of leukotrienes in the management of asthma: biology and clinical therapy.	Annu Rev Med	2001	11160764	cytosoloic by default (no detailed info) cofactors + usage still not clear unknown mechanism - biochemistry supported by PMID: 2829172, 11160764 NJ
AMACRp	3	VanVeldhoven PP, Croes K, Casteels M, Mannaerts GP	2-methylacyl racemase: a couple assay based on the use of pristanoyl-CoA oxidase/peroxidase and reinvestigation of its subcellular distribution in rat and human liver	Biochimica et Biophysica Acta	1997		Uniprot states evidence for mit, perox, and cytoplasmic, most lit refs only seemed to focus on perox and mit. This enzyme may have issues similar to other enzymes w/ ER outer membrane bound enzymes. Need to follow lit for future articles. Only in LocusLink db at present NJ
AMANK	3	Seppala R, Lehto VP, Gahl WA	Mutations in the human UDP-N-acetylglucosamine 2-epimerase gene define the disease sialuria and the allosteric site of the enzyme	Am J Hum Genet	1999	10330343	- shown as irreversible Devlin p. 672, Varki p. 74 10020: - cytoplasmic [UniProt], [Varki, p. 78] - Highest expression in liver and placenta. Also found in heart, brain, lung, kidney, skeletal muscle and pancreas. [UniProt] 55577: - GlcNAc kinase (10020) was shown to catalyze ManNAc kinase reaction when ManNAc gene was knocked out [Hinderlich, Biol Chem 2001] - ManNAc6P can be formed from ManNAc by kinase that can use either ManNAc or GlcNAc [Varki, p.77]
AMANK	3	Lucka L, Krause M, Danker K, Reutter W, Horstkorte R	Primary structure and expression analysis of human UDP-N-acetyl-glucosamine-2-epimerase/N-acetylmannosamine kinase, the bifunctional enzyme in neuraminic acid biosynthesis	FEBS Lett	1999	10431835	- shown as irreversible Devlin p. 672, Varki p. 74 10020: - cytoplasmic [UniProt], [Varki, p. 78] - Highest expression in liver and placenta. Also found in heart, brain, lung, kidney, skeletal muscle and pancreas. [UniProt] 55577: - GlcNAc kinase (10020) was shown to catalyze ManNAc kinase reaction when ManNAc gene was knocked out [Hinderlich, Biol Chem 2001] - ManNAc6P can be formed from ManNAc by kinase that can use either ManNAc or GlcNAc [Varki, p.77]
AMANK	3	Hinderlich S, Berger M, Keppler OT, Pawlita M, Reutter W	Biosynthesis of N-acetylneuraminic acid in cells lacking UDP-N-acetylglucosamine 2-epimerase/N-acetylmannosamine kinase	Biol Chem	2001	11308027	- shown as irreversible Devlin p. 672, Varki p. 74 10020: - cytoplasmic [UniProt], [Varki, p. 78] - Highest expression in liver and placenta. Also found in heart, brain, lung, kidney, skeletal muscle and pancreas. [UniProt] 55577: - GlcNAc kinase (10020) was shown to catalyze ManNAc kinase reaction when ManNAc gene was knocked out [Hinderlich, Biol Chem 2001] - ManNAc6P can be formed from ManNAc by kinase that can use either ManNAc or GlcNAc [Varki, p.77]
AMETt2m	3	Agrimi G, Di Noia MA, Marobbio CM, Fiermonte G, Lasorsa FM, Palmieri F	Identification of the human mitochondrial S-adenosylmethionine transporter: bacterial expression, reconstitution, functional characterization and tissue distribution	Biochem J	2004	14674884	- Added by KS, PV - Mitochondrial according to Agrimi G, et al. Biochem J. 2004 Apr 1;379(Pt 1):183-90. - SAM (S-adenosylmethionine) is the methyl group donor for almost all biological methylation reactions. In mitochondria, it is required for the methylation of DNA, RNA and proteins and as an intermediate in the biosynthesis of lipoic acid, and ubiquinone - As SAM is produced in the cytosol and is required inmitochondria, the primary function of SAMC is to catalyse the uptake of SAM into mitochondria. However, since SAMC functions almost exclusively by a counter-exchange mechanism, the carrier-mediated uptake of SAM requires the efflux of a counter-substrate. On the basis of transport measurements, SAHC produced from SAM in the methylation reactions and hydrolysed exclusively in the cytosol may serve as the counter-substrate of SAMC for SAM. Therefore the physiological role of the human SAMC is most probably to catalyse the uptake of SAM into the mitochondrial matrix in exchange for internal SAHC. - Very similar to yeast orthologue, except for need of a countertransport mechanism. - Expressed in all tissues. All this according to Agrimi G, et al. Biochem J. 2004 Apr 1;37
AMPDA	3	Mahnke-Zizelman DK, Sabina RL.	Cloning of human AMP deaminase isoform E cDNAs. Evidence for a third AMPD gene exhibiting alternatively spliced 5'-exons.	J Biol Chem	1992	1400401	IT 270: skeletal muscle, homotetramer 271: liver specific, homotetramer 272: erythrocytes specific

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AMPDA	3	Sabina RL, Morisaki T, Clarke P, Eddy R, Shows TB, Morton CC, Holmes EW.	Characterization of the human and rat myoadenylate deaminase genes.	J Biol Chem	1990	2345176	IT 270: skeletal muscle, homotetramer 271: liver specific, homotetramer 272: erythrocytes specific
AOBUTDsm	2	Edgar AJ.	Molecular cloning and tissue distribution of mammalian L-threonine 3-dehydrogenases.		2002	12097150	according to KEGG and MetaCyc spontaneous reaction PMID 12097150: the highly reactive intermediate, 2-amino-3-ketobutyrate, rapidly undergoes decarboxylation to form aminoacetone and CO2 MM
AP4AH1	3	Hankin S, Matthew N, Thorne H, McLennan AG.	Diadenosine 5',5'''-P1,P4-tetraphosphate hydrolase is present in human erythrocytes, leukocytes and platelets.	Int J Biochem Cell Biol	1995	7767787	IT -present in erys, leukocytes, plateles (Hankin, 1995) - requires divalemt ions (GeneCards) - compounds such as ap3a, ap4a, ap5a, ap6a have been shown to be present in and secreted from certain neurosecretory granules and the desnse granules of blood platelets. these compounds have neurotransmitter and vasomotor activity, affect platelet aggregation and may be novel regulators of blood pressure with properties distinct from ATP. In addition, virtually all cells contain submicromolar to micromolar cytosolic levels of ap3a and ap4a with the latter compound having been implicated in a number of intracellular events, including DNA replication and cellular response to metabolic stress (introduction from Hankin et al., 1995) - they are synthesized by aminoacyl tRNA-transferase (rxns not included)
AP4AH1	3	McLennan AG, Flannery AV, Morten JE, Ridanpaa M.	Chromosomal localization of the human diadenosine 5',5'''-P1,P4-tetraphosphate pyrophosphohydrolase (Ap4A hydrolase) gene (APAH1) to 9p13.	Genomics	1998	9479504	IT -present in erys, leukocytes, plateles (Hankin, 1995) - requires divalemt ions (GeneCards) - compounds such as ap3a, ap4a, ap5a, ap6a have been shown to be present in and secreted from certain neurosecretory granules and the desnse granules of blood platelets. these compounds have neurotransmitter and vasomotor activity, affect platelet aggregation and may be novel regulators of blood pressure with properties distinct from ATP. In addition, virtually all cells contain submicromolar to micromolar cytosolic levels of ap3a and ap4a with the latter compound having been implicated in a number of intracellular events, including DNA replication and cellular response to metabolic stress (introduction from Hankin et al., 1995) - they are synthesized by aminoacyl tRNA-transferase (rxns not included)
APAT2rm	1	Ohyama T, Matsuda K, Tachibana H, Fujimoto Sakata S, Mori M, Horiuchi M, Tamaki N	Purification and expression of a processing protease on beta-alanine-oxoglutarate aminotransferase from rat liver mitochondria	FEBS Lett	2004	15304357	This reaction is based on rat data as noted in the citation, so modeling data only.
ARAB-Lt	2	Lobley RW, Burrows PC, Warwick R, Dawson DJ, Holmes R	Simultaneous assessment of intestinal permeability and lactose tolerance with orally administered raffinose, lactose and L-arabinose	Clin Sci (Lond)	1990	2167807	- physiologial evidence that L-arabinose is taken up by intestine and excreted into bloodstream/urine; don't have any info on specific transport mechanism - it is generally accepted that no L-arabinose digestion occurs in humans; however, a patient with a presumed arabitol dehydrogenase deficiency has been identified [Onkenhout, Mol Genet Metab 2002] - oral arabinose loading has once been performed in man to asses intestinal permeability; median 5 h urinary excretion was 17.5% of the ingested amount indicating quite efficient absorption [Lobley 1990] - d-arabinose and d-ribose could cross cell membrane of cultured human fibroblasts [Huck 2004]
ARABR	3	Praml C, Savelyeva L, Perri P, Schwab M.	Cloning of the human aflatoxin B1-aldehyde reductase gene at 1p35-1p36.1 in a region frequently altered in human tumor cells.	Cancer Res	1998	9823300	pentoses in human fibroblasts; these reactions are most likely catalyzed by a pentose reductase [Huck, Mol Genet Metab 2004] - inferred from observations of patient with presumed deficiency (L-arabinose in diet led to high excretion of L-arabitol and arabinoic acid (-lactone)) [Onkenhout, Mol Genet Metab 2002] 8574: - gene has been cloned, 78% identical with the Rattus norvegicus aflatoxin B1 aldehyde reductase (Afar) [Praml 1998] -rat homolog appears to be Golgi-associated [Kelly 2002] based on N-terminal sequence and immunohistochemistry, however I believe these results more strongly support localization in the outer Golgi membrane (cytosolic in our model) - human AKR7A2 also has Golgi signal sequence and [GO, UniProt] list as Golgi by similarity; however, I've assumed protein is cytosolic due to lack of specific info on location of functional domain and the fact that the only (weak) evidence thus far has come from the rat homolog - NADH or NADPH [UniProt]; specificity is based on mouse homology AFAR2 [Kelly 2002]

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ARABR	3	Kelly VP, Sherratt PJ, Crouch DH, Hayes JD	Novel homodimeric and heterodimeric rat gamma-hydroxybutyrate synthases that associate with the Golgi apparatus define a distinct subclass of aldo-keto reductase 7 family proteins.	Biochem J	2002	12071861	pentoses in human fibroblasts; these reactions are most likely catalyzed by a pentose reductase [Huck, Mol Genet Metab 2004] - inferred from observations of patient with presumed deficiency (L-arabinose in diet led to high excretion of L-arabitol and arabinoic acid (-lactone)) [Onkenhout, Mol Genet Metab 2002] 8574: - gene has been cloned, 78% identical with the Rattus norvegicus aflatoxin B1 aldehyde reductase (Afar) [Praml 1998] -rat homolog appears to be Golgi-associated [Kelly 2002] based on N-terminal sequence and immunohistochemistry, however I believe these results more strongly support localization in the outer Golgi membrane (cytosolic in our model) - human AKR7A2 also has Golgi signal sequence and [GO, UniProt] list as Golgi by similarity; however, I've assumed protein is cytosolic due to lack of specific info on location of functional domain and the fact that the only (weak) evidence thus far has come from the rat homolog - NADH or NADPH [UniProt]; specificity is based on mouse homology AFAR2 [Kelly 2002]
ARABR	3	Onkenhout W, Groener JE, Verhoeven NM, Yin C, Laan LA	L-Arabinosuria: a new defect in human pentose metabolism	Mol Genet Metab	2002	12359133	pentoses in human fibroblasts; these reactions are most likely catalyzed by a pentose reductase [Huck, Mol Genet Metab 2004] - inferred from observations of patient with presumed deficiency (L-arabinose in diet led to high excretion of L-arabitol and arabinoic acid (-lactone)) [Onkenhout, Mol Genet Metab 2002] 8574: - gene has been cloned, 78% identical with the Rattus norvegicus aflatoxin B1 aldehyde reductase (Afar) [Praml 1998] -rat homolog appears to be Golgi-associated [Kelly 2002] based on N-terminal sequence and immunohistochemistry, however I believe these results more strongly support localization in the outer Golgi membrane (cytosolic in our model) - human AKR7A2 also has Golgi signal sequence and [GO, UniProt] list as Golgi by similarity; however, I've assumed protein is cytosolic due to lack of specific info on location of functional domain and the fact that the only (weak) evidence thus far has come from the rat homolog - NADH or NADPH [UniProt]; specificity is based on mouse homology AFAR2 [Kelly 2002]
ARGDCm	3	Grillo MA, Colombatto S	Metabolism and function in animal tissues of agmatine, a biogenic amine formed from arginine	Amino Acids	2004	14752610	the enzyme that catalyzes this reaction does not decarboxylate ornthine
ARGNm	3	Gotoh T, Araki M, Mori M	Chromosomal localization of the human arginase II gene and tissue distribution of its mRNA	Biochem Biophys Res Commun	1997	9144563	The physiological function of this enzyme is apparantly poorly understood, but the biochemical characterization seems legitimate.
ARGSL	3	O'Brien WE, McInnes R, Kalumuck K, Adcock M.	Cloning and sequence analysis of cDNA for human argininosuccinate lyase.	Proc Natl Acad Sci U S A	1986	3463959	- reversible according to Sampaleanu et al., 2001 - Additional information by RS/TV: - Argininosuccinate lyase catalyzes the conversion of of argininosuccinic acid into fumaric acid and arginine. The enzyme's primary physiological role is in the liver, where it functions in the urea cycle for the disposal of ingested nitrogen. (O'Brien WE, McInnes R, Kalumuck K, Adcock M. Proc Natl Acad Sci U S A. 1986 Oct;83(19):7211-5.)
ARGSL	3	Sampaleanu LM, Vallee F, Thompson GD, Howell PL.	Three-dimensional structure of the argininosuccinate lyase frequently complementing allele Q286R.		2001	11747432	- reversible according to Sampaleanu et al., 2001 - Additional information by RS/TV: - Argininosuccinate lyase catalyzes the conversion of of argininosuccinic acid into fumaric acid and arginine. The enzyme's primary physiological role is in the liver, where it functions in the urea cycle for the disposal of ingested nitrogen. (O'Brien WE, McInnes R, Kalumuck K, Adcock M. Proc Natl Acad Sci U S A. 1986 Oct;83(19):7211-5.)
ARGSL	3	Kleijer WJ et al.	Clinical, enzymatic, and molecular genetic characterization of a biochemical variant type of argininosuccinic aciduria: prenatal and postnatal diagnosis in five unrelated families.		2002	12408190	- reversible according to Sampaleanu et al., 2001 - Additional information by RS/TV: - Argininosuccinate lyase catalyzes the conversion of of argininosuccinic acid into fumaric acid and arginine. The enzyme's primary physiological role is in the liver, where it functions in the urea cycle for the disposal of ingested nitrogen. (O'Brien WE, McInnes R, Kalumuck K, Adcock M. Proc Natl Acad Sci U S A. 1986 Oct;83(19):7211-5.)
ARGSS	3	Metzler, David E	Biochemistry : the chemical reactions of living cells 2 ed vol 2		2001		- Reviewed by RS/TV

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ARGt4	2	Van Winkle LJ	Amino acid transport regulation and early embryo development	Biol Reprod	2001	11133652	From PMID 12719981: Multiple Na+-independent and Na+-dependent transport systems for amino acids have been identified by physiological and pharmacological approaches. One of these is system B0+, so named because it transports a broad array of neutral and cationic amino acids in a Na+-dependent manner, that is present in blastocysts, oocytes, intestine, pituitary, and lung [80]. Interestingly, the protein responsible for transport in this system is encoded by a member of the SLC6A gene family [69], members of which typically show more narrow substrate specificity. hATB0+ is a 645-amino acid protein, most similar to glycine and proline transporters, that is found in many non-neural tissues and which demonstrates high affinity uptake of hydrophobic amino acids. Transport is inhibited pharmacologically by 2-aminobicyclo-(2,2,1)-heptane-2-carboxylic acid (BCH). hATB0+ is highly expressed in the pituitary, where it may play a role in amino acid-induced secretion, and on the apical membrane of airway epithelia, where it may contribute to lung defense by clearing accumulated protein [70]
ARGt4	2	Chen NH, Reith ME, Quick MW	Synaptic uptake and beyond: the sodium- and chloride-dependent neurotransmitter transporter family SLC6	Pflugers Arch	2004	12719981	From PMID 12719981: Multiple Na+-independent and Na+-dependent transport systems for amino acids have been identified by physiological and pharmacological approaches. One of these is system B0+, so named because it transports a broad array of neutral and cationic amino acids in a Na+-dependent manner, that is present in blastocysts, oocytes, intestine, pituitary, and lung [80]. Interestingly, the protein responsible for transport in this system is encoded by a member of the SLC6A gene family [69], members of which typically show more narrow substrate specificity. hATB0+ is a 645-amino acid protein, most similar to glycine and proline transporters, that is found in many non-neural tissues and which demonstrates high affinity uptake of hydrophobic amino acids. Transport is inhibited pharmacologically by 2-aminobicyclo-(2,2,1)-heptane-2-carboxylic acid (BCH). hATB0+ is highly expressed in the pituitary, where it may play a role in amino acid-induced secretion, and on the apical membrane of airway epithelia, where it may contribute to lung defense by clearing accumulated protein [70]
ARSA	3	Stein C, Gieselmann V, Kreysing J, Schmidt B, Pohlmann R, Waheed A, Meyer HE, O'Brien JS, von Figura K.	Cloning and expression of human arylsulfatase A.	J Biol Chem	1989	2562955	lysosomal - uniprot + stein ref The protein encoded by this gene hydrolyzes cerebroside sulfate to cerebroside and sulfate. Defects in this gene lead to metachromatic leucodystrophy (MLD), a progressive demyelination disease which results in a variety of neurological symptoms and ultimately death. NJ
ASAH1	3	Koch J, Gartner S, Li CM, Quintern LE, Bernardo K, Levran O, Schnabel D, Desnick RJ, Schuchman EH, Sandhoff K.	Molecular cloning and characterization of a full-length complementary DNA encoding human acid ceramidase. Identification Of the first molecular lesion causing Farber disease.	J Biol Chem	1996	8955159	lysosomal - uniprot + Koch ref This gene encodes a heterodimeric protein consisting of a nonglycosylated alpha subunit and a glycosylated beta subunit that is cleaved to the mature enzyme posttranslationally. The encoded protein catalyzes the synthesis and degradation of ceramide into sphingosine and fatty acid. Mutations in this gene have been associated with a lysosomal storage disorder known as Farber disease. Two transcript variants encoding distinct isoforms have been identified for this gene. NJ
ASCBt	2	Bianchi J, Rose RC	Dehydroascorbic acid and cell membranes: possible disruptive effects	Toxicology	1986	3715892	- see [Wilson 2005], [Balll 2004] for a comprehensive review of ascorbate transport - ascorbate leaves enterocytes by soidum-independent facilitated diffusion at the basolateral membrane [Bianchi 1986] - existence of ascorbate homeoexchange system has not been demonstrated conclusively and the molecular identities of the membrane proteins responsible for homeoexchange have not been identified [Wilson 2005]

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ASCBt4	3	Rajan DP, Huang W, Dutta B, Devoe LD, Leibach FH, Ganapathy V, Prasad PD	Human placental sodium-dependent vitamin C transporter (SVCT2): molecular cloning and transport function	Biochem Biophys Res Commun	1999	10471399	- see [Wilson 2004], [Ball 2004] for a comprehensive review of ascorbate transport 9962: - cloned [Daruwala 1999], [Wang, Biochim Biophys Acta 1999], [Wang, Biochem Biophys Res Commun. 2000] - ascorbate & Na+ cotransport; does not transport dehydroascorbate [Daruwala 1999] - kidney, liver, small intestine, thymus and prostate [Wang, Biochim Biophys Acta 1999], also colon, ovary [Wang, Biochem Biophys Res Commun. 2000] - Na(+):ascorbate stoichiometry of 2:1 [Wang, Biochim Biophys Acta 1999]; electrogenic [Wang, Biochem Biophys Res Commun. 2000] 9963: - cloned [Daruwala 1999], [Wang, Biochem Biophys Res Commun. 2000] - ovary, spleen, testis, placenta, brain, prostate, WBC [Wang, Biochem Biophys Res Commun. 2000]; heart, brain, placenta [Rajan 1999] - ascorbate & Na+ cotransport; does not transport dehydroascorbate [Daruwala 1999], [Rajan 1999] - Na(+):ascorbate stoichiometry of 2:1 [Rajan 1999]; electrogenic [Wang, Biochem Biophys Res Commun. 2000]
ASCBt4	3	Wang H, Dutta B, Huang W, Devoe LD, Leibach FH, Ganapathy V, Prasad PD	Human Na(+)-dependent vitamin C transporter 1 (hSVCT1): primary structure, functional characteristics and evidence for a non-functional splice variant.	Biochim Biophys Acta	1999	10556483	- see [Wilson 2004], [Ball 2004] for a comprehensive review of ascorbate transport 9962: - cloned [Daruwala 1999], [Wang, Biochim Biophys Acta 1999], [Wang, Biochem Biophys Res Commun. 2000] - ascorbate & Na+ cotransport; does not transport dehydroascorbate [Daruwala 1999] - kidney, liver, small intestine, thymus and prostate [Wang, Biochim Biophys Acta 1999], also colon, ovary [Wang, Biochem Biophys Res Commun. 2000] - Na(+):ascorbate stoichiometry of 2:1 [Wang, Biochim Biophys Acta 1999]; electrogenic [Wang, Biochem Biophys Res Commun. 2000] 9963: - cloned [Daruwala 1999], [Wang, Biochem Biophys Res Commun. 2000] - ovary, spleen, testis, placenta, brain, prostate, WBC [Wang, Biochem Biophys Res Commun. 2000]; heart, brain, placenta [Rajan 1999] - ascorbate & Na+ cotransport; does not transport dehydroascorbate [Daruwala 1999], [Rajan 1999] - Na(+):ascorbate stoichiometry of 2:1 [Rajan 1999]; electrogenic [Wang, Biochem Biophys Res Commun. 2000]
ASCBt4	3	Daruwala R, Song J, Koh WS, Rumsey SC, Levine M	Cloning and functional characterization of the human sodium-dependent vitamin C transporters hSVCT1 and hSVCT2	FEBS Lett	1999	10556521	- see [Wilson 2004], [Ball 2004] for a comprehensive review of ascorbate transport 9962: - cloned [Daruwala 1999], [Wang, Biochim Biophys Acta 1999], [Wang, Biochem Biophys Res Commun. 2000] - ascorbate & Na+ cotransport; does not transport dehydroascorbate [Daruwala 1999] - kidney, liver, small intestine, thymus and prostate [Wang, Biochim Biophys Acta 1999], also colon, ovary [Wang, Biochem Biophys Res Commun. 2000] - Na(+):ascorbate stoichiometry of 2:1 [Wang, Biochim Biophys Acta 1999]; electrogenic [Wang, Biochem Biophys Res Commun. 2000] 9963: - cloned [Daruwala 1999], [Wang, Biochem Biophys Res Commun. 2000] - ovary, spleen, testis, placenta, brain, prostate, WBC [Wang, Biochem Biophys Res Commun. 2000]; heart, brain, placenta [Rajan 1999] - ascorbate & Na+ cotransport; does not transport dehydroascorbate [Daruwala 1999], [Rajan 1999] - Na(+):ascorbate stoichiometry of 2:1 [Rajan 1999]; electrogenic [Wang, Biochem Biophys Res Commun. 2000]

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ASCBt4	3	Wang Y, Mackenzie B, Tsukaguchi H, Weremowicz S, Morton CC, Hediger MA	Human vitamin C (L-ascorbic acid) transporter SVCT1	Biochem Biophys Res Commun	2000	10631088	- see [Wilson 2004], [Ball 2004] for a comprehensive review of ascorbate transport 9962: - cloned [Daruwala 1999], [Wang, Biochim Biophys Acta 1999], [Wang, Biochem Biophys Res Commun. 2000] - ascorbate & Na+ cotransport; does not transport dehydroascorbate [Daruwala 1999] - kidney, liver, small intestine, thymus and prostate [Wang, Biochim Biophys Acta 1999], also colon, ovary [Wang, Biochem Biophys Res Commun. 2000] - Na(+):ascorbate stoichiometry of 2:1 [Wang, Biochim Biophys Acta 1999]; electrogenic [Wang, Biochem Biophys Res Commun. 2000] 9963: - cloned [Daruwala 1999], [Wang, Biochem Biophys Res Commun. 2000] - ovary, spleen, testis, placenta, brain, prostate, WBC [Wang, Biochem Biophys Res Commun. 2000]; heart, brain, placenta [Rajan 1999] - ascorbate & Na+ cotransport; does not transport dehydroascorbate [Daruwala 1999], [Rajan 1999] - Na(+):ascorbate stoichiometry of 2:1 [Rajan 1999]; electrogenic [Wang, Biochem Biophys Res Commun. 2000]
ASNNm	2	Bush LA, Herr JC, Wolkowicz M, Sherman NE, Shore A, Flickinger CJ.	A novel asparaginase-like protein is a sperm autoantigen in rats.		2002	11984834	-only sequence has been detected -possible mitochondrial localisation (Bush et. al, 2002)
ASNS1	3	Van Heeke G, Schuster SM.	Expression of human asparagine synthetase in Escherichia coli.		1989	2564390	- necessary in the synthesis of non-essential amino acid asparagine -irreversible since it requires ATP hydrolysis Entrez Gene - The protein encoded by this gene is involved in the synthesis of asparagine. This gene complements a mutation in the temperature-sensitive hamster mutant ts11, which blocks progression through the G1 phase of the cell cycle at nonpermissive temperature. There are three alternatively spliced transcript variants encoding the same protein described for this gene.
ASNS1	3	Andrulis IL, Chen J, Ray PN.	Isolation of human cDNAs for asparagine synthetase and expression in Jensen rat sarcoma cells.		1987	2886907	- necessary in the synthesis of non-essential amino acid asparagine -irreversible since it requires ATP hydrolysis Entrez Gene - The protein encoded by this gene is involved in the synthesis of asparagine. This gene complements a mutation in the temperature-sensitive hamster mutant ts11, which blocks progression through the G1 phase of the cell cycle at nonpermissive temperature. There are three alternatively spliced transcript variants encoding the same protein described for this gene.
ASPNATm	3	Moreno A, Ross BD, Bluml S.	Direct determination of the N-acetyl-L-aspartate synthesis rate in the human brain by (13)C MRS and [1-(13)C]glucose infusion.		2001	11279290	PMID 11279290: N-Acetyl-l-aspartate (NAA) is an important amino-acid derivative in vertebrate brain and reaches its highest concentrations in neurons and axons (Tallan 1957; Passani et al. 1990). It is principally synthesized in neurons by the energy-dependent condensation of aspartate and acetyl coenzyme A (AcCoA), catalyzed by the mitochondrial enzyme NAA synthase (l-aspartate-N-acetyltransferase; EC 2.3.1.17), and is exported to the cytosolic compartment. MM
ASPTA	3	Panteghini M.	Aspartate aminotransferase isoenzymes.	Clin Biochem	1990	2225456	- Additional info added by RS/TV: Biochem text Cytosolic according to Entrez gene database Catalytic activity of protein synthesized by this gene (GPR association) according to GeneCards "Aspartate aminotransferase exists in human tissues as two distinct isozymes, one located in the cytoplasm" according to Panteghini M., Aspartate aminotransferase isoenzymes., Clin Biochem. 1990 Aug;23(4):311-9. Review. PMID: 2225456 reaction reversible according to Ford GC, Eichele G, Jansonius JN. Three-dimensional structure of a pyridoxal-phosphate-dependent enzyme, mitochondrial aspartate aminotransferase. Proc Natl Acad Sci U S A. 1980 May;77(5):2559-63. PMID: 6930651 NOTE: This reaction was classified under the Mal-Asp shuttle subsystem in the mitochondrial model.

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
ASPTA	3	Ford GC, Eichele G, Jansonius JN.	Three-dimensional structure of a pyridoxal-phosphate dependent enzyme, mitochondrial aspartate aminotransferase.	Proc Natl Acad Sci U S A	1980	6930651	- Additional info added by RS/TV: Biochem text Cytosolic according to Entrez gene database Catalytic activity of protein synthesized by this gene (GPR association) according to GeneCards "Aspartate aminotransferase exists in human tissues as two distinct isozymes, one located in the cytoplasm" according to Panteghini M., Aspartate aminotransferase isoenzymes., Clin Biochem. 1990 Aug;23(4):311-9. Review. PMID: 2225456 reaction reversible according to Ford GC, Eichele G, Jansonius JN. Three-dimensional structure of a pyridoxal-phosphate-dependent enzyme, mitochondrial aspartate aminotransferase. Proc Natl Acad Sci U S A. 1980 May;77(5):2559-63. PMID: 6930651 NOTE: This reaction was classified under the Mal-Asp shuttle subsystem in the mitochondrial model.
ATPasel	3	Forgac M.	Structure and properties of the vacuolar (H+)-ATPases	J Biol Chem	1999	10224039	Added by RS/DK - ATP is converted to ADP in the cytosol; this drives the protons from the cytosol into the lumen of the lysosome (See Figure 1 in Eskelinen, et al.) - Mechanism of lysosomal V-type ATPase is thought to be similar to F-type ATPSynthase of the mitochondrion. (See Forgac) - "The V-ATPase is composed of a peripheral V1 domain responsible for ATP hydrolysis and an integral V0 domain responsible for proton translocation" (See Forgac)
ATPasel	3	Eskelinen EL, Tanaka Y, Saftig P.	At the acidic edge: emerging functions for lysosomal membrane proteins	Trends Cell Biol	2003	12628346	Added by RS/DK - ATP is converted to ADP in the cytosol; this drives the protons from the cytosol into the lumen of the lysosome (See Figure 1 in Eskelinen, et al.) - Mechanism of lysosomal V-type ATPase is thought to be similar to F-type ATPSynthase of the mitochondrion. (See Forgac) - "The V-ATPase is composed of a peripheral V1 domain responsible for ATP hydrolysis and an integral V0 domain responsible for proton translocation" (See Forgac)
ATPH1e	3	Kaczmarek E, Koziak K, Sevigny J, Siegel JB, Anrather J, Beaudoin AR, Bach FH, Robson SC.	Identification and characterization of CD39/vascular ATP diphosphohydrolase.	J Biol Chem	1996	8955160	IT 953: expressed in placenta, lung, skeletal muscle, kidney, heart but not in brain 954: all tissues 955: brain, pancreas, spleen, prostate (no signal liver, peripheral blood leukocytes) 956: liver, kidney, prostate, testis, colon
ATPH1e	3	Chadwick BP, Frischauf AM.	The CD39-like gene family: identification of three new human members (CD39L2, CD39L3, and CD39L4), their murine homologues, and a member of the gene family from Drosophila melanogaster.	Genomics	1998	9676430	IT 953: expressed in placenta, lung, skeletal muscle, kidney, heart but not in brain 954: all tissues 955: brain, pancreas, spleen, prostate (no signal liver, peripheral blood leukocytes) 956: liver, kidney, prostate, testis, colon
B_MANNASEly	0	Alkhayat AH, Kraemer SA, Leipprandt JR, Macek M, Kleijer WJ, Friderici KH.	Human beta-mannosidase cDNA characterization and first identification of a mutation associated with human beta-mannosidosis	Hum Mol Genet	1998	9384606	MANBA encodes the final exoglycosidase in the pathway for N linked oligosaccharide catabolism. This enzyme localizes to the lysosome. Mutations in this gene cause beta-mannosidosis, a lysosomal storage disease that has a wide spectrum of neurological involvement. [RefSeq] Manbap ubiquitously expressed [Alkhayat et al, Hum Mol Genet 1998]
B3GNT11g	3	Isshiki S, Kudo T, Nishihara S, Ikehara Y, Togayachi A, Furuya A, Shitara K, Kubota T, Watanabe M, Kitajima M, Narimatsu H.	Lewis type 1 antigen synthase (beta3Gal-T5) is transcriptionally regulated by homeoproteins.		2003	12855703	localization: golgi - uniprot This gene encodes an enzyme that is a member of the beta-1,3-N-acetylglucosaminyltransferase family. This enzyme is a type II transmembrane protein. It prefers the substrate of lacto-N-neotetraose, and is involved in the biosynthesis of poly-N-acetyllactosamine chains. Two alternatively splicing transcript variants are identified from this gene and encode the same protein product. NJ

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B3GNT310g	2	Yokoyama-Kobayashi M, Yamaguchi T, Sekine S, Kato S.	Selection of cDNAs encoding putative type II membrane proteins on the cell surface from a human full-length cDNA bank.	Gene	1999	10072769	localization: golgi by uniprot This gene encodes an enzyme that is a member of the beta-1,3-N-acetylglucosaminyltransferase family. This enzyme is a type II transmembrane protein and contains a signal anchor that is not cleaved. It prefers the substrates of lacto-N-tetraose and lacto-N-neotetraose, and is involved in the biosynthesis of poly-N-acetyllactosamine chains and the biosynthesis of the backbone structure of dimeric sialyl Lewis a. It plays dominant roles in L-selectin ligand biosynthesis, lymphocyte homing and lymphocyte trafficking. NJ
B3GNT31g	3	Kolter T, Sandhoff K	Recent advances in the biochemistry of sphingolipidoses	Brain Pathology	1998		Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot This gene encodes an enzyme that is a member of the beta-1,3-N-acetylglucosaminyltransferase family. This enzyme is a type II transmembrane protein and contains a signal anchor that is not cleaved. It prefers the substrates of lacto-N-tetraose and lacto-N-neotetraose, and is involved in the biosynthesis of poly-N-acetyllactosamine chains and the biosynthesis of the backbone structure of dimeric sialyl Lewis a. It plays dominant roles in L-selectin ligand biosynthesis, lymphocyte homing and lymphocyte trafficking. NJ
B3GNT31g	3	Zhou D, Dinter A, Gutierrez Gallego R, Kamerling JP, Vliegenthart JF, Berger EG, Hennet T.	A beta-1,3-N-acetylglucosaminyltransferase with poly-N-acetyllactosamine synthase activity is structurally related to beta-1,3-galactosyltransferases.	Proc Natl Acad Sci	1999		Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot This gene encodes an enzyme that is a member of the beta-1,3-N-acetylglucosaminyltransferase family. This enzyme is a type II transmembrane protein and contains a signal anchor that is not cleaved. It prefers the substrates of lacto-N-tetraose and lacto-N-neotetraose, and is involved in the biosynthesis of poly-N-acetyllactosamine chains and the biosynthesis of the backbone structure of dimeric sialyl Lewis a. It plays dominant roles in L-selectin ligand biosynthesis, lymphocyte homing and lymphocyte trafficking. NJ
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BAAT1x	3	Falany CN, Johnson MR, Barnes S, Diasio RB	Glycine and taurine conugation of bile acids by a single enzyme	Journal of Biological Chemistry	1994		peroxisome: uniprot specificity: muscle, liver NJ
BACCL	3	Suzuki Y, Aoki Y, Ishida Y, Chiba Y, Iwamatsu A, Kishino T, Niikawa N, Matsubara Y, Narisawa K.	Isolation and characterization of mutations in the human holocarboxylase synthetase cDNA.	Nat Genet	1994	7842009	GeneCards noted that location is also mitochondrial.
BACCL	3	Pacheco-Alvarez D, Solorzano-Vargas RS, Del Rio AL.	Biotin in metabolism and its relationship to human disease.	Arch Med Res	2002	12459313	GeneCards noted that location is also mitochondrial.
BAMPPALDOX	1	White WH, Skatrud PL, Xue Z, Toyn JH	Specialization of function among aldehyde dehydrogenases: the ALD2 and ALD3 genes are required for beta-alanine biosynthesis in Saccharomyces cerevisiae	Genetics	2003	12586697	The citation clarifies this reaction in yeast, but only modeling evidence can be included here.
BBHOX	3	Vaz,F.M. , van Gool,S. , Ofman,R. , Ijlst,L. , Wanders,R.J.	Carnitine biosynthesis: identification of the cDNA encoding human gamma-butyrobetaine hydroxylase.		1998	9753662	0
BCDO	3	Wyss A, Wirtz G, Woggon W, Brugger R, Wyss M, Friedlein A, Bachmann H, Hunziker W.	Cloning and expression of beta,beta-carotene 15,15'-dioxygenase.	Biochem Biophys Res Commun	2000	10799297	IT
BCDO	3	Yan W, Jang GF, Haeseleer F, Esumi N, Chang J, Kerrigan M, Campochiaro M, Campochiaro P, Palczewski K, Zack DJ.	Cloning and characterization of a human beta,beta-carotene-15,15'-dioxygenase that is highly expressed in the retinal pigment epithelium.	Genomics	2001	11401432	IT
BDG2HCGHD	1	Daniels LB, Coyle PJ, Chiao YB, Glew RH, Labow RS.	Purification and characterization of a cytosolic broad specificity beta-glucosidase from human liver	J Biol Chem	1981	6796580	Not certain, but seems reasonable there is another enzyme or two, but localization and substrate specificity is much less certain, so they are not included

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BDHm	3	Marks AR, McIntyre JO, Duncan TM, Erdjument-Bromage H, Tempst P, Fleischer S	Molecular cloning and characterization of (R)-3-hydroxybutyrate dehydrogenase from human heart	J Biol Chem	1992	1639787	- mitochondrial inner membrane [Marks, J Biol Chem 1992], catalytic domain faces matrix [Green, Biochemistry 1996] - gene cloned [Marks J Biol Chem 1992] - has an absolute and specific requirement of phosphatidylcholine [Marks J Biol Chem 1992] - checked over by RS/TV
BDHm	3	Green D, Marks AR, Fleischer S, McIntyre JO	Wild type and mutant human heart (R)-3-hydroxybutyrate dehydrogenase expressed in insect cells	Biochemistry	1996	8679568	- mitochondrial inner membrane [Marks, J Biol Chem 1992], catalytic domain faces matrix [Green, Biochemistry 1996] - gene cloned [Marks J Biol Chem 1992] - has an absolute and specific requirement of phosphatidylcholine [Marks J Biol Chem 1992] - checked over by RS/TV
BETALDHx	3	Chern MK, Pietruszko R.	Human aldehyde dehydrogenase E3 isozyme is a betaine aldehyde dehydrogenase.		1995	7646513	NAD is preferred cofactor; cytosolic location - Chern et. Al. Biochem Biophys Res Commun. 1995 Aug 15;213(2):561-8. mitochondrial location possible PMID:15321791 MM
BETALDHx	3	Craig SA.	Betaine in human nutrition.		2004	15321791	NAD is preferred cofactor; cytosolic location - Chern et. Al. Biochem Biophys Res Commun. 1995 Aug 15;213(2):561-8. mitochondrial location possible PMID:15321791 MM
BHBt	2	Alonso de la Torre SR, Serrano MA, Medina JM.	Carrier-mediated beta-D-hydroxybutyrate transport in brush-border membrane vesicles from rat placenta.	Pediatr Res	1992	1408469	- mechanism of beta-D-hydroxybutyrate transport at brush border membrane of rat placenta is-D-hydroxybutyrate/H+ symport [Alonso de la Torre 1992] - liver mitochondria produce R-beta-hydroxybutyrate; this is released into blood and taken up by peripheral tissueswhich oxidize (R)-hydroxybutyrate back to acetoacetate [Guo 2006]
BHBt	2	Guo K, Lukacik P, Papagrigoriou E, Meier M, Lee WH, Adamski J, Oppermann U.	Characterization of Human DHRS6, an Orphan Short Chain Dehydrogenase/Reductase Enzyme: A NOVEL, CYTOSOLIC TYPE 2 R-beta-HYDROXYBUTYRATE DEHYDROGENASE.	J Biol Chem	2006	16380372	- mechanism of beta-D-hydroxybutyrate transport at brush border membrane of rat placenta is-D-hydroxybutyrate/H+ symport [Alonso de la Torre 1992] - liver mitochondria produce R-beta-hydroxybutyrate; this is released into blood and taken up by peripheral tissueswhich oxidize (R)-hydroxybutyrate back to acetoacetate [Guo 2006]
BHBtm	3	Latruffe N.	Transport of D-beta-hydroxybutyrate across rat liver mitochondrial membranes.	Comp Biochem Physiol B	1987	3427918	- Added by RS/TV - No genes found. - Inner mitochondrial membrane is largely permeable to to bhb. This process which is inhibited by several molecules indicates that this transport is carrier mediated. Two methods of transport are likely as a result of this inhibitory study: (1) like pyruvate/acetoacetate through a monocarboxylate carrier or (2) a dicarboxylate carrier. - It has also been strongly suggested that this translocation process is strongly dependent on pH medium which indicates a H+ symport or an OH- antiport. (Latruffe N. Comp Biochem Physiol B. 1987;88(3):797-802.)
BHMT	3	Sunden SL, Renduchintala MS, Park EI, Miklasz SD, Garrow TA.	Betaine-homocysteine methyltransferase expression in porcine and human tissues and chromosomal localization of the human gene.		1997	9281325	Entrez Gene - Betaine-homocysteine methyltransferase is a cytosolic enzyme that catalyzes the conversion of betaine and homocysteine to dimethylglycine and methionine, respectively. Defects in BHMT could lead to hyperhomocyst(e)inemia,but such a defect has not yet been observed.
BILIRED	0	Saito F, Yamaguchi T, Komuro A, Tobe T, Ikeuchi T, Tomita M, Nakajima H.	Mapping of the newly identified biliverdin-IX beta reductase gene (BLVRB) to human chromosome 19q13.13-->q13.2 by fluorescence in situ hybridization.	Cytogenet Cell Genet	1995	7656592	- Added by RS/TV Stefan Ryter, Free Radical Biology & Medicine, Vol 28, No2, pp. 289-309, 2000. - Cytoplasmic according to GeneCards. - Bilirubin is a lipophilic linear tetrapyrrole, abundant in blood plasma, which occurs uniquely in mammals. It is the final product of heme catabolism, as heme oxygenase (HO) cleaves the heme ring to form the water-soluble biliverdin, which is reduced by biliverdin reductase (BVR) to bilirubin. (Baranano DE, Rao M, Ferris CD, Snyder SH. Proc Natl Acad Sci U S A. 2002 Dec 10;99(25):16093-8. Epub 2002 Nov 27.) - Possible second gene: Blvrb, suspect because it claims to have two different functions. - IT - included blvrb to this GPR IT there might be the possibility that this genes also carries out biliverdin reductase activity in liver cells. Flavin depend enzyme (FMN is more effective than FAD)

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BILIRED	0	Chikuba K, Yubisui T, Shirabe K, Takeshita M.	Cloning and nucleotide sequence of a cDNA of the human erythrocyte NADPH-flavin reductase.	Biochem Biophys Res Commun	1994	8117274	- Added by RS/TV Stefan Ryter, Free Radical Biology & Medicine, Vol 28, No2, pp. 289-309, 2000. - Cytoplasmic according to GeneCards. - Bilirubin is a lipophilic linear tetrapyrrole, abundant in blood plasma, which occurs uniquely in mammals. It is the final product of heme catabolism, as heme oxygenase (HO) cleaves the heme ring to form the water-soluble biliverdin, which is reduced by biliverdin reductase (BVR) to bilirubin. (Baranano DE, Rao M, Ferris CD, Snyder SH. Proc Natl Acad Sci U S A. 2002 Dec 10;99(25):16093-8. Epub 2002 Nov 27.) - Possible second gene: Blvrb, suspect because it claims to have two different functions. - IT - included blvrb to this GPR IT there might be the possibility that this genes also carries out biliverdin reductase activity in liver cells. Flavin depend enzyme (FMN is more effective than FAD)
BILIRED	0	Baranano DE, Rao M, Ferris CD, Snyder SH.	Biliverdin reductase: a major physiologic cytoprotectant.	Proc Natl Acad Sci U S A	2002	12456881	- Added by RS/TV Stefan Ryter, Free Radical Biology & Medicine, Vol 28, No2, pp. 289-309, 2000. - Cytoplasmic according to GeneCards. - Bilirubin is a lipophilic linear tetrapyrrole, abundant in blood plasma, which occurs uniquely in mammals. It is the final product of heme catabolism, as heme oxygenase (HO) cleaves the heme ring to form the water-soluble biliverdin, which is reduced by biliverdin reductase (BVR) to bilirubin. (Baranano DE, Rao M, Ferris CD, Snyder SH. Proc Natl Acad Sci U S A. 2002 Dec 10;99(25):16093-8. Epub 2002 Nov 27.) - Possible second gene: Blvrb, suspect because it claims to have two different functions. - IT - included blvrb to this GPR IT there might be the possibility that this genes also carries out biliverdin reductase activity in liver cells. Flavin depend enzyme (FMN is more effective than FAD)
BMTer_L	3	Takahashi M, Inoue N, Ohishi K, Maeda Y, Nakamura N, Endo Y, Fujita T, Takeda J, Kinoshita T	PIG-B, a membrane protein of the endoplasmic reticulum with a large lumenal domain, is involved in transferring the third mannose of the GPI anchor	EMBO J	1996	8861954	- reaction described in Varki, pg. 136 - endoplasmic reticulum; Dol-P-Man dependent mannosyltransferases [RefSeq] - gene was cloned [Takahashi, EMBO J 1996] - transfers 3rd mannose of GPI anchor [Takahashi, EMBO J 1996]
BPNT	3	Spiegelberg BD, Xiong JP, Smith JJ, Gu RF, York JD.	Cloning and characterization of a mammalian lithium-sensitive bisphosphate 3'-nucleotidase inhibited by inositol 1,4-bisphosphate	J Biol Chem	1999	10224133	from [Spielberg 1999]: - human and mouse cDNA cloned; 92% amino acid sequence identity - human mRNA highly expressed in kidney, liver, pancreas, moderately expressed in heart, lowly expressed in placenta, kidney, brain, lung - enzyme demonstrated 3'-phosphatase activity on PAP and PAPS
BTND1	3	Stanley CM, Hymes J, Wolf B.	Identification of alternatively spliced human biotinidase mRNAs and putative localization of endogenous biotinidase.	Mol Genet Metab	2004	15059618	All this is not really clear to me. Stanley et al, 2004, reported three additional transcripts but they are not included in LocusLink yet. They localized biotinidase activity in cytoplasm. the test for mitochondrial localization seems to be negative. Probable ER localiszation. 686.1 = serum protein, outer membrane associated??? 686.2 = Stanley transcirpt 1 a, present in all tested tissues 686.3 = Stanley transcirpt 1 a, present in all tested tissues 686.4 = Stanley transcirpt 1 c, only expressed in testis due to the fact that 3 out of 5 biotin-dependent enzymes are located in mitochondria it is very likely that biotin-holocarbxylase and biotinidase are located in cytoplasm and mitochondria (Ref: Stanley et al, Pacheco-Alvarez) other splice variants were reported but not in Entrez Gene

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
BTND1n	3	Pispa J.	Animal biotinidase.	Ann Med Exp Biol Fenn	1965	5867120	IT biotin plays a role in regulation of cell cycle (Bender book) as a consequence of biotinylation of histones in the nucleus. Hymes et al (1995) discovered that human biotinidase has biotinyl transferase activity, in which biocytin serves as a biotin donor and histones serve as specific biotin acceptors. (Histones play an important role in the regulation of transcription, translation and packaging of DNA. The biotinyl transferase activity of biotinidase is not included in the model. Therefore, biocytin is only transported in nucleus and one of these two should be included in biomass function. I decided to allow transport of only biocytin since biocytin is transfered to histone (and not biotin) and released from them by proteolysis
BTND1n	3	Hymes J, Fleischhauer K, Wolf B.	Biotinylation of biotinidase following incubation with biocytin.	Clin Chim Acta	1995	7758201	IT biotin plays a role in regulation of cell cycle (Bender book) as a consequence of biotinylation of histones in the nucleus. Hymes et al (1995) discovered that human biotinidase has biotinyl transferase activity, in which biocytin serves as a biotin donor and histones serve as specific biotin acceptors. (Histones play an important role in the regulation of transcription, translation and packaging of DNA. The biotinyl transferase activity of biotinidase is not included in the model. Therefore, biocytin is only transported in nucleus and one of these two should be included in biomass function. I decided to allow transport of only biocytin since biocytin is transfered to histone (and not biotin) and released from them by proteolysis
BTND1n	3	Hymes J, Fleischhauer K, Wolf B.	Biotinylation of histones by human serum biotinidase: assessment of biotinyl-transferase activity in sera from normal individuals and children with biotinidase deficiency.	Biochem Mol Med	1995	8593541	IT biotin plays a role in regulation of cell cycle (Bender book) as a consequence of biotinylation of histones in the nucleus. Hymes et al (1995) discovered that human biotinidase has biotinyl transferase activity, in which biocytin serves as a biotin donor and histones serve as specific biotin acceptors. (Histones play an important role in the regulation of transcription, translation and packaging of DNA. The biotinyl transferase activity of biotinidase is not included in the model. Therefore, biocytin is only transported in nucleus and one of these two should be included in biomass function. I decided to allow transport of only biocytin since biocytin is transfered to histone (and not biotin) and released from them by proteolysis
BTND1n	3	Zempleni J.	Uptake, localization, and noncarboxylase roles of biotin.	Annu Rev Nutr	2005	16011464	IT biotin plays a role in regulation of cell cycle (Bender book) as a consequence of biotinylation of histones in the nucleus. Hymes et al (1995) discovered that human biotinidase has biotinyl transferase activity, in which biocytin serves as a biotin donor and histones serve as specific biotin acceptors. (Histones play an important role in the regulation of transcription, translation and packaging of DNA. The biotinyl transferase activity of biotinidase is not included in the model. Therefore, biocytin is only transported in nucleus and one of these two should be included in biomass function. I decided to allow transport of only biocytin since biocytin is transfered to histone (and not biotin) and released from them by proteolysis
BTND1n	3	Zempleni J., Mock D.M.	Biotin homeostasis during the cell cycle	Nutrition Research Reviews	2001		IT biotin plays a role in regulation of cell cycle (Bender book) as a consequence of biotinylation of histones in the nucleus. Hymes et al (1995) discovered that human biotinidase has biotinyl transferase activity, in which biocytin serves as a biotin donor and histones serve as specific biotin acceptors. (Histones play an important role in the regulation of transcription, translation and packaging of DNA. The biotinyl transferase activity of biotinidase is not included in the model. Therefore, biocytin is only transported in nucleus and one of these two should be included in biomass function. I decided to allow transport of only biocytin since biocytin is transfered to histone (and not biotin) and released from them by proteolysis
BTNDe	3	Cole H, Reynolds TR, Lockyer JM, Buck GA, Denson T, Spence JE, Hymes J, Wolf B.	Human serum biotinidase. cDNA cloning, sequence, and characterization.	J Biol Chem	1994	7509806	IT other splice variants were reported but not in Entrez Gene
BTNDe	3	Cole H, Weremowicz S, Morton CC, Wolf B.	Localization of serum biotinidase (BTD) to human chromosome 3 in band p25.	Genomics	1994	8001986	IT other splice variants were reported but not in Entrez Gene

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
BTNt2	3	Daberkow RL, White BR, Cederberg RA, Griffin JB, Zempleni J.	Monocarboxylate transporter 1 mediates biotin uptake in human peripheral blood mononuclear cells.	J Nutr	2003		6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], heart, red muscle [Halestrap 1999] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] IT It seems that the transport is reversible since Daberkow et al found that extracellular lactate stimulates efflux from biotin. However, it seems that this biotin transport mediated by MCT1 only takes in lymphoid cells but not into placental (JAr) cells, l Moreover, it might be that MCT2 is also able to transport biotin
BTNt2m	3	Daberkow RL, White BR, Cederberg RA, Griffin JB, Zempleni J.	Monocarboxylate transporter 1 mediates biotin uptake in human peripheral blood mononuclear cells.	J Nutr	2003	12949353	IT MCT1 transporter location in mito membrane has been confirmed for rat.
BTNt2m	3	Butz CE, McClelland GB, Brooks GA.	MCT1 confirmed in rat striated muscle mitochondria.	J Appl Physiol	2004	15121743	IT MCT1 transporter location in mito membrane has been confirmed for rat.
BTNt3i	3	Prasad PD, Wang H, Kekuda R, Fujita T, Fei YJ, Devoe LD, Leibach FH, Ganapathy V.	Cloning and functional expression of a cDNA encoding a mammalian sodium-dependent vitamin transporter mediating the uptake of pantothenate, biotin, and lipoate.	J Biol Chem	1998	9516450	IT
BTNt3i	3	Wang H, Huang W, Fei YJ, Xia H, Yang-Feng TL, Leibach FH, Devoe LD, Ganapathy V, Prasad PD.	Human placental Na+-dependent multivitamin transporter. Cloning, functional expression, gene structure, and chromosomal localization.	J Biol Chem	1999	10329687	IT
BTNt3i	3	Prasad PD, Wang H, Huang W, Fei YJ, Leibach FH, Devoe LD, Ganapathy V.	Molecular and functional characterization of the intestinal Na+-dependent multivitamin transporter.	Arch Biochem Biophys	1999	10334869	IT
BTNt3i	3	Balamurugan K, Ortiz A, Said HM.	Biotin uptake by human intestinal and liver epithelial cells: role of the SMVT system.	Am J Physiol Gastrointest Liver Physiol	2003	12646417	IT
BTNt3i	3	Balamurugan K, Vaziri ND, Said HM.	Biotin uptake by human proximal tubular epithelial cells: Cellular and molecular aspects.	Am J Physiol Renal Physiol	2005	15561972	IT
BTNt4i	3	Vlasova TI, Stratton SL, Wells AM, Mock NI, Mock DM.	Biotin deficiency reduces expression of SLC19A3, a potential biotin transporter, in leukocytes from human blood.	J Nutr	2005	15623830	IT i am not sure if this transport can be reversible. takes at least place in leukocytes
BUP2	3	Sakamoto T, Sakata SF, Matsuda K, Horikawa Y, Tamaki N.	Expression and properties of human liver beta-ureidopropionase.	J Nutr Sci Vitaminol (Tokyo)	2001	11508704	original rxn (BUP) added by IT changed to BUP2 by MM: 3aib in rxn BUP has been changed to 3aib-D (rxn BUP2) (see OMIM ref below) OMIM: The R enantiomer (old name D-(minus)-BAIB) derives from thymine, the S enantiomer (L-plus-BAIB) from L-valine. Human urine contains R-BAIB almost exclusively (and it is this form that is excreted in excess in the hyper-BAIB trait), whereas the plasma pool is about 80% S-BAIB.
C110CPT2m	2	Verhoeven NM, Wanders RJ, Poll-The BT, Saudubray JM, Jakobs C.	The metabolism of phytanic acid and pristanic acid in man: a review.	J Inherit Metab Dis	1998	9819701	As per Verhoeven J Inher Metab Dis 1998 dmnoncoa must be transfered into mit to undergo 1 round of beta-ox, but fate of 2,6 dimethylheptanoly-CoA is not known! NJ

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C160CPT1	3	Finocchiaro G, Taroni F, Rocchi M, Martin AL, Colombo I, Tarelli GT, DiDonato S.	cDNA cloning, sequence analysis, and chromosomal localization of the gene for human carnitine palmitoyltransferase.	Proc Natl Acad Sci U S A	1991	1988962	- Additional information added by RS/TV J Biol Chem. 2001 Jun 8;276(23):20182-5. Epub 2001 Mar 27 1)GPR association: In order to cross the mitochondrial membranes, long-chain fatty acids are first activated by coenzyme A and then reversibly conjugated with L-carnitine, a reaction that is catalyzed by the enzyme carnitine palmitoyltransferase. CPTase is split into two distinct parts: sequential action of CPT1 (addition of carnitine group to fatty-acid) followed by CPT2 (removal of carnitine group from fatty acid). 2)Differing roles of CPT1 and CPT2: CPT 1is associated with the outer mitochondrial membrane (cytosolic enzyme activity), loses activity upon exposure to strong detergents. CPT 2 is located on the inner mitochondrial membrane, insensitive to malonyl-coa inhibition and exposure to detergent. 3)Isoforms of CPT1: CPT1 exists as three isoforms, each with a different tissue expression. Isoform (a) is expressed primarily in the liver. Isoform (b) is expressed primarily in muscles. Isoform (c) is expressed primarily in the brain. 1 through 3 according to Finocchiaro G, Taroni F, Rocchi M, M Isoform(b) has four splice variants according to Entrez Gene da
C160CPT2	3	Taroni F, Verderio E, Fiorucci S, Cavadini P, Finocchiaro G, Uziel G, Lamantea E, Gellera C, DiDonato S.	Molecular characterization of inherited carnitine palmitoyltransferase II deficiency.	Proc Natl Acad Sci U S A	1992	1528846	J Biol Chem. 2001 Jun 8;276(23):20182-5. Epub 2001 Mar 27 1)GPR association: In order to cross the mitochondrial membranes, long-chain fatty acids are first activated by coenzyme A and then reversibly conjugated with L-carnitine, a reaction that is catalyzed by the enzyme carnitine palmitoyltransferase. CPTase is split into two distinct parts: sequential action of CPT1 (addition of carnitine group to fatty-acid) followed by CPT2 (removal of carnitine group from fatty acid). 2)Differing roles of CPT1 and CPT2: CPT 1is associated with the outer mitochondrial membrane (cytosolic enzyme activity), loses activity upon exposure to strong detergents. CPT 2 is located on the inner mitochondrial membrane, insensitive to malonyl-coa inhibition and exposure to detergent. 3)Isoforms of CPT1: CPT1 exists as three isoforms, each with a different tissue expression. Isoform (a) is expressed primarily in the liver. Isoform (b) is expressed primarily in muscles. Isoform (c) is expressed primarily in the brain. 4) CPT2 does not exist have any tissue-specific isoforms. It see 1 and 2 according to Finocchiaro G, Taroni F, Rocchi M, Marti 3 and 4 according to Taroni F, Verderio E, Fiorucci S, Cavadini
C226CPT2	3	Fraser F, Padovese R, Zammit VA.	Distinct kinetics of carnitine palmitoyltransferase i in contact sites and outer membranes of rat liver mitochondria.	J Biol Chem	2001	11274214	Fraser F, Padovese R, Zammit VA Distinct kinetics of carnitine palmitoyltransferase i in contact sites and outer membranes of rat liver mitochondria J Biol Chem. 2001 Jun 8;276(23):20182-5. Epub 2001 Mar 27 1)GPR association: In order to cross the mitochondrial membranes, long-chain fatty acids are first activated by coenzyme A and then reversibly conjugated with L-carnitine, a reaction that is catalyzed by the enzyme carnitine palmitoyltransferase. CPTase is split into two distinct parts: sequential action of CPT1 (addition of carnitine group to fatty-acid) followed by CPT2 (removal of carnitine group from fatty acid). 2)Differing roles of CPT1 and CPT2: CPT 1is associated with the outer mitochondrial membrane (cytosolic enzyme activity), loses activity upon exposure to strong detergents. CPT 2 is located on the inner mitochondrial membrane, insensitive to malonyl-coa inhibition and exposure to detergent. 3)Isoforms of CPT1: CPT1 exists as three isoforms, each with a different tissue expression. Isoform (a) is expressed primarily in the liver. Isoform (b) is expressed primarily in muscles. Isoform 4) CPT2 does not exist have any tissue-specific isoforms. It see 1 and 2 according to Finocchiaro G, Taroni F, Rocchi M, Marti
C3STDH1Pr	3	Caldas H, Herman GE	NSDHL, an eznyme invovled in cholesterol biosynthesis, traffics through the Golgi and accumulates on ER membranes on the surface of lipid droplets	Human Molecular Genetics	2003		ER - uniprot see refs - particularly Caldas - transport mechanism for steroids from ER -> cytosol: lipid droplets specificity: Brain, heart, liver, lung, kidney, skin and placenta. NSDHL, an enzyme involved in cholesterol synthesis, traffics through the Golgi and accumulates on ER membranes and on the surface of lipid droplets. NAD(P)H steroid dehydrogenase-like protein is localized to lipid droplets NJ

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C3STDH1Pr	3	Konig A, Happle R, Bornholdt D, Engel H, Grzeschik KH	Mutations in the NSDHL gene, encoding a 3-beta-hydroxysteroid dehydrogenase, cause CHILD syndrome	Am J Med Genet	2000		ER - uniprot see refs - particularly Caldas - transport mechanism for steroids from ER -> cytosol: lipid droplets specificity: Brain, heart, liver, lung, kidney, skin and placenta. NSDHL, an enzyme involved in cholesterol synthesis, traffics through the Golgi and accumulates on ER membranes and on the surface of lipid droplets. NAD(P)H steroid dehydrogenase-like protein is localized to lipid droplets NJ
C3STDH1Pr	3	Caldas H, Herman GE	NSDHL, an eznyme invovled in cholesterol biosynthesis, traffics through the Golgi and accumulates on ER membranes on the surface of lipid droplets	Human Molecular Genetics	2003		ER - uniprot see refs - particularly Caldas - transport mechanism for steroids from ER -> cytosol: lipid droplets specificity: Brain, heart, liver, lung, kidney, skin and placenta. NSDHL, an enzyme involved in cholesterol synthesis, traffics through the Golgi and accumulates on ER membranes and on the surface of lipid droplets. NAD(P)H steroid dehydrogenase-like protein is localized to lipid droplets NJ
CAATPS	3	Yu X, Inesi G.	Variable stoichiometric efficiency of Ca2+ and Sr2+ transport by the sarcoplasmic reticulum ATPase.	J Biol Chem	1995	7876199	More famously known for SR subtypes (some of these are also ER membrane see ATP2A family genes (GeneID 487, 488, 489). Plasma membrane CaATP include ATP2B family: 490, 491, 492, 493 Ca/ATP stoichiometry can vary see PMID: 7876199 The protein encoded by this gene belongs to the family of P-type primary ion transport ATPases characterized by the formation of an aspartyl phosphate intermediate during the reaction cycle. These enzymes remove bivalent calcium ions from eukaryotic cells against very large concentration gradients and play a critical role in intracellular calcium homeostasis. The mammalian plasma membrane calcium ATPase isoforms are encoded by at least four separate genes and the diversity of these enzymes is further increased by alternative splicing of transcripts. The expression of different isoforms and splice variants is regulated in a developmental, tissue- and cell type-specific manner, suggesting that these pumps are functionally adapted to the physiological needs of particular cells and tissues. This gene encodes the plasma membrane calcium ATPase isoform 2. Alternatively spliced transcript variants enc NJ
CAATPS	3	Hilfiker H, Strehler-Page MA, Stauffer TP, Carafoli E, Strehler EE.	Structure of the gene encoding the human plasma membrane calcium pump isoform 1.	J Biol Chem	1993	8396145	More famously known for SR subtypes (some of these are also ER membrane see ATP2A family genes (GeneID 487, 488, 489). Plasma membrane CaATP include ATP2B family: 490, 491, 492, 493 Ca/ATP stoichiometry can vary see PMID: 7876199 The protein encoded by this gene belongs to the family of P-type primary ion transport ATPases characterized by the formation of an aspartyl phosphate intermediate during the reaction cycle. These enzymes remove bivalent calcium ions from eukaryotic cells against very large concentration gradients and play a critical role in intracellular calcium homeostasis. The mammalian plasma membrane calcium ATPase isoforms are encoded by at least four separate genes and the diversity of these enzymes is further increased by alternative splicing of transcripts. The expression of different isoforms and splice variants is regulated in a developmental, tissue- and cell type-specific manner, suggesting that these pumps are functionally adapted to the physiological needs of particular cells and tissues. This gene encodes the plasma membrane calcium ATPase isoform 2. Alternatively spliced transcript variants enc NJ
CAMPt	3	Chen ZS, Lee K, Kruh GD.	Transport of cyclic nucleotides and estradiol 17-beta-D-glucuronide by multidrug resistance protein 4. Resistance to 6-mercaptopurine and 6-thioguanine.	J Biol Chem	2001	11447229	IT Chen(2001):ABCC4: cGMP: 2.0 +/- 0.3 pmol/mg/min cAMP:4.1 +/- 0.4 pmol/min/mg Estradiolglc: 102 +/- 16 pmol/min/mg
CAMPt	3	Wielinga PR, van der Heijden I, Reid G, Beijnen JH, Wijnholds J, Borst P.	Characterization of the MRP4- and MRP5-mediated transport of cyclic nucleotides from intact cells.	J Biol Chem	2003	12637526	IT Chen(2001):ABCC4: cGMP: 2.0 +/- 0.3 pmol/mg/min cAMP:4.1 +/- 0.4 pmol/min/mg Estradiolglc: 102 +/- 16 pmol/min/mg
CAMPt	3	Reid G, Wielinga P, Zelcer N, van der Heijden I, Kuil A, de Haas M, Wijnholds J, Borst P.	The human multidrug resistance protein MRP4 functions as a prostaglandin efflux transporter and is inhibited by nonsteroidal antiinflammatory drugs.	Proc Natl Acad Sci U S A	2003	12835412	IT Chen(2001):ABCC4: cGMP: 2.0 +/- 0.3 pmol/mg/min cAMP:4.1 +/- 0.4 pmol/min/mg Estradiolglc: 102 +/- 16 pmol/min/mg

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CAMPt	3	Dazert P, Meissner K, Vogelgesang S, Heydrich B, Eckel L, Bohm M, Warzok R, Kerb R, Brinkmann U, Schaeffeler E, Schwab M, Cascorbi I, Jedlitschky G, Kroemer HK.	Expression and localization of the multidrug resistance protein 5 (MRP5/ABCC5), a cellular export pump for cyclic nucleotides, in human heart.	Am J Pathol	2003	14507663	IT Chen(2001):ABCC4: cGMP: 2.0 +/- 0.3 pmol/mg/min cAMP:4.1 +/- 0.4 pmol/min/mg Estradiolglc: 102 +/- 16 pmol/min/mg
CAMPt	3	Meyer Zu Schwabedissen HE, Grube M, Heydrich B, Linnemann K, Fusch C, Kroemer HK, Jedlitschky G.	Expression, localization, and function of MRP5 (ABCC5), a transporter for cyclic nucleotides, in human placenta and cultured human trophoblasts: effects of gestational age and cellular differentiation.	Am J Pathol	2005	15631998	IT Chen(2001):ABCC4: cGMP: 2.0 +/- 0.3 pmol/mg/min cAMP:4.1 +/- 0.4 pmol/min/mg Estradiolglc: 102 +/- 16 pmol/min/mg
CAt7r	3	Schnetkamp PP.	Na-Ca or Na-Ca-K exchange in rod photoreceptors.		1989	2484986	- encodes a member of the sodium/calcium exchanger integral membrane protein family. Three mammalian isoforms in family 8 have been identified. Na+/Ca2+ exchange proteins are involved in maintaining Ca2+ homeostasis in a wide variety of cell types. - The Na+/Ca2+ exchanger is the dominant cellular Ca2+ efflux mechanism and regulates contractility. - isoform 1 is ubiquitous, 2 and 3 found in brain and skeletal muscle tissue - 6 splice variants for isoform 3 -note: Ca2+/Ca2+, Na+/Na+, Na+/Mg2+, Na+/Ba2+, Na+/Sr2+, and Na+/Ni2+ exchanges have been described, but not significant MM
CAt7r	3	Gabellini,N., Bortoluzzi,S., Danieli,G.A., Carafoli,E.,	The human SLC8A3 gene and the tissue-specific Na+/Ca2+ exchanger 3 isoforms.		2002	12406570	- encodes a member of the sodium/calcium exchanger integral membrane protein family. Three mammalian isoforms in family 8 have been identified. Na+/Ca2+ exchange proteins are involved in maintaining Ca2+ homeostasis in a wide variety of cell types. - The Na+/Ca2+ exchanger is the dominant cellular Ca2+ efflux mechanism and regulates contractility. - isoform 1 is ubiquitous, 2 and 3 found in brain and skeletal muscle tissue - 6 splice variants for isoform 3 -note: Ca2+/Ca2+, Na+/Na+, Na+/Mg2+, Na+/Ba2+, Na+/Sr2+, and Na+/Ni2+ exchanges have been described, but not significant MM
CATm	3	Radi R, Turrens JF, Chang LY, Bush KM, Crapo JD, Freeman BA.	Detection of catalase in rat heart mitochondria.	J Biol Chem	1991	1657986	Proteome Radi R, Turrens JF, Chang LY, Bush KM, Crapo JD, Freeman BA.; Detection of catalase in rat heart mitochondria.; J Biol Chem. 1991 Nov 15;266(32):22028-34. Catalytic Activity: Catalase is an enzyme which catalyzes the decomposition of hydrogen peroxide to oxygen and water. It is found in virtually all aerobic cells and is partly responsible for protecting cells from the toxic effects of hydrogen peroxide. According to F Quan. Nucleic Acids Res. 1986 July 11; 14(13): 5321 5335. Localization: In mammalian tissues found predominantly in liver, kidney, and erythrocytes, while the lowest levels are found in connective tissues. In tissues such as the liver, it is found predominantly in the peroxisome. In mature human erythrocytes, it is found in the cytosol. Catalase has also been found in the mitochondria of cardiac cells. According to F Quan. Nucleic Acids Res. 1986 July 11; 14(13): 5321 5335. Also according to Radi R. J Biol Chem. 1991 Nov 15;266(32):22028-34.

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CATm	3	Quan F, Korneluk RG, Tropak MB, Gravel RA.	Isolation and characterization of the human catalase gene.	Nucleic Acids Res	1986	3755525	Proteome Radi R, Turrens JF, Chang LY, Bush KM, Crapo JD, Freeman BA.; Detection of catalase in rat heart mitochondria.; J Biol Chem. 1991 Nov 15;266(32):22028-34. Catalytic Activity: Catalase is an enzyme which catalyzes the decomposition of hydrogen peroxide to oxygen and water. It is found in virtually all aerobic cells and is partly responsible for protecting cells from the toxic effects of hydrogen peroxide. According to F Quan. Nucleic Acids Res. 1986 July 11; 14(13): 5321 5335. Localization: In mammalian tissues found predominantly in liver, kidney, and erythrocytes, while the lowest levels are found in connective tissues. In tissues such as the liver, it is found predominantly in the peroxisome. In mature human erythrocytes, it is found in the cytosol. Catalase has also been found in the mitochondria of cardiac cells. According to F Quan. Nucleic Acids Res. 1986 July 11; 14(13): 5321 5335. Also according to Radi R. J Biol Chem. 1991 Nov 15;266(32):22028-34.
CBLATm	3	Dobson CM, Wai T, Leclerc D, Wilson A, Wu X, Dore C, Hudson T, Rosenblatt DS, Gravel RA.	Identification of the gene responsible for the cblA complementation group of vitamin B12-responsive methylmalonic acidemia based on analysis of prokaryotic gene arrangements.	Proc Natl Acad Sci U S A	2002	12438653	IT
CBLATm	3	Carmel R, Green R, Rosenblatt DS, Watkins D.	Update on cobalamin, folate, and homocysteine.	Hematology (Am Soc Hematol Educ Program)	2003	14633777	IT
CBPPer	2	Lueck JD, Nordlie RC.	The utilization of intramitochondrially generated carbamyl phosphate for microsomal glucose 6-phosphate biosynthesis.	FEBS Lett	1972	11946414	- additional activity of glucose-6-phosphate phosphatase [Orten, Human Biochem 1975] - reaction demonstrated in rat liver [Lueck 1972] - reaction produces NH3 & CO2 (not carbamate) according to [Lucius 1993]
CBPS	3	Chen KC, Vannais DB, Jones C, Patterson D, Davidson JN.	Mapping of the gene encoding the multifunctional protein carrying out the first three steps of pyrimidine biosynthesis to human chromosome 2.	Hum Genet	1989	2565865	IT
CBPS	3	Iwahana H, Fujimura M, Ii S, Kondo M, Moritani M, Takahashi Y, Yamaoka T, Yoshimoto K, Itakura M.	Molecular cloning of a human cDNA encoding a trifunctional enzyme of carbamoyl-phosphate synthetase-aspartate transcarbamoylase-dihydroorotase in de Novo pyrimidine synthesis.	Biochem Biophys Res Commun	1996	8619816	IT
CBPSam	3	Uriarte M, Marina A, Ramon-Maiques S, Fita I, Rubio V.	The carbamoyl-phosphate synthetase of Pyrococcus furiosus is enzymologically and structurally a carbamate kinase.	J Biol Chem	1999	10347186	- Infomation added by RS/TV: - Mitochondrial according to entrez Gene Database. - CPSI is highly tissue specific, with function and production limited to the liver and a lesser amount in the intestine. (Summar ML, Hall LD, Eeds AM, Hutcheson HB, Kuo AN, Willis AS, Rubio V, Arvin MK, Schofield JP, Dawson EP.Gene. 2003 Jun 5;311:51-7.) - Catalytic activity specified by GeneCards - Irreversible according to Uriarte M, Marina A, Ramon-Maiques S, Fita I, Rubio V. J Biol Chem. 1999 Jun 4;274(23):16295-303.
CBPSam	3	Summar ML, Hall LD, Eeds AM, Hutcheson HB, Kuo AN, Willis AS, Rubio V, Arvin MK, Schofield JP, Dawson EP.	Characterization of genomic structure and polymorphisms in the human carbamyl phosphate synthetase I gene.	Gene	2003	12853138	- Infomation added by RS/TV: - Mitochondrial according to entrez Gene Database. - CPSI is highly tissue specific, with function and production limited to the liver and a lesser amount in the intestine. (Summar ML, Hall LD, Eeds AM, Hutcheson HB, Kuo AN, Willis AS, Rubio V, Arvin MK, Schofield JP, Dawson EP.Gene. 2003 Jun 5;311:51-7.) - Catalytic activity specified by GeneCards - Irreversible according to Uriarte M, Marina A, Ramon-Maiques S, Fita I, Rubio V. J Biol Chem. 1999 Jun 4;274(23):16295-303.
CBPter	2	Lucius RW, Waddell ID, Burchell A, Nordlie RC	The hepatic glucose-6-phosphatase system in Ehrlich-ascites-tumour-bearing mice	Biochem J	1993	8384451	- rxn catalyzed by T2 beta translocase of glucose-6-phosphatase system [Lucius 1993], [Foster 2002]
CBPter	2	Foster JD, Nordlie RC	The biochemistry and molecular biology of the glucose-6-phosphatase system.	Exp Biol Med (Maywood)	2002	12192101	- rxn catalyzed by T2 beta translocase of glucose-6-phosphatase system [Lucius 1993], [Foster 2002]
CBR1	3	Okita RT, Okita JR.	Prostaglandin-metabolizing enzymes during pregnancy: characterization of NAD(+)-dependent prostaglandin dehydrogenase, carbonyl reductase, and cytochrome P450-dependent prostaglandin omega-hydroxylase.	Crit Rev Biochem Mol Biol	1996	8740524	cytosol - uniprot Carbonyl reductase is one of several monomeric, NADPH-dependent oxidoreductases having wide specificity for carbonyl compounds. This enzyme is widely distributed in human tissues. Another carbonyl reductase gene, CRB3, lies close to this gene on chromosome 21q. Specific rxns for CBR1 found, just general function + chrom mapping has been identified for CBR3 NJ

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CBR1	3	Watanabe K, Sugawara C, Ono A, Fukuzumi Y, Itakura S, Yamazaki M, Tashiro H, Osoegawa K, Soeda E, Nomura T.	Mapping of a novel human carbonyl reductase, CBR3, and ribosomal pseudogenes to human chromosome 21q22.2.	Genomics	1998	9740676	cytosol - uniprot Carbonyl reductase is one of several monomeric, NADPH-dependent oxidoreductases having wide specificity for carbonyl compounds. This enzyme is widely distributed in human tissues. Another carbonyl reductase gene, CRB3, lies close to this gene on chromosome 21q. Specific rxns for CBR1 found, just general function + chrom mapping has been identified for CBR3 NJ
CDIPTr	3	Lykidis A, Jackson PD, Rock CO, Jackowski S	The role of CDP-diacylglycerol synthetase and phosphatidylinositol synthase activity levels in the regulation of cellular phosphatidylinositol content	Journal of Biological Chemistry	1997		cytoplasmic side of ER - uniprot + refs Reversible reaction as per uniprot catalyzes the biosynthesis of phosphatidylinositol (PtdIns) as well as PtdIns:inositol exchange reaction [see Holub reaction PMID: 2425833]. May thus act to reduce an excessive cellular PtdIns content. The exchange activity is due to the reverse reaction. Widely expressed. Higher expression in adult liver and skeletal muscle, slightly lower levels seen in pancreas, kidney, lung, placenta, brain, heart, leucocyte, colon, small intestine, ovary, testis, prostate, thymus and spleen. In fetus, expressed in kidney, liver, lung and brain. Phosphatidylinositol breakdown products are ubiquitous second messengers that function downstream of many G protein-coupled receptors and tyrosine kinases regulating cell growth, calcium metabolism, and protein kinase C activity. Two enzymes, CDP-diacylglycerol synthase and phosphatidylinositol synthase, are involved in the biosynthesis of phosphatidylinositol. Phosphatidylinositol synthase, a member of the CDP-alcohol phosphatidyl transferase class-I family, is an integral membrane protein found on the cytoplasm NJ
CDS	3	Weeks R, Dowhan W, Shen H, Balantac N, Meengs B, Nudelman E, Leung DW	Isolation and expression of an isoform of human CDP-diacylglycerol synthease cDNA	DNA Cell Biol	1997		ER membrane - cytosolic side - UniProt Expressed in adult tissues such as placenta, brain, small intestine, ovary, testis and prostate. Highly expressed in fetal kidney, lung and brain. Lower level in fetal liver. NJ
CDS	3	Halford S, Dulai KS, Daw SC, Fitzgibbon J, Hunt DM	Isolation and chromosomal localization of two human cdp-diacylglycerol synthase genes	Genomics	1998		ER membrane - cytosolic side - UniProt Expressed in adult tissues such as placenta, brain, small intestine, ovary, testis and prostate. Highly expressed in fetal kidney, lung and brain. Lower level in fetal liver. NJ
CDS	3	Weeks R, Dowhan W, Shen H, Balantac N, Meengs B, Nudelman E, Leung DW	Isolation and expression of an isoform of human CDP-diacylglycerol synthease cDNA	DNA Cell Biol	1997		ER membrane - cytosolic side - UniProt Expressed in adult tissues such as placenta, brain, small intestine, ovary, testis and prostate. Highly expressed in fetal kidney, lung and brain. Lower level in fetal liver. NJ
CDS	3	Halford S, Dulai KS, Daw SC, Fitzgibbon J, Hunt DM	Isolation and chromosomal localization of two human cdp-diacylglycerol synthase genes	Genomics	1998		ER membrane - cytosolic side - UniProt Expressed in adult tissues such as placenta, brain, small intestine, ovary, testis and prostate. Highly expressed in fetal kidney, lung and brain. Lower level in fetal liver. NJ
CEPTC	3	Henneberry AL, McMaster CR	Cloning and expression of a human choline/ethanolaminephosphotransferase: synthesis of phosphatidylcholine and phosphatidylethanolamine	Biochem J	1999		cytosolic/integral membrane protein - Uniprot need to add additional ref for TV CHPT1 NJ
CERK	3	Sugiura M, Kono K, Liu H, Shimizugawa T, Minekura H, Spiegel S, Kohama T.	Ceramide kinase, a novel lipid kinase. Molecular cloning and functional characterization.	J Biol Chem	2002		cytoplasmic - uniprot cloning + biochem activity - see Sugiura ref Catalyzes specifically the phosphorylation of ceramide to form ceramide 1-phosphate. Acts efficiently on natural and analog ceramides (C6, C8, C16 ceramides, and C8-dihydroceramide), to a lesser extent on C2-ceramide and C6-dihydroceramide, but not on other lipids, such as various sphingosines. NJ
CERT1gt	2	van Meer G, Wolthoorn J, Degroote S.	The fate and function of glycosphingolipid glucosylceramide.	Philos Trans R Soc Lond B Biol Sci	2003	12803919	Two transcripts exist - one encodes a kinase that specifically phosphorylates the N-terminal region of the non-collagenous domain of the alpha 3 chain of type IV collagen, known as the Goodpasture antigen. The other isoform (the one encoded here) of this protein is also i Possibly ATP dependent pathway - given confidence level of 2 at this point. See also PMID: 15907394 for review. See PMID: 12803919 for proposed bidirectionality of transport. FUTURE UPDATES: For alternative substrates see PMID: 15596449 NJ

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
CERT1gt	2	Hanada K, Kumagai K, Yasuda S, Miura Y, Kawano M, Fukasawa M, Nishijima M.	Molecular machinery for non-vesicular trafficking of ceramide.	Nature	2003	14685229	Two transcripts exist - one encodes a kinase that specifically phosphorylates the N-terminal region of the non-collagenous domain of the alpha 3 chain of type IV collagen, known as the Goodpasture antigen. The other isoform (the one encoded here) of this protein is also iPossibly ATP dependent pathway - given confidence level of 2 at this point. See also PMID: 15907394 for review. See PMID: 12803919 for proposed bidirectionality of transport. FUTURE UPDATES: For alternative substrates see PMID: 15596449 NJ
CERT1gt	2	Kumagai K, Yasuda S, Okemoto K, Nishijima M, Kobayashi S, Hanada K.	CERT mediates intermembrane transfer of various molecular species of ceramides.	J Biol Chem	2005	15596449	Two transcripts exist - one encodes a kinase that specifically phosphorylates the N-terminal region of the non-collagenous domain of the alpha 3 chain of type IV collagen, known as the Goodpasture antigen. The other isoform (the one encoded here) of this protein is also iPossibly ATP dependent pathway - given confidence level of 2 at this point. See also PMID: 15907394 for review. See PMID: 12803919 for proposed bidirectionality of transport. FUTURE UPDATES: For alternative substrates see PMID: 15596449 NJ
CERT1gt	2	Perry RJ, Ridgway ND.	Molecular mechanisms and regulation of ceramide transport.	Biochim Biophys Acta	2005	15907394	Two transcripts exist - one encodes a kinase that specifically phosphorylates the N-terminal region of the non-collagenous domain of the alpha 3 chain of type IV collagen, known as the Goodpasture antigen. The other isoform (the one encoded here) of this protein is also iPossibly ATP dependent pathway - given confidence level of 2 at this point. See also PMID: 15907394 for review. See PMID: 12803919 for proposed bidirectionality of transport. FUTURE UPDATES: For alternative substrates see PMID: 15596449 NJ
CGLYt3(2)	2	Daniel H, Kottra G	The proton oligopeptide cotransporter family SLC15 in physiology and pharmacology	Pflugers Arch	2004	12905028	From PMID 12905028: In contrast, PEPT2 transports neutral substrates with a 2:1 proton to substrate stoichiometry [11] and charged substrates with variable coupling ratios. It was proposed that PEPT2 in brain astrocytes could contribute to brain glutathione metabolism by providing cysteinyl-glycine derived from extracellular glutathione for glial glutathione resynthesis [17]
CHAT	3	Dobransky T, Davis WL, Xiao GH, Rylett RJ.	Expression, purification and characterization of recombinant human choline acetyltransferase: phosphorylation of the enzyme regulates catalytic activity.	Biochem J	2000	10861222	cytosolic and nuclear - uniprot reversibility by uniprot also specificity: CNS/PNS Cholinergic systems are implicated in numerous neurologic functions. Alteration in some cholinergic neurons may account for the disturbances of Alzheimer disease. The protein encoded by this gene synthesizes the neurotransmitter acetylcholine. Alternative splice variants have been found that contain alternative 5' untranslated exons. Three of the four described splice variants encode identical 69 kDa proteins while one variant encodes both the 69 kDa and a larger 82 kDa protein. NJ
CHLP	0	Roberts SJ, Stewart AJ, Sadler PJ, Farquharson C.	Human PHOSPHO1 exhibits high specific phosphoethanolamine and phosphocholine phosphatase activities.	Biochem J	2004	15175005	cytosol - by default, no specific info in lit NJ
CHLPCTD	3	Kent C	Eukaryotic phospholipid biosynthesis	Annu Rev Biochem	1995		cytoplasmic - unitprot NJ - checked by RS/TV NOTE: This reaction was classified under the "Lipid" subsystem in the mitochondrial model.
CHLtm	2	de Ridder JJ.	The uptake of choline by rat liver mitochondria.		1976	10984	-added to allow choline transport from cytosol to mitochondria -choline is an important nutrient in diet (PMID:15640516) -choline transport by rat liver mitochondria demonstrated but mechanism is unclear (PMID:10984) MM

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CHOLATEt	3	Kullak-Ublick GA, Beuers U, Meier PJ, Domdey H, Paumgartner G.	Assignment of the human organic anion transporting polypeptide (OATP) gene to chromosome 12p12 by fluorescence in situ hybridization.	J Hepatol	1996	9007731	Tissue Specificity: SLCO2A1 - ubiquitous SLCO1A2 - brain, kidney, lung, testis, liver SLCO1B1 - liver SLCO1B3 - liver SLCO2B1 - liver, placenta, spleen, lung, kidney, heart, ovary SLCO3A1 - ubiquitously SLCO4A1 - ubiquitously SLCO1C1 - brain, testis May mediate the release of newly synthesized prostaglandins from cells, the transepithelial transport of prostaglandins, and the clearance of prostaglandins from the circulation. NJ
CHOLATEt2	3	Wong MH, Oelkers P, Dawson PA.	Identification of a mutation in the ileal sodium-dependent bile acid transporter gene that abolishes transport activity.	J Biol Chem	1995	7592981	SLC10A1: liver pancreas SLC10A2: ileum, kidney, biliary tract other SLC transporters (A3, A4, A5) have not been biochemically characterized for substrates & cotransporters. SLC10 review in PMID: 12851823 Only SLC10A1 is known to also have estrone-3-sulfate transport capabilites. Only unidirectional (uptake) activity has been shown. See CHOLATEt/GCHOLAt/TCHOLAt for bile acid excretion acitivity. Sodium/bile acid cotransporters are integral membrane glycoproteins that participate in the enterohepatic circulation of bile acids. Two homologous transporters are involved in the reabsorption of bile acids, one absorbing from the intestinal lumen, the bile duct, and the kidney with an apical localization (SLC10A2; MIM 601295), and the other being found in the basolateral membranes of hepatocytes (SLC10A1). NJ
CHOLATEt2	3	Hagenbuch B, Meier PJ.	Molecular cloning, chromosomal localization, and functional characterization of a human liver Na+/bile acid cotransporter.	J Clin Invest	1994	8132774	SLC10A1: liver pancreas SLC10A2: ileum, kidney, biliary tract other SLC transporters (A3, A4, A5) have not been biochemically characterized for substrates & cotransporters. SLC10 review in PMID: 12851823 Only SLC10A1 is known to also have estrone-3-sulfate transport capabilites. Only unidirectional (uptake) activity has been shown. See CHOLATEt/GCHOLAt/TCHOLAt for bile acid excretion acitivity. Sodium/bile acid cotransporters are integral membrane glycoproteins that participate in the enterohepatic circulation of bile acids. Two homologous transporters are involved in the reabsorption of bile acids, one absorbing from the intestinal lumen, the bile duct, and the kidney with an apical localization (SLC10A2; MIM 601295), and the other being found in the basolateral membranes of hepatocytes (SLC10A1). NJ
CHOLATEt2	3	Oelkers P, Kirby LC, Heubi JE, Dawson PA.	Primary bile acid malabsorption caused by mutations in the ileal sodium-dependent bile acid transporter gene (SLC10A2).	J Clin Invest	1997	9109432	SLC10A1: liver pancreas SLC10A2: ileum, kidney, biliary tract other SLC transporters (A3, A4, A5) have not been biochemically characterized for substrates & cotransporters. SLC10 review in PMID: 12851823 Only SLC10A1 is known to also have estrone-3-sulfate transport capabilites. Only unidirectional (uptake) activity has been shown. See CHOLATEt/GCHOLAt/TCHOLAt for bile acid excretion acitivity. Sodium/bile acid cotransporters are integral membrane glycoproteins that participate in the enterohepatic circulation of bile acids. Two homologous transporters are involved in the reabsorption of bile acids, one absorbing from the intestinal lumen, the bile duct, and the kidney with an apical localization (SLC10A2; MIM 601295), and the other being found in the basolateral membranes of hepatocytes (SLC10A1). NJ

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CHOLATEt2	3	Hagenbuch B, Dawson P.	The sodium bile salt cotransport family SLC10.	Pflugers Arch	2004	12851823	SLC10A1: liver pancreas SLC10A2: ileum, kidney, biliary tract other SLC transporters (A3, A4, A5) have not been biochemically characterized for substrates & cotransporters. SLC10 review in PMID: 12851823 Only SLC10A1 is known to also have estrone-3-sulfate transport capabilites. Only unidirectional (uptake) activity has been shown. See CHOLATEt/GCHOLAt/TCHOLAt for bile acid excretion acitivity. Sodium/bile acid cotransporters are integral membrane glycoproteins that participate in the enterohepatic circulation of bile acids. Two homologous transporters are involved in the reabsorption of bile acids, one absorbing from the intestinal lumen, the bile duct, and the kidney with an apical localization (SLC10A2; MIM 601295), and the other being found in the basolateral membranes of hepatocytes (SLC10A1). NJ
CHOLATEt3	3	Kiuchi Y, Suzuki H, Hirohashi T, Tyson CA, Sugiyama Y.	cDNA cloning and inducible expression of human multidrug resistance associated protein 3 (MRP3).	FEBS Lett	1998	9738950	This protein is a member of the MDR/TAP subfamily. Members of the MDR/TAP subfamily are involved in multidrug resistance. The protein encoded by this gene is the major canalicular bile salt export pump in man. Mutations in this gene cause a form of progressive familial intrahepatic cholestases which are a group of inherited disorders with severe cholestatic liver disease from early infancy. NJ
CHOLATEt3	3	Strautnieks SS, Bull LN, Knisely AS, Kocoshis SA, Dahl N, Arnell H, Sokal E, Dahan K, Childs S, Ling V, Tanner MS, Kagalwalla AF, Nemeth A, Pawlowska J, Baker A, Mieli-Vergani G, Freimer NB, Gardiner RM, Thompson RJ.	A gene encoding a liver-specific ABC transporter is mutated in progressive familial intrahepatic cholestasis.	Nat Genet	1998	9806540	This protein is a member of the MDR/TAP subfamily. Members of the MDR/TAP subfamily are involved in multidrug resistance. The protein encoded by this gene is the major canalicular bile salt export pump in man. Mutations in this gene cause a form of progressive familial intrahepatic cholestases which are a group of inherited disorders with severe cholestatic liver disease from early infancy. NJ
CHOLATEt3	3	Uchiumi T, Hinoshita E, Haga S, Nakamura T, Tanaka T, Toh S, Furukawa M, Kawabe T, Wada M, Kagotani K, Okumura K, Kohno K, Akiyama S, Kuwano M.	Isolation of a novel human canalicular multispecific organic anion transporter, cMOAT2/MRP3, and its expression in cisplatin-resistant cancer cells with decreased ATP-dependent drug transport.	Biochem Biophys Res Commun	1998	9813153	This protein is a member of the MDR/TAP subfamily. Members of the MDR/TAP subfamily are involved in multidrug resistance. The protein encoded by this gene is the major canalicular bile salt export pump in man. Mutations in this gene cause a form of progressive familial intrahepatic cholestases which are a group of inherited disorders with severe cholestatic liver disease from early infancy. NJ
CHOLK	3	Activity and tissue distribution of splice variants of alpha6-fucosyltransferase in human embryogenesis.	Martinez-Duncker I, Michalski JC, Bauvy C, Candelier JJ, Mennesson B, Codogno P, Oriol R, Mollicone R.	Glycobiology	2004	14514715	cytosolic - uniport CK has chol and ethal specificity according to RefSeq NJ
CHOLt4	3	Apparsundaram S, Ferguson SM, George AL Jr, Blakely RD	Molecular cloning of a human, hemicholinium-3-sensitive choline transporter	Biochem Biophys Res Commun	2000	11027560	- identified by sequence homology [Apparsundaram 2000] - cloned [Apparsundaram 2000], [Okuda 2000] - expressed in brain regions rich in cholinergic neurons [Apparsundaram 2000], [Okuda 2000] -Na(+)- and Cl(-)-dependent, high-affinity choline uptake [Okuda 2000]; Na+ is cotransported [Okuda 2003]
CHOLt4	3	Okuda T, Haga T.	Functional characterization of the human high-affinity choline transporter	FEBS Lett	2000	11068039	- identified by sequence homology [Apparsundaram 2000] - cloned [Apparsundaram 2000], [Okuda 2000] - expressed in brain regions rich in cholinergic neurons [Apparsundaram 2000], [Okuda 2000] -Na(+)- and Cl(-)-dependent, high-affinity choline uptake [Okuda 2000]; Na+ is cotransported [Okuda 2003]
CHOLt4	3	Okuda T, Haga T	High-affinity choline transporter	Neurochem Res	2003	12675135	- identified by sequence homology [Apparsundaram 2000] - cloned [Apparsundaram 2000], [Okuda 2000] - expressed in brain regions rich in cholinergic neurons [Apparsundaram 2000], [Okuda 2000] -Na(+)- and Cl(-)-dependent, high-affinity choline uptake [Okuda 2000]; Na+ is cotransported [Okuda 2003]

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CHSTEROLSULT	3	Her C, Wood TC, Eichler EE, Mohrenweiser HW, Ramagli LS, Siciliano MJ, Weinshilboum RM.	Human hydroxysteroid sulfotransferase SULT2B1: two enzymes encoded by a single chromosome 19 gene.	Genomics	1998	9799594	cytosolic by swiss-prot see PMID: 9799594, 12145317 Catalyzes the sulfate conjugation of many hormones, neurotransmitters, drugs and xenobiotic compounds. Sulfonation increases the water solubility of most compounds, and therefore their renal excretion, but it can also result in bioactivation to form active metabolites. Sulfates hydroxysteroids like DHEA. Isoform 1 preferentially sulfonates cholesterol, and isoform 2 avidly sulfonates pregnenolone but not cholesterol. NJ
CHSTEROLSULT	3	Fuda H, Lee YC, Shimizu C, Javitt NB, Strott CA.	Mutational analysis of human hydroxysteroid sulfotransferase SULT2B1 isoforms reveals that exon 1B of the SULT2B1 gene produces cholesterol sulfotransferase, whereas exon 1A yields pregnenolone sulfotransferase.	J Biol Chem	2002	12145317	cytosolic by swiss-prot see PMID: 9799594, 12145317 Catalyzes the sulfate conjugation of many hormones, neurotransmitters, drugs and xenobiotic compounds. Sulfonation increases the water solubility of most compounds, and therefore their renal excretion, but it can also result in bioactivation to form active metabolites. Sulfates hydroxysteroids like DHEA. Isoform 1 preferentially sulfonates cholesterol, and isoform 2 avidly sulfonates pregnenolone but not cholesterol. NJ
CHSTEROLt	3	Orso E, Broccardo C, Kaminski WE, Bottcher A, Liebisch G, Drobnik W, Gotz A, Chambenoit O, Diederich W, Langmann T, Spruss T, Luciani MF, Rothe G, Lackner KJ, Chimini G, Schmitz G.	Transport of lipids from golgi to plasma membrane is defective in tangier disease patients and Abc1-deficient mice.	Nat Genet	2000	10655069	IT Gene is also responsiblefor phospholipid efflux orso has shown transport of choline phospholipid
CHSTEROLt	3	Wollmer MA, Streffer JR, Lutjohann D, Tsolaki M, Iakovidou V, Hegi T, Pasch T, Jung HH, Bergmann K, Nitsch RM, Hock C, Papassotiropoulos A.	ABCA1 modulates CSF cholesterol levels and influences the age at onset of Alzheimer's disease.	Neurobiol Aging	2003	12600718	IT Gene is also responsiblefor phospholipid efflux orso has shown transport of choline phospholipid
CHSTEROLt1	2	Zhang M, Liu P, Dwyer NK, Christenson LK, Fujimoto T, Martinez F, Comly M, Hanover JA, Blanchette-Mackie EJ, Strauss JF 3rd.	MLN64 mediates mobilization of lysosomal cholesterol to steroidogenic mitochondria.	J Biol Chem	2002	12070139	Has been shown experimentally to be associated with cholesterol transport from lysosome to mitochondria. The mechanism is also likely to be proton dependent, however the stoichiometry and biochemical details have not yet been elucidated. See PMID: 12070139 for Stard3 (aka MLN64) specific for Stard3 See PMID: 12370263 and PMID: 12770731 for reviews and discussion of STAR transporters NJ
CHSTEROLt1	2	Jefcoate C.	High-flux mitochondrial cholesterol trafficking, a specialized function of the adrenal cortex.	J Clin Invest	2002	12370263	Has been shown experimentally to be associated with cholesterol transport from lysosome to mitochondria. The mechanism is also likely to be proton dependent, however the stoichiometry and biochemical details have not yet been elucidated. See PMID: 12070139 for Stard3 (aka MLN64) specific for Stard3 See PMID: 12370263 and PMID: 12770731 for reviews and discussion of STAR transporters NJ
CHSTEROLt1	2	Strauss JF 3rd, Kishida T, Christenson LK, Fujimoto T, Hiroi H.	START domain proteins and the intracellular trafficking of cholesterol in steroidogenic cells.	Mol Cell Endocrinol	2003	12770731	Has been shown experimentally to be associated with cholesterol transport from lysosome to mitochondria. The mechanism is also likely to be proton dependent, however the stoichiometry and biochemical details have not yet been elucidated. See PMID: 12070139 for Stard3 (aka MLN64) specific for Stard3 See PMID: 12370263 and PMID: 12770731 for reviews and discussion of STAR transporters NJ
CHTNASEe	3	Boot RG, Blommaart EF, Swart E, Ghauharali-van der Vlugt K, Bijl N, Moe C, Place A, Aerts JM	Identification of a novel acidic mammalian chitinase distinct from chitotriosidase	J Biol Chem	2001	11085997	- physiological function is unknown, but enzyme was shown to have chitinase activity in vitro [Boot, J Biol Chem 2001] - abundant in gastrointestinal tract and found to lesser extent in lung [Boot, J Biol Chem 2001], [UniProt] - this mRNA has a 108 bp signal sequence that suggests it is secreted (probable) [Uni-Prot]
CITL	0	Morikawa J, Nishimura Y, Uchida A, Tanaka T	Molecular cloning of novel mouse and human putative citrate lyase beta-subunit	Biochem Biophys Res Commun	2001	11741334	- putative function based on sequence similarity to murine gene [Morikawa et al, Biochem Biophys Res Commun2001] - cytosolic [Devlin, Textbook of biochemistry, 5th ed]

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CITMCOAHm	2	ADLER J, WANG SF, LARDY HA	The metabolism of itaconic acid by liver mitochondria.	J Biol Chem	1957	13502348	- reaction occurs in rat liver mitochondria [Wang, J Biol Chem 1961] - Itaconate, citraconate, and mesaconate are probably metabolized in the dog, since they could be recovered in the urine to the extent of only 24, 28, and 64 per cent, respectively (see refs in [Adler 1957]) - In guinea pig liver, itaconate is oxidized as rapidly as most members of the tricarboxylic acid cycle, methyl succinate was oxidized 1/6 as fast and mesaconate 1/8 as fast as itaconate [Adler 1957] - the major pathway of itaconate metabolism is its oxidation to pyruvate [Adler 1957]
CITt4_2	3	Pajor AM, Sun N	Functional differences between rabbit and human Na(+)-dicarboxylate cotransporters, NaDC-1 and hNaDC-1.	Am J Physiol	1996	8946005	9058: - cloned [Pajor 1996] - 78% identical to the rabbit and 42% identical to the rat orthologs [Pajor 1996] - low-affinity cotransport of Na+ w/ succinate and citrate [Pajor 1996] - kidney and intestine brush border membranes [Pajor 1996] - Hill coefficient of 2.1 [Pajor, Sun 1996]
CITt4_2	3	Pajor AM	Molecular cloning and functional expression of a sodium-dicarboxylate cotransporter from human kidney	Am J Physiol	1996	8967342	9058: - cloned [Pajor 1996] - 78% identical to the rabbit and 42% identical to the rat orthologs [Pajor 1996] - low-affinity cotransport of Na+ w/ succinate and citrate [Pajor 1996] - kidney and intestine brush border membranes [Pajor 1996] - Hill coefficient of 2.1 [Pajor, Sun 1996]
CITt4_4	3	Inoue K, Zhuang L, Maddox DM, Smith SB, Ganapathy V	Structure, function, and expression pattern of a novel sodium-coupled citrate transporter (NaCT) cloned from mammalian brain	J Biol Chem	2002	12177002	284111: - cloned [Inoue Biochem Biophys Res Commun 2002] - high in liver; moderate in brain, testis [Inoue 2002] - low affinty, cotransports Na+ w/ citrate, no succinate or malate [Inoue Biochem Biophys Res Commun 2002] - 77% sequence identity with rat ortholog [Inoue Biochem Biophys Res Commun 2002] - Na+:citrate stoichiometry couldn't be determined for human NaCT [Inoue Biochem Biophys Res Commun 2002]; assume is the same as rat ortholog (4:1) [Inohue J Biol Chem 2002]
CITt4_4	3	Inoue K, Zhuang L, Ganapathy V	Human Na+ -coupled citrate transporter: primary structure, genomic organization, and transport function	Biochem Biophys Res Commun	2002	12445824	284111: - cloned [Inoue Biochem Biophys Res Commun 2002] - high in liver; moderate in brain, testis [Inoue 2002] - low affinty, cotransports Na+ w/ citrate, no succinate or malate [Inoue Biochem Biophys Res Commun 2002] - 77% sequence identity with rat ortholog [Inoue Biochem Biophys Res Commun 2002] - Na+:citrate stoichiometry couldn't be determined for human NaCT [Inoue Biochem Biophys Res Commun 2002]; assume is the same as rat ortholog (4:1) [Inohue J Biol Chem 2002]
CK	3	Klein SC, Haas RC, Perryman MB, Billadello JJ, Strauss AW.	Regulatory element analysis and structural characterization of the human sarcomeric mitochondrial creatine kinase gene.	J Biol Chem	1991	1917943	- Additional information added by RS/TV: -proteome Mitochondrial creatine kinase comes in two isozymes: Ckmt1.1 m and Ckmt2.1-m according to Entrez gene database. (1) Catalytic Activity: Creatine kinases catalyze the reversible transfer of high energy phosphate from ATP to creatine generating ADP and phosphocreatine. (2) Tissue specificity: Ckmt2.1-m is expressed in only stirated muscle, including heart. Ckmt1.1-m on the other hand was found to be expressed in adult skeletal muscle, ventricle, and to a lesser extent in the small intestine and placenta. All this according to (1) Haas RC, Korenfeld C, Zhang ZF, Perryman B, Roman D, Strauss AW. J Biol Chem. 1989 Feb 15;264(5):2890-7. Erratum in: J Biol Chem 1989 Sep 25;264(27):16332. (2) Klein SC, Haas RC, Perryman MB, Billadello JJ, Strauss AW. J Biol Chem. 1991 Sep 25;266(27):18058-65. Runs forward in mito --SAB

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CK	3	Haas RC, Korenfeld C, Zhang ZF, Perryman B, Roman D, Strauss AW.	Isolation and characterization of the gene and cDNA encoding human mitochondrial creatine kinase.	J Biol Chem	1989	2914937	- Additional information added by RS/TV: -proteome Mitochondrial creatine kinase comes in two isozymes: Ckmt1.1 m and Ckmt2.1-m according to Entrez gene database. (1) Catalytic Activity: Creatine kinases catalyze the reversible transfer of high energy phosphate from ATP to creatine generating ADP and phosphocreatine. (2) Tissue specificity: Ckmt2.1-m is expressed in only stirated muscle, including heart. Ckmt1.1-m on the other hand was found to be expressed in adult skeletal muscle, ventricle, and to a lesser extent in the small intestine and placenta. All this according to (1) Haas RC, Korenfeld C, Zhang ZF, Perryman B, Roman D, Strauss AW. J Biol Chem. 1989 Feb 15;264(5):2890-7. Erratum in: J Biol Chem 1989 Sep 25;264(27):16332. (2) Klein SC, Haas RC, Perryman MB, Billadello JJ, Strauss AW. J Biol Chem. 1991 Sep 25;266(27):18058-65. Runs forward in mito --SAB
CKc	3	Kaye FJ, McBride OW, Battey JF, Gazdar AF, Sausville EA	Human creatine kinase-B complementary DNA. Nucleotide sequence, gene expression in lung cancer, and chromosomal assignment to two distinct loci	J Clin Invest	1987	2883200	gene and reaction are well-characterized runs backward in cytosol--SAB
CKc	3	Villarreal-Levy G, Ma TS, Kerner SA, Roberts R, Perryman MB	Human creatine kinase: isolation and sequence analysis of cDNA clones for the B subunit, development of subunit specific probes and determination of gene copy number	Biochem Biophys Res Commun	1987	3034271	gene and reaction are well-characterized runs backward in cytosol--SAB
CMPACNAtg	3	Ishida N, Miura N, Yoshioka S, Kawakita M	Molecular cloning and characterization of a novel isoform of the human UDP-galactose transporter, and of related complementary DNAs belonging to the nucleotide-sugar transporter gene family	J Biochem (Tokyo)	1996	9010752	- cloned [Ishida 1996] - Golgi membrane [Ishida 1998] - corrects CMP-Sia knockout phenotype [Ishida 1998] - ubiquitous [Ishida 2004]
CMPACNAtg	3	Ishida N, Ito M, Yoshioka S, Sun-Wada GH, Kawakita M	Functional expression of human golgi CMP-sialic acid transporter in the Golgi complex of a transporter-deficient Chinese hamster ovary cell mutant	J Biochem (Tokyo)	1998	9644260	- cloned [Ishida 1996] - Golgi membrane [Ishida 1998] - corrects CMP-Sia knockout phenotype [Ishida 1998] - ubiquitous [Ishida 2004]
CMPACNAtg	3	Ishida N, Kawakita M	Molecular physiology and pathology of the nucleotide sugar transporter family (SLC35)	Pflugers Arch	2004	12759756	- cloned [Ishida 1996] - Golgi membrane [Ishida 1998] - corrects CMP-Sia knockout phenotype [Ishida 1998] - ubiquitous [Ishida 2004]
CMPSAS	2	Munster AK, Eckhardt M, Potvin B, Muhlenhoff M, Stanley P, Gerardy-Schahn R	Mammalian cytidine 5'-monophosphate N-acetylneuraminic acid synthetase: a nuclear protein with evolutionarily conserved structural motifs	Proc Natl Acad Sci U S A	1998	9689047	- murine protein was shown to be predominantly (90%) localized to the nucleus; also localized to cytosol [Munster, PNAS 1998] - reaction shown as irreversible in Varki p. 74, Orten p. 245, Devlin p. 672
CO2tm	3	Balboni E, Lehninger AL.	Entry and exit pathways of CO2 in rat liver mitochondria respiring in a bicarbonate buffer system.	J Biol Chem	1968	3081508	- Added by RS/TV - This article states the following, "this conclusion [of the experiment] is in accord with the generally accepted view that CO2 readily traverses biological membranes, including those of the mitochondrion probably by unmediated physical diffusion". (Balboni E, Lehninger AL. J Biol Chem. 1986 Mar 15;261(8):3563-70.)
COAtl	2	Seetharam B, Alpers DH.	Absorption and transport of cobalamin (vitamin B12).	Annu Rev Nutr	1982	6313022	IT apparently, the lysosomal membrane is permeable for molecules of mol wt 400 or less.(Seetharam, p. 359).
COKECBESr	3	Pindel EV, Kedishvili NY, Abraham TL, Brzezinski MR, Zhang J, Dean RA, Bosron WF	Purification and cloning of a broad substrate specificity human liver carboxylesterase that catalyzes the hydrolysis of cocaine and heroin	J Biol Chem	1997	9169443	CES1 may catalyze a similar, but slightly different, reaction that is not in KEGG apparantly reaction may also proceed nonenzymatically
CORE2GTg	0	Yeh JC, Ong E, Fukuda M	Molecular cloning and expression of a novel beta-1, 6-N-acetylglucosaminyltransferase that forms core 2, core 4, and I branches	J Biol Chem	1999		GCNT1 is a member of the beta-1,6-N-acetylglucosaminyltransferase gene family. It is essential to the formation of Gal beta 1-3(GlcNAc beta 1-6)GalNAc structures and the core 2 O-glycan branch. The gene coding this enzyme was originally mapped to 9q21, but was later localized to 9q13 by another group. [RefSeq] C2GNT3 was identified by BLAST analysis; high levels in the thymus [Schwientek et al, J Biol Chem 2000] Gcnt1p is also referred to as C2GnT-L [Yeh et al, J Biol Chem 1999] Gcnt1p ubiquitously expressed [Yeh et al, J Biol Chem 1999] Gcnt3p expressed in colon, sm intestine, stomach, thyroid, testis, prostate, kidney, pancreas [Yeh et al, J Biol Chem 1999]

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CORE2GTg	0	Schwientek T, Yeh JC, Levery SB, Keck B, Merkx G, van Kessel AG, Fukuda M, Clausen H	Control of O-glycan branch formation. Molecular cloning and characterization of a novel thymus-associated core 2 beta1, 6-n-acetylglucosaminyltransferase	J Biol Chem	2000	10753916	GCNT1 is a member of the beta-1,6-N-acetylglucosaminyltransferase gene family. It is essential to the formation of Gal beta 1-3(GlcNAc beta 1-6)GalNAc structures and the core 2 O-glycan branch. The gene coding this enzyme was originally mapped to 9q21, but was later localized to 9q13 by another group. [RefSeq] C2GNT3 was identified by BLAST analysis; high levels in the thymus [Schwientek et al, J Biol Chem 2000] Gcnt1p is also referred to as C2GnT-L [Yeh et al, J Biol Chem 1999] Gcnt1p ubiquitously expressed [Yeh et al, J Biol Chem 1999] Gcnt3p expressed in colon, sm intestine, stomach, thyroid, testis, prostate, kidney, pancreas [Yeh et al, J Biol Chem 1999]
CORE3GTg	0	Iwai T, Inaba N, Naundorf A, Zhang Y, Gotoh M, Iwasaki H, Kudo T, Togayachi A, Ishizuka Y, Nakanishi H, Narimatsu H	Molecular cloning and characterization of a novel UDP-GlcNAc:GalNAc-peptide beta1,3-N-acetylglucosaminyltransferase (beta 3Gn-T6), an enzyme synthesizing the core 3 structure of O-glycans	J Biol Chem	2002	11821425	transcript is expressed in stomach, colon, sm intestine, sk muscle, testis [Iwai et al, J Biol Chem 2002]
CPPPGO	2	Kohno H, Furukawa T, Yoshinaga T, Tokunaga R, Taketani S.	Coproporphyrinogen oxidase. Purification, molecular cloning, and induction of mRNA during erythroid differentiation.	J Biol Chem	1995	8407975	- Added by RS/TV Proteome - Coproporphyrinogen III oxidase is a soluble mitochondrial protein that is localized in the intermembrance space within mammalian cells. It catalyzes the sixth step in heme biosynthesis, the conversion of the two propionate groups at positions 2 and 4 of coproporphyrinogen III to two vinyl groups, thus producing protoporphyrinogen IX. - Located in liver cells, also found in the cytosol. (Kohno H, Furukawa T, Yoshinaga T, Tokunaga R, Taketani S. J Biol Chem. 1993 Oct 5;268(28):21359-63.)
CREATtmdiffir	2	Bessman SP, Carpenter CL	The creatine-creatine phosphate energy shuttle	Annu Rev Biochem	1985	3896131	Added to make creatine shuttle. Need to add Biochemical Pathways book citation.
CREATtmdiffir	2	Speer O, Neukomm LJ, Murphy RM, Zanolla E, Schlattner U, Henry H, Snow RJ, Wallimann T	Creatine transporters: a reappraisal	Mol Cell Biochem	2004	14977199	Added to make creatine shuttle. Need to add Biochemical Pathways book citation.
CREATtmdiffir	2		Biochemical pathways : an atlas of biochemistry and molecular biology		1999		Added to make creatine shuttle. Need to add Biochemical Pathways book citation.
CRMPte	2	Baumruker T, Bornancin F, Billich A.	The role of sphingosine and ceramide kinases in inflammatory responses.	Immunol Lett	2005	15585321	Unknown mechanism - may or may not be energy dependent. However these metabolites must be able to be transported intracellularly and exported outside of the cell. NJ
CRNt	3	Verhaagh S, Schweifer N, Barlow DP, Zwart R.	Cloning of the mouse and human solute carrier 22a3 (Slc22a3/SLC22A3) identifies a conserved cluster of three organic cation transporters on mouse chromosome 17 and human 6q26-q27.	Genomics	1999	9933568	specificity: skeletal muscle, kidney, prostate, lung, pancreas, heart, small intestine, adrenal gland, thyroid gland, liver - Koepsell 2004 paper - PMID: 12883891 Km and Ki listed in Koepsell 2003 (in addition to detailed info about localization and other members of organic cation transporters) PMID: 12827517 Polyspecific organic cation transporters in the liver, kidney, intestine, and other organs are critical for elimination of many endogenous small organic cations as well as a wide array of drugs and environmental toxins. The encoded protein is a plasma integral membrane protein which functions both as an organic cation transporter and as a sodium-dependent high affinity carnitine transporter. The encoded protein is involved in the active cellular uptake of carnitine. Mutations in this gene are the cause of systemic primary carnitine deficiency (CDSP), an autosomal recessive disorder manifested early in life by hypoketotic hypoglycemia and acute metabolic decompensation, and later in life by skeletal myopathy or cardiomyopathy. NJ

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
CRNt	3	Koepsell H, Schmitt BM, Gorboulev V.	Organic cation transporters.	Rev Physiol Biochem Pharmacol	2003	12827517	specificity: skeletal muscle, kidney, prostate, lung, pancreas, heart, small intestine, adrenal gland, thyroid gland, liver - Koepsell 2004 paper - PMID: 12883891 Km and Ki listed in Koepsell 2003 (in addition to detailed info about localization and other members of organic cation transporters) PMID: 12827517 Polyspecific organic cation transporters in the liver, kidney, intestine, and other organs are critical for elimination of many endogenous small organic cations as well as a wide array of drugs and environmental toxins. The encoded protein is a plasma integral membrane protein which functions both as an organic cation transporter and as a sodium-dependent high affinity carnitine transporter. The encoded protein is involved in the active cellular uptake of carnitine. Mutations in this gene are the cause of systemic primary carnitine deficiency (CDSP), an autosomal recessive disorder manifested early in life by hypoketotic hypoglycemia and acute metabolic decompensation, and later in life by skeletal myopathy or cardiomyopathy. NJ
CRNt	3	Koepsell H, Endou H	The SLC22 drug transporter family	Pflugers Arch	2004	12883891	specificity: skeletal muscle, kidney, prostate, lung, pancreas, heart, small intestine, adrenal gland, thyroid gland, liver - Koepsell 2004 paper - PMID: 12883891 Km and Ki listed in Koepsell 2003 (in addition to detailed info about localization and other members of organic cation transporters) PMID: 12827517 Polyspecific organic cation transporters in the liver, kidney, intestine, and other organs are critical for elimination of many endogenous small organic cations as well as a wide array of drugs and environmental toxins. The encoded protein is a plasma integral membrane protein which functions both as an organic cation transporter and as a sodium-dependent high affinity carnitine transporter. The encoded protein is involved in the active cellular uptake of carnitine. Mutations in this gene are the cause of systemic primary carnitine deficiency (CDSP), an autosomal recessive disorder manifested early in life by hypoketotic hypoglycemia and acute metabolic decompensation, and later in life by skeletal myopathy or cardiomyopathy. NJ
CRNtx	2	Mukherji M, Schofield CJ, Wierzbicki AS, Jansen GA, Wanders RJ, Lloyd MD.	The chemical biology of branched-chain lipid metabolism.	Prog Lipid Res	2003	12814641	Transport of branched chain derivatives from peroxisome to mitochondria. See Mukherji and Wanders for more details. NJ
CSNAT2x	3	Wanders RJ.	Peroxisomes, lipid metabolism, and peroxisomal disorders.	Mol Genet Metab	2004	15464416	For transport of branched fatty acids (initial alpha and beta ox in peroxisome -> partial continuation in mit). See Wanders PMID: 15464416 for further details. -- REVISED GPR, dmnon has specific crn transferase gene and protein. see PMID: 10486279 and PMID: 9469587. Carnitine acetyltransferase (CRAT) is a key enzyme in the metabolic pathway in mitochondria, peroxisomes and endoplasmic reticulum. CRAT catalyzes the reversible transfer of acyl groups from an acyl-CoA thioester to carnitine and regulates the ratio of acylCoA/CoA in the subcellular compartments. Different subcellular localizations of the CRAT mRNAs are thought to result from alternative splicing of the CRAT gene suggested by the divergent sequences in the 5' region of peroxisomal and mitochondrial CRAT cDNAs and the location of an intron where the sequences diverge. Cloning in PMID: 7829107 NJ
CSNATm	3	Lysiak W, Toth PP, Suelter CH, Bieber LL.	Quantitation of the efflux of acylcarnitines from rat heart, brain, and liver mitochondria.	J Biol Chem	1986	3759988	- Additional information added by RS/TV: - CRAT is located in the mitochondria, endoplasmic reticulum, and Kinetic Information: Lysiak W, Toth PP, Suelter CH, Bieber LL. J Biol Chem. 1986 Oct 15;261(29):13698-703 Quantitation of the efflux of acylcarnitines from rat heart, brain, and liver mitochondria Poirier M, Vincent G, Reszko AE, Bouchard B, Kelleher JK, Brunengraber H, Des Rosiers C. Am J Physiol Heart Circ Physiol. 2002 Oct;283(4):H1379-86; Probing the link between citrate and malonyl-CoA in perfused rat hearts
CSNATr	3	Corti O, Finocchiaro G, Rossi E, Zuffardi O, DiDonato S.	Molecular cloning of cDNAs encoding human carnitine acetyltransferase and mapping of the corresponding gene to chromosome 9q34.1.	Genomics	1994	7829107	for reversible cytosolic interconversion between acrn and accoa (for part of peroxisomal FA degradation). see PMID: 11257506 NJ

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CSNATr	3	Ramsay, Gandour, van der Leij	Molecular enzymology of carnitine transfer and transport	Biochim Biophys Acta	2001	11257506	for reversible cytosolic interconversion between acrn and accoa (for part of peroxisomal FA degradation). see PMID: 11257506 NJ
CTPS1	2	Takahashi E, Yamauchi M, Tsuji H, Hitomi A, Meuth M, Hori T.	Chromosome mapping of the human cytidine-5'-triphosphate synthetase (CTPS) gene to band 1p34.1-p34.3 by fluorescence in situ hybridization.	Hum Genet	1991	1959918	IT
CTPS2	3	van Kuilenburg AB, Meinsma R, Vreken P, Waterham HR, van Gennip AH.	Identification of a cDNA encoding an isoform of human CTP synthetase.	Biochim Biophys Acta	2000	10899599	Entrez genes said that reactions is with glutamine as well as orten IT
CYANSTm	3	Horowitz PM, Butler M, McClure GD Jr.	Reducing sugars can induce the oxidative inactivation of rhodanese.		1992	1429701	Entrez Gene - The product of this gene is a mitochondrial matrix enzyme that is encoded by the nucleus. It may play roles in cyanide detoxification, the formation of iron-sulfur proteins, and the modification of sulfur-containing enzymes. The gene product contains two highly conservative domains (rhodanese homology domains), suggesting these domains have a common evolutionary origin. MM
CYANSTm	3	Aita N, Ishii K, Akamatsu Y, Ogasawara Y, Tanabe S.	Cloning and expression of human liver rhodanese cDNA.		1997	9070219	Entrez Gene - The product of this gene is a mitochondrial matrix enzyme that is encoded by the nucleus. It may play roles in cyanide detoxification, the formation of iron-sulfur proteins, and the modification of sulfur-containing enzymes. The gene product contains two highly conservative domains (rhodanese homology domains), suggesting these domains have a common evolutionary origin. MM
CYSGLTH	3	Oshima RG, Rhead WJ, Thoene JG, Schneider JA.	Cystine metabolism in human fibroblasts. Comparison of normal, cystinotic, and gamma-glutamylcysteine synethetase-deficient cells.		1976	932033	- enzyme has been identified, but not gene - reaction known to physiologically exist in order for cystine to be converted to cysteine (see references) MM
CYSO	3	Millard J, Parsons RB, Waring RH, Williams AC, Ramsden DB	Expression of cysteine dioxygenase (EC 1.13.11.20) and sulfite oxidase in the human lung: a potential role for sulfate production in the protection from airborne xenobiotica	Mol Pathol	2003	14514920	Reaction and gene are characterized.
CYSTA	2	Akagi R.	Purification and characterization of cysteine aminotransferase from rat liver cytosol.		1982	7113743	- reaction required in cysteine metabolism according to Stipanuk (2004) - aspartate aminotransferase gene associated based on KEGG and rat evidence indicating its activity is similar to cysteine aminotransferase MM
CYSTAm	2	Ubuka T, Umemura S, Yuasa S, Kinuta M, Watanabe K.	Purification and characterization of mitochondrial cysteine aminotransferase from rat liver.		1978	754189	- reaction required in cysteine metabolism according to Stipanuk (2004) - aspartate aminotransferase gene associated based on KEGG and rat evidence indicating its activity is similar to cysteine aminotransferase
CYSTGL	3	Levonen AL, Lapatto R, Saksela M, Raivio KO.	Human cystathionine gamma-lyase: developmental and in vitro expression of two isoforms.		2000	10727430	0
CYSTGL	3	Stipanuk MH	Sulfur amino acid metabolism: pathways for production and removal of homocysteine and cysteine	Annu Rev Nutr	2004	15189131	0
CYSTS	3	Kraus J, Packman S, Fowler B, Rosenberg LE.	Purification and properties of cystathionine beta-synthase from human liver. Evidence for identical subunits.		1978	681363	Entrez Gene - The protein encoded by this gene is involved in the transsulfuration pathway. The first step of this pathway, from homocysteine to cystathionine, is catalyzed by this protein. CBS deficiency can cause homocystinuria which affects many organs and tissues, including the eyes and the skeletal, vascular and central nervous systems. -in the adult strongly expressed in liver and pancreas, some expression in heart and brain, weak expression in lung and kidney. in the fetus, expressed in brain, liver and kidney. -MM
CYSTS	3	Kraus JP, Oliveriusova J, Sokolova J, Kraus E, Vlcek C, de Franchis R	The human cystathionine beta-synthase (CBS) gene: complete sequence, alternative splicing, and polymorphisms.		1998	9790750	Entrez Gene - The protein encoded by this gene is involved in the transsulfuration pathway. The first step of this pathway, from homocysteine to cystathionine, is catalyzed by this protein. CBS deficiency can cause homocystinuria which affects many organs and tissues, including the eyes and the skeletal, vascular and central nervous systems. -in the adult strongly expressed in liver and pancreas, some expression in heart and brain, weak expression in lung and kidney. in the fetus, expressed in brain, liver and kidney. -MM

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CYTD	2	Laliberte J, Momparler RL.	Human cytidine deaminase: purification of enzyme, cloning, and expression of its complementary DNA.	Cancer Res	1994	7923172	IT mRNA found n myoblase and leukemia cell line, bone marrow cells, monocytes, macrophages I did find any report of measurement of enzyme activity AICDA(57379):Activation-induced cysodine deaminase (AID) is a cytosine deaminase that is critical to immunoglobulin hypermutation, class switch recombination, and gene conversion.In the context of hypermutating B cells, AID deaminates cytosine in the DNA of immunoglobulin genes, leading to the accumulation of mutations in the variable regions. --> most enzyme is in cytoplasm but can also be found in nucleus.
CYTD	2	Saccone S, Besati C, Andreozzi L, Della Valle G, Garattini E, Terao M.	Assignment of the human cytidine deaminase (CDA) gene to chromosome 1 band p35-p36.2.	Genomics	1994	8001985	IT mRNA found n myoblase and leukemia cell line, bone marrow cells, monocytes, macrophages I did find any report of measurement of enzyme activity AICDA(57379):Activation-induced cysodine deaminase (AID) is a cytosine deaminase that is critical to immunoglobulin hypermutation, class switch recombination, and gene conversion.In the context of hypermutating B cells, AID deaminates cytosine in the DNA of immunoglobulin genes, leading to the accumulation of mutations in the variable regions. --> most enzyme is in cytoplasm but can also be found in nucleus.
CYTD	2	Kuhn K, Bertling WM, Emmrich F.	Cloning of a functional cDNA for human cytidine deaminase (CDD) and its use as a marker of monocyte/macrophage differentiation.	Biochem Biophys Res Commun	1993	8422236	IT mRNA found n myoblase and leukemia cell line, bone marrow cells, monocytes, macrophages I did find any report of measurement of enzyme activity AICDA(57379):Activation-induced cysodine deaminase (AID) is a cytosine deaminase that is critical to immunoglobulin hypermutation, class switch recombination, and gene conversion.In the context of hypermutating B cells, AID deaminates cytosine in the DNA of immunoglobulin genes, leading to the accumulation of mutations in the variable regions. --> most enzyme is in cytoplasm but can also be found in nucleus.
CYTD	2	Ge Y, Jensen TL, Stout ML, Flatley RM, Grohar PJ, Ravindranath Y, Matherly LH, Taub JW.	The role of cytidine deaminase and GATA1 mutations in the increased cytosine arabinoside sensitivity of Down syndrome myeloblasts and leukemia cell lines.	Cancer Res	2004	14744791	IT mRNA found n myoblase and leukemia cell line, bone marrow cells, monocytes, macrophages I did find any report of measurement of enzyme activity AICDA(57379):Activation-induced cysodine deaminase (AID) is a cytosine deaminase that is critical to immunoglobulin hypermutation, class switch recombination, and gene conversion.In the context of hypermutating B cells, AID deaminates cytosine in the DNA of immunoglobulin genes, leading to the accumulation of mutations in the variable regions. --> most enzyme is in cytoplasm but can also be found in nucleus.
CYTD	2	Brar SS, Watson M, Diaz M.	Activation-induced cytosine deaminase (AID) is actively exported out of the nucleus but retained by the induction of DNA breaks.	J Biol Chem	2004	15087440	IT mRNA found n myoblase and leukemia cell line, bone marrow cells, monocytes, macrophages I did find any report of measurement of enzyme activity AICDA(57379):Activation-induced cysodine deaminase (AID) is a cytosine deaminase that is critical to immunoglobulin hypermutation, class switch recombination, and gene conversion.In the context of hypermutating B cells, AID deaminates cytosine in the DNA of immunoglobulin genes, leading to the accumulation of mutations in the variable regions. --> most enzyme is in cytoplasm but can also be found in nucleus.
CYTDt4	3	Gray JH, Owen RP, Giacomini KM.	The concentrative nucleoside transporter family, SLC28.		2004	12856181	IT Tissue distribution: SLC28A1 (CNT1): Liver, kidney, small intestine, (epitheloa apical membrane) SLC28A2 (CNT2): kidney (apical membrane), liver, heart, brain, plcaenta, pancreas, skeletal muscle, colon, rectum, small intestine SLC28A3 (CNT3): pancreas, trachea, bone marrow and mammary gland, intestine, lung, placenta, prostrate, testis, liver
CYTK1	3	Van Rompay AR, Johansson M, Karlsson A.	Phosphorylation of deoxycytidine analog monophosphates by UMP-CMP kinase: molecular characterization of the human enzyme.	Mol Pharmacol	1999	10462544	IT
CYTK1	3	Liou JY, Dutschman GE, Lam W, Jiang Z, Cheng YC.	Characterization of human UMP/CMP kinase and its phosphorylation of D- and L-form deoxycytidine analogue monophosphates.	Cancer Res	2002	11912132	IT

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D3AIBTm	3	Kakimoto Y, Taniguchi K, Sano I.	D-beta-aminoisobutyrate:pyruvate aminotransferase in mammalian liver and excretion of beta-aminoisobutyrate by man.		1969	5773299	OMIM: Human urine contains R-BAIB almost exclusively (and it is this form that is excreted in excess in the hyper-BAIB trait), whereas the plasma pool is about 80% S-BAIB. As reviewed by Scriver and Perry (1989), the 'defect' is an impairment of R-BAIB catabolism due to deficient activity of a pyruvate-requiring transaminase, D-beta-aminoisobutyrate:pyruvate aminotransferase MM
DADNK	3	Hurley MC, Palella TD, Fox IH.	Human placental deoxyadenosine and deoxyguanosine phosphorylating activity.	J Biol Chem	1983	6317685	reaction was included during gap filling. i could not identify a gene for this reaction, however, its activity has been observed/studied in/from human placenta cells IT
DAGK_hs	3	Tang W, Bunting M, Zimmerman GA, McIntyre TM, Prescott SM	Molecular cloning of a novel human diacylglycerol kinase highly selective for arachidonate-containing substrates	Journal of Biological Chemistry	1996		cytoplasmic and nuclear - uniport many different genes, some have different isoforms, some are ubiquitous and others are tissue specific - dependent on substrate (different gene products have different specificities different fatty acid chains. NJ
DARGOp	3	D'Aniello A, Vetere A, Petrucelli L	Further study on the specificity of D-amino acid oxidase and D-aspartate oxidase and time course for complete oxidation of D-amino acids	Comp Biochem Physiol B	1993	8103425	Slow rate according to second citation.
DARGOp	3	Vanoni MA, Cosma A, Mazzeo D, Mattevi A, Todone F, Curti B	Limited proteolysis and X-ray crystallography reveal the origin of substrate specificity and of the rate-limiting product release during oxidation of D-amino acids catalyzed by mammalian D-amino acid oxidase	Biochemistry	1997	9153402	Slow rate according to second citation.
DASCBR	3	Lundberg M, Johansson C, Chandra J, Enoksson M, Jacobsson G, Ljung J, Johansson M, Holmgren A.	Cloning and expression of a novel human glutaredoxin (Grx2) with mitochondrial and nuclear isoforms	J Biol Chem	2001	11297543	TV (6/1/2005) Grx1 and Grx2 appear to primarily act on cys residue of proteins, but Lundberg also found that they can reduce dehydroascobic acid. It's not clear what the enzyme uses as reducing agent.
DASPO1p	3	Amery L, Brees C, Baes M, Setoyama C, Miura R, Mannaerts GP, Van Veldhoven PP.	C-terminal tripeptide Ser-Asn-Leu (SNL) of human D-aspartate oxidase is a functional peroxisome-targeting signal.		1998	9820813	Entrez Gene - The protein encoded by this gene is a peroxisomal flavoprotein that catalyzes the oxidative deamination of D-aspartate and N-methyl D-aspartate. Flavin adenine dinucleotide or 6-hydroxyflavin adenine dinucleotide can serve as the cofactor in this reaction. Two transcript variants encoding different isoforms have been found for this gene. **did not include deamination of N-methyl D-aspartate. Could not find another reaction that either transports or synthesizes it. Found references stating that there is a receptor for this compound, but not metabolism MM
DASPO1p	3	Zaar K, Kost HP, Schad A, Volkl A, Baumgart E, Fahimi HD.	Cellular and subcellular distribution of D-aspartate oxidase in human and rat brain.		2002	12209855	Entrez Gene - The protein encoded by this gene is a peroxisomal flavoprotein that catalyzes the oxidative deamination of D-aspartate and N-methyl D-aspartate. Flavin adenine dinucleotide or 6-hydroxyflavin adenine dinucleotide can serve as the cofactor in this reaction. Two transcript variants encoding different isoforms have been found for this gene. **did not include deamination of N-methyl D-aspartate. Could not find another reaction that either transports or synthesizes it. Found references stating that there is a receptor for this compound, but not metabolism MM
DCIm	2	Partanen ST, Novikov DK, Popov AN, Mursula AM, Hiltunen JK, Wierenga RK.	The 1.3 A crystal structure of human mitochondrial Delta3-Delta2-enoyl-CoA isomerase shows a novel mode of binding for the fatty acyl group.	J Mol Biol	2004	15351645	mitochondrial matrix: Uniprot (RefSeq) Function: Partanen et al (PMID: 15351645) This gene encodes a member of the hydratase/isomerase superfamily. The protein encoded is a key mitochondrial enzyme involved in beta-oxidation of unsaturated fatty acids. It catalyzes the transformation of 3-cis and 3-trans-enoyl-CoA esters arising during the stepwise degradation of cis-, mono-, and polyunsaturated fatty acids to the 2-trans-enoyl-CoA intermediates. Alternatively spliced transcript variants have been described, but their full-length nature has not been determined. NJ
DCK1m	3	Cheng YC, Domin B, Lee LS.	Human deoxycytidine kinase. Purification and characterization of the cytoplasmic and mitochondrial isozymes derived from blast cells of acute myelocytic leukemia patients.	Biochim Biophys Acta	1977	265735	reaction was included during gap filling process. could not find gene for mitochondrial recation (1633 is only responsible for nuclear protein based on gene cards). I could only get abstract of reference - but since they used cells from leukemia patient it should be nicely human data mitochondrial version acts on deoxycytidine and deoxythymidine. only with ATP highest activity observed. IT

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DCK1n	3	Turk B, Awad R, Usova EV, Bjork I, Eriksson S.	A pre-steady-state kinetic analysis of substrate binding to human recombinant deoxycytidine kinase: a model for nucleoside kinase action.	Biochemistry	1999	10387103	seems also react with dADO but much slower --> rxns not included yet IT
DCMPDA	2	Weiner KX, Ciesla J, Jaffe AB, Ketring R, Maley F, Maley GF.	Chromosomal location and structural organization of the human deoxycytidylate deaminase gene.	J Biol Chem	1995	7642519	IT
DCMPDA	2	Weiner KX, Weiner RS, Maley F, Maley GF.	Primary structure of human deoxycytidylate deaminase and overexpression of its functional protein in Escherichia coli.	J Biol Chem	1993	7685356	IT
DCT	3	Yokoyama K, Suzuki H, Yasumoto K, Tomita Y, Shibahara S	Molecular cloning and functional analysis of a cDNA coding for human DOPAchrome tautomerase/tyrosinase-related protein-2	Biochim Biophys Acta	1994	8148378	Textbook reaction.
DDPGAm	2	MAITRA U, DEKKER EE	PURIFICATION AND PROPERTIES OF RAT LIVER 2-KETO-4-HYDROXYGLUTARATE ALDOLASE	J Biol Chem	1964	14193832	Reaction noted in citation, but no gene information could be found. - 4-hydroxy-2-ketoglutarate is cleaved by the mitochondrial enzyme 4-hydroxy-2-ketoglutarate aldolase to produce pyruvate and glyoxylate [Holmes 1998] - in rat, this enzyme is found in liver and kidney [Maitra 1964] - isolated mouse liver mitochondria produced glyoxylate from hydroxyproline [Knight 2005]
DESAT16_2	3	Wang Y, Kurdi-Haidar B, Oram JF.	LXR-mediated activation of macrophage stearoyl-CoA desaturase generates unsaturated fatty acids that destabilize ABCA1.	J Lipid Res	2004	14967823	ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). Acts on 16C and 18C see PMID: 14967823 (Wang Y, Kurdi-Haidar B, Oram JF.) Electron carrier is NAD as per texts (future articles in primary literature may provide more specifics. NJ
DESAT16_2	3	Leonard AE, Bobik EG, Dorado J, Kroeger PE, Chuang LT, Thurmond JM, Parker-Barnes JM, Das T, Huang YS, Mukerji P	Cloning of a human cDNA encoding a novel enzyme involved in the elongation of long-chain polyunsaturated fatty acids	Biochem J	2000		ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). Acts on 16C and 18C see PMID: 14967823 (Wang Y, Kurdi-Haidar B, Oram JF.) Electron carrier is NAD as per texts (future articles in primary literature may provide more specifics. NJ
DESAT16_2	3	Pereira SL, Leonard AE, Mukerji P	Recent advances in the study of fatty acid desaturases from animals and lower eukaryotes	Prostaglandins Leukotrienes and Essential Fatty Acids	2003		ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). Acts on 16C and 18C see PMID: 14967823 (Wang Y, Kurdi-Haidar B, Oram JF.) Electron carrier is NAD as per texts (future articles in primary literature may provide more specifics. NJ
DESAT16_2	3	Leonard AE, Bobik EG, Dorado J, Kroeger PE, Chuang LT, Thurmond JM, Parker-Barnes JM, Das T, Huang YS, Mukerji P	Cloning of a human cDNA encoding a novel enzyme involved in the elongation of long-chain polyunsaturated fatty acids	Biochem J	2000		ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). Acts on 16C and 18C see PMID: 14967823 (Wang Y, Kurdi-Haidar B, Oram JF.) Electron carrier is NAD as per texts (future articles in primary literature may provide more specifics. NJ
DESAT16_2	3	Horrobin DF	Omega-6 Essential Fatty Acids		1990		ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). Acts on 16C and 18C see PMID: 14967823 (Wang Y, Kurdi-Haidar B, Oram JF.) Electron carrier is NAD as per texts (future articles in primary literature may provide more specifics. NJ
DESAT16_2	3	Pereira SL, Leonard AE, Mukerji P	Recent advances in the study of fatty acid desaturases from animals and lower eukaryotes	Prostaglandins Leukotrienes and Essential Fatty Acids	2003		ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). Acts on 16C and 18C see PMID: 14967823 (Wang Y, Kurdi-Haidar B, Oram JF.) Electron carrier is NAD as per texts (future articles in primary literature may provide more specifics. NJ

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DESAT16_2	3	Zhang L, Ge L, Parimoo S, Stenn K, Prouty SM	Human stearoyl-CoA desaturase: alternative transcripts generated from a single gene by usage of tandem polyadenylation sites	Biochem J	1999		ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). Acts on 16C and 18C see PMID: 14967823 (Wang Y, Kurdi-Haidar B, Oram JF.) Electron carrier is NAD as per texts (future articles in primary literature may provide more specifics. NJ
DESAT16_2	3	Leonard AE, Bobik EG, Dorado J, Kroeger PE, Chuang LT, Thurmond JM, Parker-Barnes JM, Das T, Huang YS, Mukerji P	Cloning of a human cDNA encoding a novel enzyme involved in the elongation of long-chain polyunsaturated fatty acids	Biochem J	2000		ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). Acts on 16C and 18C see PMID: 14967823 (Wang Y, Kurdi-Haidar B, Oram JF.) Electron carrier is NAD as per texts (future articles in primary literature may provide more specifics. NJ
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DESAT18_6	3	Cho HP, Nakamura MT, Clarke SD	Cloning, expression, and nutritional regulation of the mammalian delta-6 desaturase	The Journal of Biological Chemistry	1999		ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). NJ
DESAT20_1	3	Cho HP, Nakamura M, Clarke SD	Cloning, expression, and fatty acid regulation of the human delta-5 desaturase	The Journal of Biological Chemistry	1999		ER localization: outer membrane. Not well characterized in literature (poor data to specifiy inner vs outer membrane). NJ
DESAT22_1p	3	Sprecher H	An update on the pathways of polyunsaturated fatty acid metabolism	Curr Opn in Clinical Nutrition and Metabolic Care	1999		no gene identified specifically yet for delta 4 desaturase activity peroxisomal localization (inner side) - see mx refs: Sprecher "An update ... pathways of polyunsaturated fatty acid metabolism" (PMID: 8847474) adrn and dcsptn1 are n-6 fatty acids NJ
DESAT22_1p	3	Ferdinandusse S, Denis S, Mooijer PAW, Zhang Z, Reddy JK, Spector AA, Wanders RJA	Identification of the peroxisomal beta-oxidation enzymes involved in the biosynthesis of docosahexaenoic acid	Journal of Lipid Research	2001		no gene identified specifically yet for delta 4 desaturase activity peroxisomal localization (inner side) - see mx refs: Sprecher "An update ... pathways of polyunsaturated fatty acid metabolism" (PMID: 8847474) adrn and dcsptn1 are n-6 fatty acids NJ
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DGNSKm	3	Johansson M, Karlsson A.	Cloning and expression of human deoxyguanosine kinase cDNA.	Proc Natl Acad Sci U S A	1996	8692979	IT

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DGSNtm	2	Watkins LF, Lewis RA.	The metabolism of deoxyguanosine in mitochondria. Characterization of the uptake process.	Mol Cell Biochem	1987	3696164	transport has been characterized in rat but not in human. however, has to take place since deoxynucleoside precursor have to be transported in mito for mt DNA synthesis IT
DHAAt1r	3	Wilson JX	Regulation of Vitamin C Transport	Annu Rev Nutr	2004	15705056	see [Wilson 2004], [Ball 2004] for comprehensive review of DHAA transport 6513: - major substrates are Glc, Gal, Man, GlcN [Uldry, 2004], [Uldry, 2002], DHAA [Ball 2004] - facilitated diffusion [Uldry, 2004] - primarily expressed in erythrocytes, brain, blood brain barrier, blood-tissue barrier [Uldry, 2004], [Maher, 1994] - cloned, cDNA isolated [Mueckler, 1985] - not expressed in normal hepatocytes, but induced during cancer [Flier, 1987] 6515: - major substrates are Glc (high affinity), Gal, Man, Xyl, maltose, dehydroascorbic acid [Uldry, Pflugers Arch 2004] - protein localized to brain (neurons), testis (spermatozoa) [Haber, Endocrinology 1993], [Uldry, Pflugers Arch 2004], sk muscle (slow twitch fibers) [Stuart, Metabolism 1999], platelets (alpha-granules) [Heijen, J Cell Biol 1997] - cDNA was cloned [Kayano, J Biol Chem 1988] 6517: - transports Glc, GlcN, dehydroascorbic acid [Uldry, Pflugers Arch 2004] - protein localized to adipose tissue (brown and white), sk and cardiac muscle [Uldry, Pflugers Arch 2004] - insulin binding, exercise stimulate translocation of GLUT4 ve - cDNA was cloned [Fukumoto, J Biol Chem 1989]
DHAPA	3	Thai TP, Heid H, Rackwitz HR, Hunziker A, Gorgas K, Just WW.	Ether lipid biosynthesis: isolation and molecular characterization of human dihydroxyacetonephosphate acyltransferase.	FEBS Lett	1997	9459311	mitochondrial and ER localization. ER localization on outer membrane -> effectively cytosolic. NJ
DHCR71r	3	Moebius FF, Fitzky BU, Lee, JN, Paik YK, Glossmann H	Molecular cloning and expression of the human delta-7-sterol reductase	PNAS	1998		ER - specificity: Most abundant in adrenal gland, liver, testis, and brain. NJ
DHCR72r	3	Correa-Cerro LS, Porter FD.	3beta-hydroxysterol Delta7-reductase and the Smith-Lemli-Opitz syndrome.	Mol Genet Metab	2005	15670717	ER - specificity: Most abundant in adrenal gland, liver, testis, and brain. Moebius ref (seq and cloning) see PMID: 9465114 Marijanovic ref PMID: 12829805 see also PMID: 15670717 for review NJ
DHCRD1	3	Cadena DL, Kurten RC, Gill GN.	The product of the MLD gene is a member of the membrane fatty acid desaturase family: overexpression of MLD inhibits EGF receptor biosynthesis.	Biochemistry	1997	9188692	cytoplasmic (assumed cyt side of ER) - uniprot Varki supports cytoplasmic side of ER or Golgi - chpa 9, p116 NJ
DHDPBMTm	3	Jonassen T, Clarke CF.	Isolation and functional expression of human COQ3, a gene encoding a methyltransferase required for ubiquinone biosynthesis.	J Biol Chem	2000	10777520	IT
DHEASULT	3	Otterness DM, Wieben ED, Wood TC, Watson WG, Madden BJ, McCormick DJ, Weinshilboum RM.	Human liver dehydroepiandrosterone sulfotransferase: molecular cloning and expression of cDNA.	Mol Pharmacol	1992	1588921	cytosolic - uniport and refs TISSUE SPECIFICITY: Liver, adrenal and at lower level in the kidney. Is present in human fetus in higher level in the adrenal than the liver and the kidney. Estrogens present in maternal circulation is predominantly derived from fetal dehydroepiandosterone sulfate which is hydrolyzed and metabolized to estrogens in placenta. Sulfotransferase enzymes catalyze the sulfate conjugation of many hormones, neurotransmitters, drugs, and xenobiotic compounds. These cytosolic enzymes are different in their tissue distributions and substrate specificities. The gene structure (number and length of exons) is similar among family members. This gene is primarily expressed in liver and adrenal tissues where the encoded protein sulfates steroids and bile acids. - It is also known that Sult1e1 catalyzes this reaction according to Table 2 in Glatt H, et al. Mutat Res. 2001 Oct 1;482(1-2):27-40. Review. (RS/TV) NJ

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DHEASULT	3	Glatt H, Boeing H, Engelke CE, Ma L, Kuhlow A, Pabel U, Pomplun D, Teubner W, Meinl W.	Human cytosolic sulphotransferases: genetics, characteristics, toxicological aspects.	Mutat Res	2001	11535246	cytosolic - uniport and refs TISSUE SPECIFICITY: Liver, adrenal and at lower level in the kidney. Is present in human fetus in higher level in the adrenal than the liver and the kidney. Estrogens present in maternal circulation is predominantly derived from fetal dehydroepiandosterone sulfate which is hydrolyzed and metabolized to estrogens in placenta. Sulfotransferase enzymes catalyze the sulfate conjugation of many hormones, neurotransmitters, drugs, and xenobiotic compounds. These cytosolic enzymes are different in their tissue distributions and substrate specificities. The gene structure (number and length of exons) is similar among family members. This gene is primarily expressed in liver and adrenal tissues where the encoded protein sulfates steroids and bile acids. - It is also known that Sult1e1 catalyzes this reaction according to Table 2 in Glatt H, et al. Mutat Res. 2001 Oct 1;482(1-2):27-40. Review. (RS/TV) NJ
DHFR	2	Funanage VL, Myoda TT, Moses PA, Cowell HR.	Assignment of the human dihydrofolate reductase gene to the q11----q22 region of chromosome 5.	Mol Cell Biol	1984	6504041	IT cyto? Ball: p 359
DHFR	2	Ball, G.F.M	Vitamins: Their Role in the Human Body		2004		IT cyto? Ball: p 359
DHORD9	3	Bader B, Knecht W, Fries M, Loffler M.	Expression, purification, and characterization of histidine-tagged rat and human flavoenzyme dihydroorotate dehydrogenase.	Protein Expr Purif	1998	9693067	01-27-05 IT based on Bader et al, 1998, Prot. Expr. Purif.,, 13,414-422 (I guess it is ubiquinone but i am not sure) (needs FMN as cofactor) v(enzyme is located in inner-membrane of mitochondria, reaction take place in cytosol, Fig. 8 Rawls)
DHORD9	3	Rawls J, Knecht W, Diekert K, Lill R, Loffler M.	Requirements for the mitochondrial import and localization of dihydroorotate dehydrogenase.	Eur J Biochem	2005	10727948	01-27-05 IT based on Bader et al, 1998, Prot. Expr. Purif.,, 13,414-422 (I guess it is ubiquinone but i am not sure) (needs FMN as cofactor) v(enzyme is located in inner-membrane of mitochondria, reaction take place in cytosol, Fig. 8 Rawls)
DHORD9	3	Loffler M, Jockel J, Schuster G, Becker C.	Dihydroorotat-ubiquinone oxidoreductase links mitochondria in the biosynthesis of pyrimidine nucleotides.	Mol Cell Biochem	1997		01-27-05 IT based on Bader et al, 1998, Prot. Expr. Purif.,, 13,414-422 (I guess it is ubiquinone but i am not sure) (needs FMN as cofactor) v(enzyme is located in inner-membrane of mitochondria, reaction take place in cytosol, Fig. 8 Rawls)
DHPM1	3	Hamajima N, Matsuda K, Sakata S, Tamaki N, Sasaki M, Nonaka M.	A novel gene family defined by human dihydropyrimidinase and three related proteins with differential tissue distribution.	Gene	1996	8973361	IT
DMGDHm	3	Binzak BA, Wevers RA, Moolenaar SH	Cloning of dimethylglycine dehydrogenase and a new human inborn error of metabolism, dimethylglycine dehydrogenase deficiency.		2001	1123190	irreversibility according to cited paper Entrez Gene -This gene encodes an enzyme involved in the catabolism of choline, catalyzing the oxidative demethylation of dimethylglycine to form sarcosine. The enzyme is found as a monomer in the mitochondrial matrix, and uses flavin adenine dinucleotide and folate as cofactors. Mutation in this gene causes dimethylglycine dehydrogenase deficiency, characterized by a fishlike body odor, chronic muscle fatigue, and elevated levels of the muscle form of creatine kinase in serum. MM
DMGDHm	3	Binzak,B.A. , Vockley,J.G. , Jenkins,R.B. , Vockley,J.	Structure and analysis of the human dimethylglycine dehydrogenase gene.		2000	10767172	irreversibility according to cited paper Entrez Gene -This gene encodes an enzyme involved in the catabolism of choline, catalyzing the oxidative demethylation of dimethylglycine to form sarcosine. The enzyme is found as a monomer in the mitochondrial matrix, and uses flavin adenine dinucleotide and folate as cofactors. Mutation in this gene causes dimethylglycine dehydrogenase deficiency, characterized by a fishlike body odor, chronic muscle fatigue, and elevated levels of the muscle form of creatine kinase in serum. MM
DMHPTCRNte	2	Libert R, Van Hoof F, Thillaye-M, Vincent MF, Nassogne MC, Stroobant V, de Hoffmann E, Schanck A.	Identification of new medium-chain acylcarnitines present in normal human urine	Anal Biochem	1997	9299016	See PMID 9299016. dmhptcrn found in urine. Metabolic fate of corresponding coa (dmhptcoa) is unknown - see PMID 9469587 NJ
DNADDP	2	Funakoshi I, Kato H, Horie K, Yano T, Hori Y, Kobayashi H, Inoue T, Suzuki H, Fukui S, Tsukahara M, et al.	Molecular cloning of cDNAs for human fibroblast nucleotide pyrophosphatase.	Arch Biochem Biophys	1992	1315502	homodimer; membrane protein; broad specificity; in general: a dinucleotide +h2o--> 2 mononucleotides IT
DNAMTn	3	Yen RW, Vertino PM, Nelkin BD, Yu JJ, el-Deiry W, Cumaraswamy A, Lennon GG, Trask BJ, Celano P, Baylin SB.	Isolation and characterization of the cDNA encoding human DNA methyltransferase.		1992	1594447	DNA methylation patterns (cytosine-specific)

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DNAMTn	3	Deloukas,P. , Matthews,L.H. , Ashurst,J.	The DNA sequence and comparative analysis of human chromosome 20.		2001		DNA methylation patterns (cytosine-specific)
DNDPt1m	3	Dolce V, Fiermonte G, Runswick MJ, Palmieri F, Walker JE.	The human mitochondrial deoxynucleotide carrier and its role in the toxicity of nucleoside antivirals.	Proc Natl Acad Sci U S A	2001	11226231	IT
DOGULNO1	2	Simpson GL, Ortwerth BJ	The non-oxidative degradation of ascorbic acid at physiological conditions	Biochim Biophys Acta	2000	10727845	- rxn shown in Fig 2 of [Banhegyi, Fee Radic Mol Biol 1997] - rxn described in [Simpson and Ortwerth, 2000]
DOLASNT_Uer	0	Nakashima T, Sekiguchi T, Kuraoka A, Fukushima K, Shibata Y, Komiyama S, Nishimoto T.	Molecular cloning of a human cDNA encoding a novel protein, DAD1, whose defect causes apoptotic cell death in hamster BHK21 cells.	Mol Cell Biol	1993	8413235	Dad1p ubiquitously expressed [Nakashima et al, Mol Cell Biol 1993] Ddostp ubiquitously expressed [Yamagata et al, Genomics 1997]
DOLASNT_Uer	0	Yamagata T, Tsuru T, Momoi MY, Suwa K, Nozaki Y, Mukasa T, Ohashi H, Fukushima Y, Momoi T	Genome organization of human 48-kDa oligosaccharyltransferase (DDOST)	Genomics	1997	9367678	Dad1p ubiquitously expressed [Nakashima et al, Mol Cell Biol 1993] Ddostp ubiquitously expressed [Yamagata et al, Genomics 1997]
DOLDPP_Ler	0	Helenius J, Aebi M.	Transmembrane movement of dolichol linked carbohydrates during N-glycoprotein biosynthesis in the endoplasmic reticulum	Semin Cell Dev Biol	2002	12137737	occurs in ER lumen Helenius J, Aebi M. Semin Cell Dev Biol. 2002 Jun;13(3):171-8. Putative gene assignment was result of LocusLink search for additional N-glycosylation genes not associated with KEGG maps.
DOLPH_Ler	0	Schenk B, Fernandez F, Waechter CJ.	The ins(ide) and out(side) of dolichyl phosphate biosynthesis and recycling in the endoplasmic reticulum.	Glycobiology	2001	11425794	occurs in ER lumen Schenk B, Fernandez F, Waechter CJ. Glycobiology. 2001 11(5):61-70.
DOLPMT_L	0	Manos EJ, Kim ML, Kassis J, Chang PY, Wells A, Jones DA.	Dolichol-phosphate-mannose-3 (DPM3)/prostin-1 is a novel phospholipase C-gamma regulated gene negatively associated with prostate tumor invasion.	Oncogene	2001	11420690	Dpm3 transcript ubiquitously expressed [Manos et al, Oncogene 2001]
DOPAMT	3	Bertocci B, Miggiano V, Da Prada M, Dembic Z, Lahm HW, Malherbe P	Human catechol-O-methyltransferase: cloning and expression of the membrane-associated form	Proc Natl Acad Sci U S A	1991	1847521	from kegg map
DOPASULT	3	Chapman E, Best MD, Hanson SR, Wong CH.	Sulfotransferases: structure, mechanism, biological activity, inhibition, and synthetic utility.	Angew Chem Int Ed Engl	2004	15293241	- Added by RS/TV - Sulfotransferases (SULT) catalyze the transfer of a sulfuryl group from a donor molecule, PAPS, to a variety of amine and hydroxy substrates as nucleophiles; - All cytosolic according to ref. - Family 1A of the SULT family is known to prefer phenols as substrates: 1) Sult1a1: 4-nitrophenol, dopamine, iodothyronines (e.g. T3), tyramine 2) Sult1a2: 4-nitrophenol, dopamine, 3) Sult1a3: Dopamine, 4-nitrophenol, norepinephrine, tyramine, Based on Table 2 in Glatt H, Boeing H, Engelke CE, Ma L, Kuhlow A, Pabel U, Pomplun D, Teubner W, Meinl W. Mutat Res. 2001 Oct 1;482(1-2):27-40. Review. & Chapman E, Best MD, Hanson SR, Wong CH.Angew Chem Int Ed Engl. 2004 Jul 5;43(27):3526-48. Review. - Tissue Specificity info: 1)Sult1a1: High in liver; present in numerous other tissues such as platelets, placenta, adrenal gland, colon, brain, leukocytes, endometrium, jejunum. 2) Sult1a2: Liver, some bladder tumors 3) Sult1a3: Very high in jejunum and colon; also present in platelets, placenta, brain, leukocytes; neligible in liver.
DPMVDx	3	Iglesias J, Gonzalez-Pacanowska D, Caamano G, Garcia-Peregrin E	Mevalonate 5-pyrophosphate decarboxylase in isolated villus and crypt cells of chick intestine	Lipids	1988		peroxisomal (not cytosolic, contrary to uniprot etc) specificity: Expressed in heart, skeletal muscle, lung, liver, brain, pancreas, kidney and placenta. The enzyme mevalonate pyrophosphate decarboxylase catalyzes the conversion of mevalonate pyrophosphate into isopentenyl pyrophosphate in one of the early steps in cholesterol biosynthesis. It decarboxylates and dehydrates its substrate while hydrolyzing ATP. NJ
DPMVDx	3	Bonanno JB, Edo C, Eswar N, Pieper U, Romanowski MJ, Ilyin V, Gerchman SE, Kycia H, Studier W, Sali A, Burley SK	Structural genomics of enzymes involved in sterol/isoprenoid biosynthesis	Proceedings of the National Academy of Sciences	2001		peroxisomal (not cytosolic, contrary to uniprot etc) specificity: Expressed in heart, skeletal muscle, lung, liver, brain, pancreas, kidney and placenta. The enzyme mevalonate pyrophosphate decarboxylase catalyzes the conversion of mevalonate pyrophosphate into isopentenyl pyrophosphate in one of the early steps in cholesterol biosynthesis. It decarboxylates and dehydrates its substrate while hydrolyzing ATP. NJ
DTMPK	3	Su JY, Sclafani RA.	Molecular cloning and expression of the human deoxythymidylate kinase gene in yeast.	Nucleic Acids Res	1991	2017365	IT in kegg enzyme acts also on dUMP --> not included yet needs Mg2+
DTMPK	3	Huang SH, Tang A, Drisco B, Zhang SQ, Seeger R, Li C, Jong A.	Human dTMP kinase: gene expression and enzymatic activity coinciding with cell cycle progression and cell growth.	DNA Cell Biol	1994	8024690	IT in kegg enzyme acts also on dUMP --> not included yet needs Mg2+

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DURAD	3	Lu ZH, Zhang R, Diasio RB.	Purification and characterization of dihydropyrimidine dehydrogenase from human liver.	J Biol Chem	1992	1512248	01-26-05 IT based on Lu et al., JBC,1992,24,17102-17109 IT homodimer, need 2 FMN, 2 FAD, 33 iron atoms per molecule enzyme It has not been observed that FAD is converted to FMN
DURAD	3	Lu Z, Zhang R, Diasio RB.	Dihydropyrimidine dehydrogenase activity in human peripheral blood mononuclear cells and liver: population characteristics, newly identified deficient patients, and clinical implication in 5-fluorouracil chemotherapy.	Cancer Res	1993	8221682	01-26-05 IT based on Lu et al., JBC,1992,24,17102-17109 IT homodimer, need 2 FMN, 2 FAD, 33 iron atoms per molecule enzyme It has not been observed that FAD is converted to FMN
DUTPDPm	3	Ladner RD, McNulty DE, Carr SA, Roberts GD, Caradonna SJ.	Characterization of distinct nuclear and mitochondrial forms of human deoxyuridine triphosphate nucleotidohydrolase.	J Biol Chem	1996	8631816	Ladner et al, 1996 report that there are to isoforms originating from the same gene but use the 5'exon differently - however in locuslink there is only one transcript noted IT
DUTPDPm	3	Cohen D, Heng HH, Shi XM, McIntosh EM, Tsui LC, Pearlman RE.	Assignment of the human dUTPase gene (DUT) to chromosome 15q15-q21. 1 by fluorescence in situ hybridization.	Genomics	1997	9070952	Ladner et al, 1996 report that there are to isoforms originating from the same gene but use the 5'exon differently - however in locuslink there is only one transcript noted IT
DUTPDPm	3	Tinkelenberg BA, Fazzone W, Lynch FJ, Ladner RD.	Identification of sequence determinants of human nuclear dUTPase isoform localization.	Exp Cell Res	2003	12799180	Ladner et al, 1996 report that there are to isoforms originating from the same gene but use the 5'exon differently - however in locuslink there is only one transcript noted IT
EBP1r	3	Braverman N, Lin P, Moebius FF, Obie C, Moser A, Glossmann H, Wilcox WR, Rimoin DL, Smith M, Kratz L, Kelley RI, Valle D	Mutations in the gene encoding 3-beta-hydroxysteroid delta8, delta7- isomerase cause X-linked dominant Conradi-Hunermann Syndrome	Nature Genetics	1999		ER - uniprot + ref Emopamil-binding protein (EBP) is an integral membrane protein of the endoplasmic reticulum. It is a high affinity binding protein for the antiischemic phenylalkylamine Ca2+ antagonist [3H]emopamil and the photoaffinity label [3H]azidopamil. It is similar to sigma receptors and may be a member of a superfamily of high affinity drug-binding proteins in the endoplasmic reticulum of different tissues. EBP shares structural features with bacterial and eukaryontic drug transporting proteins. It has four putative transmembrane segments and contains two conserved glutamate residues which may be involved in the transport of cationic amphiphilics. Another prominent feature of EBP is its high content of aromatic amino acid residues (>23%) in its transmembrane segments. These aromatic amino acid residues have been suggested to be involved in the drug transport by the P-glycoprotein. Mutations in this gene cause Chondrodysplasia punctata 2 (CDPX2; also known as Conradi-Hunermann syndrome). NJ
ECOAH1m	2	FitzPatrick DR, Germain-Lee E, Valle D.	Isolation and characterization of rat and human cDNAs encoding a novel putative peroxisomal enoyl-CoA hydratase.		1995	7558027	Additional comments by NJ: localization: peroxiosme an mitochondria by similarity - uniprot see PMID: 7558027 for characterization
EGMESTr	2	Probst MR, Beer M, Beer D, Jeno P, Meyer UA, Gasser R	Human liver arylacetamide deacetylase. Molecular cloning of a novel esterase involved in the metabolic activation of arylamine carcinogens with high sequence similarity to hormone-sensitive lipase	J Biol Chem	1994	8063807	this reaction is reasonable, but the gene association is questionable and without good evidence
ENGASE	0	Suzuki T, Yano K, Sugimoto S, Kitajima K, Lennarz WJ, Inoue S, Inoue Y, Emori Y	Endo-beta-N-acetylglucosaminidase, an enzyme involved in processing of free oligosaccharides in the cytosol	Proc Natl Acad Sci U S A	2002	12114544	Suzuki suggests that FLJ21865p is localized to the cytosol because it lacks a signal sequence. [Suzuki et al, PNAS 99(15):9691-6 (2002)] Flj21865 expressed in thymus, spleen, pancreas, kidney, heart, sk muscle, placenta, brain [Suzuki et al, PNAS 2002]
ENMAN3g	0	Roth J, Ziak M, Zuber C.	The role of glucosidase II and endomannosidase in glucose trimming of asparagine-linked oligosaccharides.	Biochimie	2003	12770767	catalyzed by endomannosidase Fig 4 in [Spiro. J. Biol Chem 275(46): pp. 35637-35660 (2000)] Fig 4 in [Roth, Ziak, Zuber. Biochimie 85: pp. 287-294 (2003).
EPCTX	3	Tan SA, Tan LG	Distribution of ciliatine(2-aminoethylphosphonic acid) and phosphonalanine (2-amino-3-phosphonopropionic acid) in human tissues	Clin Physiol Biochem	1989		EPCTX addition by SAB scant evidence of 2ameph in humans - NJ PETHCT: cyt - uniport NJ
EPCTX	3	Lykidis A, Murtis KG, Jackowski S	Cloning and characterization of a second human CTP:Phosphocholine cytidylyltransferase	Journal of Biological Chemistry	1998		EPCTX addition by SAB scant evidence of 2ameph in humans - NJ PETHCT: cyt - uniport NJ
EPCTX	3	Tan SA, Tan LG	Distribution of ciliatine(2-aminoethylphosphonic acid) and phosphonalanine (2-amino-3-phosphonopropionic acid) in human tissues	Clin Physiol Biochem	1989		EPCTX addition by SAB scant evidence of 2ameph in humans - NJ PETHCT: cyt - uniport NJ
EPCTX	3	Lykidis A, Murtis KG, Jackowski S	Cloning and characterization of a second human CTP:Phosphocholine cytidylyltransferase	Journal of Biological Chemistry	1998		EPCTX addition by SAB scant evidence of 2ameph in humans - NJ PETHCT: cyt - uniport NJ

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ESTRONEGLCt	3	Scheffer GL, Kool M, de Haas M, de Vree JM, Pijnenborg AC, Bosman DK, Elferink RP, van der Valk P, Borst P, Scheper RJ.	Tissue distribution and induction of human multidrug resistant protein 3.	Lab Invest	2002	11850532	ABCC3 - also in hepatocyte and cholangiocytes. a/w pancreatic carcinomas, liver neoplasms. Similar to ABCB11, except also shown to transport estradiol-17-beta-d-glucuronide. Down- and up-regulation of MRP1 (and MRP3) expression can influence cellular folate homeostasis, in particular when cellular retention by polyglutamylation of folates is attenuated. NJ
ESTSULT	3	Aksoy IA, Wood TC, Weinshilboum R.	Human liver estrogen sulfotransferase: identification by cDNA cloning and expression.	Biochem Biophys Res Commun	1994	8185618	cytosolic - uniport and refs TISSUE SPECIFICITY: Liver, intestine and at lower level in the kidney. May also sulfate dehydroepiandrosterone, pregnenolone, ethinylestradiol, equalenin, diethylstilbesterol and 1-naphthol to much smaller degree Sulfotransferase enzymes catalyze the sulfate conjugation of many hormones, neurotransmitters, drugs, and xenobiotic compounds. These cytosolic enzymes are different in their tissue distributions and substrate specificities. The gene structure (number and length of exons) is similar among family members. This gene encodes a protein that transfers a sulfo moiety to and from estrone, which may control levels of estrogen receptors. - It is also known that Sult1a1 is known to catalyze this reaction according to Table 2 in Glatt H, et al. Mutat Res. 2001 Oct 1;482(1-2):27-40. Review. (RS/TV) NJ
ETF	3	Finocchiaro G, Ito M, Ikeda Y, Tanaka K.	Molecular cloning and nucleotide sequence of cDNAs encoding the alpha-subunit of human electron transfer flavoprotein.	J Biol Chem	1988	3170610	- Added by RS/TV - a heterodimer consisting of alpha and beta subunits (Finocchiaro G, Ito M, Ikeda Y, Tanaka K. J Biol Chem. 1988 Oct 25;263(30):15773-80.) - electron-transferring flavoprotein is the physiological electron acceptor from the primary mitochondrial dehydronages such as acyl-CoA dehydrogenases. (Thorpe C, Kim JJ. FASEB J. 1995 Jun;9(9):718-25. Review.) - mitochondrial according to entrez. - beta subunit has two transcriptional variants.
ETF	3	Thorpe C, Kim JJ.	Structure and mechanism of action of the acyl-CoA dehydrogenases.	FASEB J	1995	7601336	- Added by RS/TV - a heterodimer consisting of alpha and beta subunits (Finocchiaro G, Ito M, Ikeda Y, Tanaka K. J Biol Chem. 1988 Oct 25;263(30):15773-80.) - electron-transferring flavoprotein is the physiological electron acceptor from the primary mitochondrial dehydronages such as acyl-CoA dehydrogenases. (Thorpe C, Kim JJ. FASEB J. 1995 Jun;9(9):718-25. Review.) - mitochondrial according to entrez. - beta subunit has two transcriptional variants.
ETFQO	3	Spector EB, Seltzer WK, Goodman SI.	Assignment of electron transfer flavoprotein-ubiquinone oxidoreductase (ETF-QO) to human chromosome 4q33 by fluorescence in situ hybridization and somatic cell hybridization.	Mol Genet Metab	1997	10444348	- Added by RS/TV - Electron transfer flavoprotein-uniquinone oxidoreductase is an iron-sulphur flavoprotein and a component of an electron-transfer system that links 10 different mitochondrial flavoprotein dehydrogenases to the mitochondrial bc1 complex via electron transfer flavoprotein and ubiquinone.(Simkovic M, Degala GD, Eaton SS, Frerman FE. Biochem J. 2002 Jun 15;364(Pt 3):659-67.) - Etfdh.1 is mitochondrial accoridng to Entrez. 84883: - putative function based on significant homology with NADH oxidoreductases/flavoproteins from bacteria to mammalian species (discovered through BLAST searches of the GenBank database). - AMID is associated with the outer membrane of mitochondria and localized in the cytosol.

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ETFQO	3	Simkovic M, Degala GD, Eaton SS, Frerman FE.	Expression of human electron transfer flavoprotein-ubiquinone oxidoreductase from a baculovirus vector: kinetic and spectral characterization of the human protein.	Biochem J	2002	12049629	- Added by RS/TV - Electron transfer flavoprotein-uniquinone oxidoreductase is an iron-sulphur flavoprotein and a component of an electron-transfer system that links 10 different mitochondrial flavoprotein dehydrogenases to the mitochondrial bc1 complex via electron transfer flavoprotein and ubiquinone.(Simkovic M, Degala GD, Eaton SS, Frerman FE. Biochem J. 2002 Jun 15;364(Pt 3):659-67.) - Etfdh.1 is mitochondrial accoridng to Entrez. 84883: - putative function based on significant homology with NADH oxidoreductases/flavoproteins from bacteria to mammalian species (discovered through BLAST searches of the GenBank database). - AMID is associated with the outer membrane of mitochondria and localized in the cytosol.
FA120ACPH	3	Chirala SS, Huang WY, Jayakumar A, Sakai K, Wakil SJ.	Animal fatty acid synthase: functional mapping and cloning and expression of the domain I constituent activities.	Proc Natl Acad Sci U S A	1997	9159116	Lumped FAS reactions in current reconstruction, so these reactions won't be used. In future versions if synthesis is unlumped these will be used in simulations. see PMID: 9159116 NJ
FACOAL160i	3	Stanczak H, Stanczak JJ, Singh I.	Chromosomal localization of the human gene for palmitoyl-CoA ligase (FACL1).	Cytogenet Cell Genet	1992	1531127	mitochondrial membrane and peroxisomal membrane. Assuming outer mitochondrial membrane is effectively cytosolic (reaction product will need to be transported into mit if it is to undergo any reactions there). - UniProt - Additional information by RS/TV J Biol Chem. 2001 Jun 8;276(23):20182-5. Epub 2001 Mar 27 ACS results in the conversion of free fatty acid into fatty acyl-CoA esters according to Cao Y. Genomics. 1998 Apr 15;49(2):327-30. Tissue expression and substrate specificity: 1) Acsl1 is highly expressed in highly in liver, adipose tissue, and heart. Acsl6 is a brain-specific subtype of Acsl1. Acsl1 (and consequently Acsl6 which is nearly identical to Acsl2) prefers C10-C18 saturated fatty acids and C16-C20 unsaturated fatty acids. 2) Acsl3 is also highly expressed in the brain. Acsl3 on the other hand prefers C8-C22 saturated fatty acids and C16-C20 unsaturated fatty acids. 3) Acsl4 prefers archidonic acid (20:4) and eicosapentaenoic acid(20:5) as substrates. Human placenta, brain, testis, ovary, spleen, and adrenal cortex all expressed high levels of Acsl4. 4) It is suggested that Acsl5 tends to be expressed in human sm Subcellular localizations: All are expressed to some extent in m Subcellular localizations according to Lewin TM. J Biol Chem. Tissue expression and substrate specificity according to
FACOAL161	3	Mashek DG, Bornfeldt KE, Coleman RA, Berger J, Bernlohr DA, Black P, DiRusso CC, Farber SA, Guo W, Hashimoto N, Khodiyar V, Kuypers FA, Maltais LJ, Nebert DW, Renieri A, Schaffer JE, Stahl A, Watkins PA, Vasiliou V, Yamamoto TT.	Revised nomenclature for the mammalian long-chain acyl-CoA synthetase gene family.	J Lipid Res	2004	15292367	Additional set only added to Acsl1.1 for the time being. For updated nomenclature of Acsl see PMID: 15292367. Reversible reaction due to Keq ~1 - see Reich: Energy metabolism of the cell. ACSL1,3,4,5 noted to occur in Microsomes, outer mitochondrial membrane and peroxisomal membrane. Assuming outer mitochondrial membrane is effectively cytosolic (reaction product will need to be transported into mit if it is to undergo any reactions there). - UniProt NJ
FACOAL161	3	Reich, Sel'kov	Energy metabolism of the cell : a theoretical treatise		1981		Additional set only added to Acsl1.1 for the time being. For updated nomenclature of Acsl see PMID: 15292367. Reversible reaction due to Keq ~1 - see Reich: Energy metabolism of the cell. ACSL1,3,4,5 noted to occur in Microsomes, outer mitochondrial membrane and peroxisomal membrane. Assuming outer mitochondrial membrane is effectively cytosolic (reaction product will need to be transported into mit if it is to undergo any reactions there). - UniProt NJ
FACOAL206	3	Steinberg SJ, Wang SJ, Kim DG, Mihalik J, Watkins PA	Human very-long-chain-acyl-CoA synthetase	Biochemical and biophysical research communications	1999		The protein encoded by this gene is an isozyme of long-chain fatty-acid-coenzyme A ligase family. Although differing in substrate specificity, subcellular localization, and tissue distribution, all isozymes of this family convert free long-chain fatty acids into fatty acyl-CoA esters, and thereby play a key role in lipid biosynthesis and fatty acid degradation. This isozyme activates long-chain, branched-chain and very-long-chain fatty acids containing 22 or more carbons to their CoA derivatives. It is expressed primarily in liver and kidney, and is present in both endoplasmic reticulum and peroxisomes but not in mitochondria. Its decreased peroxisomal enzyme activity is in part responsible for the biochemical pathology in X-linked adrenoleukodystrophy. NJ

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FACOAL40im	3	Bugaut M	Occurrence, absorption and metabolism of short chain fatty acids in the digestive tract of mammals	Comp Biochem Physiol B	1987	3297476	- identification of gene [Fujino, J Biol Chem 2001] - mouse gene was characterized and shown to be localized to mitochondrial matrix [Fujino, J Biol Chem 2001] - reaction described in [Bugaut, Comp Biochem Physiol B 1987]
FACOAL40im	3	Fujino T, Takei YA, Sone H, Ioka RX, Kamataki A, Magoori K, Takahashi S, Sakai J, Yamamoto TT	Molecular identification and characterization of two medium-chain acyl-CoA synthetases, MACS1 and the Sa gene product	J Biol Chem	2001	11470804	- identification of gene [Fujino, J Biol Chem 2001] - mouse gene was characterized and shown to be localized to mitochondrial matrix [Fujino, J Biol Chem 2001] - reaction described in [Bugaut, Comp Biochem Physiol B 1987]
FAEL183	3	Zhang K, Kniazeva M, Han M, Li W, Yu Z, Yang Z, Li Y, Metzker ML, Allikmets R, Zack DJ, Kakuk LE, Lagali PS, Wong PW, MacDonald IM, Sieving PA, Figueroa DJ, Austin CP, Gould RJ, Ayyagari R, Petrukhin K.	A 5-bp deletion in ELOVL4 is associated with two related forms of autosomal dominant macular dystrophy.	Nat Genet	2001	11138005	localization: microsomal - specificty (inner vs outer membrane) not determined so it is assumed to take place on the outer membrane - part of the microsomal elongase/desaturase reactions. Set of 7 elongase genes (ELOVL1-7), the other genes (ELOVL1,2,3,5,6,7) are not as well characterized so gene associations have not yet been made. see PMID: 11138005 for dz association (macular dystrophy) NJ
FAEL183	3	Nakamura MT, Nara TY.	Structure, function, and dietary regulation of delta6, delta5, and delta9 desaturases.	Annu Rev Nutr	2004	15189125	localization: microsomal - specificty (inner vs outer membrane) not determined so it is assumed to take place on the outer membrane - part of the microsomal elongase/desaturase reactions. Set of 7 elongase genes (ELOVL1-7), the other genes (ELOVL1,2,3,5,6,7) are not as well characterized so gene associations have not yet been made. see PMID: 11138005 for dz association (macular dystrophy) NJ
FAH1	3	Palmer CN, Richardson TH, Griffin KJ, Hsu MH, Muerhoff AS, Clark JE, Johnson EF.	Characterization of a cDNA encoding a human kidney, cytochrome P-450 4A fatty acid omega-hydroxylase and the cognate enzyme expressed in Escherichia coli.	Biochim Biophys Acta	1993	7679927	ER integral membrane - assumed to face cytosol This protein localizes to the endoplasmic reticulum and hydroxylates medium-chain fatty acids such as laurate and myristate. NJ
FAH1	3	Imaoka S, Ogawa H, Kimura S, Gonzalez FJ.	Complete cDNA sequence and cDNA-directed expression of CYP4A11, a fatty acid omega-hydroxylase expressed in human kidney.	DNA Cell Biol	1993	8274222	ER integral membrane - assumed to face cytosol This protein localizes to the endoplasmic reticulum and hydroxylates medium-chain fatty acids such as laurate and myristate. NJ
FALDH	3	Uotila L, Koivusalo M	Purification and properties of S-formylglutathione hydrolase from human liver	J Biol Chem	1974	4436331	The degradation of fald is relatively certain. The exact mechanism appears to be well established, but is less certain.
FALDH	3	Engeland K, Hoog JO, Holmquist B, Estonius M, Jornvall H, Vallee BL	Mutation of Arg-115 of human class III alcohol dehydrogenase: a binding site required for formaldehyde dehydrogenase activity and fatty acid activation	Proc Natl Acad Sci U S A	1993	8460164	The degradation of fald is relatively certain. The exact mechanism appears to be well established, but is less certain.
FAOXC160	3	Naito E, Ozasa H, Ikeda Y, Tanaka K.	Molecular cloning and nucleotide sequence of complementary DNAs encoding human short chain acyl-coenzyme A dehydrogenase and the study of the molecular basis of human short chain acyl-coenzyme A dehydrogenase deficiency.	J Clin Invest	1985	2565344	- Added by RS/TV 1) Acyl-Coa dehydrogenases all catalyze a,b-dehydrogenation of acyl-CoA esters and transfer electrons to electron transfer flavoprotein 2) Acads-m and Acadm-m catalyze the first step of B-oxidation cycles for fatty acids with various chain length 3) Acads-m & Acadm-m are located in the mitochondrial matrix 4) Acadm-m is active with acyl chain lengths of C4 to C16. However, Acadm-m and Acads-m exhibits overlap in substrate range (length of fatty acyl chains) 1 through 4 according to Naito E, Ozasa H, Ikeda Y, Tanaka K. Molecular cloning and nucleotide sequence of complementary DNAs encoding human short chain acyl-coenzyme A dehydrogenase and the study of the molecular basis of human short chain acyl-coenzyme A dehydrogenase deficiency. J Clin Invest. 1989 May;83(5):1605-13. PMID: 2565344 Also, Kelly DP, Kim JJ, Billadello JJ, Hainline BE, Chu TW, Strauss AW. Nucleotide sequence of medium-chain acyl-CoA dehydrogenase mRNA and its expression in enzyme-deficient human tissue. Proc Natl Acad Sci U S A. 1987 Jun;84(12):4068-72. PMID: 3035565

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FAOXC160	3	Kelly DP, Kim JJ, Billadello JJ, Hainline BE, Chu TW, Strauss AW	Nucleotide sequence of medium-chain acyl-CoA dehydrogenase mRNA and its expression in enzyme-deficient human tissue.	Proc Natl Acad Sci U S A	1987	3035565	- Added by RS/TV 1) Acyl-Coa dehydrogenases all catalyze a,b-dehydrogenation of acyl-CoA esters and transfer electrons to electron transfer flavoprotein 2) Acads-m and Acadm-m catalyze the first step of B-oxidation cycles for fatty acids with various chain length 3) Acads-m & Acadm-m are located in the mitochondrial matrix 4) Acadm-m is active with acyl chain lengths of C4 to C16. However, Acadm-m and Acads-m exhibits overlap in substrate range (length of fatty acyl chains) 1 through 4 according to Naito E, Ozasa H, Ikeda Y, Tanaka K. Molecular cloning and nucleotide sequence of complementary DNAs encoding human short chain acyl-coenzyme A dehydrogenase and the study of the molecular basis of human short chain acyl-coenzyme A dehydrogenase deficiency. J Clin Invest. 1989 May;83(5):1605-13. PMID: 2565344 Also, Kelly DP, Kim JJ, Billadello JJ, Hainline BE, Chu TW, Strauss AW. Nucleotide sequence of medium-chain acyl-CoA dehydrogenase mRNA and its expression in enzyme-deficient human tissue. Proc Natl Acad Sci U S A. 1987 Jun;84(12):4068-72. PMID: 3035565
FAOXC204	3	Weiping Le, Azfar S. Abbas, Howard Sprecher, Jerry Vockley and Horst Schulz	Long-chain acyl-CoA dehydrogenase is a key enzyme in the mitochondrial [beta]-oxidation of unsaturated fatty acids	Biochimica et Biophysica Acta (BBA) - Molecular and Cell Biology of Lipids	2000		- Added by RS/TV 1) Acyl-Coa dehydrogenases all catalyze a,b-dehydrogenation of acyl-CoA esters and transfer electrons to electron transfer flavoprotein 2) Acads-m and Acadm-m catalyze the first step of B-oxidation cycles for fatty acids with various chain length 3) Acads-m & Acadm-m are located in the mitochondrial matrix 4) Acadm-m is active with acyl chain lengths of C4 to C16. However, Acadm-m and Acads-m exhibits overlap in substrate range (length of fatty acyl chains) 5) Acadl-m is important for the -oxidation of regular fatty acid intermediates that have acyl chains with 10 to 14 carbons, although it accpets substrates with long carbon chains. There is some overlap of the type of substrate it will accept with Acadm-m 1 through 4 according to Naito E, Ozasa H, Ikeda Y, Tanaka K. Molecular cloning and nucleotide sequence of complementary DNAs encoding human short chain acyl-coenzyme A dehydrogenase and the study of the molecular basis of human short chain acyl-coenzyme A dehydrogenase deficiency. J Clin Invest. 1989 May;83(5):1605-13. PMID: 2565344 Also, Kelly DP, Kim JJ, Billadello JJ, Hainline BE, Chu TW, Strauss Proc Natl Acad Sci U S A. 1987 Jun;84(12):4068-72.
FAS100COA	3	Wakil SJ.	Fatty acid synthase, a proficient multifunctional enzyme.	Biochemistry	1989	2669958	cytosol: uniprot Fatty acid synthetase catalyzes the formation of long- chain fatty acids from acetyl-CoA, malonyl-CoA and NADPH. This multifunctional protein has 7 catalytic activities and an acyl carrier protein. See PMID: 2669958 for lumped reaction stoichiometry. Specificity: all tissues, but prominent expression in brain, lung, and liver. Structure and substrate specificity: PMID: 15507492. The enzyme encoded by this gene is a multifunctional protein. Its main function is to catalyze the synthesis of palmitate from acetyl-CoA and malonyl-CoA, in the presence of NADPH, into long-chain saturated fatty acids. In some cancer cell lines, this protein has been found to be fused with estrogen receptor-alpha (ER-alpha), in which the N-terminus of FAS is fused in-frame with the C-terminus of ER-alpha. Most tissues produce C16, some tissues produce smaller chains (e.g. C12), hence lumped reaction in addition to stepwise reactions included (see closing sentences of PMID: 2669958). NJ

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
FAS100COA	3	Brink J, Ludtke SJ, Yang CY, Gu ZW, Wakil SJ, Chiu W.	Quaternary structure of human fatty acid synthase by electron cryomicroscopy.	Proc Natl Acad Sci	2002	11756679	cytosol: uniprot Fatty acid synthetase catalyzes the formation of long- chain fatty acids from acetyl-CoA, malonyl-CoA and NADPH. This multifunctional protein has 7 catalytic activities and an acyl carrier protein. See PMID: 2669958 for lumped reaction stoichiometry. Specificity: all tissues, but prominent expression in brain, lung, and liver. Structure and substrate specificity: PMID: 15507492. The enzyme encoded by this gene is a multifunctional protein. Its main function is to catalyze the synthesis of palmitate from acetyl-CoA and malonyl-CoA, in the presence of NADPH, into long-chain saturated fatty acids. In some cancer cell lines, this protein has been found to be fused with estrogen receptor-alpha (ER-alpha), in which the N-terminus of FAS is fused in-frame with the C-terminus of ER-alpha. Most tissues produce C16, some tissues produce smaller chains (e.g. C12), hence lumped reaction in addition to stepwise reactions included (see closing sentences of PMID: 2669958). NJ
FAS100COA	3	Chakravarty B, Gu Z, Chirala SS, Wakil SJ, Quiocho FA.	Human fatty acid synthase: structure and substrate selectivity of the thioesterase domain.		2004	15507492	cytosol: uniprot Fatty acid synthetase catalyzes the formation of long- chain fatty acids from acetyl-CoA, malonyl-CoA and NADPH. This multifunctional protein has 7 catalytic activities and an acyl carrier protein. See PMID: 2669958 for lumped reaction stoichiometry. Specificity: all tissues, but prominent expression in brain, lung, and liver. Structure and substrate specificity: PMID: 15507492. The enzyme encoded by this gene is a multifunctional protein. Its main function is to catalyze the synthesis of palmitate from acetyl-CoA and malonyl-CoA, in the presence of NADPH, into long-chain saturated fatty acids. In some cancer cell lines, this protein has been found to be fused with estrogen receptor-alpha (ER-alpha), in which the N-terminus of FAS is fused in-frame with the C-terminus of ER-alpha. Most tissues produce C16, some tissues produce smaller chains (e.g. C12), hence lumped reaction in addition to stepwise reactions included (see closing sentences of PMID: 2669958). NJ
FATP1t	3	Fitscher BA, Riedel HD, Young KC, Stremmel W.	Tissue distribution and cDNA cloning of a human fatty acid transport protein (hsFATP4).	Biochim Biophys Acta	1998	9878842	SLC27A1: heart SLC27A2: kidney SLC27A3: lung SLC27A4: all tissues (ubiquitous) SLC27A5: liver SLC27A6: not well known Specific mechanism unknown - added Na cotransport to make electroneutral transport (mechanism likely involves active prot channels, in contrast to diffusional transport of fatty acids). Specificity of fatty acids not very well known (FATP5t/SLC27A5 transports "very long chain FA"). SLC27A2: The protein encoded by this gene is an isozyme of long-chain fatty-acid-coenzyme A ligase family. Although differing in substrate specificity, subcellular localization, and tissue distribution, all isozymes of this family convert free long-chain fatty acids into fatty acyl-CoA esters, and thereby play a key role in lipid biosynthesis and fatty acid degradation. This isozyme activates long-chain, branched-chain and very-long-chain fatty acids containing 22 or more carbons to their CoA derivatives. It is expressed primarily in liver and kidney, and is present in both endoplasmic reticulum and peroxisomes but not in mitochondria. Its decreased peroxisomal enzyme activity is i SLC27A5: The protein encoded by this gene is an isozyme of v FATP6 is involved in heart LCFA uptake, in which it may play

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
FATP1t	3	Stahl A.	A current review of fatty acid transport proteins (SLC27).	Pflugers Arch	2004	12856180	SLC27A1: heart SLC27A2: kidney SLC27A3: lung SLC27A4: all tissues (ubiquitous) SLC27A5: liver SLC27A6: not well known Specific mechanism unknown - added Na cotransport to make electroneutral transport (mechanism likely involves active prot channels, in contrast to diffusional transport of fatty acids). Specificity of fatty acids not very well known (FATP5t/SLC27A5 transports "very long chain FA"). SLC27A2: The protein encoded by this gene is an isozyme of long-chain fatty-acid-coenzyme A ligase family. Although differing in substrate specificity, subcellular localization, and tissue distribution, all isozymes of this family convert free long- chain fatty acids into fatty acyl-CoA esters, and thereby play a key role in lipid biosynthesis and fatty acid degradation. This isozyme activates long-chain, branched-chain and very-long- chain fatty acids containing 22 or more carbons to their CoA derivatives. It is expressed primarily in liver and kidney, and is present in both endoplasmic reticulum and peroxisomes but not in mitochondria. Its decreased peroxisomal enzyme activity is i SLC27A5: The protein encoded by this gene is an isozyme of v FATP6 is involved in heart LCFA uptake, in which it may play
FATP1t	3	Pohl J, Ring A, Hermann T, Stremmel W.	Role of FATP in parenchymal cell fatty acid uptake.	Biochim Biophys Acta	2004	15522816	SLC27A1: heart SLC27A2: kidney SLC27A3: lung SLC27A4: all tissues (ubiquitous) SLC27A5: liver SLC27A6: not well known Specific mechanism unknown - added Na cotransport to make electroneutral transport (mechanism likely involves active prot channels, in contrast to diffusional transport of fatty acids). Specificity of fatty acids not very well known (FATP5t/SLC27A5 transports "very long chain FA"). SLC27A2: The protein encoded by this gene is an isozyme of long-chain fatty-acid-coenzyme A ligase family. Although differing in substrate specificity, subcellular localization, and tissue distribution, all isozymes of this family convert free long- chain fatty acids into fatty acyl-CoA esters, and thereby play a key role in lipid biosynthesis and fatty acid degradation. This isozyme activates long-chain, branched-chain and very-long- chain fatty acids containing 22 or more carbons to their CoA derivatives. It is expressed primarily in liver and kidney, and is present in both endoplasmic reticulum and peroxisomes but not in mitochondria. Its decreased peroxisomal enzyme activity is i SLC27A5: The protein encoded by this gene is an isozyme of v FATP6 is involved in heart LCFA uptake, in which it may play
FATP4t	3	Steinberg SJ, Wang SJ, McGuinness MC, Watkins PA.	Human liver-specific very-long-chain acyl-coenzyme A synthetase: cDNA cloning and characterization of a second enzymatically active protein.	Mol Genet Metab	1999	10479480	SLC27A1: heart SLC27A2: kidney SLC27A3: lung SLC27A4: all tissues (ubiquitous) SLC27A5: liver SLC27A6: not well known Specific mechanism unknown - added Na cotransport to make electroneutral transport (mechanism likely involves active prot channels, in contrast to diffusional transport of fatty acids). Specificity of fatty acids not very well known (FATP5t/SLC27A5 transports "very long chain FA"). SLC27A2: The protein encoded by this gene is an isozyme of long-chain fatty-acid-coenzyme A ligase family. Although differing in substrate specificity, subcellular localization, and tissue distribution, all isozymes of this family convert free long- chain fatty acids into fatty acyl-CoA esters, and thereby play a key role in lipid biosynthesis and fatty acid degradation. This isozyme activates long-chain, branched-chain and very-long- chain fatty acids containing 22 or more carbons to their CoA derivatives. It is expressed primarily in liver and kidney, and is present in both endoplasmic reticulum and peroxisomes but not in mitochondria. Its decreased peroxisomal enzyme activity is i SLC27A5: The protein encoded by this gene is an isozyme of v FATP6 is involved in heart LCFA uptake, in which it may play
FBP	0	Tillmann H, Eschrich K	Isolation and characterization of an allelic cDNA for human muscle fructose-1,6-bisphosphatase	Gene	1998	9678974	2203: - expressed in liver [Genatlas] 8789: - expressed in muscle [Tillmann and Eschrich, Gene 1998]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
FCLTm	2	Tugores A, Magness ST, Brenner DA.	A single promoter directs both housekeeping and erythroid preferential expression of the human ferrochelatase gene.	J Biol Chem	1994	7983009	- Added by RS/TV Proteome - Subcellular location: Bound to the mitochondrial inner membrane in eukaryotic cells with its active site on the matrix side of the membrane, according to Gene Cards. - Added by RS/TV - Catalytic Activity according to Gene Cards: Catalyzes the ferrous insertion into protoporphyrin IX. - Ferrocheletase, the last enzyme of the heme biosynthetic pathway, catalyzes the chelation of ferrous iron and protoporphyrin to form heme. Enzymes participating the heme biosynthetic pathway must be expressed in all cell types to supply the heme required as a co-factor for the respiratory cytochromes. (Tugores A, Magness ST, Brenner DA. J Biol Chem. 1994 Dec 9;269(49):30789-97.)
FCOAH	2	Jansen GA, van den Brink DM, Ofman R, Draghici O, Dacremont G, Wanders RJ.	Identification of pristanal dehydrogenase activity in peroxisomes: conclusive evidence that the complete phytanic acid alpha-oxidation pathway is localized in peroxisomes.	Biochem Biophys Res Commun	2001	11341778	Verhoeven 11591435 and clayton 11356171 See Jansen et al (PMID 11341778) Localization unclear - may be peroxisomal or cytoplasmic - peroxisomal chosen since the next step (oxidation of formate - FDH) is catalyzed by an cytoplasmic enzyme. NJ
FE2t	3	Goswami T, Rolfs A, Hediger MA	Iron transport: emerging roles in health and disease	Biochem Cell Biol	2002	12440707	- this is a lumped reaction representing: (1) iron absorption at the apical membrane intestinal mucosal cells, (2) its secretion and oxidation at the basolateral membrane, (3) binding to transferrin (Tf) and transport in the bloodstream, (4) uptake into heme-producing cells and release of Fe2+ from Tf [Goswami 2002], [Orten 1978] - uptake across apical membrane is mediated by SLC11 family (DCT1, Nramp2) in a voltage-dependent, proton coupled mechanism [Goswami 2002] - export at the basolateral membrane is mediated by IREG1; subsquent pxidation is by hephaestin [Goswami 2002]
FE2tm	2	Lange H, Kispal G, Lill R.	Mechanism of iron transport to the site of heme synthesis inside yeast mitochondria.	J Biol Chem	1999	10383398	- Added by RS/TV - No genes found. - Iron uptake is driven energetically by a membrane potential across the inner membrane but does not require ATP. (Lange H, Kispal G, Lill R. Biol Chem. 1999 Jul 2;274(27):18989-96.) - ABCB10 (ABC-me) appears to play a role in the transport of heme or heme intermediates across the mitochondrial membrane based on indirect experimental observations. See [Chung 2003] for refs.
FE2tm	2	Chung J, Wessling-Resnick M	Molecular mechanisms and regulation of iron transport.	Crit Rev Clin Lab Sci	2003	12755454	- Added by RS/TV - No genes found. - Iron uptake is driven energetically by a membrane potential across the inner membrane but does not require ATP. (Lange H, Kispal G, Lill R. Biol Chem. 1999 Jul 2;274(27):18989-96.) - ABCB10 (ABC-me) appears to play a role in the transport of heme or heme intermediates across the mitochondrial membrane based on indirect experimental observations. See [Chung 2003] for refs.
FE3R2e	3	McKie AT, Barrow D, Latunde-Dada GO, Rolfs A, Sager G, Mudaly E, Mudaly M, Richardson C, Barlow D, Bomford A, Peters TJ, Raja KB, Shirali S, Hediger MA, Farzaneh F, Simpson RJ.	An iron-regulated ferric reductase associated with the absorption of dietary iron	Science	2001	11230685	- stoichiometry uncertain; inferred from EC 1.16.1.7 - Cybrd1 has ascrobic acid binding site; ascorbate also enhances iron absroption; rabbit protein showm to catazlye iron reduction in presence of ascorbate; see refs in [Chung 2003] - Fe3+ reducing avitivity has been isolated from duodenal microvilli membranes and intestinal cell lines; see refs in [Chung 2003] 79901: - cloned [McKie 2001] - reductase activity observed in Xenopus oocytes [McKie 2001] - expressed in brush border membrane of duodenum [Chung 2003]
FPGS	3	McGuire JJ, Russell CA, Balinska M.	Human cytosolic and mitochondrial folylpolyglutamate synthetase are electrophoretically distinct. Expression in antifolate-sensitive and -resistant human cell lines.	J Biol Chem	2000	10777604	IT

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FRDPtr	2	Kovacs WJ, Olivier LM, Krisans SK.	Central role of peroxisomes in isoprenoid biosynthesis.	Prog Lipid Res	2002	12121718	Evidence of flip-flop as mechanism for intracellular transport (PMID: 12840657) discussed in Hamilton. Signifiance of FRDP in particular for IC transport, Krisans et al (PMID: 14713247, PMID: 12121718). NJ
FRDPtr	2	Hamilton JA.	Fast flip-flop of cholesterol and fatty acids in membranes: implications for membrane transport proteins.	Curr Opin Lipidol	2003	12840657	Evidence of flip-flop as mechanism for intracellular transport (PMID: 12840657) discussed in Hamilton. Signifiance of FRDP in particular for IC transport, Krisans et al (PMID: 14713247, PMID: 12121718). NJ
FRUt1r	3	Burant CF, Takeda J, Brot-Laroche E, Bell GI, Davidson NO	Fructose transporter in human spermatozoa and small intestine is GLUT5	J Biol Chem	1992	1634504	6514: - major substrates are Glc (low affinity [Johnson, 1990]), Gal, Fru, Man, GlcN [Uldry, 2004], [Uldry, 2002] - facilitated diffusion [Uldry, Pflugers Arch 2004] - intestine and kidney [Thorens, 1990], liver, pancreatic B cells, islet of Langerhans, brain [Uldry, 2004] - cDNA was cloned [Fukumoto, 1988] - glucose release from hepatocytes has two major pathways: 1) faciliated diffusion through GLUT2 and 2) membrane traffic pathway from ER to extracellular space. There is also a third minor route inwhich glucose re-enters the cytosol to form a pool that slowly diffuses out of the cells [Hosokawa, 2002] - release of glucose by transporters other than GLUT2 is low [Guillam, 1998] 6515: - major substrates are Glc (high affinity), Gal, Man, Xyl, maltose, dehydroascorbic acid [Uldry, 2004] - brain (neurons), testis (spermatozoa) [Haber, 1993], [Uldry, 2004], sk muscle (slow twitch fibers) [Stuart, 1999], platelets (alpha-granules) [Heijen, 1997] - cDNA was cloned [Kayano, 1988] 6518: - transports Fru [Uldry, Pflugers Arch 2004], but not Glc [Burant, 1992] - sm intestine (jejunal region), kidney, sk muscle, adipocytes [U - cDNA was isolated [Kayano,1990] 155184: - gene identified [Joost, 2001] and cloned [Li, 2004] - transports Glc, Fru [Li, 2004]
FRUt1r	3	Kayano T, Burant CF, Fukumoto H, Gould GW, Fan YS, Eddy RL, Byers MG, Shows TB, Seino S, Bell GI	Human facilitative glucose transporters.	J Biol Chem	1990	1695905	6514: - major substrates are Glc (low affinity [Johnson, 1990]), Gal, Fru, Man, GlcN [Uldry, 2004], [Uldry, 2002] - facilitated diffusion [Uldry, Pflugers Arch 2004] - intestine and kidney [Thorens, 1990], liver, pancreatic B cells, islet of Langerhans, brain [Uldry, 2004] - cDNA was cloned [Fukumoto, 1988] - glucose release from hepatocytes has two major pathways: 1) faciliated diffusion through GLUT2 and 2) membrane traffic pathway from ER to extracellular space. There is also a third minor route inwhich glucose re-enters the cytosol to form a pool that slowly diffuses out of the cells [Hosokawa, 2002] - release of glucose by transporters other than GLUT2 is low [Guillam, 1998] 6515: - major substrates are Glc (high affinity), Gal, Man, Xyl, maltose, dehydroascorbic acid [Uldry, 2004] - brain (neurons), testis (spermatozoa) [Haber, 1993], [Uldry, 2004], sk muscle (slow twitch fibers) [Stuart, 1999], platelets (alpha-granules) [Heijen, 1997] - cDNA was cloned [Kayano, 1988] 6518: - transports Fru [Uldry, Pflugers Arch 2004], but not Glc [Burant, 1992] - sm intestine (jejunal region), kidney, sk muscle, adipocytes [U - cDNA was isolated [Kayano,1990] 155184: - gene identified [Joost, 2001] and cloned [Li, 2004] - transports Glc, Fru [Li, 2004]

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FRUt1r	3	Mantych GJ, James DE, Devaskar SU	Jejunal/kidney glucose transporter isoform (Glut-5) is expressed in the human blood-brain barrier	Endocrinology	1993	8419132	6514: - major substrates are Glc (low affinity [Johnson, 1990]), Gal, Fru, Man, GlcN [Uldry, 2004], [Uldry, 2002] - facilitated diffusion [Uldry, Pflugers Arch 2004] - intestine and kidney [Thorens, 1990], liver, pancreatic B cells, islet of Langerhans, brain [Uldry, 2004] - cDNA was cloned [Fukumoto, 1988] - glucose release from hepatocytes has two major pathways: 1) faciliated diffusion through GLUT2 and 2) membrane traffic pathway from ER to extracellular space. There is also a third minor route inwhich glucose re-enters the cytosol to form a pool that slowly diffuses out of the cells [Hosokawa, 2002] - release of glucose by transporters other than GLUT2 is low [Guillam, 1998] 6515: - major substrates are Glc (high affinity), Gal, Man, Xyl, maltose, dehydroascorbic acid [Uldry, 2004] - brain (neurons), testis (spermatozoa) [Haber, 1993], [Uldry, 2004], sk muscle (slow twitch fibers) [Stuart, 1999], platelets (alpha-granules) [Heijen, 1997] - cDNA was cloned [Kayano, 1988] 6518: - transports Fru [Uldry, Pflugers Arch 2004], but not Glc [Burant, 1992] - sm intestine (jejunal region), kidney, sk muscle, adipocytes [U - cDNA was isolated [Kayano,1990] 155184: - gene identified [Joost, 2001] and cloned [Li, 2004] - transports Glc, Fru [Li, 2004]
FTHFCL	3	Anguera MC, Suh JR, Ghandour H, Nasrallah IM, Selhub J, Stover PJ.	Methenyltetrahydrofolate synthetase regulates folate turnover and accumulation.	J Biol Chem	2003	12764149	IT cytoplasm based on GeneCards
FTHFDH	3	Johlin FC, Swain E, Smith C, Tephly TR.	Studies on the mechanism of methanol poisoning: purification and comparison of rat and human liver 10-formyltetrahydrofolate dehydrogenase.	Mol Pharmacol	1989	2733692	IT
FTHFLm	3	Prasannan P, Pike S, Peng K, Shane B, Appling DR	Human mitochondrial C1-tetrahydrofolate synthase: gene structure, tissue distribution of the mRNA, and immunolocalization in Chinese hamster ovary calls	J Biol Chem	2003	12937168	- encodes the mitochondrial isozyme of C1-tetrahydrofolate (THF) synthase, a trifunctional enzyme containing formyl-THF synthetase, methenyl-THF cyclohydrolase, and methylene-THF dehydrogenase activities [Prasannan, J Biol Chem 2003]
FUCASE2e	3	Hopfer RL, Johnson SW, Masserini M, Giuliani A, Alhadeff JA	Hydrolysis of fucosyl-GM1 ganglioside by purified pellet-associated human brain and human liver alpha-L-fucosidases without activator proteins or detergents	Biochem J	1990	2317201	2519: - integral membrane protein on cell surface [Cordero 2001] - accounts for 10-20% of total cellular fucosidase activity [Cordero 2001] - hematopoietic, epithelial, & mesenchimal cells expressed cell surface protein with alpha-l-fucosidase activity [Cordero 2001]; also found in membrane-assoc fraction from brain [Hopfer 1990] and plasma membrane of sperm [Alhadeff 1999] - purified and kinetically characterized [Khunsook 2003]
FUCASE2e	3	Alhadeff JA, Khunsook S, Choowongkomon K, Baney T, Heredia V, Tweedie A, Bean B	Characterization of human semen alpha-L-fucosidases	Mol Hum Reprod	1999	10460218	2519: - integral membrane protein on cell surface [Cordero 2001] - accounts for 10-20% of total cellular fucosidase activity [Cordero 2001] - hematopoietic, epithelial, & mesenchimal cells expressed cell surface protein with alpha-l-fucosidase activity [Cordero 2001]; also found in membrane-assoc fraction from brain [Hopfer 1990] and plasma membrane of sperm [Alhadeff 1999] - purified and kinetically characterized [Khunsook 2003]
FUCASE2e	3	Cordero OJ, Merino A, Paez de la Cadena M, Bugia B, Nogueira M, Vinuela JE, Martinez-Zorzano VS, de Carlos A, Rodriguez-Berrocal FJ	Cell surface human alpha-L-fucosidase	Eur J Biochem	2001	11389735	2519: - integral membrane protein on cell surface [Cordero 2001] - accounts for 10-20% of total cellular fucosidase activity [Cordero 2001] - hematopoietic, epithelial, & mesenchimal cells expressed cell surface protein with alpha-l-fucosidase activity [Cordero 2001]; also found in membrane-assoc fraction from brain [Hopfer 1990] and plasma membrane of sperm [Alhadeff 1999] - purified and kinetically characterized [Khunsook 2003]
FUCASE2e	3	Khunsook S, Bean BS, McGowan SR, Alhadeff JA	Purification and characterization of plasma membrane-associated human sperm alpha-L-fucosidase	Biol Reprod	2003	12604617	2519: - integral membrane protein on cell surface [Cordero 2001] - accounts for 10-20% of total cellular fucosidase activity [Cordero 2001] - hematopoietic, epithelial, & mesenchimal cells expressed cell surface protein with alpha-l-fucosidase activity [Cordero 2001]; also found in membrane-assoc fraction from brain [Hopfer 1990] and plasma membrane of sperm [Alhadeff 1999] - purified and kinetically characterized [Khunsook 2003]

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FUCASE2ly	3	Johnson SW, Alhadeff JA.	Mammalian alpha-L-fucosidases	Comp Biochem Physiol B	1991	1769200	- keratan sulfate chains are released extracellularly by proteolysis; mammalian cells do not have endoglycosidases activity towards keratan sulfate, so chains are broken down sequentially by exoglycosidases and sulfatases in the lysosome [Winchester 19996]; NOTE: extracellular proteolysis is not modeled here, chains are assumed to be linked to only ASn-X-Ser/Thr peptide - the linkage region of keratan sulfate type I is probably degraded by the same set of enzymes used for N-glycan degradation [Winchester 1996] -glycoconjugates contain L-fucose in various alpha-glycosic linkages (a-1,2, a-1,3, a-1,4, a-1,6) primarily to Gal and GlcNAc; appears that only one a-L-fucosidase is responsible for hydrolysis of all linkages [Johnson 1991] 2517: - a-L-fucosidases have been isolated from human brain, kidney, spleen, placenta, liver, epidermis, leucocytes, cultured fibroblasts, serum, and amniotic fluid (see refs in [Johnson 1991]) and seminal plasma [Khunsook 2002] - summary of kinetic properties and substrate specificities of purified enzymes [Johnson 1991], [Khunsook 2002] - cDNA cloned [Occhiodoro 1989], (partial seq cloned by [Fuk
FUCASE2ly	3	Occhiodoro T, Beckmann KR, Morris CP, Hopwood JJ	Human alpha-L-fucosidase: complete coding sequence from cDNA clones	Biochem Biophys Res Commun	1989	2803312	- keratan sulfate chains are released extracellularly by proteolysis; mammalian cells do not have endoglycosidases activity towards keratan sulfate, so chains are broken down sequentially by exoglycosidases and sulfatases in the lysosome [Winchester 19996]; NOTE: extracellular proteolysis is not modeled here, chains are assumed to be linked to only ASn-X-Ser/Thr peptide - the linkage region of keratan sulfate type I is probably degraded by the same set of enzymes used for N-glycan degradation [Winchester 1996] -glycoconjugates contain L-fucose in various alpha-glycosic linkages (a-1,2, a-1,3, a-1,4, a-1,6) primarily to Gal and GlcNAc; appears that only one a-L-fucosidase is responsible for hydrolysis of all linkages [Johnson 1991] 2517: - a-L-fucosidases have been isolated from human brain, kidney, spleen, placenta, liver, epidermis, leucocytes, cultured fibroblasts, serum, and amniotic fluid (see refs in [Johnson 1991]) and seminal plasma [Khunsook 2002] - summary of kinetic properties and substrate specificities of purified enzymes [Johnson 1991], [Khunsook 2002] - cDNA cloned [Occhiodoro 1989], (partial seq cloned by [Fuk
FUCASE2ly	3	Fukushima H, de Wet JR, O'Brien JS	Molecular cloning of a cDNA for human alpha-L-fucosidase	Proc Natl Acad Sci U S A	1985	2983333	- keratan sulfate chains are released extracellularly by proteolysis; mammalian cells do not have endoglycosidases activity towards keratan sulfate, so chains are broken down sequentially by exoglycosidases and sulfatases in the lysosome [Winchester 19996]; NOTE: extracellular proteolysis is not modeled here, chains are assumed to be linked to only ASn-X-Ser/Thr peptide - the linkage region of keratan sulfate type I is probably degraded by the same set of enzymes used for N-glycan degradation [Winchester 1996] -glycoconjugates contain L-fucose in various alpha-glycosic linkages (a-1,2, a-1,3, a-1,4, a-1,6) primarily to Gal and GlcNAc; appears that only one a-L-fucosidase is responsible for hydrolysis of all linkages [Johnson 1991] 2517: - a-L-fucosidases have been isolated from human brain, kidney, spleen, placenta, liver, epidermis, leucocytes, cultured fibroblasts, serum, and amniotic fluid (see refs in [Johnson 1991]) and seminal plasma [Khunsook 2002] - summary of kinetic properties and substrate specificities of purified enzymes [Johnson 1991], [Khunsook 2002] - cDNA cloned [Occhiodoro 1989], (partial seq cloned by [Fuk
FUCASE2ly	3	Khunsook S, Alhadeff JA, Bean BS	Purification and characterization of human seminal plasma alpha-L-fucosidase.	Mol Hum Reprod	2002	11870229	- keratan sulfate chains are released extracellularly by proteolysis; mammalian cells do not have endoglycosidases activity towards keratan sulfate, so chains are broken down sequentially by exoglycosidases and sulfatases in the lysosome [Winchester 19996]; NOTE: extracellular proteolysis is not modeled here, chains are assumed to be linked to only ASn-X-Ser/Thr peptide - the linkage region of keratan sulfate type I is probably degraded by the same set of enzymes used for N-glycan degradation [Winchester 1996] -glycoconjugates contain L-fucose in various alpha-glycosic linkages (a-1,2, a-1,3, a-1,4, a-1,6) primarily to Gal and GlcNAc; appears that only one a-L-fucosidase is responsible for hydrolysis of all linkages [Johnson 1991] 2517: - a-L-fucosidases have been isolated from human brain, kidney, spleen, placenta, liver, epidermis, leucocytes, cultured fibroblasts, serum, and amniotic fluid (see refs in [Johnson 1991]) and seminal plasma [Khunsook 2002] - summary of kinetic properties and substrate specificities of purified enzymes [Johnson 1991], [Khunsook 2002] - cDNA cloned [Occhiodoro 1989], (partial seq cloned by [Fuk

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
FUMtm	3	Passarella S, Atlante A, Barile M, Quagliariello E.	Anion transport in rat brain mitochondria: fumarate uptake via the dicarboxylate carrier.	Neurochem Res	1987	3587497	- Added by RS/TV - fumarate and malate share a single carrier to enter mitochondria according to Passarella S, Atlante A, Barile M, Quagliariello E. Related Articles, Links Anion transport in rat brain mitochondria: fumarate uptake via the dicarboxylate carrier. Neurochem Res. 1987 Mar;12(3):255-64.
FUMTSULtm	3	Crompton M, Palmieri F, Capano M, Quagliariello E.	The transport of sulphate and sulphite in rat liver mitochondria.		1974	4441366	SLC25A10 (DIC) transporter facilitates the entry of thiosulphate into mitochondria, where rhodanase and thiosulphate reductase are found (PMID:14598172) biochemical evidence found in rat mitochondria MM
FUT12g	3	Larsen RD, Ernst LK, Nair RP, Lowe JB.	Molecular cloning, sequence, and expression of a human GDP-L-fucose:beta-D-galactoside 2-alpha-L-fucosyltransferase cDNA that can form the H blood group antigen.	Proc Natl Acad Sci U S A	1990	2118655	localization: golgi see PMID 2118655 and swiss-prot. specificity: There are two genes (FUT1 and FUT2) which encode galactoside 2-L-fucosyltransferase. They are expressed in a tissue-specific manner with expression restricted to cells of mesodermal or endodermal origin respectively. Creates a soluble precursor oligosaccharide FuC-alpha ((1,2)Galbeta-) called the H antigen which is an essential substrate for the final step in the soluble A and B antigen synthesis pathway. H and Se enzymes fucosylate the same acceptor substrates but exhibit different Km values. sequence and function: PMID 2118655 NJ
FUT31g	3	Kukowska-Latallo JF, Larsen RD, Nair RP, Lowe JB.	A cloned human cDNA determines expression of a mouse stage-specific embryonic antigen and the Lewis blood group alpha(1,3/1,4)fucosyltransferase.	Genes Dev	1990	1977660	localization: Golgi and PMID: 1977660 and PMID: 12493760 specificity: Highly expressed in stomach, colon, small intestine, lung and kidney and to a lesser extent in salivary gland, bladder, uterus and liver. Swiss-Prot: May catalyze alpha-1,3 and alpha-1,4 glycosidic linkages involved in the expression of Vim-2, Lewis A, Lewis B, sialyl Lewis X and Lewis X/SSEA-1 antigens. May be involved in blood group Lewis determination; Lewis-positive (Le(+)) individuals have an active enzyme while Lewis-negative (Le(-)) individuals have an inactive enzyme. For sequence and cloning see PMID: 1977660. Interesting genotyping and allele freq ref: PMID: 12424536 NJ
FUT31g	3	Cakir B, Pankow JS, Salomaa V, Couper D, Morris TL, Brantley KR, Hiller KM, Heiss G, Weston BW.	Distribution of Lewis (FUT3)genotype and allele: frequencies in a biethnic United States population.	Ann Hematol	2002	12424536	localization: Golgi and PMID: 1977660 and PMID: 12493760 specificity: Highly expressed in stomach, colon, small intestine, lung and kidney and to a lesser extent in salivary gland, bladder, uterus and liver. Swiss-Prot: May catalyze alpha-1,3 and alpha-1,4 glycosidic linkages involved in the expression of Vim-2, Lewis A, Lewis B, sialyl Lewis X and Lewis X/SSEA-1 antigens. May be involved in blood group Lewis determination; Lewis-positive (Le(+)) individuals have an active enzyme while Lewis-negative (Le(-)) individuals have an inactive enzyme. For sequence and cloning see PMID: 1977660. Interesting genotyping and allele freq ref: PMID: 12424536 NJ

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
FUT31g	3	Sousa VL, Brito C, Costa T, Lanoix J, Nilsson T, Costa J.	Importance of Cys, Gln, and Tyr from the transmembrane domain of human alpha 3/4 fucosyltransferase III for its localization and sorting in the Golgi of baby hamster kidney cells.	J Biol Chem	2003	12493760	localization: Golgi and PMID: 1977660 and PMID: 12493760 specificity: Highly expressed in stomach, colon, small intestine, lung and kidney and to a lesser extent in salivary gland, bladder, uterus and liver. Swiss-Prot: May catalyze alpha-1,3 and alpha-1,4 glycosidic linkages involved in the expression of Vim-2, Lewis A, Lewis B, sialyl Lewis X and Lewis X/SSEA-1 antigens. May be involved in blood group Lewis determination; Lewis-positive (Le(+)) individuals have an active enzyme while Lewis-negative (Le(-)) individuals have an inactive enzyme. For sequence and cloning see PMID: 1977660. Interesting genotyping and allele freq ref: PMID: 12424536 NJ
FUT91g	3	Kaneko M, Kudo T, Iwasaki H, Ikehara Y, Nishihara S, Nakagawa S, Sasaki K, Shiina T, Inoko H, Saitou N, Narimatsu H.	Alpha1,3-fucosyltransferase IX (Fuc-TIX) is very highly conserved between human and mouse; molecular cloning, characterization and tissue distribution of human Fuc-TIX.	FEBS Lett	1999	10386598	localization: golgi - uniprot specificity: Strongly expressed in forebrain and stomach, lower expression in spleen and peripheral blood leukocytes, and no expression in small intestine, colon, liver, lung, kidney, adrenal cortex or uterus in PMID: 10386598 FUT9 is one of several alpha-3-fucosyltransferases that can catalyze the last step in the biosynthesis of Lewis antigen, the addition of a fucose to precursor polysaccharides. FUT9 synthesizes the LeX oligosaccharide, which is expressed in organ buds progressing in mesenchyma during human embryogenesis.[supplied by OMIM] Transfers a fucose to lacto-N-neotetraose but not to either alpha2,3-sialyl lacto-N-neotetraose or lacto-N-tetraose. Can catalyze the last step in the biosynthesis of Lewis antigen, the addition of a fucose to precursor polysaccharides. NJ
G14T2g	3	Mengle-Gaw L, McCoy-Haman MF, Tiemeier DC	Genomic structure and expression of human beta-1,4-galactosyltransferase	Biochem Biophys Res Commun	1991	1903938	- sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] - scheme for poly-NAcLac extension of core 4 proposed in [Ujita, J Biol Chem 2000] - all have exclusive specificity for UDP-galactose; Golgi apparatus [RefSeq], [UniProt] 2683: - cDNA was isolated [Appert, Biochem Biophys Res Commun 1986], sequence identified [Masri, Biochem Biophys Res Commun 1988], and expressed [Mengle-Gaw, Biochem Biophys Res Commun 1991] - homodimer [UniProt] 8703: - enzyme mainly involved in synthesis of first N-acetyllactosamine unit [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 8704: - synthesizes N-acetyllactosamine in glycolipids and glycoproteins [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 9334: - cDNA was isolated; 37% identity w/ beta-1,4-GalT and 28% with Lymnaea stagnalis beta-1,4-GlcNAcT [Sato, PNAS 1998] B4galt1p expressed in all tissues except brain; B4galt2p heart, muscle, pancreas; B4galt3 and B4galt5 ubiquitously expressed [Lo et al, Glycobiology 1998], [UniProt]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
G14T2g	3	Appert HE, Rutherford TJ, Tarr GE, Wiest JS, Thomford NR, McCorquodale DJ	Isolation of a cDNA coding for human galactosyltransferase	Biochem Biophys Res Commun	1986	3094506	- sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] - scheme for poly-NAcLac extension of core 4 proposed in [Ujita, J Biol Chem 2000] - all have exclusive specificity for UDP-galactose; Golgi apparatus [RefSeq], [UniProt] 2683: - cDNA was isolated [Appert, Biochem Biophys Res Commun 1986], sequence identified [Masri, Biochem Biophys Res Commun 1988], and expressed [Mengle-Gaw, Biochem Biophys Res Commun 1991] - homodimer [UniProt] 8703: - enzyme mainly involved in synthesis of first N-acetyllactosamine unit [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 8704: - synthesizes N-acetyllactosamine in glycolipids and glycoproteins [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 9334: - cDNA was isolated; 37% identity w/ beta-1,4-GalT and 28% with Lymnaea stagnalis beta-1,4-GlcNAcT [Sato, PNAS 1998] B4galt1p expressed in all tissues except brain; B4galt2p heart, muscle, pancreas; B4galt3 and B4galt5 ubiquitously expressed [Lo et al, Glycobiology 1998], [UniProt]
G14T2g	3	Masri KA, Appert HE, Fukuda MN	Identification of the full-length coding sequence for human galactosyltransferase (beta-N-acetylglucosaminide: beta 1,4-galactosyltransferase)	Biochem Biophys Res Commun	1988	3144273	- sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] - scheme for poly-NAcLac extension of core 4 proposed in [Ujita, J Biol Chem 2000] - all have exclusive specificity for UDP-galactose; Golgi apparatus [RefSeq], [UniProt] 2683: - cDNA was isolated [Appert, Biochem Biophys Res Commun 1986], sequence identified [Masri, Biochem Biophys Res Commun 1988], and expressed [Mengle-Gaw, Biochem Biophys Res Commun 1991] - homodimer [UniProt] 8703: - enzyme mainly involved in synthesis of first N-acetyllactosamine unit [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 8704: - synthesizes N-acetyllactosamine in glycolipids and glycoproteins [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 9334: - cDNA was isolated; 37% identity w/ beta-1,4-GalT and 28% with Lymnaea stagnalis beta-1,4-GlcNAcT [Sato, PNAS 1998] B4galt1p expressed in all tissues except brain; B4galt2p heart, muscle, pancreas; B4galt3 and B4galt5 ubiquitously expressed [Lo et al, Glycobiology 1998], [UniProt]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
G14T2g	3	Almeida R, Amado M, David L, Levery SB, Holmes EH, Merkx G, van Kessel AG, Rygaard E, Hassan H, Bennett E, Clausen H	A family of human beta4-galactosyltransferases. Cloning and expression of two novel UDP-galactose:beta-n-acetylglucosamine beta1, 4-galactosyltransferases, beta4Gal-T2 and beta4Gal-T3	J Biol Chem	1997	9405390	- sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] - scheme for poly-NAcLac extension of core 4 proposed in [Ujita, J Biol Chem 2000] - all have exclusive specificity for UDP-galactose; Golgi apparatus [RefSeq], [UniProt] 2683: - cDNA was isolated [Appert, Biochem Biophys Res Commun 1986], sequence identified [Masri, Biochem Biophys Res Commun 1988], and expressed [Mengle-Gaw, Biochem Biophys Res Commun 1991] - homodimer [UniProt] 8703: - enzyme mainly involved in synthesis of first N-acetyllactosamine unit [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 8704: - synthesizes N-acetyllactosamine in glycolipids and glycoproteins [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 9334: - cDNA was isolated; 37% identity w/ beta-1,4-GalT and 28% with Lymnaea stagnalis beta-1,4-GlcNAcT [Sato, PNAS 1998] B4galt1p expressed in all tissues except brain; B4galt2p heart, muscle, pancreas; B4galt3 and B4galt5 ubiquitously expressed [Lo et al, Glycobiology 1998], [UniProt]
G14T2g	3	Sato T, Furukawa K, Bakker H, Van den Eijnden DH, Van Die I.	Molecular cloning of a human cDNA encoding beta-1,4-galactosyltransferase with 37% identity to mammalian UDP-Gal:GlcNAc beta-1,4-galactosyltransferase	Proc Natl Acad Sci U S A	1998	9435216	- sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] - scheme for poly-NAcLac extension of core 4 proposed in [Ujita, J Biol Chem 2000] - all have exclusive specificity for UDP-galactose; Golgi apparatus [RefSeq], [UniProt] 2683: - cDNA was isolated [Appert, Biochem Biophys Res Commun 1986], sequence identified [Masri, Biochem Biophys Res Commun 1988], and expressed [Mengle-Gaw, Biochem Biophys Res Commun 1991] - homodimer [UniProt] 8703: - enzyme mainly involved in synthesis of first N-acetyllactosamine unit [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 8704: - synthesizes N-acetyllactosamine in glycolipids and glycoproteins [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 9334: - cDNA was isolated; 37% identity w/ beta-1,4-GalT and 28% with Lymnaea stagnalis beta-1,4-GlcNAcT [Sato, PNAS 1998] B4galt1p expressed in all tissues except brain; B4galt2p heart, muscle, pancreas; B4galt3 and B4galt5 ubiquitously expressed [Lo et al, Glycobiology 1998], [UniProt]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
G14T2g	3	Funderburgh JL	Keratan sulfate biosynthesis	IUBMB Life	2002	12512857	- sequence of biosynthetic pathway reviewed in [Funderburgh, IUBMB Life2002] - scheme for poly-NAcLac extension of core 4 proposed in [Ujita, J Biol Chem 2000] - all have exclusive specificity for UDP-galactose; Golgi apparatus [RefSeq], [UniProt] 2683: - cDNA was isolated [Appert, Biochem Biophys Res Commun 1986], sequence identified [Masri, Biochem Biophys Res Commun 1988], and expressed [Mengle-Gaw, Biochem Biophys Res Commun 1991] - homodimer [UniProt] 8703: - enzyme mainly involved in synthesis of first N-acetyllactosamine unit [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 8704: - synthesizes N-acetyllactosamine in glycolipids and glycoproteins [RefSeq] - gene was cloned and expressed [Almeida, J Biol Chem 1997] 9334: - cDNA was isolated; 37% identity w/ beta-1,4-GalT and 28% with Lymnaea stagnalis beta-1,4-GlcNAcT [Sato, PNAS 1998] B4galt1p expressed in all tissues except brain; B4galt2p heart, muscle, pancreas; B4galt3 and B4galt5 ubiquitously expressed [Lo et al, Glycobiology 1998], [UniProt]
G14Tg	3	Lo NW, Shaper JH, Pevsner J, Shaper NL.	The expanding beta 4-galactosyltransferase gene family: messages from the databanks	Glycobiology	1998	9597550	membrane-bound glycoproteins that appear to have exclusive specificity for the donor substrate UDP-galactose; all transfer galactose in a beta1,4 linkage to similar acceptor sugars: GlcNAc, Glc, and Xyl. As type II membrane proteins, they have an N-terminal hydrophobic signal sequence that directs the protein to the Golgi apparatus and which then remains uncleaved to function as a transmembrane anchor. By sequence similarity, the beta4GalTs form four groups: beta4GalT1 and beta4GalT2, beta4GalT3 and beta4GalT4, beta4GalT5 and beta4GalT6, and beta4GalT7. B4GALT1 is unique among the beta4GalT genes because it encodes an enzyme that participates both in glycoconjugate and lactose biosynthesis. For the first activity, the enzyme adds galactose to N-acetylglucosamine residues that are either monosaccharides or the nonreducing ends of glycoprotein carbohydrate chains. The two enzymatic forms result from alternate transcription initiation sites and post-translational processing. Two transcripts, which differ only at the 5' end, wit B4GALT3 encodes an enzyme that may be mainly involved in The enzyme encoded by B4GALT2 synthesizes N-acetyllactosa
G14Tg	3	Amado M, Almeida R, Carneiro F, Levery ST, Holmes EH, Nomoto M, Hollingsworth MA, Hassan H, Schwientek T, Nielsen PA, Bennett EP, Clausen H	A family of human beta3-galactosyltransferases	The Journal of Biological Chemistry	1998		membrane-bound glycoproteins that appear to have exclusive specificity for the donor substrate UDP-galactose; all transfer galactose in a beta1,4 linkage to similar acceptor sugars: GlcNAc, Glc, and Xyl. As type II membrane proteins, they have an N-terminal hydrophobic signal sequence that directs the protein to the Golgi apparatus and which then remains uncleaved to function as a transmembrane anchor. By sequence similarity, the beta4GalTs form four groups: beta4GalT1 and beta4GalT2, beta4GalT3 and beta4GalT4, beta4GalT5 and beta4GalT6, and beta4GalT7. B4GALT1 is unique among the beta4GalT genes because it encodes an enzyme that participates both in glycoconjugate and lactose biosynthesis. For the first activity, the enzyme adds galactose to N-acetylglucosamine residues that are either monosaccharides or the nonreducing ends of glycoprotein carbohydrate chains. The two enzymatic forms result from alternate transcription initiation sites and post-translational processing. Two transcripts, which differ only at the 5' end, wit B4GALT3 encodes an enzyme that may be mainly involved in The enzyme encoded by B4GALT2 synthesizes N-acetyllactosa
G3PD1	3	Menaya J, Gonzalez-Manchon C, Parrilla R, Ayuso MS.	Molecular cloning, sequencing and expression of a cDNA encoding a human liver NAD-dependent alpha-glycerol-3-phosphate dehydrogenase.	Biochim Biophys Acta	1995	7772607	cytoplasm - uniprot cloning, seq, biochem act - Menaya ref NJ NCD pointed out that GLYC3P -> DHAP directionality needed for glycogenolysis in the cytosol. Rev direction supported by Salway text and Brisson review (PMID: 11385633).

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
G3PD1	3	Brisson D, Vohl MC, St-Pierre J, Hudson TJ, Gaudet D.	Glycerol: a neglected variable in metabolic processes?	Bioessays	2001	11385633	cytoplasm - uniprot cloning, seq, biochem act - Menaya ref NJ NCD pointed out that GLYC3P -> DHAP directionality needed for glycogenolysis in the cytosol. Rev direction supported by Salway text and Brisson review (PMID: 11385633).
G3PD1	3	Salway, JG	Metabolism at a Glance, 2nd ed		1999		cytoplasm - uniprot cloning, seq, biochem act - Menaya ref NJ NCD pointed out that GLYC3P -> DHAP directionality needed for glycogenolysis in the cytosol. Rev direction supported by Salway text and Brisson review (PMID: 11385633).
G3PD2m	2	Ferrer J, Aoki M, Behn P, Nestorowicz A, Riggs A, Permutt MA.	Mitochondrial glycerol-3-phosphate dehydrogenase. Cloning of an alternatively spliced human islet-cell cDNA, tissue distribution, physical mapping, and identification of a polymorphic genetic marker.	Diabetes	1996	8549872	- Added by RS/TV - mitochondrial according to GeneCards - FAD dependent according to GeneCards - Tissue Expression: human pancreatic islets and other tissues (Ferrer J, Aoki M, Behn P, Nestorowicz A, Riggs A, Permutt MA. Diabetes. 1996 Feb;45(2):262-6.) Compartmentation of FAD/FADH2 in mit matrix, but glyc3p/dhap in cytosol noted by NCD as per Brisson (11385633).
G5SADs	2	Hu CA, Lin WW, Obie C, Valle D	Molecular enzymology of mammalian Delta1-pyrroline-5-carboxylate synthase. Alternative splice donor utilization generates isoforms with different sensitivity to ornithine inhibition	J Biol Chem	1999	10037775	KEGG says "non enzymatic" under R03314 citation describes it as "nonenzymatic equibibrium"
G6PDA	3	Shevchenko V, Hogben M, Ekong R, Parrington J, Lai FA	The human glucosamine-6-phosphate deaminase gene: cDNA cloning and expression, genomic organization and chromosomal localization	Gene	1998	9714720	- cloning and expression of 10007 [Shevchenko, Gene 1998] - shown as irreversible in Figure 6.1 on p74 of Varki - high-energy requiring tissues such as neurona and transporting epithelial cells in kidney and intestine use this enzyme to generate f6p for glycolysis; enzyme is essentially absent from liver where other sources of energy are available [Varki, p. 77-8]
G6PDA	3	Varki, Cummings, Esko, Freeze, Hart, Marth	Essentials of Glycobiology		1999		- cloning and expression of 10007 [Shevchenko, Gene 1998] - shown as irreversible in Figure 6.1 on p74 of Varki - high-energy requiring tissues such as neurona and transporting epithelial cells in kidney and intestine use this enzyme to generate f6p for glycolysis; enzyme is essentially absent from liver where other sources of energy are available [Varki, p. 77-8]
G6PDH1er	3	Barash V, Erlich T, Bashan N.	Microsomal hexose-6-phosphate and 6-phosphogluconate dehydrogenases in extrahepatic tissues: human placenta and pig kidney cortex.	Biochem Int	1990	2156506	9563: - can use NAD or NADP [UniProt] - oxidizes glucose-6-phosphate and glucose, as well as other hexose-6-phosphates [UniProt] -shows activity with other hexose-6-phosphates, especially galactose-6-phosphate [RefSeq] - not found in red cells [RefSeq] - endoplasmic reticulum lumen [UniProt], [Clarke 2003] - cDNA isolated and cloned [Mason 1999] - hexose-6-phosphate dehydrogenase isolated from human placental microsomes [Barash 1990]
G6PDH1er	3	Mason PJ, Stevens D, Diez A, Knight SW, Scopes DA, Vulliamy TJ.	Human hexose-6-phosphate dehydrogenase (glucose 1-dehydrogenase) encoded at 1p36: coding sequence and expression.	Blood Cells Mol Dis	1999	10349511	9563: - can use NAD or NADP [UniProt] - oxidizes glucose-6-phosphate and glucose, as well as other hexose-6-phosphates [UniProt] -shows activity with other hexose-6-phosphates, especially galactose-6-phosphate [RefSeq] - not found in red cells [RefSeq] - endoplasmic reticulum lumen [UniProt], [Clarke 2003] - cDNA isolated and cloned [Mason 1999] - hexose-6-phosphate dehydrogenase isolated from human placental microsomes [Barash 1990]
G6PDH1er	3	Clarke JL, Mason PJ	Murine hexose-6-phosphate dehydrogenase: a bifunctional enzyme with broad substrate specificity and 6-phosphogluconolactonase activity.	Arch Biochem Biophys	2003	12831846	9563: - can use NAD or NADP [UniProt] - oxidizes glucose-6-phosphate and glucose, as well as other hexose-6-phosphates [UniProt] -shows activity with other hexose-6-phosphates, especially galactose-6-phosphate [RefSeq] - not found in red cells [RefSeq] - endoplasmic reticulum lumen [UniProt], [Clarke 2003] - cDNA isolated and cloned [Mason 1999] - hexose-6-phosphate dehydrogenase isolated from human placental microsomes [Barash 1990]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
G6PPer	3	Lei KJ, Shelly LL, Pan CJ, Sidbury JB, Chou JY	Mutations in the glucose-6-phosphatase gene that cause glycogen storage disease type 1a	Science	1993	8211187	- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]
G6PPer	3	Shelly LL, Lei KJ, Pan CJ, Sakata SF, Ruppert S, Schutz G, Chou JY.	Isolation of the gene for murine glucose-6-phosphatase, the enzyme deficient in glycogen storage disease type 1A	J Biol Chem	1993	8407995	- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]
G6PPer	3	Pan CJ, Lei KJ, Annabi B, Hemrika W, Chou JY	Transmembrane topology of glucose-6-phosphatase.	J Biol Chem	1998	9497333	- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]

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G6PPer	3	Arden SD, Zahn T, Steegers S, Webb S, Bergman B, O'Brien RM, Hutton JC.	Molecular cloning of a pancreatic islet-specific glucose-6-phosphatase catalytic subunit-related protein.	Diabetes	1999	10078553	- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]
G6PPer	3	Martin CC, Bischof LJ, Bergman B, Hornbuckle LA, Hilliker C, Frigeri C, Wahl D, Svitek CA, Wong R, Goldman JK, Oeser JK, Lepretre F, Froguel P, O'Brien RM, Hutton JC.	Cloning and characterization of the human and rat islet-specific glucose-6-phosphatase catalytic subunit-related protein (IGRP) genes.	J Biol Chem	2001	11297555	- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]
G6PPer	3	Martin CC, Oeser JK, Svitek CA, Hunter SI, Hutton JC, O'Brien RM	Identification and characterization of a human cDNA and gene encoding a ubiquitously expressed glucose-6-phosphatase catalytic subunit-related protein.	J Mol Endocrinol	2002	12370122	- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]

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G6PPer	3	Guionie O, Clottes E, Stafford K, Burchell A.	Identification and characterisation of a new human glucose-6-phosphatase isoform	FEBS Lett	2003	12965222	- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]
G6PPer	3	Shieh JJ, Pan CJ, Mansfield BC, Chou JY.	A glucose-6-phosphate hydrolase, widely expressed outside the liver, can explain age-dependent resolution of hypoglycemia in glycogen storage disease type Ia.	J Biol Chem	2003	13129915	- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]
G6PPer	3	Shieh JJ, Pan CJ, Mansfield BC, Chou JY	The islet-specific glucose-6-phosphatase-related protein, implicated in diabetes, is a glycoprotein embedded in the endoplasmic reticulum membrane	FEBS Lett	2004	15044018	- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]

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G6PPer	3	Nordlie, R.C. and Sukalski, K.A.	The Enzymes of Biological Membranes		1985		- enzyme is found in liver, kidney, intestine, but NOT muscle [Orten, Human Biochem 1975] 2538: -integral to ER membrane [LocusLink] - may be hydrolase and translocase [UniProt] - active site faces ER lumen [Pan et al, J Biol Chem 1998] - also known as G6Pase alpha [Lei, Science 1993], [Shelly, J Biol Chem 1993] - expressed in liver, kidney, intestine (gluconeogenic tissues) [Nordlie and Sukalski, The Enzymes of Biological Membranes 1985], [Orten, Human Biochem 1975] 57818: - also known as islet-specific glucose-6-phosphatase-related protein (IGRP) -specific to pancreatic B islet cells [Arden, Diabetes 1999], [Martin, J Biol Chem 2001] - active site in ER [Shieh, FEBS Lett 2004] 92579: - also known as G6Pase-related protein - ubiquitously distributed [Martin, J Mol Endocrinol 2002], [Shieh, J Biol Chem 2003], [Guione, FEBS Lett 2003]
G6Pter	3	Gerin I, Veiga-da-Cunha M, Achouri Y, Collet JF, Van Schaftingen E	Sequence of a putative glucose 6-phosphate translocase, mutated in glycogen storage disease type Ib	FEBS Lett	1997	9428641	2542: - encodes a reversible glucose-6-phosphate transporter [van Schaftingen and Gerin, Biochem J 2002] - cloned [Gerin 1997] - highly expressed in human liver, kidney, haematopoietcs prgenitor cells [Ihara 2000] - 20-26% identity to E. coli G6P transporters [Gerin 1997] - ER target sequence [Gerin 1997] - brain/heart/sk muscle isoform transports G6P into ER lumen [Lin 2000]
G6Pter	3	Ihara K, Nomura A, Hikino S, Takada H, Hara T	Quantitative analysis of glucose-6-phosphate translocase gene expression in various human tissues and haematopoietic progenitor cells	J Inherit Metab Dis	2000	11032333	2542: - encodes a reversible glucose-6-phosphate transporter [van Schaftingen and Gerin, Biochem J 2002] - cloned [Gerin 1997] - highly expressed in human liver, kidney, haematopoietcs prgenitor cells [Ihara 2000] - 20-26% identity to E. coli G6P transporters [Gerin 1997] - ER target sequence [Gerin 1997] - brain/heart/sk muscle isoform transports G6P into ER lumen [Lin 2000]
G6Pter	3	Lin B, Pan CJ, Chou JY	Human variant glucose-6-phosphate transporter is active in microsomal transport	Hum Genet	2000	11140953	2542: - encodes a reversible glucose-6-phosphate transporter [van Schaftingen and Gerin, Biochem J 2002] - cloned [Gerin 1997] - highly expressed in human liver, kidney, haematopoietcs prgenitor cells [Ihara 2000] - 20-26% identity to E. coli G6P transporters [Gerin 1997] - ER target sequence [Gerin 1997] - brain/heart/sk muscle isoform transports G6P into ER lumen [Lin 2000]
G6Pter	3	van Schaftingen E, Gerin I	The glucose-6-phopsphatase system	Biochem J	2002	11879177	2542: - encodes a reversible glucose-6-phosphate transporter [van Schaftingen and Gerin, Biochem J 2002] - cloned [Gerin 1997] - highly expressed in human liver, kidney, haematopoietcs prgenitor cells [Ihara 2000] - 20-26% identity to E. coli G6P transporters [Gerin 1997] - ER target sequence [Gerin 1997] - brain/heart/sk muscle isoform transports G6P into ER lumen [Lin 2000]
GACMTRc	3	Pegg AE, Nagarajan S, Naficy S, Ganem B	Role of unsaturated derivatives of spermidine as substrates for spermine synthase and in supporting growth of SV-3T3 cells	Biochem J	1991	2001229	Well-established function
GAL3ST11	3	Honke K, Tsuda M, Hirahara Y, Ishii A, Makita A, Wada Y.	Molecular cloning and expression of cDNA encoding human 3'-phosphoadenylylsulfate:galactosylceramide 3'-sulfotransferase.	J Biol Chem	1997	9030544	Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot Sulfonation, an important step in the metabolism of many drugs, xenobiotics, hormones, and neurotransmitters, is catalyzed by sulfotransferases. The product of this gene is galactosylceramide sulfotransferase which catalyzes the conversion between 3'-phosphoadenylylsulfate + a galactosylceramide to adenosine 3',5'-bisphosphate + galactosylceramide sulfate. Activity of this sulfotransferase is enhanced in renal cell carcinoma. NJ

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GALASE1ly	3	Glossl J, Truppe W, Kresse H	Purification and properties of N-acetylgalactosamine 6-sulphate sulphatase from human placenta	Biochem J	1979	39554	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]
GALASE1ly	3	Tomatsu S, Fukuda S, Masue M, Sukegawa K, Fukao T, Yamagishi A, Hori T, Iwata H, Ogawa T, Nakashima Y, et al	Morquio disease: isolation, characterization and expression of full-length cDNA for human N-acetylgalactosamine-6-sulfate sulfatase	Biochem Biophys Res Commun	1991	1755850	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]
GALASE1ly	3	Masue M, Sukegawa K, Orii T, Hashimoto T	N-acetylgalactosamine-6-sulfate sulfatase in human placenta: purification and characteristics	J Biochem (Tokyo)	1991	1794986	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]

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GALASE1ly	3	Bielicki J, Hopwood JJ	Human liver N-acetylgalactosamine 6-sulphatase. Purification and characterization	Biochem J	1991	1953646	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]
GALASE1ly	3	Yamamoto Y, Hake CA, Martin BM, Kretz KA, Ahern-Rindell AJ, Naylor SL, Mudd M, O'Brien JS	Isolation, characterization, and mapping of a human acid beta-galactosidase cDNA	DNA Cell Biol	1990	2111707	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]
GALASE1ly	3	Oshima A, Tsuji A, Nagao Y, Sakuraba H, Suzuki Y.	Cloning, sequencing, and expression of cDNA for human beta-galactosidase.	Biochem Biophys Res Commun	1988	3143362	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]

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GALASE1ly	3	Lim CT, Horwitz AL	Purification and properties of human N-acetylgalactosamine-6-sulfate sulfatase	Biochim Biophys Acta	1981	7213753	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]
GALASE1ly	3	Bielicki J, Fuller M, Guo XH, Morris CP, Hopewood JJ, Anson DS	Expression, purification and characterization of recombinant human N-acetylgalactosamine-6-sulphatase	Biochem J	1995	7575473	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]
GALASE1ly	3	Bonten E, van der Spoel A, Fornerod M, Grosveld G, d'Azzo A	Characterization of human lysosomal neuraminidase defines the molecular basis of the metabolic storage disorder sialidosis.	Genes Dev	1996	8985184	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]

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GALASE1ly	3	Milner CM, Smith SV, Carrillo MB, Taylor GL, Hollinshead M, Campbell RD	Identification of a sialidase encoded in the human major histocompatibility complex	J Biol Chem	1997	9020182	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]
GALASE1ly	3	Pshezhetsky AV, Richard C, Michaud L, Igdoura S, Wang S, Elsliger MA, Qu J, Leclerc D, Gravel R, Dallaire L, Potier M	Cloning, expression and chromosomal mapping of human lysosomal sialidase and characterization of mutations in sialidosis	Nat Genet	1997	9054950	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]
GALASE1ly	3	Pshezhetsky AV, Ashmarina M	Lysosomal multienzyme complex: biochemistry, genetics, and molecular pathophysiology.	Prog Nucleic Acid Res Mol Biol	2001	11550799	- two distinct forms of lysosomal complex purified from human placenta: CathA/GAL/SIAL/GALNS and CathA/GAL [Pshezhetsky 2001], [Ostrowska 2003]; average ratio btwn CathA/GAL/SIAL/GALNS and CathA/GAL complexes is 1:10 [Pshezhetsky 2001] 2720: - cleaves terminal beta-1,4- or beta-1,3- linked galactose from GM1-ganglioside, GA1 ganglioside, lactosylceramide, asialofetuin, galactose-containing oligosaccharides, keratan sulfate [Pshezhetsky 2001] - pH optimum is 4.5-4.75 [Pshezhetsky 2001] - most abundant in liver, kidney, skin, blood cells [Pshezhetsky 2001] - cloned and expressed in COS cells [Oshima 1988] - cDNA isolated [Yamamoto 1990] 4758: - hydrolyzes terminal sialic acids of glycolipids and glycopeptides [Pshezhetsky 2001] - pH optimum 4.2 [Pshezhetsky 2001] - cloned an expressed in fibroblasts [Pshezhetsky 1997],[Bonten 1996] and COS-7 cells [Milner 1997] 2588: - purified from human placenta [Glossl 1979], [Lim & Horwitz 1981], [Masue1991] and human liver [Bielicki 1991], recombinant protein purified from CHO cells [Bielicki 1995] - kinetic characterization [Bielicki 1991], [Masue1991], [Bielic - could desulfate acetylgalactosamine 6-sulphate (from chondroitin-6-sulfate) or galactose 6-sulphate glucosamine 6-sulphate residues in keratan sulfate or heparan su - cloned and expressed in deficient fibroblasts [Tomatsu 1991]

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GALC	3	Luzi P, Rafi MA, Wenger DA.	Structure and organization of the human galactocerebrosidase (GALC) gene.	Genomics	1995	7601472	lysosomal - uniprot and luzi ref biochem act + localization by luzi ref Galactosylceramidase (GALC) is a lysosomal enzyme that hydrolyzes several galactolipids. GALC deficiency is associated with Krabbe disease (globoid cell leukodystrophy). The gene is about 60 kb in length and consists of 17 exons. This gene contains ten GC-box-like sequences within the promoter region but no typical TATA box. This feature is also characteristic of other lysosomal protein encoding genes. Highest level of activity in testes compared to brain, kidney, placenta and liver. Can also be found in urine. NJ
GALGT1	3	Nagata Y, Yamashiro S, Yodoi J, Lloyd KO, Shiku H, Furukawa K.	Expression cloning of beta 1,4 N-acetylgalactosaminyltransferase cDNAs that determine the expression of GM2 and GD2 gangliosides.	J Biol Chem	1992	1601877	Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot Involved in the biosynthesis of gangliosides GM2, GD2 and GA2. GM2 and GD2 gangliosides are sialic acid-containing glycosphingolipids expressed in some normal tissues such as brain and in various tumors such as neuroblastomas, astrocytomas, and malignant melanomas. NJ
GALGT1	3	Furukawa K, Soejima H, Niikawa N, Shiku H.	Genomic organization and chromosomal assignment of the human beta1, 4-N-acetylgalactosaminyltransferase gene. Identification of multiple transcription units.	J Biol Chem	1996	8702839	Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot Involved in the biosynthesis of gangliosides GM2, GD2 and GA2. GM2 and GD2 gangliosides are sialic acid-containing glycosphingolipids expressed in some normal tissues such as brain and in various tumors such as neuroblastomas, astrocytomas, and malignant melanomas. NJ
GALNACT1g	3	Uyama T, Kitagawa H, Tamura Ji J, Sugahara K	Molecular cloning and expression of human chondroitin N-acetylgalactosaminyltransferase: the key enzyme for chain initiation and elongation of chondroitin/dermatan sulfate on the protein linkage region tetrasaccharide shared by heparin/heparan sulfate	J Biol Chem	2002	11788602	- Golgi localization [Silbert, IUBMB LIfe 2002] 55454: - exhibits both GalNAcT-I and GalNAcT-II activity, but mainly participates in elongation, not initiation [Sato, J Biol Chem 2003] - gene identified by BLAST; cDNA isolated [Sato, J Biol Chem 2003] - ubiquitous, but most highly expressed in the small intestine, leukocytes, and spleen [Sato, J Biol Chem 2003] 55790: - identified based on sequence homology [Uyama, J Biol Chem 2002], [Gotoh, J Biol Chem 2002] - exhibits both GalNAcT-I and GalNAcT-II activity, but mainly participates in initiation, not elongation [Uyama, J Biol Chem 2002], [Sato, J Biol Chem 2003], [Gotoh, J Biol Chem 2002] - ubiquitously expressed [Uyama, J Biol Chem 2002], but highly expressed in thyroid and placenta [Gotoh, J Biol Chem 2002]
GALNACT1g	3	Gotoh M, Sato T, Akashima T, Iwasaki H, Kameyama A, Mochizuki H, Yada T, Inaba N, Zhang Y, Kikuchi N, Kwon YD, Togayachi A, Kudo T, Nishihara S, Watanabe H, Kimata K, Narimatsu H	Enzymatic synthesis of chondroitin with a novel chondroitin sulfate N-acetylgalactosaminyltransferase that transfers N-acetylgalactosamine to glucuronic acid in initiation and elongation of chondroitin sulfate synthesis	J Biol Chem	2002	12163485	- Golgi localization [Silbert, IUBMB LIfe 2002] 55454: - exhibits both GalNAcT-I and GalNAcT-II activity, but mainly participates in elongation, not initiation [Sato, J Biol Chem 2003] - gene identified by BLAST; cDNA isolated [Sato, J Biol Chem 2003] - ubiquitous, but most highly expressed in the small intestine, leukocytes, and spleen [Sato, J Biol Chem 2003] 55790: - identified based on sequence homology [Uyama, J Biol Chem 2002], [Gotoh, J Biol Chem 2002] - exhibits both GalNAcT-I and GalNAcT-II activity, but mainly participates in initiation, not elongation [Uyama, J Biol Chem 2002], [Sato, J Biol Chem 2003], [Gotoh, J Biol Chem 2002] - ubiquitously expressed [Uyama, J Biol Chem 2002], but highly expressed in thyroid and placenta [Gotoh, J Biol Chem 2002]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
GALNACT1g	3	Sato T, Gotoh M, Kiyohara K, Akashima T, Iwasaki H, Kameyama A, Mochizuki H, Yada T, Inaba N, Togayachi A, Kudo T, Asada M, Watanabe H, Imamura T, Kimata K, Narimatsu H	Differential roles of two N-acetylgalactosaminyltransferases, CSGalNAcT-1, and a novel enzyme, CSGalNAcT-2. Initiation and elongation in synthesis of chondroitin sulfate	J Biol Chem	2003	12446672	- Golgi localization [Silbert, IUBMB LIfe 2002] 55454: - exhibits both GalNAcT-I and GalNAcT-II activity, but mainly participates in elongation, not initiation [Sato, J Biol Chem 2003] - gene identified by BLAST; cDNA isolated [Sato, J Biol Chem 2003] - ubiquitous, but most highly expressed in the small intestine, leukocytes, and spleen [Sato, J Biol Chem 2003] 55790: - identified based on sequence homology [Uyama, J Biol Chem 2002], [Gotoh, J Biol Chem 2002] - exhibits both GalNAcT-I and GalNAcT-II activity, but mainly participates in initiation, not elongation [Uyama, J Biol Chem 2002], [Sato, J Biol Chem 2003], [Gotoh, J Biol Chem 2002] - ubiquitously expressed [Uyama, J Biol Chem 2002], but highly expressed in thyroid and placenta [Gotoh, J Biol Chem 2002]
GALNTg	0	Cheng L, Tachibana K, Zhang Y, Guo J, Kahori Tachibana K, Kameyama A, Wang H, Hiruma T, Iwasaki H, Togayachi A, Kudo T, Narimatsu H	Characterization of a novel human UDP-GalNAc transferase, pp-GalNAc-T10	FEBS Lett	2002		Golgi apparatus by catalyzing the transfer of GalNAc to serine and threonine residues on target proteins. They are characterized by an N-terminal transmembrane domain, a stem region, a lumenal catalytic domain containing a GT1 motif and Gal/GalNAc transferase motif, and a C-terminal ricin/lectin-like domain. GalNAc-Ts have different, but overlapping, substrate specificities and patterns of expression. GALNT6p is capable of glycosylating fibronectin peptide in vitro and is expressed in a fibroblast cell line, indicating that it may be involved in the synthesis of oncofetal fibronectin. Transcript variants of GALNT1 utilize alternative polyA signals which have been described in the literature. GALNT9 is expressed specifically in the brain, with highest expression in the cerebellum. GALNT8 has two main isoforms, the larger of which is ubiquitously distributed, and the smaller of which is primarily expressed in th e heart, kidney, liver, and placenta. [White et al. Gene 246 (1-2): 347-356 (2000)] Following expression in insect cells, recombinant GALNT10 sh GALNT7p shows exclusive specificity for partially GalNAc-gly References for tissue distributions:
GALNTg	0	Ten Hagen KG, Hagen FK, Balys MM, Beres TM, Van Wuyckhuyse B, Tabak LA	Cloning and expression of a novel, tissue specifically expressed member of the UDP-GalNAc:polypeptide N-acetylgalactosaminyltransferase family.	J Biol Chem	1998	9765313	Golgi apparatus by catalyzing the transfer of GalNAc to serine and threonine residues on target proteins. They are characterized by an N-terminal transmembrane domain, a stem region, a lumenal catalytic domain containing a GT1 motif and Gal/GalNAc transferase motif, and a C-terminal ricin/lectin-like domain. GalNAc-Ts have different, but overlapping, substrate specificities and patterns of expression. GALNT6p is capable of glycosylating fibronectin peptide in vitro and is expressed in a fibroblast cell line, indicating that it may be involved in the synthesis of oncofetal fibronectin. Transcript variants of GALNT1 utilize alternative polyA signals which have been described in the literature. GALNT9 is expressed specifically in the brain, with highest expression in the cerebellum. GALNT8 has two main isoforms, the larger of which is ubiquitously distributed, and the smaller of which is primarily expressed in th e heart, kidney, liver, and placenta. [White et al. Gene 246 (1-2): 347-356 (2000)] Following expression in insect cells, recombinant GALNT10 sh GALNT7p shows exclusive specificity for partially GalNAc-gly References for tissue distributions:

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GALNTg	0	White KE, Lorenz B, Evans WE, Meitinger T, Strom TM, Econs MJ	Molecular cloning of a novel human UDP-GalNAc:polypeptide N-acetylgalactosaminyltransferase, GalNAc-T8, and analysis as a candidate autosomal dominant hypophosphatemic rickets (ADHR) gene.	Gene	2000	10767557	Golgi apparatus by catalyzing the transfer of GalNAc to serine and threonine residues on target proteins. They are characterized by an N-terminal transmembrane domain, a stem region, a lumenal catalytic domain containing a GT1 motif and Gal/GalNAc transferase motif, and a C-terminal ricin/lectin-like domain. GalNAc-Ts have different, but overlapping, substrate specificities and patterns of expression. GALNT6p is capable of glycosylating fibronectin peptide in vitro and is expressed in a fibroblast cell line, indicating that it may be involved in the synthesis of oncofetal fibronectin. Transcript variants of GALNT1 utilize alternative polyA signals which have been described in the literature. GALNT9 is expressed specifically in the brain, with highest expression in the cerebellum. GALNT8 has two main isoforms, the larger of which is ubiquitously distributed, and the smaller of which is primarily expressed in th e heart, kidney, liver, and placenta. [White et al. Gene 246 (1-2): 347-356 (2000)] Following expression in insect cells, recombinant GALNT10 sh GALNT7p shows exclusive specificity for partially GalNAc-gly References for tissue distributions:
GALNTg	0	Wang H, Tachibana K, Zhang Y, Iwasaki H, Kameyama A, Cheng L, Guo J, Hiruma T, Togayachi A, Kudo T, Kikuchi N, Narimatsu H	Cloning and characterization of a novel UDP-GalNAc:polypeptide N-acetylgalactosaminyltransferase, pp-GalNAc-T14.	Biochem Biophys Res Commun	2003	12507512	Golgi apparatus by catalyzing the transfer of GalNAc to serine and threonine residues on target proteins. They are characterized by an N-terminal transmembrane domain, a stem region, a lumenal catalytic domain containing a GT1 motif and Gal/GalNAc transferase motif, and a C-terminal ricin/lectin-like domain. GalNAc-Ts have different, but overlapping, substrate specificities and patterns of expression. GALNT6p is capable of glycosylating fibronectin peptide in vitro and is expressed in a fibroblast cell line, indicating that it may be involved in the synthesis of oncofetal fibronectin. Transcript variants of GALNT1 utilize alternative polyA signals which have been described in the literature. GALNT9 is expressed specifically in the brain, with highest expression in the cerebellum. GALNT8 has two main isoforms, the larger of which is ubiquitously distributed, and the smaller of which is primarily expressed in th e heart, kidney, liver, and placenta. [White et al. Gene 246 (1-2): 347-356 (2000)] Following expression in insect cells, recombinant GALNT10 sh GALNT7p shows exclusive specificity for partially GalNAc-gly References for tissue distributions:
GALNTg	0	Ten Hagen KG, Fritz TA, Tabak LA.	All in the family: the UDP-GalNAc:polypeptide N-acetylgalactosaminyltransferases	Glycobiology	2003	12634319	Golgi apparatus by catalyzing the transfer of GalNAc to serine and threonine residues on target proteins. They are characterized by an N-terminal transmembrane domain, a stem region, a lumenal catalytic domain containing a GT1 motif and Gal/GalNAc transferase motif, and a C-terminal ricin/lectin-like domain. GalNAc-Ts have different, but overlapping, substrate specificities and patterns of expression. GALNT6p is capable of glycosylating fibronectin peptide in vitro and is expressed in a fibroblast cell line, indicating that it may be involved in the synthesis of oncofetal fibronectin. Transcript variants of GALNT1 utilize alternative polyA signals which have been described in the literature. GALNT9 is expressed specifically in the brain, with highest expression in the cerebellum. GALNT8 has two main isoforms, the larger of which is ubiquitously distributed, and the smaller of which is primarily expressed in th e heart, kidney, liver, and placenta. [White et al. Gene 246 (1-2): 347-356 (2000)] Following expression in insect cells, recombinant GALNT10 sh GALNT7p shows exclusive specificity for partially GalNAc-gly References for tissue distributions:

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GALNTg	0	Cheng L, Tachibana K, Iwasaki H, Kameyama A, Zhang Y, Kubota T, Hiruma T, Tachibana K, Kudo T, Guo JM, Narimatsu H	Characterization of a novel human UDP-GalNAc transferase, pp-GalNAc-T15	FEBS Lett	2004	15147861	Golgi apparatus by catalyzing the transfer of GalNAc to serine and threonine residues on target proteins. They are characterized by an N-terminal transmembrane domain, a stem region, a lumenal catalytic domain containing a GT1 motif and Gal/GalNAc transferase motif, and a C-terminal ricin/lectin-like domain. GalNAc-Ts have different, but overlapping, substrate specificities and patterns of expression. GALNT6p is capable of glycosylating fibronectin peptide in vitro and is expressed in a fibroblast cell line, indicating that it may be involved in the synthesis of oncofetal fibronectin. Transcript variants of GALNT1 utilize alternative polyA signals which have been described in the literature. GALNT9 is expressed specifically in the brain, with highest expression in the cerebellum. GALNT8 has two main isoforms, the larger of which is ubiquitously distributed, and the smaller of which is primarily expressed in th e heart, kidney, liver, and placenta. [White et al. Gene 246 (1-2): 347-356 (2000)] Following expression in insect cells, recombinant GALNT10 sh GALNT7p shows exclusive specificity for partially GalNAc-gly References for tissue distributions:
GALOR	2	Leslie ND	Insights into the pathogenesis of galactosemia.	Annu Rev Nutr	2003	12704219	- galactitol production is a dead end pathway, and the product is poorly diffusable [Leslie, Annu Rev Nutr 2003], elevated levels can cause cataracts [Champe, Biochemistry 2005] - enzyme is present in liver, kidney, retina, lens, nerve tissue, seminal vesicles, and ovaries [Champe, Biochemistry 2005] - reaction is physiologically unimportant unless galactose levels are high [Champe, Biochemistry 2005]
GALT	0	Leslie ND, Immerman EB, Flach JE, Florez M, Fridovich-Keil JL, Elsas LJ	The human galactose-1-phosphate uridyltransferase gene	Genomics	1992	1427861	- UTP-hexose-1-phosphate uridylyltransferase activity [Leslie et al, Genomics 1992]
GALT2g	3	Bai X, Zhou D, Brown JR, Crawford BE, Hennet T, Esko JD	Biosynthesis of the linkage region of glycosaminoglycans: cloning and activity of galactosyltransferase II, the sixth member of the beta 1,3-galactosyltransferase family (beta 3GalT6)	J Biol Chem	2001	11551958	- Golgi lumen [Bai, J Biol Chem 2001], [Silbert, IUBMB LIfe 2002] - identification of galactosyltransferase II activity [Bai, J Biol Chem 2001] - cDNA was cloned and expressed [Bai, J Biol Chem 2001] - broad expression in human tissues [Bai, J Biol Chem 2001]
GALTg	3	Almeida R, Levery SB, Mandel U, Kresse H, Schwientek T, Bennett EP, Clausen H	Cloning and expression of a proteoglycan UDP-galactose:beta-xylose beta1,4-galactosyltransferase I. A seventh member of the human beta4-galactosyltransferase gene family	J Biol Chem	1999	1073568	- attaches first Gal in tetrasaccharide linkage region of chondroitin / heparan sulfate [RefSeq] - Golgi membrane protein [UniProt], [Silbert, IUBMB Life 2002] - High expression in heart, pancreas and liver, medium in placenta and kidney, low in brain, skeletal muscle and lung [UniProt] - cDNA identified by BLAST and expressed [Okajima, J Biol Chem 1999], [Almeida, J Biol Chem 1999] - gene has 38% homology to C. elegans SQV-3 [Okajima, J Biol Chem 1999]
GALTg	3	Okajima T, Yoshida K, Kondo T, Furukawa K	Human homolog of Caenorhabditis elegans sqv-3 gene is galactosyltransferase I involved in the biosynthesis of the glycosaminoglycan-protein linkage region of proteoglycans	J Biol Chem	1999	10438455	- attaches first Gal in tetrasaccharide linkage region of chondroitin / heparan sulfate [RefSeq] - Golgi membrane protein [UniProt], [Silbert, IUBMB Life 2002] - High expression in heart, pancreas and liver, medium in placenta and kidney, low in brain, skeletal muscle and lung [UniProt] - cDNA identified by BLAST and expressed [Okajima, J Biol Chem 1999], [Almeida, J Biol Chem 1999] - gene has 38% homology to C. elegans SQV-3 [Okajima, J Biol Chem 1999]
GAO1	3	Kanamori A, Nakayama J, Fukuda MN, Stallcup WB, Sasaki K, Fukuda M, Hirabayashi Y	Expression cloning and characterization of a cDNA encoding a novel membrane protein required for the formation of O-acetylated ganglioside: A putative acetyl-CoA-transporter	Proc Natl Acad Sci	1997		activity in cytoplasm - refseq expression in ER (probable) Kanamori et al ref - assumed outer membrane of ER seq homology initially determined as transporter, also found to have O-acetyl activity on gangliosides. NJ
GASNASE3ly	3	Ikonen E, Baumann M, Gron K, Syvanen AC, Enomaa N, Halila R, Aula P, Peltonen L	Aspartylglucosaminuria: cDNA encoding human aspartylglucosaminidase and the missense mutation causing the disease.	EMBO J	1991	1703489	175: - isolated from human placenta [Fisher 1990], fetal liver cDNA library [Ikonen 1991] - cloned [Fisher 1990], [Ikonen 1991] - expressed in monkey COS-1 cells [Ikonen 1991] - requires a free a-amino and carboxyl group on the Asn and any L-fucose at the 6-position of GlcNAc must be removed prior to GlcNAc-Asn hydrolysis [Fisher 1990] - 86% of residues identical in overlapping regions of human and mouse cDNA clones [Fisher 1990] - native protein is a heterodimer; single gene encodes both heavy and light chains [Tollersrud 1994]

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GASNASE3ly	3	Fisher KJ, Tollersrud OK, Aronson NN Jr	Cloning and sequence analysis of a cDNA for human glycosylasparaginase. A single gene encodes the subunits of this lysosomal amidase	FEBS Lett	1990	2401370	175: - isolated from human placenta [Fisher 1990], fetal liver cDNA library [Ikonen 1991] - cloned [Fisher 1990], [Ikonen 1991] - expressed in monkey COS-1 cells [Ikonen 1991] - requires a free a-amino and carboxyl group on the Asn and any L-fucose at the 6-position of GlcNAc must be removed prior to GlcNAc-Asn hydrolysis [Fisher 1990] - 86% of residues identical in overlapping regions of human and mouse cDNA clones [Fisher 1990] - native protein is a heterodimer; single gene encodes both heavy and light chains [Tollersrud 1994]
GASNASE3ly	3	Tollersrud OK, Heiskanen T, Peltonen L	Human leucocyte glycosylasparaginase is an alpha/beta-heterodimer of 19 kDa alpha-subunit and 17 and 18 kDa beta-subunit.	Biochem J	1994	8002961	175: - isolated from human placenta [Fisher 1990], fetal liver cDNA library [Ikonen 1991] - cloned [Fisher 1990], [Ikonen 1991] - expressed in monkey COS-1 cells [Ikonen 1991] - requires a free a-amino and carboxyl group on the Asn and any L-fucose at the 6-position of GlcNAc must be removed prior to GlcNAc-Asn hydrolysis [Fisher 1990] - 86% of residues identical in overlapping regions of human and mouse cDNA clones [Fisher 1990] - native protein is a heterodimer; single gene encodes both heavy and light chains [Tollersrud 1994]
GBA	0	Schmitz M, Alfalah M, Aerts JM, Naim HY, Zimmer KP.	Impaired trafficking of mutants of lysosomal glucocerebrosidase in Gaucher's disease.	Int J Biochem Cell Biol	2005	15982918	This gene encodes a lysosomal membrane protein that cleaves the beta-glucosidic linkage of glycosylceramide, an intermediate in glycolipid metabolism. Mutations in this gene cause Gaucher disease, a lysosomal storage disease characterized by an accumulation of glucocerebrosides. A related pseudogene is approximately 12 kb downstream of this gene on chromosome 1. Alternative splicing results in multiple transcript variants encoding the same protein. NJ
GBAl	3	Orvisky E, Park JK, Parker A, Walker JM, Martin BM, Stubblefield BK, Uyama E, Tayebi N, Sidransky E.	The identification of eight novel glucocerebrosidase (GBA) mutations in patients with Gaucher disease.	Hum Mutat	2002	11933202	lysosomal - uniprot cytosolic form also exists - berrin ref: Substrate (aglycone) specificity of human cytosolic beta-glucosidase. Biochem J. 2003 Jul 1;373(Pt 1):41-8. This gene encodes a lysosomal membrane protein that cleaves the beta-glucosidic linkage of glycosylceramide, an intermediate in glycolipid metabolism. Mutations in this gene cause Gaucher disease, a lysosomal storage disease characterized by an accumulation of glucocerebrosides. A related pseudogene is approximately 12 kb downstream of this gene on chromosome 1. Alternative splicing results in multiple transcript variants encoding the same protein. NJ
GBAl	3	Berrin JG, Czjzek M, Kroon PA, McLauchlan WR, Puigserver A, Williamson G, Juge N.	Substrate (aglycone) specificity of human cytosolic beta-glucosidase.	Biochem J	2003	12667141	lysosomal - uniprot cytosolic form also exists - berrin ref: Substrate (aglycone) specificity of human cytosolic beta-glucosidase. Biochem J. 2003 Jul 1;373(Pt 1):41-8. This gene encodes a lysosomal membrane protein that cleaves the beta-glucosidic linkage of glycosylceramide, an intermediate in glycolipid metabolism. Mutations in this gene cause Gaucher disease, a lysosomal storage disease characterized by an accumulation of glucocerebrosides. A related pseudogene is approximately 12 kb downstream of this gene on chromosome 1. Alternative splicing results in multiple transcript variants encoding the same protein. NJ
GBAl	3	Tettamanti G, Bassi R, Viani P, Riboni L	Salvage pathways in glycosphingolipid metabolism	Biochimie	2003		lysosomal - uniprot cytosolic form also exists - berrin ref: Substrate (aglycone) specificity of human cytosolic beta-glucosidase. Biochem J. 2003 Jul 1;373(Pt 1):41-8. This gene encodes a lysosomal membrane protein that cleaves the beta-glucosidic linkage of glycosylceramide, an intermediate in glycolipid metabolism. Mutations in this gene cause Gaucher disease, a lysosomal storage disease characterized by an accumulation of glucocerebrosides. A related pseudogene is approximately 12 kb downstream of this gene on chromosome 1. Alternative splicing results in multiple transcript variants encoding the same protein. NJ

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GBGT1	3	Xu H, Storch T, Yu M, Elliott SP, Haslam DB.	haracterization of the human Forssman synthetase gene. An evolving association between glycolipid synthesis and host-microbial interactions.	J Biol Chem	1999	10506200	Golgi - lumen side - see ref Kolter and Sandhoff; also noted in uniprot no direct biochem evidence - see xu ref This gene encodes a member of the histo-blood group ABO gene family that encodes glycosyltransferases with related but distinct substrate specificity. This protein plays a role in synthesizing Forssman glycolipid (FG), a member of the globoseries glycolipid family. Human cells do not normally produce FG but produce the precursor glycolipids globotriaosylceramide and globoside. This protein may be involved in the tropism and binding of pathogenic organisms. NJ
GCALDD	2	Brent J	Current management of ethylene glycol poisoning	Drugs	2001	11434452	- reaction catalyzed by aldehyde dehydrogenase [Brent, Drugs 2001] - reaction is essentially irreversible in vivo [Poore 1998] - several liver enzymes can catalyze this rxn (see refs in [Poore 1998]) - this rxn is primarily catalyzed by the mitochondrial isozyme, particularly at low levels of acetaldehyde (see refs in [Poore 1998])
GCCcm	3	Lee HH, Kim do J, Ahn HJ, Ha JY, Suh SW.	Crystal structure of T-protein of the glycine cleavage system. Cofactor binding, insights into H-protein recognition, and molecular basis for understanding nonketotic hyperglycinemia.		2004	15355973	0
GCNTg	3	Bierhuizen MF, Mattei MG, Fukuda M.	Expression of the developmental I antigen by a cloned human cDNA encoding a member of a beta-1,6-N-acetylglucosaminyltransferase gene family.	Genes Dev	1993	8449405	specificity: In the adult, highly expressed in prostate and to a lesser extent in small intestine and colon. Barely detected in heart, brain, kidkey and pancreas. No expression in placenta, lung, liver, skeletal muscle, spleen, thymus, testis, ovary and peripheral blood leukocytes. In fetus, highly expressed in brain and to a lesser extent in lung and kidney. Barely detected in liver. The enzyme encoded by this gene is responsible for the formation of the blood group I antigen. The i and I antigens are determined by linear and branched poly-N-acetyllactosaminoglycans, respectively. During embryonic development in human erythrocytes, the fetal i antigen is replaced by the adult I antigen as the result of the appearance of a beta-1,6-N-acetylglucosaminyltransferase, the I-branching enzyme. This gene encodes the I-branching enzyme that converts the linear form into the branched form. Defects in this gene have been associated with adult i blood group phenotype. Several transcript variants encoding different isoforms have been described. Branching enzyme that converts linear into branched poly-N-ac Introduces the blood group I antigen during embryonic develop
GCNTg	3	Pras E, Raz J, Yahalom V, Frydman M, Garzozi HJ, Pras E, Hejtmancik JF.	A nonsense mutation in the glucosaminyl (N-acetyl) transferase 2 gene (GCNT2): association with autosomal recessive congenital cataracts.	Invest Ophthalmol Vis Sci	2004	15161861	specificity: In the adult, highly expressed in prostate and to a lesser extent in small intestine and colon. Barely detected in heart, brain, kidkey and pancreas. No expression in placenta, lung, liver, skeletal muscle, spleen, thymus, testis, ovary and peripheral blood leukocytes. In fetus, highly expressed in brain and to a lesser extent in lung and kidney. Barely detected in liver. The enzyme encoded by this gene is responsible for the formation of the blood group I antigen. The i and I antigens are determined by linear and branched poly-N-acetyllactosaminoglycans, respectively. During embryonic development in human erythrocytes, the fetal i antigen is replaced by the adult I antigen as the result of the appearance of a beta-1,6-N-acetylglucosaminyltransferase, the I-branching enzyme. This gene encodes the I-branching enzyme that converts the linear form into the branched form. Defects in this gene have been associated with adult i blood group phenotype. Several transcript variants encoding different isoforms have been described. Branching enzyme that converts linear into branched poly-N-ac Introduces the blood group I antigen during embryonic develop
GDPFUCtg	3	Luhn K, Wild MK, Eckhardt M, Gerardy-Schahn R, Vestweber D	The gene defective in leukocyte adhesion deficiency II encodes a putative GDP-fucose transporter	Nat Genet	2001	11326279	- isolated; restored fucosylation in mutant cells [Lubke 2001] - 55% identity to C. elegans ortholog [Luhn 2001] - cloned [Luhn 2001] - Golgi [Luhn 2001]
GDPFUCtg	3	Lubke T, Marquardt T, Etzioni A, Hartmann E, von Figura K, Korner C	Complementation cloning identifies CDG-IIc, a new type of congenital disorders of glycosylation, as a GDP-fucose transporter deficiency	Nat Genet	2001	11326280	- isolated; restored fucosylation in mutant cells [Lubke 2001] - 55% identity to C. elegans ortholog [Luhn 2001] - cloned [Luhn 2001] - Golgi [Luhn 2001]
GF6PTA	3	McKnight GL, Mudri SL, Mathewes SL, Traxinger RR, Marshall S, Sheppard PO, O'Hara PJ.	Molecular cloning, cDNA sequence, and bacterial expression of human glutamine:fructose-6-phosphate amidotransferase.		1992	1460020	- shown as irreversible in Devlin p. 672, 676 and Orten p. 243, Varki p. 74 [however, transaminase reactions are typically reversible] - sequence data - GFPT1 and GFPT2 - biochemical data - GFPT1 -cytosol based on GeneCards

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GF6PTA	3	Oki T, Yamazaki K, Kuromitsu J, Okada M, Tanaka I.	cDNA cloning and mapping of a novel subtype of glutamine:fructose-6-phosphate amidotransferase (GFAT2) in human and mouse.		1999	10198162	- shown as irreversible in Devlin p. 672, 676 and Orten p. 243, Varki p. 74 [however, transaminase reactions are typically reversible] - sequence data - GFPT1 and GFPT2 - biochemical data - GFPT1 -cytosol based on GeneCards
GF6PTA	3	Broschat KO, Gorka C, Page JD, Martin-Berger CL, Davies MS, Huang Hc HC, Gulve EA, Salsgiver WJ, Kasten TP.	Kinetic characterization of human glutamine-fructose-6-phosphate amidotransferase I: potent feedback inhibition by glucosamine 6-phosphate.		2002	11842094	- shown as irreversible in Devlin p. 672, 676 and Orten p. 243, Varki p. 74 [however, transaminase reactions are typically reversible] - sequence data - GFPT1 and GFPT2 - biochemical data - GFPT1 -cytosol based on GeneCards
GGH-7THFl	3	Chandler CJ, Wang TT, Halsted CH.	Pteroylpolyglutamate hydrolase from human jejunal brush borders. Purification and characterization.	J Biol Chem	1986	2867095	IT
GGH-7THFl	3	Rhee MS, Lindau-Shepard B, Chave KJ, Galivan J, Ryan TJ.	Characterization of human cellular gamma-glutamyl hydrolase.	Mol Pharmacol	1998	9614206	IT
GGLUCT	3	York MJ, Crossley MJ, Hyslop SJ, Fisher ML, Kuchel PW	gamma-Glutamylcyclotransferase: inhibition by D-beta-aminoglutaryl-L-alanine and analysis of the solvent kinetic isotope effect	Eur J Biochem	1989	2570694	also may work with other amino acids probably the reaction is standard, although there is little information about the gene
GGNG	3	Barbetti F, Rocchi M, Bossolasco M, Cordera R, Sbraccia P, Finelli P, Consalez GG.	The human skeletal muscle glycogenin gene: cDNA, tissue expression and chromosomal localization	Biochem Biophys Res Commun	1996	8602861	- glycogenin self glucosylates, forming a primer for glycogen synthesis [Devlin, Texbook of Biochem, 2001] -two known forms of glycogenin, expressed in different tissues [Mu, J Biol Chem 1997], [Mu, J Biol Chem 1998] -pathway reviewed in Lomako, Biochim Biophys Acta 2004
GGNG	3	Mu J, Roach PJ	Characterization of human glycogenin-2, a self-glucosylating initiator of liver glycogen metabolism	J Biol Chem	1998	9857012	- glycogenin self glucosylates, forming a primer for glycogen synthesis [Devlin, Texbook of Biochem, 2001] -two known forms of glycogenin, expressed in different tissues [Mu, J Biol Chem 1997], [Mu, J Biol Chem 1998] -pathway reviewed in Lomako, Biochim Biophys Acta 2004
GGNG	3	Mu J, Skurat AV, Roach PJ	Glycogenin-2, a novel self-glucosylating protein involved in liver glycogen biosynthesis	J Biol Chem	1997		- glycogenin self glucosylates, forming a primer for glycogen synthesis [Devlin, Texbook of Biochem, 2001] -two known forms of glycogenin, expressed in different tissues [Mu, J Biol Chem 1997], [Mu, J Biol Chem 1998] -pathway reviewed in Lomako, Biochim Biophys Acta 2004
GHMT2rm	3	Garrow TA, Brenner AA, Whitehead VM, Chen XN, Duncan RG, Korenberg JR, Shane B.	Cloning of human cDNAs encoding mitochondrial and cytosolic serine hydroxymethyltransferases and chromosomal localization.		1993	8505317	reversible according to Lehninger (4th ed., p.844) - humans have a cytosolic and mitochondrial isoform of serine (glycine) hydroxymethyltransferase [Poore 1998] - experiments w/ CHO cells indicate that the mitochondrial enzyme participates in the conversion of serine to glycine whereas the cytoplasmic enzyme may primarily act in the reverse reaction [Narkewicz 1996] - in the liver, the conversion is largely in the direction of glycine to serine (see refs in [Poore 1998])
GHMT2rm	3	Narkewicz MR, Sauls SD, Tjoa SS, Teng C, Fennessey PV	Evidence for intracellular partitioning of serine and glycine metabolism in Chinese hamster ovary cells	Biochem J	1996	8611185	reversible according to Lehninger (4th ed., p.844) - humans have a cytosolic and mitochondrial isoform of serine (glycine) hydroxymethyltransferase [Poore 1998] - experiments w/ CHO cells indicate that the mitochondrial enzyme participates in the conversion of serine to glycine whereas the cytoplasmic enzyme may primarily act in the reverse reaction [Narkewicz 1996] - in the liver, the conversion is largely in the direction of glycine to serine (see refs in [Poore 1998])
GHMT2rm	3	Poore RE, Hurst CH, Assimos DG, Holmes RP	Pathways of hepatic oxalate synthesis and their regulation	Am J Physiol	1997	9038835	reversible according to Lehninger (4th ed., p.844) - humans have a cytosolic and mitochondrial isoform of serine (glycine) hydroxymethyltransferase [Poore 1998] - experiments w/ CHO cells indicate that the mitochondrial enzyme participates in the conversion of serine to glycine whereas the cytoplasmic enzyme may primarily act in the reverse reaction [Narkewicz 1996] - in the liver, the conversion is largely in the direction of glycine to serine (see refs in [Poore 1998])
GK1	3	Brady WA, Kokoris MS, Fitzgibbon M, Black ME.	Cloning, characterization, and modeling of mouse and human guanylate kinases.	J Biol Chem	1996	8663313	IT

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GLACO	2	Marsh CA	Biosynthesis of D-glucaric acid in mammals: a free-radical mechanism?	Carbohydr Res	1986	3779687	- A cytosolic liver enzyme, with NAD+ as co-substrate, has been found12 to produce D-glucaric acid from D-ghicurono-6,3-lactone. However, studies of the enzyme, purified from rat liver, established that this glucuronolactone dehydrogenase was an aldehyde dehydrogenase of wide specificity [Marsh, Carb Res 1986] - glucaric acid seems to be a dead end metabolite [Marsh, Carb Res 1986]
GLAl	3	Bishop DF, Kornreich R, Desnick RJ	Structural organization of the human alpha-galactosidase A gene: further evidence for the absence of a 3' untranslated region	Proc Natl Acad Sci U S A	1988	2836863	lysosomal - uniprot Look -> great article: [Fabry's disease (alpha-galactosidase-A deficiency): physiopathology, clinical signs, and genetic aspects] J Soc Biol. 2002;196(2):161-73. Review. --> can't read because it's French (consult IT) Lam ref - SNP study w/ pts w/ Fabry's dz NJ - cloned [Bishop 1998] - alpha-galactosidase has been purified from human placenta, [11.] liver cells, [12.] spleen cells, [13.] plasma, [13.] and fibroblasts; [14.] recombinant enzyme has been produced in Escherichia coli bacterial cells,[15.] COS monkey cells, [16.] CHO cells, [17.] baculovirus-infected Sf9 insect cells, [18. and 19.] Pichia pastoris yeast cells,[20.] transduced human bone marrow cells, [21.] (refs are from [Garman 2004])
GLAl	3	Lam CW, Mak YT, Lo YM, Tong SF, To KF, Lai FM.	Molecular genetic analysis of a Chinese patient with Fabry disease.	Chin Med J (Engl)	2000	11775551	lysosomal - uniprot Look -> great article: [Fabry's disease (alpha-galactosidase-A deficiency): physiopathology, clinical signs, and genetic aspects] J Soc Biol. 2002;196(2):161-73. Review. --> can't read because it's French (consult IT) Lam ref - SNP study w/ pts w/ Fabry's dz NJ - cloned [Bishop 1998] - alpha-galactosidase has been purified from human placenta, [11.] liver cells, [12.] spleen cells, [13.] plasma, [13.] and fibroblasts; [14.] recombinant enzyme has been produced in Escherichia coli bacterial cells,[15.] COS monkey cells, [16.] CHO cells, [17.] baculovirus-infected Sf9 insect cells, [18. and 19.] Pichia pastoris yeast cells,[20.] transduced human bone marrow cells, [21.] (refs are from [Garman 2004])
GLAl	3	Ishii S, Nakao S, Minamikawa-Tachino R, Desnick RJ, Fan JQ.	Alternative splicing in the alpha-galactosidase A gene: increased exon inclusion results in the Fabry cardiac phenotype.	Am J Hum Genet	2002	11828341	lysosomal - uniprot Look -> great article: [Fabry's disease (alpha-galactosidase-A deficiency): physiopathology, clinical signs, and genetic aspects] J Soc Biol. 2002;196(2):161-73. Review. --> can't read because it's French (consult IT) Lam ref - SNP study w/ pts w/ Fabry's dz NJ - cloned [Bishop 1998] - alpha-galactosidase has been purified from human placenta, [11.] liver cells, [12.] spleen cells, [13.] plasma, [13.] and fibroblasts; [14.] recombinant enzyme has been produced in Escherichia coli bacterial cells,[15.] COS monkey cells, [16.] CHO cells, [17.] baculovirus-infected Sf9 insect cells, [18. and 19.] Pichia pastoris yeast cells,[20.] transduced human bone marrow cells, [21.] (refs are from [Garman 2004])
GLAl	3	Garman SC, Garboczi DN.	The molecular defect leading to Fabry disease: structure of human alpha-galactosidase.	J Mol Biol	2004	15003450	lysosomal - uniprot Look -> great article: [Fabry's disease (alpha-galactosidase-A deficiency): physiopathology, clinical signs, and genetic aspects] J Soc Biol. 2002;196(2):161-73. Review. --> can't read because it's French (consult IT) Lam ref - SNP study w/ pts w/ Fabry's dz NJ - cloned [Bishop 1998] - alpha-galactosidase has been purified from human placenta, [11.] liver cells, [12.] spleen cells, [13.] plasma, [13.] and fibroblasts; [14.] recombinant enzyme has been produced in Escherichia coli bacterial cells,[15.] COS monkey cells, [16.] CHO cells, [17.] baculovirus-infected Sf9 insect cells, [18. and 19.] Pichia pastoris yeast cells,[20.] transduced human bone marrow cells, [21.] (refs are from [Garman 2004])
GLBRAN	3	Thon VJ, Khalil M, Cannon JF	Isolation of human glycogen branching enzyme cDNAs by screening complementation in yeast.	J Biol Chem	1993	8463281	- forms alpha 1,6- glucosidic linkages [RefSeq] -most highly expressed in liver and muscle [RefSeq] - see Devlin p. 650

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GLCAASE1ly	3	Oshima A, Kyle JW, Miller RD, Hoffmann JW, Powell PP, Grubb JH, Sly WS, Tropak M, Guise KS, Gravel RA	Cloning, sequencing, and expression of cDNA for human beta-glucuronidase	Proc Natl Acad Sci U S A	1987	3468507	2990: - isolated and expressed in E. coli [Guise 1985] - isolation from human liver and kinetic characterization [Ho 1985] -cloned and expressed in COS cells [Oshima 1987]
GLCAASE1ly	3	Guise KS, Korneluk RG, Waye J, Lamhonwah AM, Quan F, Palmer R, Ganschow RE, Sly WS, Gravel RA	Isolation and expression in Escherichia coli of a cDNA clone encoding human beta-glucuronidase	Gene	1985	3924735	2990: - isolated and expressed in E. coli [Guise 1985] - isolation from human liver and kinetic characterization [Ho 1985] -cloned and expressed in COS cells [Oshima 1987]
GLCAASE1ly	3	Ho YC, Ho LH, Ho KJ	Human hepatic beta-glucuronidase: an enzyme kinetic study	Enzyme	1985	3987656	2990: - isolated and expressed in E. coli [Guise 1985] - isolation from human liver and kinetic characterization [Ho 1985] -cloned and expressed in COS cells [Oshima 1987]
GLCAE2g	3	Li JP, Gong F, El Darwish K, Jalkanen M, Lindahl U	Characterization of the D-glucuronyl C5-epimerase involved in the biosynthesis of heparin and heparan sulfate	J Biol Chem	2001	11274177	- has > 96% identity to mouse and bovine cDNAs [Li, J Biol Chem 2001] - gene identified by BLAST and cloned [Crawford, J Biol Chem 2001] - reaction is effectively irreversible in vivo [Hagner-McWhirter, J Biol Chem 2004] - ubiquitously expressed [Sugahara, IUBMB Life 2002] - Golgi (based on mouse protein's localization) [UniProt], [Crawford, J Biol Chem 2001]
GLCAE2g	3	Crawford BE, Olson SK, Esko JD, Pinhal MA	Cloning, Golgi localization, and enzyme activity of the full-length heparin/heparan sulfate-glucuronic acid C5-epimerase	J Biol Chem	2001	11279150	- has > 96% identity to mouse and bovine cDNAs [Li, J Biol Chem 2001] - gene identified by BLAST and cloned [Crawford, J Biol Chem 2001] - reaction is effectively irreversible in vivo [Hagner-McWhirter, J Biol Chem 2004] - ubiquitously expressed [Sugahara, IUBMB Life 2002] - Golgi (based on mouse protein's localization) [UniProt], [Crawford, J Biol Chem 2001]
GLCAE2g	3	Hagner-McWhirter A, Li JP, Oscarson S, Lindahl U	Irreversible glucuronyl C5-epimerization in the biosynthesis of heparan sulfate.	J Biol Chem	2004	14718527	- has > 96% identity to mouse and bovine cDNAs [Li, J Biol Chem 2001] - gene identified by BLAST and cloned [Crawford, J Biol Chem 2001] - reaction is effectively irreversible in vivo [Hagner-McWhirter, J Biol Chem 2004] - ubiquitously expressed [Sugahara, IUBMB Life 2002] - Golgi (based on mouse protein's localization) [UniProt], [Crawford, J Biol Chem 2001]
GLCAT2g	3	Kitagawa H, Uyama T, Sugahara K	Molecular cloning and expression of a human chondroitin synthase	J Biol Chem	2001	11514575	- Golgi localization [Silbert, IUBMB LIfe 2002] 54480: - gene was cloned and characterized [Gotoh, J Biol Chem 2002] - has only GlcA-T activity [Gotoh, J Biol Chem 2002] - ubiquitously expressed, most highly expressed in the placenta, small intestine, and pancreas [Gotoh, J Biol Chem 2002] 22856: - gene was cloned and expressed [Kitagawa, J Biol Chem 2001] - has GlcA-T and GalNAc-T activities [Kitagawa, J Biol Chem 2001] - ubiquitously expressed [Kitagawa, J Biol Chem 2001] - requires concomitant expression of chondroitin polymerizing factor (79586) [Kitagawa, J Biol Chem 2003] - 79586 & 22856 form a protein complex [Kitagawa, J Biol Chem 2003] 79586: - gene was cloned and expressed [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed, but highly expressed in the pancreas, ovary, placenta, small intestine, and stomach [Yada, J Biol Chem 2003] 337876: - cloned and characterized [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed [Yada, J Biol Chem 2003]

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GLCAT2g	3	Gotoh M, Yada T, Sato T, Akashima T, Iwasaki H, Mochizuki H, Inaba N, Togayachi A, Kudo T, Watanabe H, Kimata K, Narimatsu H	Molecular cloning and characterization of a novel chondroitin sulfate glucuronyltransferase that transfers glucuronic acid to N-acetylgalactosamine	J Biol Chem	2002	12145278	- Golgi localization [Silbert, IUBMB LIfe 2002] 54480: - gene was cloned and characterized [Gotoh, J Biol Chem 2002] - has only GlcA-T activity [Gotoh, J Biol Chem 2002] - ubiquitously expressed, most highly expressed in the placenta, small intestine, and pancreas [Gotoh, J Biol Chem 2002] 22856: - gene was cloned and expressed [Kitagawa, J Biol Chem 2001] - has GlcA-T and GalNAc-T activities [Kitagawa, J Biol Chem 2001] - ubiquitously expressed [Kitagawa, J Biol Chem 2001] - requires concomitant expression of chondroitin polymerizing factor (79586) [Kitagawa, J Biol Chem 2003] - 79586 & 22856 form a protein complex [Kitagawa, J Biol Chem 2003] 79586: - gene was cloned and expressed [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed, but highly expressed in the pancreas, ovary, placenta, small intestine, and stomach [Yada, J Biol Chem 2003] 337876: - cloned and characterized [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed [Yada, J Biol Chem 2003]
GLCAT2g	3	Kitagawa H, Izumikawa T, Uyama T, Sugahara K	Molecular cloning of a chondroitin polymerizing factor that cooperates with chondroitin synthase for chondroitin polymerization	J Biol Chem	2003	12716890	- Golgi localization [Silbert, IUBMB LIfe 2002] 54480: - gene was cloned and characterized [Gotoh, J Biol Chem 2002] - has only GlcA-T activity [Gotoh, J Biol Chem 2002] - ubiquitously expressed, most highly expressed in the placenta, small intestine, and pancreas [Gotoh, J Biol Chem 2002] 22856: - gene was cloned and expressed [Kitagawa, J Biol Chem 2001] - has GlcA-T and GalNAc-T activities [Kitagawa, J Biol Chem 2001] - ubiquitously expressed [Kitagawa, J Biol Chem 2001] - requires concomitant expression of chondroitin polymerizing factor (79586) [Kitagawa, J Biol Chem 2003] - 79586 & 22856 form a protein complex [Kitagawa, J Biol Chem 2003] 79586: - gene was cloned and expressed [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed, but highly expressed in the pancreas, ovary, placenta, small intestine, and stomach [Yada, J Biol Chem 2003] 337876: - cloned and characterized [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed [Yada, J Biol Chem 2003]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
GLCAT2g	3	Yada T, Gotoh M, Sato T, Shionyu M, Go M, Kaseyama H, Iwasaki H, Kikuchi N, Kwon YD, Togayachi A, Kudo T, Watanabe H, Narimatsu H, Kimata K	Chondroitin sulfate synthase-2. Molecular cloning and characterization of a novel human glycosyltransferase homologous to chondroitin sulfate glucuronyltransferase, which has dual enzymatic activities	J Biol Chem	2003	12761225	- Golgi localization [Silbert, IUBMB LIfe 2002] 54480: - gene was cloned and characterized [Gotoh, J Biol Chem 2002] - has only GlcA-T activity [Gotoh, J Biol Chem 2002] - ubiquitously expressed, most highly expressed in the placenta, small intestine, and pancreas [Gotoh, J Biol Chem 2002] 22856: - gene was cloned and expressed [Kitagawa, J Biol Chem 2001] - has GlcA-T and GalNAc-T activities [Kitagawa, J Biol Chem 2001] - ubiquitously expressed [Kitagawa, J Biol Chem 2001] - requires concomitant expression of chondroitin polymerizing factor (79586) [Kitagawa, J Biol Chem 2003] - 79586 & 22856 form a protein complex [Kitagawa, J Biol Chem 2003] 79586: - gene was cloned and expressed [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed, but highly expressed in the pancreas, ovary, placenta, small intestine, and stomach [Yada, J Biol Chem 2003] 337876: - cloned and characterized [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed [Yada, J Biol Chem 2003]
GLCAT2g	3	Yada T, Sato T, Kaseyama H, Gotoh M, Iwasaki H, Kikuchi N, Kwon YD, Togayachi A, Kudo T, Watanabe H, Narimatsu H, Kimata K	Chondroitin sulfate synthase-3. Molecular cloning and characterization	J Biol Chem	2003	12907687	- Golgi localization [Silbert, IUBMB LIfe 2002] 54480: - gene was cloned and characterized [Gotoh, J Biol Chem 2002] - has only GlcA-T activity [Gotoh, J Biol Chem 2002] - ubiquitously expressed, most highly expressed in the placenta, small intestine, and pancreas [Gotoh, J Biol Chem 2002] 22856: - gene was cloned and expressed [Kitagawa, J Biol Chem 2001] - has GlcA-T and GalNAc-T activities [Kitagawa, J Biol Chem 2001] - ubiquitously expressed [Kitagawa, J Biol Chem 2001] - requires concomitant expression of chondroitin polymerizing factor (79586) [Kitagawa, J Biol Chem 2003] - 79586 & 22856 form a protein complex [Kitagawa, J Biol Chem 2003] 79586: - gene was cloned and expressed [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed, but highly expressed in the pancreas, ovary, placenta, small intestine, and stomach [Yada, J Biol Chem 2003] 337876: - cloned and characterized [Yada, J Biol Chem 2003] - has GlcA-T and GalNAc-T activities [Yada, J Biol Chem 2003] - ubiquitously expressed [Yada, J Biol Chem 2003]
GLCAT6g	3	Ahn J, Ludecke HJ, Lindow S, Horton WA, Lee B, Wagner MJ, Horsthemke B, Wells DE	Cloning of the putative tumour suppressor gene for hereditary multiple exostoses (EXT1)	Nat Genet	1995	7550340	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt]

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GLCAT6g	3	Stickens D, Clines G, Burbee D, Ramos P, Thomas S, Hogue D, Hecht JT, Lovett M, Evans GA	The EXT2 multiple exostoses gene defines a family of putative tumour suppressor genes	Nat Genet	1996	8782816	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt]
GLCAT6g	3	McCormick C, Leduc Y, Martindale D, Mattison K, Esford LE, Dyer AP, Tufaro F	The putative tumour suppressor EXT1 alters the expression of cell-surface heparan sulfate	Nat Genet	1998	9620772	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt]
GLCAT6g	3	Lind T, Tufaro F, McCormick C, Lindahl U, Lidholt K	The putative tumor suppressors EXT1 and EXT2 are glycosyltransferases required for the biosynthesis of heparan sulfate	J Biol Chem	1998	9756849	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt]
GLCAT6g	3	McCormick C, Duncan G, Goutsos KT, Tufaro F	The putative tumor suppressors EXT1 and EXT2 form a stable complex that accumulates in the Golgi apparatus and catalyzes the synthesis of heparan sulfate	Proc Natl Acad Sci U S A	2000	10639137	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt]
GLCAT6g	3	Kobayashi S, Morimoto K, Shimizu T, Takahashi M, Kurosawa H, Shirasawa T	Association of EXT1 and EXT2, hereditary multiple exostoses gene products, in Golgi apparatus	Biochem Biophys Res Commun	2000	10679296	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt]

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GLCATg	3	Kitagawa H, Tone Y, Tamura J, Neumann KW, Ogawa T, Oka S, Kawasaki T, Sugahara K	Molecular cloning and expression of glucuronyltransferase I involved in the biosynthesis of the glycosaminoglycan-protein linkage region of proteoglycans	J Biol Chem	1998	9506957	- Golgi localization [Silbert, IUBMB LIfe 2002] 135152: - transfers a GlcA to terminal Gal residue in linkage region of chondroitin / heparan sulfate [RefSeq], [UniProt] - Golgi [UniProt] - Expressed in the trachea, retina, spinal cord, hippocampus and other brain regions, and, at lower levels, in testis and ovary [UniProt], [Marcus, J Hum Genet 2002] - gene identified via BLAST; 89% homology to rat protein [Marcus, J Hum Genet 2002] 26229: - transfers a GlcA to terminal Gal residue in linkage region of chondroitin / heparan sulfate [RefSeq], [UniProt], [Kitagawa, J Biol Chem 1998] - Golgi [UniProt] - Ubiquitous (but weakly expresed in all tissues examined) [UniProt] - gene was cloned and expressed [Kitagawa, J Biol Chem 1998] - 43% identity to rat protein [Kitagawa, J Biol Chem 1998] 27087: - Golgi [UniProt] - transfers a GlcA to terminal Gal residue in linkage region of chondroitin / heparan sulfate [UniProt] - mainly expressed in the brain [UniProt], [Mitsumoto, Genomics 2000] - cDNA was isolated [Mitsumoto, Genomics 2000] - 98% identical to rat protein [Mitsumoto, Genomics 2000]
GLCATg	3	Mitsumoto Y, Oka S, Sakuma H, Inazawa J, Kawasaki T	Cloning and chromosomal mapping of human glucuronyltransferase involved in biosynthesis of the HNK-1 carbohydrate epitope	Genomics	2000	10783264	- Golgi localization [Silbert, IUBMB LIfe 2002] 135152: - transfers a GlcA to terminal Gal residue in linkage region of chondroitin / heparan sulfate [RefSeq], [UniProt] - Golgi [UniProt] - Expressed in the trachea, retina, spinal cord, hippocampus and other brain regions, and, at lower levels, in testis and ovary [UniProt], [Marcus, J Hum Genet 2002] - gene identified via BLAST; 89% homology to rat protein [Marcus, J Hum Genet 2002] 26229: - transfers a GlcA to terminal Gal residue in linkage region of chondroitin / heparan sulfate [RefSeq], [UniProt], [Kitagawa, J Biol Chem 1998] - Golgi [UniProt] - Ubiquitous (but weakly expresed in all tissues examined) [UniProt] - gene was cloned and expressed [Kitagawa, J Biol Chem 1998] - 43% identity to rat protein [Kitagawa, J Biol Chem 1998] 27087: - Golgi [UniProt] - transfers a GlcA to terminal Gal residue in linkage region of chondroitin / heparan sulfate [UniProt] - mainly expressed in the brain [UniProt], [Mitsumoto, Genomics 2000] - cDNA was isolated [Mitsumoto, Genomics 2000] - 98% identical to rat protein [Mitsumoto, Genomics 2000]
GLCATg	3	Marcos I, Galan JJ, Borrego S, Antinolo G	Cloning, characterization, and chromosome mapping of the human GlcAT-S gene	J Hum Genet	2002	12522689	- Golgi localization [Silbert, IUBMB LIfe 2002] 135152: - transfers a GlcA to terminal Gal residue in linkage region of chondroitin / heparan sulfate [RefSeq], [UniProt] - Golgi [UniProt] - Expressed in the trachea, retina, spinal cord, hippocampus and other brain regions, and, at lower levels, in testis and ovary [UniProt], [Marcus, J Hum Genet 2002] - gene identified via BLAST; 89% homology to rat protein [Marcus, J Hum Genet 2002] 26229: - transfers a GlcA to terminal Gal residue in linkage region of chondroitin / heparan sulfate [RefSeq], [UniProt], [Kitagawa, J Biol Chem 1998] - Golgi [UniProt] - Ubiquitous (but weakly expresed in all tissues examined) [UniProt] - gene was cloned and expressed [Kitagawa, J Biol Chem 1998] - 43% identity to rat protein [Kitagawa, J Biol Chem 1998] 27087: - Golgi [UniProt] - transfers a GlcA to terminal Gal residue in linkage region of chondroitin / heparan sulfate [UniProt] - mainly expressed in the brain [UniProt], [Mitsumoto, Genomics 2000] - cDNA was isolated [Mitsumoto, Genomics 2000] - 98% identical to rat protein [Mitsumoto, Genomics 2000]

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GLCNACASE1ly	3	Sasaki T, Sukegawa K, Masue M, Fukuda S, Tomatsu S, Orii T	Purification and partial characterization of alpha-N-acetylglucosaminidase from human liver	J Biochem (Tokyo)	1991	1783617	- cleaves terminal N-acetylglucosamine in heparan sulfate [Winchester 1996] 4669: - purified from human liver [Sasaki 1991] - cloned and expressed in CHO cells [Weber 1996]
GLCNACASE1ly	3	Weber B, Blanch L, Clements PR, Scott HS, Hopwood JJ	Cloning and expression of the gene involved in Sanfilippo B syndrome (mucopolysaccharidosis III B)	Hum Mol Genet	1996	8776591	- cleaves terminal N-acetylglucosamine in heparan sulfate [Winchester 1996] 4669: - purified from human liver [Sasaki 1991] - cloned and expressed in CHO cells [Weber 1996]
GLCNACASE1ly	3	Winchester BG	Lysosomal metabolism of glycoconjugates	Subcell Biochem	1996	8993162	- cleaves terminal N-acetylglucosamine in heparan sulfate [Winchester 1996] 4669: - purified from human liver [Sasaki 1991] - cloned and expressed in CHO cells [Weber 1996]
GLCNACDASg	3	Dixon J, Loftus SK, Gladwin AJ, Scambler PJ, Wasmuth JJ, Dixon MJ	Cloning of the human heparan sulfate-N-deacetylase/N-sulfotransferase gene from the Treacher Collins syndrome candidate region at 5q32-q33.1	Genomics	1995	7601448	- N-deacetylase/ N-sulfotransferase acts on a subset of GlcNAc residues in a cluster along the chain [Varki, Glycobiology 1999] - enzyme rapidly deacetylates then sulfates, but some of the deacetylated GlcN residues can escape sulfation [Varki, Glycobiology 1999] 3340: - gene was cloned [Dixon, Genomics 1995], [Humphries, Biochem J 1997] - ubiquitous, but expression most abundant in heart, liver, pancreas [UniProt], [Humphries, Biochem J 1997] - Golgi [UniProt], [Humphries, Biochem J 1997] - N-sulfation and N-deacetylation of glucosamine in heparan sulfate [UniProt] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - NDST1 & NDST2 have similar ratios of deactylase and sulfotransferase activities [Aikawa, J Biol Chem 2001] 8509: - gene was cloned [Humphries, Biochem J 1998] - 94% similar to mouse homolog [Humphries, Biochem J 1998] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - Golgi [UniProt] - ubiquitously expressed [Sugahara, IUBMB Liife 2002] 9348: - gene was cloned and expressed [Aikawa, J Biol Chem 1999] - N-sulfation and N-deacetylation of glucosamine in heparan su - highly expressed in brain, liver, kidney [Aikawa, J Biol Chem
GLCNACDASg	3	Humphries DE, Sullivan BM, Aleixo MD, Stow JL	Localization of human heparan glucosaminyl N-deacetylase/N-sulphotransferase to the trans-Golgi network.	Biochem J	1997	9230113	- N-deacetylase/ N-sulfotransferase acts on a subset of GlcNAc residues in a cluster along the chain [Varki, Glycobiology 1999] - enzyme rapidly deacetylates then sulfates, but some of the deacetylated GlcN residues can escape sulfation [Varki, Glycobiology 1999] 3340: - gene was cloned [Dixon, Genomics 1995], [Humphries, Biochem J 1997] - ubiquitous, but expression most abundant in heart, liver, pancreas [UniProt], [Humphries, Biochem J 1997] - Golgi [UniProt], [Humphries, Biochem J 1997] - N-sulfation and N-deacetylation of glucosamine in heparan sulfate [UniProt] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - NDST1 & NDST2 have similar ratios of deactylase and sulfotransferase activities [Aikawa, J Biol Chem 2001] 8509: - gene was cloned [Humphries, Biochem J 1998] - 94% similar to mouse homolog [Humphries, Biochem J 1998] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - Golgi [UniProt] - ubiquitously expressed [Sugahara, IUBMB Liife 2002] 9348: - gene was cloned and expressed [Aikawa, J Biol Chem 1999] - N-sulfation and N-deacetylation of glucosamine in heparan su - highly expressed in brain, liver, kidney [Aikawa, J Biol Chem

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GLCNACDASg	3	Humphries DE, Lanciotti J, Karlinsky JB	cDNA cloning, genomic organization and chromosomal localization of human heparan glucosaminyl N-deacetylase/N-sulphotransferase-2	Biochem J	1998	9601056	- N-deacetylase/ N-sulfotransferase acts on a subset of GlcNAc residues in a cluster along the chain [Varki, Glycobiology 1999] - enzyme rapidly deacetylates then sulfates, but some of the deacetylated GlcN residues can escape sulfation [Varki, Glycobiology 1999] 3340: - gene was cloned [Dixon, Genomics 1995], [Humphries, Biochem J 1997] - ubiquitous, but expression most abundant in heart, liver, pancreas [UniProt], [Humphries, Biochem J 1997] - Golgi [UniProt], [Humphries, Biochem J 1997] - N-sulfation and N-deacetylation of glucosamine in heparan sulfate [UniProt] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - NDST1 & NDST2 have similar ratios of deactylase and sulfotransferase activities [Aikawa, J Biol Chem 2001] 8509: - gene was cloned [Humphries, Biochem J 1998] - 94% similar to mouse homolog [Humphries, Biochem J 1998] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - Golgi [UniProt] - ubiquitously expressed [Sugahara, IUBMB Liife 2002] 9348: - gene was cloned and expressed [Aikawa, J Biol Chem 1999] - N-sulfation and N-deacetylation of glucosamine in heparan su - highly expressed in brain, liver, kidney [Aikawa, J Biol Chem
GLCNACDASg	3	Aikawa J, Esko JD	Molecular cloning and expression of a third member of the heparan sulfate/heparin GlcNAc N-deacetylase/ N-sulfotransferase family	J Biol Chem	1999	9915799	- N-deacetylase/ N-sulfotransferase acts on a subset of GlcNAc residues in a cluster along the chain [Varki, Glycobiology 1999] - enzyme rapidly deacetylates then sulfates, but some of the deacetylated GlcN residues can escape sulfation [Varki, Glycobiology 1999] 3340: - gene was cloned [Dixon, Genomics 1995], [Humphries, Biochem J 1997] - ubiquitous, but expression most abundant in heart, liver, pancreas [UniProt], [Humphries, Biochem J 1997] - Golgi [UniProt], [Humphries, Biochem J 1997] - N-sulfation and N-deacetylation of glucosamine in heparan sulfate [UniProt] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - NDST1 & NDST2 have similar ratios of deactylase and sulfotransferase activities [Aikawa, J Biol Chem 2001] 8509: - gene was cloned [Humphries, Biochem J 1998] - 94% similar to mouse homolog [Humphries, Biochem J 1998] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - Golgi [UniProt] - ubiquitously expressed [Sugahara, IUBMB Liife 2002] 9348: - gene was cloned and expressed [Aikawa, J Biol Chem 1999] - N-sulfation and N-deacetylation of glucosamine in heparan su - highly expressed in brain, liver, kidney [Aikawa, J Biol Chem
GLCNACDASg	3	Aikawa J, Grobe K, Tsujimoto M, Esko JD	Multiple isozymes of heparan sulfate/heparin GlcNAc N-deacetylase/GlcN N-sulfotransferase. Structure and activity of the fourth member, NDST4	J Biol Chem	2001	11087757	- N-deacetylase/ N-sulfotransferase acts on a subset of GlcNAc residues in a cluster along the chain [Varki, Glycobiology 1999] - enzyme rapidly deacetylates then sulfates, but some of the deacetylated GlcN residues can escape sulfation [Varki, Glycobiology 1999] 3340: - gene was cloned [Dixon, Genomics 1995], [Humphries, Biochem J 1997] - ubiquitous, but expression most abundant in heart, liver, pancreas [UniProt], [Humphries, Biochem J 1997] - Golgi [UniProt], [Humphries, Biochem J 1997] - N-sulfation and N-deacetylation of glucosamine in heparan sulfate [UniProt] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - NDST1 & NDST2 have similar ratios of deactylase and sulfotransferase activities [Aikawa, J Biol Chem 2001] 8509: - gene was cloned [Humphries, Biochem J 1998] - 94% similar to mouse homolog [Humphries, Biochem J 1998] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - Golgi [UniProt] - ubiquitously expressed [Sugahara, IUBMB Liife 2002] 9348: - gene was cloned and expressed [Aikawa, J Biol Chem 1999] - N-sulfation and N-deacetylation of glucosamine in heparan su - highly expressed in brain, liver, kidney [Aikawa, J Biol Chem

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GLCNACDASg	3	van den Born J, Pikas DS, Pisa BJ, Eriksson I, Kjellen L, Berden JH	Antibody-based assay for N-deacetylase activity of heparan sulfate/heparin N-deacetylase/N-sulfotransferase (NDST): novel characteristics of NDST-1 and -2	Glycobiology	2003	12634318	- N-deacetylase: N-sulfotransferase acts on a subset of GlcNAc residues in a cluster along the chain [Varki, Glycobiology 1999] - enzyme rapidly deacetylates then sulfates, but some of the deacetylated GlcN residues can escape sulfation [Varki, Glycobiology 1999] 3340: - gene was cloned [Dixon, Genomics 1995], [Humphries, Biochem J 1997] - ubiquitous, but expression most abundant in heart, liver, pancreas [UniProt], [Humphries, Biochem J 1997] - Golgi [UniProt], [Humphries, Biochem J 1997] - N-sulfation and N-deacetylation of glucosamine in heparan sulfate [UniProt] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - NDST1 & NDST2 have similar ratios of deactylase and sulfotransferase activities [Aikawa, J Biol Chem 2001] 8509: - gene was cloned [Humphries, Biochem J 1998] - 94% similar to mouse homolog [Humphries, Biochem J 1998] - characterization of N-deacetylase activity [van den Born, Glycobiology 2003] - Golgi [UniProt] - ubiquitously expressed [Sugahara, IUBMB Liife 2002] 9348: - gene was cloned and expressed [Aikawa, J Biol Chem 1999] - N-sulfation and N-deacetylation of glucosamine in heparan su - highly expressed in brain, liver, kidney [Aikawa, J Biol Chem
GLCNACT1g	3	Wuyts W, Van Hul W, Hendrickx J, Speleman F, Wauters J, De Boulle K, Van Roy N, Van Agtmael T, Bossuyt P, Willems PJ	Identification and characterization of a novel member of the EXT gene family, EXTL2	Eur J Hum Genet	1997	9450183	- 2135 and 2137 both have GlcNAc-T1 activity 2135: - listed as ER localization, but has been modeled as Golgi since synthesis of linkage region and most other reactions occur there [UniProt],[Sugahara, IUBMB Life 2002] - transfers GalNAc in beta-1,4- linkages [UniProt],[Sugahara, IUBMB Life 2002] - ubiquitous [UniProt], [Wuyts, Eur J Hum Genet 1997], [Saito, Biochem Biophy Res Commun 1998] - gene was identified [Wuyts, Eur J Hum Genet 1997], [Saito, Biochem Biophy Res Commun 1998] - isolated enzyme; recombinantly expressed, identified alpha1, 4-N-acetylhexosaminyltransferase activity [Kitagawa, J Biol Chem 1999] 2317: - listed as ER localization, but has been modeled as Golgi since synthesis of linkage region and most other reactions occur there [UniProt],[Sugahara, IUBMB Life 2002] - transfers GalNAc in beta-1,4- linkages [UniProt],[Sugahara, IUBMB Life 2002] - has both GlcNAc-TI and TII activities (initiation and elongation) [Sugahara, IUBMB Life 2002], [Kim, PNAS 2001] - ubiquitous [UniProt], [Saito, Biochem Biophy Res Commun 1998] - gene was identified [Van Hul, Genomics 1998], [Saito, Bioch
GLCNACT1g	3	Kitagawa H, Shimakawa H, Sugahara K.	The tumor suppressor EXT-like gene EXTL2 encodes an alpha1, 4-N-acetylhexosaminyltransferase that transfers N-acetylgalactosamine and N-acetylglucosamine to the common glycosaminoglycan protein linkage region.	J Biol Chem	1999	10318803	- 2135 and 2137 both have GlcNAc-T1 activity 2135: - listed as ER localization, but has been modeled as Golgi since synthesis of linkage region and most other reactions occur there [UniProt],[Sugahara, IUBMB Life 2002] - transfers GalNAc in beta-1,4- linkages [UniProt],[Sugahara, IUBMB Life 2002] - ubiquitous [UniProt], [Wuyts, Eur J Hum Genet 1997], [Saito, Biochem Biophy Res Commun 1998] - gene was identified [Wuyts, Eur J Hum Genet 1997], [Saito, Biochem Biophy Res Commun 1998] - isolated enzyme; recombinantly expressed, identified alpha1, 4-N-acetylhexosaminyltransferase activity [Kitagawa, J Biol Chem 1999] 2317: - listed as ER localization, but has been modeled as Golgi since synthesis of linkage region and most other reactions occur there [UniProt],[Sugahara, IUBMB Life 2002] - transfers GalNAc in beta-1,4- linkages [UniProt],[Sugahara, IUBMB Life 2002] - has both GlcNAc-TI and TII activities (initiation and elongation) [Sugahara, IUBMB Life 2002], [Kim, PNAS 2001] - ubiquitous [UniProt], [Saito, Biochem Biophy Res Commun 1998] - gene was identified [Van Hul, Genomics 1998], [Saito, Bioch

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
GLCNACT2g	3	Wise CA, Clines GA, Massa H, Trask BJ, Lovett M	Identification and localization of the gene for EXTL, a third member of the multiple exostoses gene family	Genome Res	1997	9037597	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt] 2317: - listed as ER localization, but has been modeled as Golgi since syhtnesis of linkage region and most other reactions occur there [UniProt],[Sugahara, IUBMB Life 2002] - transfers GalNAc in beta-1,4- linkages [UniProt],[Sugahara, IUBMB Life 2002] - has both GlcNAc-TI and TII activities (initiation and elongation) [Sugahara, IUBMB Life 2002], [Kim, PNAS 2001] - ubiquitous [UniProt], [Saito, Biochem Biophy Res Commun 1998] - gene was identified [Van Hul, Genomics 1998], [Saito, Bioch 2134: - only has GlcNAcT-II activity (elongation) [Sugahara, IUBMB - gene was identified and characterized [Wise, Genome Res 19
GLCNACT2g	3	Saito T, Seki N, Yamauchi M, Tsuji S, Hayashi A, Kozuma S, Hori T	Structure, chromosomal location, and expression profile of EXTR1 and EXTR2, new members of the multiple exostoses gene family	Biochem Biophys Res Commun	1998	9473480	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt] 2317: - listed as ER localization, but has been modeled as Golgi since syhtnesis of linkage region and most other reactions occur there [UniProt],[Sugahara, IUBMB Life 2002] - transfers GalNAc in beta-1,4- linkages [UniProt],[Sugahara, IUBMB Life 2002] - has both GlcNAc-TI and TII activities (initiation and elongation) [Sugahara, IUBMB Life 2002], [Kim, PNAS 2001] - ubiquitous [UniProt], [Saito, Biochem Biophy Res Commun 1998] - gene was identified [Van Hul, Genomics 1998], [Saito, Bioch 2134: - only has GlcNAcT-II activity (elongation) [Sugahara, IUBMB - gene was identified and characterized [Wise, Genome Res 19
GLCNACT2g	3	Van Hul W, Wuyts W, Hendrickx J, Speleman F, Wauters J, De Boulle K, Van Roy N, Bossuyt P, Willems PJ	Identification of a third EXT-like gene (EXTL3) belonging to the EXT gene family	Genomics	1998	9479495	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt] 2317: - listed as ER localization, but has been modeled as Golgi since syhtnesis of linkage region and most other reactions occur there [UniProt],[Sugahara, IUBMB Life 2002] - transfers GalNAc in beta-1,4- linkages [UniProt],[Sugahara, IUBMB Life 2002] - has both GlcNAc-TI and TII activities (initiation and elongation) [Sugahara, IUBMB Life 2002], [Kim, PNAS 2001] - ubiquitous [UniProt], [Saito, Biochem Biophy Res Commun 1998] - gene was identified [Van Hul, Genomics 1998], [Saito, Bioch 2134: - only has GlcNAcT-II activity (elongation) [Sugahara, IUBMB - gene was identified and characterized [Wise, Genome Res 19

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
GLCNACT2g	3	Kim BT, Kitagawa H, Tamura J, Saito T, Kusche-Gullberg M, Lindahl U, Sugahara K	Human tumor suppressor EXT gene family members EXTL1 and EXTL3 encode alpha 1,4- N-acetylglucosaminyltransferases that likely are involved in heparan sulfate/ heparin biosynthesis	Proc Natl Acad Sci U S A	2001	11390981	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt] 2317: - listed as ER localization, but has been modeled as Golgi since syhtnesis of linkage region and most other reactions occur there [UniProt],[Sugahara, IUBMB Life 2002] - transfers GalNAc in beta-1,4- linkages [UniProt],[Sugahara, IUBMB Life 2002] - has both GlcNAc-TI and TII activities (initiation and elongation) [Sugahara, IUBMB Life 2002], [Kim, PNAS 2001] - ubiquitous [UniProt], [Saito, Biochem Biophy Res Commun 1998] - gene was identified [Van Hul, Genomics 1998], [Saito, Biochem 2134: - only has GlcNAcT-II activity (elongation) [Sugahara, IUBMB - gene was identified and characterized [Wise, Genome Res 199
GLCNACT2g	3	Sugahara K, Kitagawa H	Heparin and heparan sulfate biosynthesis	IUBMB Life	2002	12512855	- 2131, 2132 form a hetero-oligomeric complex in the Golgi [UniProt],[McCormick, PNAS 2000], [Kobayashi, Biochem Biophys Res Commun 2000]; complex has significantly higher activity in vitro than EXT1 or EXT2 alone, suggesting it is the predominant form in vivo [McCormick, PNAS 2000] 2131: - gene was cloned [Ahn, Nat Genet 1995] - human EXT1 was used to complement mouse knockout [McCormick, Nat Genet 1998] - ubiquitous [UniProt] 2132: - gene was isolated and characterized [Stickens, Nat Genet 1996] - bovine protein has both GlcNAc and GlcA transferase activities [Lind, J Biol Chem 1998] - ubiquitous [UniProt] 2317: - listed as ER localization, but has been modeled as Golgi since syhtnesis of linkage region and most other reactions occur there [UniProt],[Sugahara, IUBMB Life 2002] - transfers GalNAc in beta-1,4- linkages [UniProt],[Sugahara, IUBMB Life 2002] - has both GlcNAc-TI and TII activities (initiation and elongation) [Sugahara, IUBMB Life 2002], [Kim, PNAS 2001] - ubiquitous [UniProt], [Saito, Biochem Biophy Res Commun 1998] - gene was identified [Van Hul, Genomics 1998], [Saito, Biochem 2134: - only has GlcNAcT-II activity (elongation) [Sugahara, IUBMB - gene was identified and characterized [Wise, Genome Res 199
GLCt1r	3	Thorens B, Cheng ZQ, Brown D, Lodish HF	Liver glucose transporter: a basolateral protein in hepatocytes and intestine and kidney cells	Am J Physiol	1990	1701966	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Johnson JH, Newgard CB, Milburn JL, Lodish HF, Thorens B	The high Km glucose transporter of islets of Langerhans is functionally similar to the low affinity transporter of liver and has an identical primary sequence	J Biol Chem	1990	2182619	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Fukumoto H, Kayano T, Buse JB, Edwards Y, Pilch PF, Bell GI, Seino S	Cloning and characterization of the major insulin-responsive glucose transporter expressed in human skeletal muscle and other insulin-responsive tissues	J Biol Chem	1989	2656669	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Flier JS, Mueckler MM, Usher P, Lodish HF	Elevated levels of glucose transport and transporter messenger RNA are induced by ras or src oncogenes	Science	1987	3103217	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Fukumoto H, Seino S, Imura H, Seino Y, Eddy RL, Fukushima Y, Byers MG, Shows TB, Bell GI	Sequence, tissue distribution, and chromosomal localization of mRNA encoding a human glucose transporter-like protein	Proc Natl Acad Sci U S A	1988	3399500	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Mueckler M, Caruso C, Baldwin SA, Panico M, Blench I, Morris HR, Allard WJ, Lienhard GE, Lodish HF	Sequence and structure of a human glucose transporter	Science	1985	3839598	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Maher F, Vannucci SJ, Simpson IA	Glucose transporter proteins in brain	FASEB J	1994	7926364	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Guillam MT, Burcelin R, Thorens B	Normal hepatic glucose production in the absence of GLUT2 reveals an alternative pathway for glucose release from hepatocytes	Proc Natl Acad Sci U S A	1998	9770484	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Doege H, Schurmann A, Bahrenberg G, Brauers A, Joost HG	GLUT8, a novel member of the sugar transport facilitator family with glucose transport activity	J Biol Chem	2000	10821868	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Phay JE, Hussain HB, Moley JF	Cloning and expression analysis of a novel member of the facilitative glucose transporter family, SLC2A9 (GLUT9)	Genomics	2000	10860667	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Carayannopoulos MO, Chi MM, Cui Y, Pingsterhaus JM, McKnight RA, Mueckler M, Devaskar SU, Moley KH	GLUT8 is a glucose transporter responsible for insulin-stimulated glucose uptake in the blastocyst	Proc Natl Acad Sci U S A	2000	10860996	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Doege H, Bocianski A, Joost HG, Schurmann A	Activity and genomic organization of human glucose transporter 9 (GLUT9), a novel member of the family of sugar-transport facilitators predominantly expressed in brain and leucocytes	Biochem J	2000	10970791	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	McVie-Wylie AJ, Lamson DR, Chen YT	Molecular cloning of a novel member of the GLUT family of transporters, SLC2a10 (GLUT10), localized on chromosome 20q13.1: a candidate gene for NIDDM susceptibility	Genomics	2001	11247674	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
GLCt1r	3	Doege H, Bocianski A, Scheepers A, Axer H, Eckel J, Joost HG, Schurmann A	Characterization of human glucose transporter (GLUT) 11 (encoded by SLC2A11), a novel sugar-transport facilitator specifically expressed in heart and skeletal muscle	Biochem J	2001	11583593	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Dawson PA, Mychaleckyj JC, Fossey SC, Mihic SJ, Craddock AL, Bowden DW	Sequence and functional analysis of GLUT10: a glucose transporter in the Type 2 diabetes-linked region of chromosome 20q12-13.1	Mol Genet Metab	2001	11592815	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Joost HG, Thorens B	The extended GLUT-family of sugar/polyol transport facilitators: nomenclature, sequence characteristics, and potential function of its novel members (review)	Mol Membr Biol	2001	11780753	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Rogers S, Macheda ML, Docherty SE, Carty MD, Henderson MA, Soeller WC, Gibbs EM, James DE, Best JD	Identification of a novel glucose transporter-like protein-GLUT-12	Am J Physiol Endocrinol Metab	2002	11832379	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Hosokawa M, Thorens B	Glucose release from GLUT2-null hepatocytes: characterization of a major and a minor pathway	Am J Physiol Endocrinol Metab	2002	11882499	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Uldry M, Ibberson M, Hosokawa M, Thorens B	GLUT2 is a high affinity glucosamine transporter	FEBS Lett	2002	12135767	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Macheda ML, Williams ED, Best JD, Wlodek ME, Rogers S	Expression and localisation of GLUT1 and GLUT12 glucose transporters in the pregnant and lactating rat mammary gland	Cell Tissue Res	2003	12483288	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Wu X, Freeze HH	GLUT14, a duplicon of GLUT3, is specifically expressed in testis as alternative splice forms	Genomics	2002	12504846	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Rogers S, Chandler JD, Clarke AL, Petrou S, Best JD	Glucose transporter GLUT12-functional characterization in Xenopus laevis oocytes	Biochem Biophys Res Commun	2003	12914765	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt1r	3	Augustin R, Carayannopoulos MO, Dowd LO, Phay JE, Moley JF, Moley KH	Identification and characterization of human glucose transporter-like protein-9 (GLUT9): alternative splicing alters trafficking	J Biol Chem	2004	14739288	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]

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GLCt1r	3	Li Q, Manolescu A, Ritzel M, Yao S, Slugoski M, Young JD, Chen XZ, Cheeseman CI	Cloning and functional characterization of the human GLUT7 isoform SLC2A7 from the small intestine	Am J Physiol Gastrointest Liver Physiol	2004	15033637	simple diffusion 6513: - transports Glc, Gal, Man, GlcN [Uldry 2004], [Uldry 2002] - RBC, brain [Maher 1994] - cloned [Mueckler 1985] - not expressed in normal hepatocytes; induced during cancer [Flier 1987] 6514: - transports Glc (low affinity [Johnson 1990]), Gal, Fru, Man, GlcN [Uldry 2004], [Uldry 2002] - intest and kidney [Thorens 1990], liver, panc B cells, Langerhans, brain [Uldry 2004] - cloned [Fukumoto 1988] - primary GLUT for Glc release in hepatocytes [Hosokawa 2002], [Guillam 1998] 6515: - transports Glc (high), Gal, Man, Xyl, Malt, DHA [Uldry 2004] - brain, testis [Haber 1993], [Uldry 2004], sk muscle [Stuart 1999], platelets [Heijen 1997] - cloned [Kayano 1988] 6517: - cDNA was cloned [Fukumoto 1989] - transports Glc, GlcN, DHA; adipose (brown & white), sk and cardiac muscle; insulin, exercise stimulate translocation of GLUT4 vesicles to cell surface [Uldry 2004] 11182: - cDNA was cloned [Doege 2000] - transports Glc; brain, spleen, WBC; activity only demonstrated in liposomes [Udry 2004] 155184: - gene identified [Joost 2001]
GLCt2r	3	Wells RG, Pajor AM, Kanai Y, Turk E, Wright EM, Hediger MA	Cloning of a human kidney cDNA with similarity to the sodium-glucose cotransporter	Am J Physiol	1992	1415574	0
GLCt2r	3	Kanai Y, Lee WS, You G, Brown D, Hediger MA	The human kidney low affinity Na+/glucose cotransporter SGLT2. Delineation of the major renal reabsorptive mechanism for D-glucose	J Clin Invest	1994	8282810	0
GLCt2r	3	Wright EM	Renal Na(+)-glucose cotransporters	Am J Physiol Renal Physiol	2001	11133510	0
GLCt4	3	van den Heuvel LP, Assink K, Willemsen M, Monnens L	Autosomal recessive renal glucosuria attributable to a mutation in the sodium glucose cotransporter (SGLT2).	Hum Genet	2002	12436245	6524: - cloned [Wells 1992] - transports Glc/Na+ (low affinity) at a 1:1 ratio [Kanai 1994] - renal cortex [Wright 2001] - major transporter involved in reabsorption of glc from glomrtular filtrate [Van de Heuvel 2002] 6526: - cloned [Kwon 1992], [Berry 1995] - kidney, brain, placenta, pancreas, heart, skeletal muscle, lung [Berry 1995] - Na+/myo-inositol cotransport; also transports other sugars (incl glc) with low affinity [Hager 1995], [Kwon 1992] - plasma membrane; see [Wright 2004] for refs - transports Na+ in the absence of sugar [Wright, Physiology 2004] 125206: - function inferred from electronic annotation [GO] - kidney (rabbit ortholog); see [Wright 2004] for refs 159963: - function inferred from electronic annotation [GO]
GLCt4_2	3	Tazawa S, Yamato T, Fujikura H, Hiratochi M, Itoh F, Tomae M, Takemura Y, Maruyama H, Sugiyama T, Wakamatsu A, Isogai T, Isaji M	SLC5A9/SGLT4, a new Na+-dependent glucose transporter, is an essential transporter for mannose, 1,5-anhydro-D-glucitol, and fructose	Life Sci	2005	15607332	- glc is transported against concentration gradient, typically into the cell [Champe, Biochemistry 2005] - occurs in epithelial cels of the intestine, renal tubules, and choroid plexus [Champe, Biochemistry 2005] 6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 200010: - cloned and characterized [Tazawa 2005] - mainly sm intestine & kidney, also liver, lung, brain [Tazawa 2005] - transports D-mannose (Man) >> D-glucose (Glc) > D-fructose (Fru) = 1,5-anhydro-D-glucitol (1,5AG) > D-galactose (Gal) [Tazawa 2005]

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GLCtg	3	Haney PM	Glucose transport in lactation	Adv Exp Med Biol	2004	15384581	Glc passes across Golgi membrane into Golgi lumen by GLUT 1 [Haney 2004] - GLUT1 is the only known glucose transporter isoform expressed in mammary gland [Haney 2004] original ref was WL Hurley's website on Lactation Biology, Dept of Animal Sciences, Univ of Illinois, Urbana-Champaign. He cites Kuhn, Ch 6 in Mepham, 1983, Biochem of Lactation, Elsevier http://classes.aces.uiuc.edu/AnSci308/lactosesynthesis.html
GLCtly	2	Pisoni RL, Thoene JG	The transport systems of mammalian lysosomes	Biochim Biophys Acta	1991	1751541	- function has only been characterized in rat liver lysosomes (see refs in [Pisoni 1991]) - D-glucose, D-galactose, D-mannose, and L-fucose are known substrates [Lloyd 1996] - D-xylose is also recognized by the carrier, but passive diffusion may be the predominant mode of efflux [Lloyd 1996]
GLCtly	2	Lloyd JB	Metabolite efflux and influx across the lysosome membrane	Subcell Biochem	1996	8993166	- function has only been characterized in rat liver lysosomes (see refs in [Pisoni 1991]) - D-glucose, D-galactose, D-mannose, and L-fucose are known substrates [Lloyd 1996] - D-xylose is also recognized by the carrier, but passive diffusion may be the predominant mode of efflux [Lloyd 1996]
GLCURtly	3	Mancini GM, de Jonge HR, Galjaard H, Verheijen FW.	Characterization of a proton-driven carrier for sialic acid in the lysosomal membrane. Evidence for a group-specific transport system for acidic monosaccharides.	J Biol Chem	1989	2768261	H symport with sialic acid (precursors) into lysososome. See PMID: 2768261for biochem characterization and PMID: 10581036 for discussion of particular SNPs with sialic acid storage diseases. Sialic acid storage disorders (due to transporter mutations) require import and export e.g. PMID: 2768266 - hence made reversible. NJ
GLCURtly	3	Verheijen FW, Verbeek E, Aula N, Beerens CE, Havelaar AC, Joosse M, Peltonen L, Aula P, Galjaard H, van der Spek PJ, Mancini GM.	A new gene, encoding an anion transporter, is mutated in sialic acid storage diseases.	Nat Genet	1999	10581036	H symport with sialic acid (precursors) into lysososome. See PMID: 2768261for biochem characterization and PMID: 10581036 for discussion of particular SNPs with sialic acid storage diseases. Sialic acid storage disorders (due to transporter mutations) require import and export e.g. PMID: 2768266 - hence made reversible. NJ
GLDBRAN	3	Yang BZ, Ding JH, Enghild JJ, Bao Y, Chen YT	Molecular cloning and nucleotide sequence of cDNA encoding human muscle glycogen debranching enzyme	J Biol Chem	1992	1374391	- see Devlin p. 648 - Variants 1, 5, and 6 are present in both liver and muscle, whereas variants 2, 3, and 4 occur in muscle [RefSeq]
GLGNS1	2	Lomako J, Lomako WM, Whelan WJ	Glycogenin: the primer for mammalian and yeast glycogen synthesis	Biochim Biophys Acta	2004	15238248	-glycogenin and glycogen synthase form a complex [Lomako, Biochim Biophys Acta 2004] -see pathway in Fig 2 of Lomako, Biochim Biophys Acta 2004 - see Devlin p.652
GLNS	2	Gibbs CS, Campbell KE, Wilson RH.	Sequence of a human glutamine synthetase cDNA.		1987	2888076	cytosolic - Reactome irreversible reaction - Reactome and "common knowledge" that synthetase reactions are typically irreversible
GLPASE1	3	Newgard CB, Littman DR, van Genderen C, Smith M, Fletterick RJ	Human brain glycogen phosphorylase. Cloning, sequence analysis, chromosomal mapping, tissue expression, and comparison with the human liver and muscle isozymes	J Biol Chem	1988	3346228	- see Devlin p. 648 5834: - found predominantly in the brain [RefSeq]
GLUCYS	3	Gipp JJ, Bailey HH, Mulcahy RT.	Cloning and sequencing of the cDNA for the light subunit of human liver gamma-glutamylcysteine synthetase and relative mRNA levels for heavy and light subunits in human normal tissues.	Biochem Biophys Res Commun	1995	7826375	- Added by RS/TV Meister, A, Mitochondrial changes associated with glutathione deficiency, Biochimica et Biophysica Acta 1271 (1995) 35-42. 1) Catalytic activity specified by GeneCards. 2) Glutamate-cysteine ligase (GCL) is a rate-limiting enzyme for GSH synthesis (cytosol) Intracellular gamma-glutamylcystein synthetase can be dissociated into two subunits: a heavy(Gclc.1), catalytic subunit and a light(Gclm.1), regulatory subunit. 3) While enzymatic activity was found to some extent in all tissues, gamma-glutamylcystein synthetase activity has been reported with highest levels of expression commonly being found in liver and kidney. All this according to Gipp JJ, Bailey HH, Mulcahy RT. Biochem Biophys Res Commun. 1995 Jan 17;206(2):584-9.

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GLUDC	3	Karlsen AE, Hagopian WA, Grubin CE, Dube S, Disteche CM, Adler DA, Barmeier H, Mathewes S, Grant FJ, Foster D, et al.	Cloning and primary structure of a human islet isoform of glutamic acid decarboxylase from chromosome 10.		1991	1924293	Entrez gene - This gene encodes one of several forms of glutamic acid decarboxylase, identified as a major autoantigen in insulin-dependent diabetes. The enzyme encoded is responsible for catalyzing the production of gamma-aminobutyric acid from L-glutamic acid. A pathogenic role for this enzyme has been identified in the human pancreas since it has been identified as an autoantigen and an autoreactive T cell target in insulin-dependent diabetes. This gene may also play a role in the stiff man syndrome. Deficiency in this enzyme has been shown to lead to pyridoxine dependency with seizures. Alternative splicing of this gene results in two products, the predominant 67-kD form and a less-frequent 25-kD form
GLUDC	3	Bu DF, Tobin AJ.	The exon-intron organization of the genes (GAD1 and GAD2) encoding two human glutamate decarboxylases (GAD67 and GAD65) suggests that they derive from a common ancestral GAD.		1994	8088791	Entrez gene - This gene encodes one of several forms of glutamic acid decarboxylase, identified as a major autoantigen in insulin-dependent diabetes. The enzyme encoded is responsible for catalyzing the production of gamma-aminobutyric acid from L-glutamic acid. A pathogenic role for this enzyme has been identified in the human pancreas since it has been identified as an autoantigen and an autoreactive T cell target in insulin-dependent diabetes. This gene may also play a role in the stiff man syndrome. Deficiency in this enzyme has been shown to lead to pyridoxine dependency with seizures. Alternative splicing of this gene results in two products, the predominant 67-kD form and a less-frequent 25-kD form
GLUDxm	3	Mavrothalassitis G, Tzimagiorgis G, Mitsialis A, Zannis V, Plaitakis A, Papamatheakis J, Moschonas N.	Isolation and characterization of cDNA clones encoding human liver glutamate dehydrogenase: evidence for a small gene family.	Proc Natl Acad Sci U S A	1988	3368458	Reversible reaction, Mitochondrial matrix - Fang et al. Biochem. J. (2002) 363 (81 87) - Additional information added by RS/TV: mitochondrial according to GeneCards 1) Glutamate dehydrogenase is known to catalyze the reversible oxidative deamination of L-glutamate to akg using NAD and/or NADP as cofactors according to G Mavrothalassitis, G Tzimagiorgis, A Mitsialis, V Zannis, A Plaitakis, J Papamatheakis, and N Moschonas. Isolation and characterization of cDNA clones encoding human liver glutamate dehydrogenase: evidence for a small gene family. Proc Natl Acad Sci U S A. 1988 May;85(10):3494-8. PMID: 3368458 2) Glutamate dehydrogenase exists in two isoforms. Glud1.1-m is considered to be housekeeping (localization description) according to Zaganas I, Spanaki C, Karpusas M, Plaitakis A. Substitution of Ser for Arg-443 in the regulatory domain of human housekeeping (GLUD1) glutamate dehydrogenase virtually abolishes basal activity and markedly alters the activation of the enzyme by ADP and L-leucine. J Biol Chem. 2002 Nov 29;277(48):46552-8. Epub 2002 Sep 24. PMID: 12324473
GLUDxm	3	Zaganas I, Spanaki C, Karpusas M, Plaitakis A.	Substitution of Ser for Arg-443 in the regulatory domain of human housekeeping (GLUD1) glutamate dehydrogenase virtually abolishes basal activity and markedly alters the activation of the enzyme by ADP and L-leucine.	J Biol Chem	2002	12324473	Reversible reaction, Mitochondrial matrix - Fang et al. Biochem. J. (2002) 363 (81 87) - Additional information added by RS/TV: mitochondrial according to GeneCards 1) Glutamate dehydrogenase is known to catalyze the reversible oxidative deamination of L-glutamate to akg using NAD and/or NADP as cofactors according to G Mavrothalassitis, G Tzimagiorgis, A Mitsialis, V Zannis, A Plaitakis, J Papamatheakis, and N Moschonas. Isolation and characterization of cDNA clones encoding human liver glutamate dehydrogenase: evidence for a small gene family. Proc Natl Acad Sci U S A. 1988 May;85(10):3494-8. PMID: 3368458 2) Glutamate dehydrogenase exists in two isoforms. Glud1.1-m is considered to be housekeeping (localization description) according to Zaganas I, Spanaki C, Karpusas M, Plaitakis A. Substitution of Ser for Arg-443 in the regulatory domain of human housekeeping (GLUD1) glutamate dehydrogenase virtually abolishes basal activity and markedly alters the activation of the enzyme by ADP and L-leucine. J Biol Chem. 2002 Nov 29;277(48):46552-8. Epub 2002 Sep 24. PMID: 12324473

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GLUDxm	3	Jie FANG , Betty Y. L. HSU , Courtney M. MACMULLEN , Mortimer PONCZ , Thomas J. SMITH and Charles A. STANLEY	Expression, purification and characterization of human glutamate dehydrogenase (GDH) allosteric regulatory mutations		2002		Reversible reaction, Mitochondrial matrix - Fang et al. Biochem. J. (2002) 363 (81 87) - Additional information added by RS/TV: mitochondrial according to GeneCards 1) Glutamate dehydrogenase is known to catalyze the reversible oxidative deamination of L-glutamate to akg using NAD and/or NADP as cofactors according to G Mavrothalassitis, G Tzimagiorgis, A Mitsialis, V Zannis, A Plaitakis, J Papamatheakis, and N Moschonas. Isolation and characterization of cDNA clones encoding human liver glutamate dehydrogenase: evidence for a small gene family. Proc Natl Acad Sci U S A. 1988 May;85(10):3494-8. PMID: 3368458 2) Glutamate dehydrogenase exists in two isoforms. Glud1.1-m is considered to be housekeeping (localization description) according to Zaganas I, Spanaki C, Karpusas M, Plaitakis A. Substitution of Ser for Arg-443 in the regulatory domain of human housekeeping (GLUD1) glutamate dehydrogenase virtually abolishes basal activity and markedly alters the activation of the enzyme by ADP and L-leucine. J Biol Chem. 2002 Nov 29;277(48):46552-8. Epub 2002 Sep 24. PMID: 12324473
GluForTx	3	Hilton JF, Christensen KE, Watkins D, Raby BA, Renaud Y, de la Luna S, Estivill X, MacKenzie RE, Hudson TJ, Rosenblatt DS	The molecular basis of glutamate formiminotransferase deficiency	Hum Mutat	2003	12815595	This enzyme is likely responsible for the second most common inborn error of folate metabolism.
GLUNm	3	Holcomb T, Taylor L, Trohkimoinen J, Curthoys NP.	Isolation, characterization and expression of a human brain mitochondrial glutaminase cDNA.		2000	10719215	-2 isozymes, one expressed mainly in kidney and brain, the other in the liver - Aledo et. al. Mamm Genome. 2000 Dec;11(12):1107-10. -mitochondrial matrix- Holcomb et al. Brain Res Mol Brain Res. 2000 Mar 10;76(1):56-63. and Reactome -irreversible according to Reactome -genetic data - Zacharias et al. Cell Biochem Funct. 2003 Mar;21(1):81-4.
GLUNm	3	Zacharias DP, Lima MM, Souza AL Jr, de Abranches Oliveira Santos ID, Enokiara M, Michalany N, Curi R.	Human cutaneous melanoma expresses a significant phosphate-dependent glutaminase activity: a comparison with the surrounding skin of the same patient.		2003	12579526	-2 isozymes, one expressed mainly in kidney and brain, the other in the liver - Aledo et. al. Mamm Genome. 2000 Dec;11(12):1107-10. -mitochondrial matrix- Holcomb et al. Brain Res Mol Brain Res. 2000 Mar 10;76(1):56-63. and Reactome -irreversible according to Reactome -genetic data - Zacharias et al. Cell Biochem Funct. 2003 Mar;21(1):81-4.
GLUNm	3	Aledo JC, Gomez-Fabre PM, Olalla L, Marquez J.	Identification of two human glutaminase loci and tissue-specific expression of the two related genes.		2005		-2 isozymes, one expressed mainly in kidney and brain, the other in the liver - Aledo et. al. Mamm Genome. 2000 Dec;11(12):1107-10. -mitochondrial matrix- Holcomb et al. Brain Res Mol Brain Res. 2000 Mar 10;76(1):56-63. and Reactome -irreversible according to Reactome -genetic data - Zacharias et al. Cell Biochem Funct. 2003 Mar;21(1):81-4.
GLUPRT	3	Iwahana H, Oka J, Mizusawa N, Kudo E, Ii S, Yoshimoto K, Holmes EW, Itakura M.	Molecular cloning of human amidophosphoribosyltransferase.	Biochem Biophys Res Commun	1993	8380692	I could not find any infos about compartiment, however, the HPRD database said nucleus. This info is based on paper of Chen et al, 1997. But they found only a nuclear protein that regulates the expression of PPAT but they did not mentioned where PPAt is located. IT
GLUPRT	3	Chen S, Nagy PL, Zalkin H	Role of NRF-1 in bidirectional transcription of the human GPAT-AIRC purine biosynthesis locus.	Nucleic Acids Res	1997	9108165	I could not find any infos about compartiment, however, the HPRD database said nucleus. This info is based on paper of Chen et al, 1997. But they found only a nuclear protein that regulates the expression of PPAT but they did not mentioned where PPAt is located. IT
GLUt6	3	Kanai Y, Hediger MA	The glutamate/neutral amino acid transporter family SLC1: molecular, physiological and pharmacological aspects	Pflugers Arch	2004	14530974	The high affinity glutamate transporters mediate transport of l-Glu, l-Asp and d-Asp, accompanied by the cotransport of 3 Na+ and 1 H+, and the countertransport of 1 K+ from PMID 14530974

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GLUVESSEC	3	Bai L, Xu H, Collins JF, Ghishan FK	Molecular and functional analysis of a novel neuronal vesicular glutamate transporter	J Biol Chem	2001	11432869	This is a lumped reaction. The most precise description is that h is transported from the cytosol into a vesicle with an ATP-dependent transporter, and then the h gradient used with an antiport to drive glutamate transport from the cytosol into the vesicle. The vesicle fuses with the membrane and dumps the glutamate outside the cell. From PMID 12811560: The three VGLUT isoforms exhibit saturable glutamate transport with a Km~1 mM that is driven primarily by the electrical component (Dy) of the proton electrochemical gradient across the vesicle membrane: valinomycin, a K+ ionophore that dissipates Dy reduces transport to a greater extent than nigericin, a H+-K+- exchanging ionophore that dissipates the pH gradient [6, 9, 17, 18, 19, 24, 37, 40, 41, 42, 44]. From PMID 15383652: In addition to this intrinsic dependency on the transmembrane electrochemical gradient, the transport rate can also be modulated by alterations in the rate of ATP hydrolysis and its coupling to H+ translocation. A recent detailed analysis of current-voltage relationships in the absence and presence of sev
GLUVESSEC	3	Wagner CA, Finberg KE, Breton S, Marshansky V, Brown D, Geibel JP	Renal vacuolar H+-ATPase	Physiol Rev	2004	15383652	This is a lumped reaction. The most precise description is that h is transported from the cytosol into a vesicle with an ATP-dependent transporter, and then the h gradient used with an antiport to drive glutamate transport from the cytosol into the vesicle. The vesicle fuses with the membrane and dumps the glutamate outside the cell. From PMID 12811560: The three VGLUT isoforms exhibit saturable glutamate transport with a Km~1 mM that is driven primarily by the electrical component (Dy) of the proton electrochemical gradient across the vesicle membrane: valinomycin, a K+ ionophore that dissipates Dy reduces transport to a greater extent than nigericin, a H+-K+- exchanging ionophore that dissipates the pH gradient [6, 9, 17, 18, 19, 24, 37, 40, 41, 42, 44]. From PMID 15383652: In addition to this intrinsic dependency on the transmembrane electrochemical gradient, the transport rate can also be modulated by alterations in the rate of ATP hydrolysis and its coupling to H+ translocation. A recent detailed analysis of current-voltage relationships in the absence and presence of sev
GLXO1	3	Holmes RP	Pharmacological approaches in the treatment of primary hyperoxaluria	J Nephrol	1998	9604807	- described in Devlin, p. 795, Orten p. 316 -catalyzed by lactate dehydrogenase [Holmes, J Urol 1998], [Holmes, J Nerphol 1998], [Pirulli, J Nephrol 2003] - cytoplasm [Holmes, J Urol 1998] - lactate dehydrogenase catalyzes the bulk of glyoxylate -> oxalate conversion in vivo as the concentrations of glycolate and lactate in hepatocytes will inhibit the glycolate oxidase catalyzed reaction [Poore 1997]
GLXO1	3	Holmes RP, Assimos DG	Glyoxylate synthesis, and its modulation and influence on oxalate synthesis	J Urol	1998	9783918	- described in Devlin, p. 795, Orten p. 316 -catalyzed by lactate dehydrogenase [Holmes, J Urol 1998], [Holmes, J Nerphol 1998], [Pirulli, J Nephrol 2003] - cytoplasm [Holmes, J Urol 1998] - lactate dehydrogenase catalyzes the bulk of glyoxylate -> oxalate conversion in vivo as the concentrations of glycolate and lactate in hepatocytes will inhibit the glycolate oxidase catalyzed reaction [Poore 1997]
GLXO1	3	Pirulli D, Marangella M, Amoroso A	Primary hyperoxaluria: genotype-phenotype correlation	J Nephrol	2003	12768081	- described in Devlin, p. 795, Orten p. 316 -catalyzed by lactate dehydrogenase [Holmes, J Urol 1998], [Holmes, J Nerphol 1998], [Pirulli, J Nephrol 2003] - cytoplasm [Holmes, J Urol 1998] - lactate dehydrogenase catalyzes the bulk of glyoxylate -> oxalate conversion in vivo as the concentrations of glycolate and lactate in hepatocytes will inhibit the glycolate oxidase catalyzed reaction [Poore 1997]
GLXtp	1	Baker PR, Cramer SD, Kennedy M, Assimos DG, Holmes RP.	Glycolate and glyoxylate metabolism in HepG2 cells.		2004	15240345	- reaction proposed in Fig 6 of [Baker 2004]
GLYAMDTRc	3	Isbrandt D, von Figura K	Cloning and sequence analysis of human guanidinoacetate N-methyltransferase cDNA	Biochim Biophys Acta	1995	8547310	Introduction of first citation states function from another paper from 1973.
GLYATm	2	Edgar,A.J. , Polak,J.M.	Molecular cloning of the human and murine 2-amino-3-ketobutyrate coenzyme A ligase cDNAs.		2000	10712613	0

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GLYBtm	2	Porter RK, Scott JM, Brand MD.	Characterization of betaine efflux from rat liver mitochondria.		1993	8443213	-added to allow glyb transport from mitochondria to cytosol -glyb diffuses across rat liver mitochondrial membrane to cytosol (PMID:8443213) -needed this rxn to fill gap since glyb is used by rxn BHMT in the cytosol MM
GLYCLTDy	3	Taylor SW, Fahy E, Zhang B, Glenn GM, Warnock DE, Wiley S, Murphy AN, Gaucher SP, Capaldi RA, Gibson BW, Ghosh SS	Characterization of the human heart mitochondrial proteome	Nat Biotechnol	2003	1252411	- ubiquitous [RefSeq], [UniProt] - has hydroxy-pyruvate reductase, glyoxylate reductase and D-glycerate dehydrogenase enzymatic activities [RefSeq], [UniProt] - uses NADPH as coenzyme [Van Schaftingen, Eur J Biochem 1989] - cellular localization uncertain, but glyoxylate reductase activity seems to be present in both cytosol and mitochondria - mitochondrial glyoxylate dehydrogenase was identified in human heart proteomic data [Taylor 2003] - HepG2 mitochondria have glyoxylate reductase acticity similar to that found in cell homogenates, suggesting that the cytosol and mitochondria have equivalent activities [Baker 2004]
GLYCLTDy	3	Van Schaftingen E, Draye JP, Van Hoof F	Coenzyme specificity of mammalian liver D-glycerate dehydrogenase	Eur J Biochem	1989	2689175	- ubiquitous [RefSeq], [UniProt] - has hydroxy-pyruvate reductase, glyoxylate reductase and D-glycerate dehydrogenase enzymatic activities [RefSeq], [UniProt] - uses NADPH as coenzyme [Van Schaftingen, Eur J Biochem 1989] - cellular localization uncertain, but glyoxylate reductase activity seems to be present in both cytosol and mitochondria - mitochondrial glyoxylate dehydrogenase was identified in human heart proteomic data [Taylor 2003] - HepG2 mitochondria have glyoxylate reductase acticity similar to that found in cell homogenates, suggesting that the cytosol and mitochondria have equivalent activities [Baker 2004]
GLYC-St	1	Petrarulo M, Vitale C, Facchini P, Marangella M	Biochemical approach to diagnosis and differentiation of primary hyperoxalurias: an update	J Nephrol	1998	9604805	- L-glycerate is virtually absent from urine of healthy individuals [Petrarulo 1998] (presumably because it is not produced in detectable quantities) - Patients with primary hyperoxaluria type 2 overproduce L-glycerate (mutation in glyoxylate reductase/hydroxypyruvate reductase gene leaves lactate dehydrogenase as only enzyme available to metabolize hydroxypyruvate) and as a result L-glycerate is detectable in plasma and urine [Petrarulo 1998] - Note that this rxn should not be necessary in healthy people since the rxn producing L-glycerate (HPYRR2x) should essentially be "off"
GLYCt	2	Kuriyama H, Kawamoto S, Ishida N, Ohno I, Mita S, Matsuzawa Y, Matsubara K, Okubo K	Molecular cloning and expression of a novel human aquaporin from adipose tissue with glycerol permeability	Biochem Biophys Res Commun	1997	9405233	- glycerol is released during hydrolysis of triacylglycerols in adipose tissue and delivered to the liver by the blood [Champe, Biochemistry 2005] - is is taken up aquaglyceroporins that transport glycerol as well as water [Biochem Biophys Res Commun 1997]
GLYCTO1p	3	Ushijima Y	Identity of aliphatic L- -hydroxyacid oxidase and glycolate oxidase from rat livers	Arch Biochem Biophys	1973	4705431	- reaction is essentially irreversible under in vivo conditions [Poore 1998] - rat liver glycolate oxidase has 10x higher affinity for glycolate than glyoxylate [Ushijima 1973] 51179: - peroxisome [RefSeq], [UniProt], [Jones, J Biol Chem 2000] - liver and kidney [RefSeq], [UniProt], [Jones, J Biol Chem 2000] 54363: - peroxisome [RefSeq], [UniProt], [Jones, J Biol Chem 2000] - liver and pancreas [RefSeq], [UniProt], [Jones, J Biol Chem 2000] - most active on glycolate [RefSeq], [UniProt], [Jones, J Biol Chem 2000]

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GLYCTO1p	3	Jones JM, Morrell JC, Gould SJ	Identification and characterization of HAOX1, HAOX2, and HAOX3, three human peroxisomal 2-hydroxy acid oxidases	J Biol Chem	2000	10777549	- reaction is essentially irreversible under in vivo conditions [Poore 1998] - rat liver glycolate oxidase has 10x higher affinity for glycolate than glyoxylate [Ushijima 1973] 51179: - peroxisome [RefSeq], [UniProt], [Jones, J Biol Chem 2000] - liver and kidney [RefSeq], [UniProt], [Jones, J Biol Chem 2000] 54363: - peroxisome [RefSeq], [UniProt], [Jones, J Biol Chem 2000] - liver and pancreas [RefSeq], [UniProt], [Jones, J Biol Chem 2000] - most active on glycolate [RefSeq], [UniProt], [Jones, J Biol Chem 2000]
GLYK	3	Sargent CA, Young C, Marsh S, Ferguson-Smith MA, Affara NA.	The glycerol kinase gene family: structure of the Xp gene, and related intronless retroposons.	Hum Mol Genet	1994	7987308	Bound to the mitochondrial surface or cytoplasmic. In sperm and fetal tissues, the majority of the enzyme is bound to mitochondria, but in adult tissues, such as liver found in the cytoplasm. see Sargent refs for different types Highly expressed in the liver, kidney and testis. Isoforms 2 (GK2) and 3(GKP3) are expressed specifically in testis and fetal liver, but not in the adult liver. NJ - adipocytes lack glycerol kinase [Champe, Biochemistry 2005] - ~50% similarity btwn human and E. coli GK proteins [Hurley 1993]
GLYK	3	Hurley JH, Faber HR, Worthylake D, Meadow ND, Roseman S, Pettigrew DW, Remington SJ	Structure of the regulatory complex of Escherichia coli IIIGlc with glycerol kinase	Science	1993	8430315	Bound to the mitochondrial surface or cytoplasmic. In sperm and fetal tissues, the majority of the enzyme is bound to mitochondria, but in adult tissues, such as liver found in the cytoplasm. see Sargent refs for different types Highly expressed in the liver, kidney and testis. Isoforms 2 (GK2) and 3(GKP3) are expressed specifically in testis and fetal liver, but not in the adult liver. NJ - adipocytes lack glycerol kinase [Champe, Biochemistry 2005] - ~50% similarity btwn human and E. coli GK proteins [Hurley 1993]
GLYK	3	Voet D, Voet JG	Biochemistry		1995		Bound to the mitochondrial surface or cytoplasmic. In sperm and fetal tissues, the majority of the enzyme is bound to mitochondria, but in adult tissues, such as liver found in the cytoplasm. see Sargent refs for different types Highly expressed in the liver, kidney and testis. Isoforms 2 (GK2) and 3(GKP3) are expressed specifically in testis and fetal liver, but not in the adult liver. NJ - adipocytes lack glycerol kinase [Champe, Biochemistry 2005] - ~50% similarity btwn human and E. coli GK proteins [Hurley 1993]

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GLYK	3	Murray RK, Granner DK, Mayes PA, Rodwell VW	Harper's Biochemistry		1999		Bound to the mitochondrial surface or cytoplasmic. In sperm and fetal tissues, the majority of the enzyme is bound to mitochondria, but in adult tissues, such as liver found in the cytoplasm. see Sargent refs for different types Highly expressed in the liver, kidney and testis. Isoforms 2 (GK2) and 3(GKP3) are expressed specifically in testis and fetal liver, but not in the adult liver. NJ - adipocytes lack glycerol kinase [Champe, Biochemistry 2005] - ~50% similarity btwn human and E. coli GK proteins [Hurley 1993]
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GLYK	3	Guo W, Worley K, Adams V, Mason J, Sylvester-Jackson D, Zhang Y, Towbin JA, Fogt DD, Madu S, Wheeler DA, McCabe ERB	Genomic scanning for expressed sequences in Xp21 identifies the glycerol kinase gene	Nature Genetics	1993		Bound to the mitochondrial surface or cytoplasmic. In sperm and fetal tissues, the majority of the enzyme is bound to mitochondria, but in adult tissues, such as liver found in the cytoplasm. see Sargent refs for different types Highly expressed in the liver, kidney and testis. Isoforms 2 (GK2) and 3(GKP3) are expressed specifically in testis and fetal liver, but not in the adult liver. NJ - adipocytes lack glycerol kinase [Champe, Biochemistry 2005] - ~50% similarity btwn human and E. coli GK proteins [Hurley 1993]
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GLYOX	3	Ridderstrom M, Saccucci F, Hellman U, Bergman T, Principato G, Mannervik B	Molecular cloning, heterologous expression, and characterization of human glyoxalase II	J Biol Chem	1996	8550579	3029: - hydrolyzes S-D- lactoyl-glutathione to form glutathione and D-lactic acid [UniProt] - cloned & expressed in E. coli; recombinant enzyme had full catalytic activity and kinetic parameters indistinguishable from those of the native enzyme purified from human erythrocytes [Ridderstrom 1996] - gene encodes both a cytosolic enzyme and a mitochondrial matrix enzyme [Cordell 2004] 84264: - hydrolyzes S-D- lactoyl-glutathione to form glutathione and D-lactic acid [UniProt] - function inferred from electronic annotation [GO]; protein has been assigned EC number
GLYOX	3	Cordell PA, Futers TS, Grant PJ, Pease RJ	The Human hydroxyacylglutathione hydrolase (HAGH) gene encodes both cytosolic and mitochondrial forms of glyoxalase II.	J Biol Chem	2004	15117945	3029: - hydrolyzes S-D- lactoyl-glutathione to form glutathione and D-lactic acid [UniProt] - cloned & expressed in E. coli; recombinant enzyme had full catalytic activity and kinetic parameters indistinguishable from those of the native enzyme purified from human erythrocytes [Ridderstrom 1996] - gene encodes both a cytosolic enzyme and a mitochondrial matrix enzyme [Cordell 2004] 84264: - hydrolyzes S-D- lactoyl-glutathione to form glutathione and D-lactic acid [UniProt] - function inferred from electronic annotation [GO]; protein has been assigned EC number
GLYtm	3	Benavides J, Garcia ML, Lopez-Lahoya J, Ugarte M, Valdivieso F.	Glycine transport in rat brain and liver mitochondria.	Biochim Biophys Acta	1980	7388024	- Added by RS/TV - No genes found. - The data presented in this report demonstrate that glycine is taken up by brain and liver mitochondria by a carrier-mediated process without requiring energy. (Benavides J, Garcia ML, Lopez-Lahoya J, Ugarte M, Valdivieso F. Biochim Biophys Acta. 1980 Jun 6;598(3):588-94.)

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GLYVESSEC	3	Gasnier B	The SLC32 transporter, a key protein for the synaptic release of inhibitory amino acids	Pflugers Arch	2004	12750892	This represents the vesiclar secretion of the compound. The reaction is written as a net secretion reaction--the ATP would actually drive a proton pump, creating a gradient that is used to concentrate a compound in a secretory vesicle. from PMID 12750892: There is evidence that glycine is also translocated [3, 19], in agreement with previous studies showing that GABA and glycine compete for uptake into synaptic vesicles [2, 5]. Similar data suggest that b-alanine is another substrate [5]. Although not proven, a 1:1 stoichiometry of H+ and amino acid is usually postulated and would concentrate enough transmitter in the vesicles because of the high cytosolic concentration of GABA and glycine at nerve terminals. This abundance is also consistent with the low affinity of VIAAT for its substrates (~5 and ~25 mM for GABA and glycine, respectively [15]). From PMID 15383652: In addition to this intrinsic dependency on the transmembrane electrochemical gradient, the transport rate can also be modulated by alterations in the rate of ATP hydrolysis and its coupling to H+ translocation. A recent detailed analysis of curr
GMPR	3	Murano I, Tsukahara M, Kajii T, Yoshida A.	Mapping of the human guanosine monophosphate reductase gene (GMPR) to chromosome 6p23 by fluorescence in situ hybridization.	Genomics	1994	8188226	0
GMPR	3	Deng Y, Wang Z, Ying K, Gu S, Ji C, Huang Y, Gu X, Wang Y, Xu Y, Li Y, Xie Y, Mao Y.	NADPH-dependent GMP reductase isoenzyme of human (GMPR2). Expression, purification, and kinetic properties.	Int J Biochem Cell Biol	2002	12009299	0
GMPS2	3	Nakamura J, Lou L.	Biochemical characterization of human GMP synthetase.	J Biol Chem	1995	7706277	IT GeneCards: needs MG2+ homodimer
GND	3	Tsui SK, Chan JY, Waye MM, Fung KP, Lee CY	Identification of a cDNA encoding 6-phosphogluconate dehydrogenase from a human heart cDNA library.	Biochem Genet	1996	8978909	5226: - cDNA isolated from adult heart library [Tsui 1996] - 94.2% identity to sheep aa seq [Tsui 1996]
GNMT	3	Chen YM, Chen LY, Wong FH, Lee CM, Chang TJ, Yang-Feng TL.	Genomic structure, expression, and chromosomal localization of the human glycine N-methyltransferase gene.		2000	10843803	abundant in liver cytosolic according to GeneCards
GNMT	3	Luka Z, Cerone R, Phillips JA 3rd, Mudd HS, Wagner C.	Mutations in human glycine N-methyltransferase give insights into its role in methionine metabolism.		2002	11810299	abundant in liver cytosolic according to GeneCards
GPIAT	3	Murakami Y, Siripanyapinyo U, Hong Y, Kang JY, Ishihara S, Nakakuma H, Maeda Y, Kinoshita T	PIG-W is critical for inositol acylation but not for flipping of glycosylphosphatidylinositol-anchor	Mol Biol Cell	2003	14517336	- identification, cloning and expression of gene [Murakami, Mol Biol Cell 2003] - knockout was complemented by S. cerevisiae and S. pombe homologs [Murakami, Mol Biol Cell 2003] - enzyme is localized in ER membrane but reaction takes place on cytosolic side [Murakami, Mol Biol Cell 2003]
GPIDAer	3	Tanaka S, Maeda Y, Tashima Y, Kinoshita T	Inositol deacylation of glycosylphosphatidylinositol-anchored proteins is mediated by mammalian PGAP1 and yeast Bst1p	J Biol Chem	2004	14734546	- gene was cloned, encodes an ER-associated GPI inositol-deacylase [Tanaka, J Biol Chem 2004] - inositol deacylation occurs in the ER soon after GPI-anchor attachment; as a result, the acyl group is usually absent from GPI-anchored proteins on the cell surface [Tanaka, J Biol Chem 2004] - this is an important step for efficient transport of GPI-anchored proteins from the ER to the Golgi [Tanaka, J Biol Chem 2004] - note: the inositol remains acylated in human erythrocytes [Tanaka, J Biol Chem 2004]
GPIMTer_L	3	Maeda Y, Watanabe R, Harris CL, Hong Y, Ohishi K, Kinoshita K, Kinoshita T	PIG-M transfers the first mannose to glycosylphosphatidylinositol on the lumenal side of the ER	EMBO J	2001	11226175	93183: - transfers the first mannose to GPI on the lumenal side of the endoplasmic reticulum [RefSeq] - identification of gene for cell line defective in activity [Maeda, EMBO J 2001] - gene has 38 and 35% similarity w/ C. elegans and S. cerevisiae, respectively [Maeda, EMBO J 2001] - catalytic domain on luminal side [Maeda, EMBO J 2001] 54965: - gene was cloned; has 77% identify to rat homolog [Ashida, Mol Biol Cell 2005] - associates w/ and stabilizes PIG-M [Ashida, Mol Biol Cell 2005]
GPIMTer_L	3	Ashida H, Hong Y, Murakami Y, Shishioh N, Sugimoto N, Kim YU, Maeda Y, Kinoshita T	Mammalian PIG-X and yeast Pbn1p are the essential components of glycosylphosphatidylinositol-mannosyltransferase I	Mol Biol Cell	2005	15635094	93183: - transfers the first mannose to GPI on the lumenal side of the endoplasmic reticulum [RefSeq] - identification of gene for cell line defective in activity [Maeda, EMBO J 2001] - gene has 38 and 35% similarity w/ C. elegans and S. cerevisiae, respectively [Maeda, EMBO J 2001] - catalytic domain on luminal side [Maeda, EMBO J 2001] 54965: - gene was cloned; has 77% identify to rat homolog [Ashida, Mol Biol Cell 2005] - associates w/ and stabilizes PIG-M [Ashida, Mol Biol Cell 2005]

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GTHO	2	Kelner MJ, Montoya MA.	Structural organization of the human glutathione reductase gene: determination of correct cDNA sequence and identification of a mitochondrial leader sequence.	Biochem Biophys Res Commun	2000	10708558	-biochemically shown to have both cytosolic and mitochondrial forms (see citation)
GTHOm	2	Tramer F, Caponecchia L, Sgro P, Martinelli M, Sandri G, Panfili E, Lenzi A, Gandini L.	Native specific activity of glutathione peroxidase (GPx-1), phospholipid hydroperoxide glutathione peroxidase (PHGPx) and glutathione reductase (GR) does not differ between normo- and hypomotile human sperm samples.	Int J Androl	2004	15149466	-biochemically shown to have both cytosolic and mitochondrial forms (see citation) - additional information added by RS/TV 1) Mitochondrial & cytosol enzyme. 2) Glutathione reductase is a major component of cellular defense mechanisms against oxidative injury. according to Kelner MJ, Montoya MA. Biochem Biophys Res Commun. 2000 Mar 16;269(2):366-8. 3) Catalytic activity: Glutathione reductase is often considered the 'ancillary' enzyme necessary for restoring oxidized glutathione at the expense of the NADPH/NADP+ couple according to Tramer F, Caponecchia L, Sgro P, Martinelli M, Sandri G, Panfili E, Lenzi A, Gandini L. Int J Androl. 2004 Apr;27(2):88-93.
GTHP	3	Metzler, David E	Biochemistry : the chemical reactions of living cells 2 ed vol 1		2001		TV (6/1/2005) Rabilloud, 2002 Peroxiredoxins are a class of enzymes, similar to glutathione peroxidase, reduces perodixes to either alcohol or water. Its substrates are not specific, and since we don't have other ROS other than h2o2, I used h2o2 as a representative. Additional unique characteristic of this enz is that its cys residue gets oxidized, and later is supposedly reduced by thioredoxin. The reduction by thioredoxin is speculative based on yeast.
GTHPe	3	Takahashi K, Avissar N, Whitin J, Cohen H.	Purification and characterization of human plasma glutathione peroxidase: a selenoglycoprotein distinct from the known cellular enzyme	Arch Biochem Biophys	1987	3619451	0
GTHPm	3	Borchert A, Savaskan NE, Kuhn H.	Regulation of expression of the phospholipid hydroperoxide/sperm nucleus glutathione peroxidase gene. Tissue-specific expression pattern and identification of functional cis- and trans-regulatory elements.	J Biol Chem	2003	12427732	- Additional information added by RS/TV: 1) Shlafer M, Myers CL, Adkins S.; Mitochondrial hydrogen peroxide generation and activities of glutathione peroxidase and superoxide dismutase following global ischemia.; J Mol Cell Cardiol. 1987 Dec;19(12):1195-206. 2) Mitochondrial & cytoplasmic according to gene cards. 3) The production of ROS is regulated by a number of antioxidant enzymes within the mitochondria. Which include phospholipid hydroperoxide glutathione peroxidase. (Mitochondrial) 4) Glutathione peroxidases (GPx)1 constitute a family of antioxidative enzymes that are capable of reducing organic and inorganic hydroperoxides to the corresponding hydroxy compounds utilizing glutathione or other hydrogen donors as reducing equivalents. 5) Gpx4 is expressed in small small amounts in many cells and tissues, but at much higher levels in testis. 6) Gpx 4 is also known to reside in the cytoplasm. All this according to Borchert A, Savaskan NE, Kuhn H.J Biol Chem. 2003 Jan 24;278(4):2571-80. Epub 2002 Nov 08.
GTHRDt	2	J Mårtensson, J C Lai, and A Meister	High-affinity transport of glutathione is part of a multicomponent system essential for mitochondrial function.	Proc Natl Acad Sci U S A	1990		- Added by RS/TV - No genes found as of yet for this transporter. - findings strongly indicate a multicomponent transport system that includes a high-affinity component that functions at very low external GSH levels (J Mårtensson, J C Lai, and A Meister. Proc Natl Acad Sci U S A. 1990 September; 87(18): 7185 7189.)
GTHS	3	Dinescu A, Cundari TR, Bhansali VS, Luo JL, Anderson ME.	Function of conserved residues of human glutathione synthetase: implications for the ATP-grasp enzymes.	J Biol Chem	2004	14990577	- Extra information added by RS/TV: -Meister, A, Mitochondrial changes associated with glutathione deficiency, Biochimica et Biophysica Acta 1271 (1995) 35-42. - Gluthione synthetase catalyzes the second and final step in the biosynthesis of gluthione from gamma-glutamylcysteine and glycine in an ATP dependent manner. - Cytosolic enzyme (gluthione is predominantly found in the cytosol) All according to Dinescu A, Cundari TR, Bhansali VS, Luo JL, Anderson ME. J Biol Chem. 2004 May 21;279(21):22412-21. Epub 2004 Feb 27.

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GTMLTe	3	Hultberg B, Hultberg M	High glutathione turnover in human cell lines revealed by acivicin inhibition of gamma-glutamyltranspeptidase and the effects of thiol-reactive metals during acivicin inhibition	Clin Chim Acta	2004	15469854	parallel reaction and transport may work with other amino acids also intracellular ala-L assumed but is the least certain part of this reaction
GTPCI	3	Thony B, Auerbach G, Blau N.	Tetrahydrobiopterin biosynthesis, regeneration and functions.	Biochem J	2000	10727395	IT
GUACYC	3	Giuili G, Scholl U, Bulle F, Guellaen G.	Molecular cloning of the cDNAs coding for the two subunits of soluble guanylyl cyclase from human brain.	FEBS Lett	1992	1352257	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Arden KC, Viars CS, Weiss S, Argentin S, Nemer M.	Localization of the human B-type natriuretic peptide precursor (NPPB) gene to chromosome 1p36.	Genomics	1995	7601467	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Sugawara T, Holt JA, Driscoll D, Strauss JF 3rd, Lin D, Miller WL, Patterson D, Clancy KP, Hart IM, Clark BJ, et al.	Human steroidogenic acute regulatory protein: functional activity in COS-1 cells, tissue-specific expression, and mapping of the structural gene to 8p11.2 and a pseudogene to chromosome 13.	Proc Natl Acad Sci U S A	1995	7761400	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Lowe DG, Dizhoor AM, Liu K, Gu Q, Spencer M, Laura R, Lu L, Hurley JB.	Cloning and expression of a second photoreceptor-specific membrane retina guanylyl cyclase (RetGC), RetGC-2.	Proc Natl Acad Sci U S A	1995	7777544	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Pardhasaradhi K, Kutty RK, Gentleman S, Krishna G.	Expression of mRNA for atrial natriuretic peptide receptor guanylate cyclase (ANPRA) in human retina.	Cell Mol Neurobiol	1994	7954658	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Yang RB, Fulle HJ, Garbers DL.	Chromosomal localization and genomic organization of genes encoding guanylyl cyclase receptors expressed in olfactory sensory neurons and retina.	Genomics	1996	8838319	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)

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GUACYC	3	Kelsell RE, Evans K, Gregory CY, Moore AT, Bird AC, Hunt DM.	Localisation of a gene for dominant cone-rod dystrophy (CORD6) to chromosome 17p.	Hum Mol Genet	1997	9097965	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Zabel U, Weeger M, La M, Schmidt HH.	Human soluble guanylate cyclase: functional expression and revised isoenzyme family.	Biochem J	1998	9742212	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Malterer A, Gupta G, Danziger RS.	Assignment of GUCY1B2, the human homologue of a candidate gene for hypertension, to chromosome bands 13q14.2-->q14.3 by in situ hybridization.	Cytogenet Cell Genet	1999	10449911	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Behrends S, Vehse K.	The beta(2) subunit of soluble guanylyl cyclase contains a human-specific frameshift and is expressed in gastric carcinoma.	Biochem Biophys Res Commun	2000	10777682	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Payne AM, Morris AG, Downes SM, Johnson S, Bird AC, Moore AT, Bhattacharya SS, Hunt DM.	Clustering and frequency of mutations in the retinal guanylate cyclase (GUCY2D) gene in patients with dominant cone-rod dystrophies.	J Med Genet	2001	11565546	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Koptides M, Mean R, Stavrou C, Pierides A, Demetriou K, Nakayama T, Hildebrandt F, Fuchshuber A, Deltas CC.	Novel NPR1 polymorphic variants and its exclusion as a candidate gene for medullary cystic kidney disease (ADMCKD) type 1.	Mol Cell Probes	2001	11851379	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)

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GUACYC	3	Ghanekar Y, Chandrashaker A, Tatu U, Visweswariah SS.	Glycosylation of the receptor guanylate cyclase C: role in ligand binding and catalytic activity.	Biochem J	2004	14748740	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUACYC	3	Potthast R, Abbey-Hosch SE, Antos LK, Marchant JS, Kuhn M, Potter LR.	Calcium-dependent dephosphorylation mediates the hyperosmotic and lysophosphatidic acid-dependent inhibition of natriuretic peptide receptor-B/guanylyl cyclase-B.	J Biol Chem	2004	15371450	Potthast et al 2004 has measured activity for 4882 4881: retina, heart, kindey, others? Gucy1A3B2.1 less active than Gucy1A3B3.1 but functional heterodimer (Zaebl et al, 1998) from 2977 there is an alternative, non-functional spliceform that can inhibit the function of the heterodimer. cDNA 2977 isolated from fetal brain 2984: testis, ovary, lower expression in kidney (sugaware et al 1995) - has a extracellular domain - precursor has been found in ER (Ghanekar et al, 2004) 2986:olfactory sensory neurons and retina (Yang et al, 1996)
GUAD	3	Yuan G, Bin JC, McKay DJ, Snyder FF.	Cloning and characterization of human guanine deaminase. Purification and partial amino acid sequence of the mouse protein.	J Biol Chem	1999	10075721	homodimer binds 1 zinc per subunit IT
GULLACter	1	Banhegyi G, Mandl J	The hepatic glycogenoreticular system	Pathol Oncol Res	2001	11458272	- gulonolactone may possibly be transported into the ER; suggested by [Banhegyi 2001]
H2CO3D	0	Sly WS, Hu PY	Human carbonic anhydrases and carbonic anhydrase deficiencies	Annu Rev Biochem	1995	7574487	- reversible (Entrez) [759.1]: cytosolic; 5 to 6 times more abundant than CA2 in human erythrocytes; expressed in epithelium of the large intestine, corneal epithelium, lens, ciliary body epithelium, sweat glands, adipose tissue, myoepithelial cells, neutrophils and zona glomerulosa cells of adrenal gland (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [760.1]: cytosolic; expression - see paper, has very specific expression data (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [761.1]: cytosolic; expression: skeletal muscle (type I fiber red skeletal muscle), adipose tissue (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [762.1]: CA IV is glycosylphosphatidyl-inositol-anchored membrane isozyme (GPI- linked); expressed: kidney, lung, colon, brain; (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) & (Fleming RE, et al. J Clin Invest. 1995 Dec;96(6):2907-13) [766.1, 766.2]: CA7 is a cytosolic enzyme. The mRNA is primarily expressed in the cytosol of the salivary glands. Functional role remains speculative. (Sly WS, Hu PY. Annu Re [768.1]: CA 9 is a transmembrane protein, with the carbonic an [771.1, 771.2]: Type-1 membrane protein; Expressed in normal [23632.1]: Type-1 transmembrane protein; CA 14 is expressed
H2CO3D	0	Parkkila S, Parkkila AK, Saarnio J, Kivela J, Karttunen TJ, Kaunisto K, Waheed A, Sly WS, Tureci O, Virtanen I, Rajaniemi H.	Expression of the membrane-associated carbonic anhydrase isozyme XII in the human kidney and rena tumors	J Histochem Cytochem	2000	11101628	- reversible (Entrez) [759.1]: cytosolic; 5 to 6 times more abundant than CA2 in human erythrocytes; expressed in epithelium of the large intestine, corneal epithelium, lens, ciliary body epithelium, sweat glands, adipose tissue, myoepithelial cells, neutrophils and zona glomerulosa cells of adrenal gland (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [760.1]: cytosolic; expression - see paper, has very specific expression data (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [761.1]: cytosolic; expression: skeletal muscle (type I fiber red skeletal muscle), adipose tissue (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [762.1]: CA IV is glycosylphosphatidyl-inositol-anchored membrane isozyme (GPI- linked); expressed: kidney, lung, colon, brain; (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) & (Fleming RE, et al. J Clin Invest. 1995 Dec;96(6):2907-13) [766.1, 766.2]: CA7 is a cytosolic enzyme. The mRNA is primarily expressed in the cytosol of the salivary glands. Functional role remains speculative. (Sly WS, Hu PY. Annu Re [768.1]: CA 9 is a transmembrane protein, with the carbonic an [771.1, 771.2]: Type-1 membrane protein; Expressed in normal [23632.1]: Type-1 transmembrane protein; CA 14 is expressed

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
H2CO3D	0	Parkkila S, Parkkila AK, Rajaniemi H, Shah GN, Grubb JH, Waheed A, Sly WS.	Expression of membrane-associated carbonic anhydrase XIV on neurons and axons in mouse and human brain	Proc Natl Acad Sci U S A	2001	11172051	[759.1]: cytosolic; 5 to 6 times more abundant than CA2 in human erythrocytes; expressed in epithelium of the large intestine, corneal epithelium, lens, ciliary body epithelium, sweat glands, adipose tissue, myoepithelial cells, neutrophils and zona glomerulosa cells of adrenal gland (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [760.1]: cytosolic; expression - see paper, has very specific expression data (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [761.1]: cytosolic; expression: skeletal muscle (type I fiber red skeletal muscle), adipose tissue (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [762.1]: CA IV is glycosylphosphatidyl-inositol-anchored membrane isozyme (GPI- linked); expressed: kidney, lung, colon, brain; (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) & (Fleming RE, et al. J Clin Invest. 1995 Dec;96(6):2907-13) [766.1, 766.2]: CA7 is a cytosolic enzyme. The mRNA is primarily expressed in the cytosol of the salivary glands. Functional role remains speculative. (Sly WS, Hu PY. Annu Re [768.1]: CA 9 is a transmembrane protein, with the carbonic an [771.1, 771.2]: Type-1 membrane protein; Expressed in normal [23632.1]: Type-1 transmembrane protein; CA 14 is expressed
H2CO3D	0	Wingo T, Tu C, Laipis PJ, Silverman DN.	The catalytic properties of human carbonic anhydrase IX	Biochem Biophys Res Commun	2001	11676494	[759.1]: cytosolic; 5 to 6 times more abundant than CA2 in human erythrocytes; expressed in epithelium of the large intestine, corneal epithelium, lens, ciliary body epithelium, sweat glands, adipose tissue, myoepithelial cells, neutrophils and zona glomerulosa cells of adrenal gland (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [760.1]: cytosolic; expression - see paper, has very specific expression data (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [761.1]: cytosolic; expression: skeletal muscle (type I fiber red skeletal muscle), adipose tissue (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [762.1]: CA IV is glycosylphosphatidyl-inositol-anchored membrane isozyme (GPI- linked); expressed: kidney, lung, colon, brain; (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) & (Fleming RE, et al. J Clin Invest. 1995 Dec;96(6):2907-13) [766.1, 766.2]: CA7 is a cytosolic enzyme. The mRNA is primarily expressed in the cytosol of the salivary glands. Functional role remains speculative. (Sly WS, Hu PY. Annu Re [768.1]: CA 9 is a transmembrane protein, with the carbonic an [771.1, 771.2]: Type-1 membrane protein; Expressed in normal [23632.1]: Type-1 transmembrane protein; CA 14 is expressed
H2CO3D	0	Lehtonen J, Shen B, Vihinen M, Casini A, Scozzafava A, Supuran CT, Parkkila AK, Saarnio J, Kivela AJ, Waheed A, Sly WS, Parkkila S.	Characterization of CA XIII, a novel member of the carbonic anhydrase isozyme family.	J Biol Chem	2004	14600151	[759.1]: cytosolic; 5 to 6 times more abundant than CA2 in human erythrocytes; expressed in epithelium of the large intestine, corneal epithelium, lens, ciliary body epithelium, sweat glands, adipose tissue, myoepithelial cells, neutrophils and zona glomerulosa cells of adrenal gland (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [760.1]: cytosolic; expression - see paper, has very specific expression data (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [761.1]: cytosolic; expression: skeletal muscle (type I fiber red skeletal muscle), adipose tissue (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [762.1]: CA IV is glycosylphosphatidyl-inositol-anchored membrane isozyme (GPI- linked); expressed: kidney, lung, colon, brain; (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) & (Fleming RE, et al. J Clin Invest. 1995 Dec;96(6):2907-13) [766.1, 766.2]: CA7 is a cytosolic enzyme. The mRNA is primarily expressed in the cytosol of the salivary glands. Functional role remains speculative. (Sly WS, Hu PY. Annu Re [768.1]: CA 9 is a transmembrane protein, with the carbonic an [771.1, 771.2]: Type-1 membrane protein; Expressed in normal [23632.1]: Type-1 transmembrane protein; CA 14 is expressed

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H2CO3D	0	Taniuchi K, Nishimori I, Takeuchi T, Fujikawa-Adachi K, Ohtsuki Y, Onishi S.	Developmental expression of carbonic anhydrase-related proteins VIII, X, and XI in the human brain.	Neuroscience	2002		- reversible (Entrez) [759.1]: cytosolic; 5 to 6 times more abundant than CA2 in human erythrocytes; expressed in epithelium of the large intestine, corneal epithelium, lens, ciliary body epithelium, sweat glands, adipose tissue, myoepithelial cells, neutrophils and zona glomerulosa cells of adrenal gland (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [760.1]: cytosolic; expression - see paper, has very specific expression data (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [761.1]: cytosolic; expression: skeletal muscle (type I fiber red skeletal muscle), adipose tissue (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [762.1]: CA IV is glycosylphosphatidyl-inositol-anchored membrane isozyme (GPI- linked); expressed: kidney, lung, colon, brain; (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) & (Fleming RE, et al. J Clin Invest. 1995 Dec;96(6):2907-13) [766.1, 766.2]: CA7 is a cytosolic enzyme. The mRNA is primarily expressed in the cytosol of the salivary glands. Functional role remains speculative. (Sly WS, Hu PY. Annu Re [768.1]: CA 9 is a transmembrane protein, with the carbonic an [771.1, 771.2]: Type-1 membrane protein; Expressed in normal [23632.1]: Type-1 transmembrane protein; CA 14 is expressed
H2CO3D	0	Fleming RE, Parkkila S, Parkkila AK, Rajaniemi H, Waheed A, Sly WS.	Carbonic anhydrase IV expression in rat and human gastrointestinal tract regional, cellular, and subcellular localization.	J Clin Invest	1995		- reversible (Entrez) [759.1]: cytosolic; 5 to 6 times more abundant than CA2 in human erythrocytes; expressed in epithelium of the large intestine, corneal epithelium, lens, ciliary body epithelium, sweat glands, adipose tissue, myoepithelial cells, neutrophils and zona glomerulosa cells of adrenal gland (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [760.1]: cytosolic; expression - see paper, has very specific expression data (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [761.1]: cytosolic; expression: skeletal muscle (type I fiber red skeletal muscle), adipose tissue (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) [762.1]: CA IV is glycosylphosphatidyl-inositol-anchored membrane isozyme (GPI- linked); expressed: kidney, lung, colon, brain; (Sly WS, Hu PY. Annu Rev Biochem. 1995;64:375-401. Review.) & (Fleming RE, et al. J Clin Invest. 1995 Dec;96(6):2907-13) [766.1, 766.2]: CA7 is a cytosolic enzyme. The mRNA is primarily expressed in the cytosol of the salivary glands. Functional role remains speculative. (Sly WS, Hu PY. Annu Re [768.1]: CA 9 is a transmembrane protein, with the carbonic an [771.1, 771.2]: Type-1 membrane protein; Expressed in normal [23632.1]: Type-1 transmembrane protein; CA 14 is expressed
H2CO3Dm	3	Shah GN, Hewett-Emmett D, Grubb JH, Migas MC, Fleming RE, Waheed A, Sly WS.	Mitochondrial carbonic anhydrase CA VB: differences in tissue distribution and pattern of evolution from those of CA VA suggest distinct physiological roles.	Proc Natl Acad Sci U S A	2000	10677517	- Added by RS/TV Catalytic Activity: The carbonic anhydrases (CAs) are a family of zinc metalloenzymes that catalyze the reversible hydration of CO2 in the reaction CO2 + H2O<--> HCO3- + H+. Both Ca5a.1-m and Ca5b.1-m catalyze this reaction. Tissue Distribution: Ca5a.1-m: Found exclusively in liver tissue. Ca5b.1-m: Found in kidney, liver, muscle, and brain. All according to Shah GN. Proc Natl Acad Sci U S A. 2000 Feb 15;97(4):1677-82.
H2ETer	2	Hong Y, Maeda Y, Watanabe R, Ohishi K, Mishkind M, Riezman H, Kinoshita T	Pig-n, a mammalian homologue of yeast Mcd4p, is involved in transferring phosphoethanolamine to the first mannose of the glycosylphosphatidylinositol	J Biol Chem	1999	10574991	- reaction described in Varki, pg. 136 - expressed in the endoplasmic reticulum and transfers phosphoethanolamine (EtNP) to the first mannose of the GPI anchor [RefSeq] - mouse gene has been cloned [Hong, J Biol Chem 1999]
H2MTer_L	3	Kang JY, Hong Y, Ashida H, Shishioh N, Murakami Y, Morita YS, Maeda Y, Kinoshita T	IG-V involved in transferring the second mannose in glycosylphosphatidylinositol	J Biol Chem	2005	15623507	- reaction described in Varki, pg. 136 - gene was identified and cloned [Kang, J Biol Chem 2005] - Man-GlcN-acylPI inferred as preferred substrate (based on knockout); may also mannosylate EtNP-Man-GlcN-acylPI [Kang, J Biol Chem 2005]
H2O2syn	2	Forteza R, Salathe M, Miot F, Forteza R, Conner GE	Regulated hydrogen peroxide production by Duox in human airway epithelial cells	Am J Respir Cell Mol Biol	2005	15677770	This reaction is supported by at least two references. However, it is not absolutely certain that this is the mechanism that these genes use to make H2O2, so physiological evidence is assigned. These genes make H2O2 that is used in the synthesis of the thyroid hormones T3 and T4.
H2O2syn	2	Ameziane-El-Hassani R, Morand S, Boucher JL, Frapart YM, Apostolou D, Agnandji D, Gnidehou S, Ohayon R, Noel-Hudson MS, Francon J, Lalaoui K, Virion A, Dupuy C	Dual Oxidase-2 Has an Intrinsic Ca2+-dependent H2O2-generating Activity	J Biol Chem	2005	15972824	This reaction is supported by at least two references. However, it is not absolutely certain that this is the mechanism that these genes use to make H2O2, so physiological evidence is assigned. These genes make H2O2 that is used in the synthesis of the thyroid hormones T3 and T4.

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H2O2tp	2	Ohno Y, Gallin JI.	Diffusion of extracellular hydrogen peroxide into intracellular compartments of human neutrophils. Studies utilizing the inactivation of myeloperoxidase by hydrogen peroxide and azide.	J Biol Chem	1985	2989289	- extracellular hydrogen peroxide diffuses into intracellular compartments of human neutrophils [Ohno 1985]
H2Ot	3	Hediger MA, Turk E, Wright EM	Homology of the human intestinal Na+/glucose and Escherichia coli Na+/proline cotransporters	Proc Natl Acad Sci U S A	1989	2490366	6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel; approximately 260 water molecules are transported per 2 Na+:1 Glc [Loo 1999] NOTE: it's a point of depate whether water is transported by SLC5A1 or by osmosis - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs
H2Ot	3	Hirayama BA, Loo DD, Wright EM.	Protons drive sugar transport through the Na+/glucose cotransporter (SGLT1)	J Biol Chem	1994	8063771	6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel; approximately 260 water molecules are transported per 2 Na+:1 Glc [Loo 1999] NOTE: it's a point of depate whether water is transported by SLC5A1 or by osmosis - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs
H2Ot	3	Quick M, Loo DD, Wright EM	Neutralization of a conserved amino acid residue in the human Na+/glucose transporter (hSGLT1) generates a glucose-gated H+ channel	J Biol Chem	2001	11024018	6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel; approximately 260 water molecules are transported per 2 Na+:1 Glc [Loo 1999] NOTE: it's a point of depate whether water is transported by SLC5A1 or by osmosis - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs
H2Ot	3	Leung DW, Loo DD, Hirayama BA, Zeuthen T, Wright EM	Urea transport by cotransporters	J Physiol	2000	11034615	6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel; approximately 260 water molecules are transported per 2 Na+:1 Glc [Loo 1999] NOTE: it's a point of depate whether water is transported by SLC5A1 or by osmosis - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs
H2Ot	3	Wright EM, Turk E	The sodium/glucose cotransport family SLC5.	Pflugers Arch	2004	12748858	6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel; approximately 260 water molecules are transported per 2 Na+:1 Glc [Loo 1999] NOTE: it's a point of depate whether water is transported by SLC5A1 or by osmosis - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs
H2Ot	3	Wright EM, Hirayama BA, Loo DDF, Turk E, Hager K	Intestinal sugar transport		1994		6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel; approximately 260 water molecules are transported per 2 Na+:1 Glc [Loo 1999] NOTE: it's a point of depate whether water is transported by SLC5A1 or by osmosis - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs

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H2Otm	3	Calamita G, Ferri D, Gena P, Liquori GE, Cavalier A, Thomas D, Svelto M.	The inner mitochondrial membrane has aquaporin-8 water channels and is highly permeable to water.	J Biol Chem	2005	15749715	- Added by RS/TV - It is suggested that AQP8-mediated water transport may be particularly important for rapid expansions of mitochondrial volume. - Immunoblotting, electron microscopy and biophysical studies show the presence of AQP8. - In this study was found in the rat liver, but also found in the mitochondria of other organs. (Calamita G, Ferri D, Gena P, Liquori GE, Cavalier A, Thomas D, Svelto M. J Biol Chem. 2005 Mar 4)
H4ET3er	3	Inoue N, Kinoshita T, Orii T, Takeda J	Cloning of a human gene, PIG-F, a component of glycosylphosphatidylinositol anchor biosynthesis, by a novel expression cloning strategy	J Biol Chem	1993	8463218	- reaction described in Varki, pg. 136 - PIGF and PIGO transfer ethanolaminephosphate to the third mannose in GPI [RefSeq] - PIGF and PIGO form a complex [UniProt], [Hong, J Biol Chem 2000] 5281: - gene was cloned [Inoue, J Biol Chem 1993] 84720: - mouse gene was cloned [Hong, J Biol Chem 2000]
H4ET3er	3	Hong Y, Maeda Y, Watanabe R, Inoue N, Ohishi K, Kinoshita T	Requirement of PIG-F and PIG-O for transferring phosphoethanolamine to the third mannose in glycosylphosphatidylinositol	J Biol Chem	2000	10781593	- reaction described in Varki, pg. 136 - PIGF and PIGO transfer ethanolaminephosphate to the third mannose in GPI [RefSeq] - PIGF and PIGO form a complex [UniProt], [Hong, J Biol Chem 2000] 5281: - gene was cloned [Inoue, J Biol Chem 1993] 84720: - mouse gene was cloned [Hong, J Biol Chem 2000]
H6'ET2er	3	Shishioh N, Hong Y, Ohishi K, Ashida H, Maeda Y, Kinoshita T	GPI7 is the second partner of PIG-F and involved in modification of glycosylphosphatidylinositol	J Biol Chem	2005	15632136	- reaction described in Varki, pg. 136 - gene was cloned, function inferred from knockout [Shishioh, J Biol Chem 2005] - enzyme complex consisting of GPI7 and PIG-F is involved in the conversion of EtNP-Man-Man-(EtNP)Man-GlcN-(acyl)PI to EtNP-Man-(EtNP)Man-(EtNP)Man-GlcN-(acyl)PI [Shishioh, J Biol Chem 2005]
H8TAer	3	Yu J, Nagarajan S, Knez JJ, Udenfriend S, Chen R, Medof ME	The affected gene underlying the class K glycosylphosphatidylinositol (GPI) surface protein defect codes for the GPI transamidase	Proc Natl Acad Sci U S A	1997	9356492	- minor structure transfered to proteins as GPI anchor [Shishioh, J Biol Chem 2005] - transamidase is complex with GPAA1, PIGK/GPI8, PIGT, PIGU and PIGS [UniProt] 8733: - functions in GPI transfer [RefSeq] - uniquitous expression [UniProt], [Hiroi, FEBS Lett 1998] - gene has been cloned [Hiroi, FEBS Lett 1998] - 25% identity to yeast homolog [Hiroi, FEBS Lett 1998] 10026: - gene was identified [Yu, PNAS 1997] - active site characterized [Meyer, Biochemistry 2000] 94005, 51604: - gene was cloned, demonstrated to be essential to complex [Ohishi, EMBO J 2001] 128869: - gene was cloned, demonstrated to be part of complex [Hong, Mol Biol Cell 2003]
H8TAer	3	Hiroi Y, Komuro I, Chen R, Hosoda T, Mizuno T, Kudoh S, Georgescu SP, Medof ME, Yazaki Y	Molecular cloning of human homolog of yeast GAA1 which is required for attachment of glycosylphosphatidylinositols to proteins	FEBS Lett	1998	9468317	- minor structure transfered to proteins as GPI anchor [Shishioh, J Biol Chem 2005] - transamidase is complex with GPAA1, PIGK/GPI8, PIGT, PIGU and PIGS [UniProt] 8733: - functions in GPI transfer [RefSeq] - uniquitous expression [UniProt], [Hiroi, FEBS Lett 1998] - gene has been cloned [Hiroi, FEBS Lett 1998] - 25% identity to yeast homolog [Hiroi, FEBS Lett 1998] 10026: - gene was identified [Yu, PNAS 1997] - active site characterized [Meyer, Biochemistry 2000] 94005, 51604: - gene was cloned, demonstrated to be essential to complex [Ohishi, EMBO J 2001] 128869: - gene was cloned, demonstrated to be part of complex [Hong, Mol Biol Cell 2003]

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H8TAer	3	Meyer U, Benghezal M, Imhof I, Conzelmann A	Active site determination of Gpi8p, a caspase-related enzyme required for glycosylphosphatidylinositol anchor addition to proteins	Biochemistry	2000	10727241	- minor structure transfered to proteins as GPI anchor [Shishioh, J Biol Chem 2005] - transamidase is complex with GPAA1, PIGK/GPI8, PIGT, PIGU and PIGS [UniProt] 8733: - functions in GPI transfer [RefSeq] - uniquitous expression [UniProt], [Hiroi, FEBS Lett 1998] - gene has been cloned [Hiroi, FEBS Lett 1998] - 25% identity to yeast homolog [Hiroi, FEBS Lett 1998] 10026: - gene was identified [Yu, PNAS 1997] - active site characterized [Meyer, Biochemistry 2000] 94005, 51604: - gene was cloned, demonstrated to be essential to complex [Ohishi, EMBO J 2001] 128869: - gene was cloned, demonstrated to be part of complex [Hong, Mol Biol Cell 2003]
H8TAer	3	Ohishi K, Inoue N, Kinoshita T	PIG-S and PIG-T, essential for GPI anchor attachment to proteins, form a complex with GAA1 and GPI8	EMBO J	2001	11483512	- minor structure transfered to proteins as GPI anchor [Shishioh, J Biol Chem 2005] - transamidase is complex with GPAA1, PIGK/GPI8, PIGT, PIGU and PIGS [UniProt] 8733: - functions in GPI transfer [RefSeq] - uniquitous expression [UniProt], [Hiroi, FEBS Lett 1998] - gene has been cloned [Hiroi, FEBS Lett 1998] - 25% identity to yeast homolog [Hiroi, FEBS Lett 1998] 10026: - gene was identified [Yu, PNAS 1997] - active site characterized [Meyer, Biochemistry 2000] 94005, 51604: - gene was cloned, demonstrated to be essential to complex [Ohishi, EMBO J 2001] 128869: - gene was cloned, demonstrated to be part of complex [Hong, Mol Biol Cell 2003]
H8TAer	3	Hong Y, Ohishi K, Kang JY, Tanaka S, Inoue N, Nishimura J, Maeda Y, Kinoshita T	Human PIG-U and yeast Cdc91p are the fifth subunit of GPI transamidase that attaches GPI-anchors to proteins	Mol Biol Cell	2003	12802054	- minor structure transfered to proteins as GPI anchor [Shishioh, J Biol Chem 2005] - transamidase is complex with GPAA1, PIGK/GPI8, PIGT, PIGU and PIGS [UniProt] 8733: - functions in GPI transfer [RefSeq] - uniquitous expression [UniProt], [Hiroi, FEBS Lett 1998] - gene has been cloned [Hiroi, FEBS Lett 1998] - 25% identity to yeast homolog [Hiroi, FEBS Lett 1998] 10026: - gene was identified [Yu, PNAS 1997] - active site characterized [Meyer, Biochemistry 2000] 94005, 51604: - gene was cloned, demonstrated to be essential to complex [Ohishi, EMBO J 2001] 128869: - gene was cloned, demonstrated to be part of complex [Hong, Mol Biol Cell 2003]

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HAS2	3	Itano N, Kimata K.	Molecular cloning of human hyaluronan synthase	Biochem Biophys Res Commun	1996	8651928	from [Varki 1999]: - GlcNAc and GlcA are copolymerized from UDP-GlcNAC and UDP-GlcA - synthesis occurs at plasma membrane; assembles from reducing end, causing its extrusion from cell surface - HA synthesizes can polymerize ~100 monosaccharides/sec in vitro from [Itano 2002]: - each HAS protein can independently synthesize HA - HAS2 synthesizes very large HA molecules; smaller molecules synthesized by HAS1, HAS3 - recombinant HAS1 has higher Km for UDP-GlcNAc and UDP-GlcA than HAS2 and HAS3 3036: - clone isolated from human fetal cDNA library [Itano 1996] - 84.4% and 96.0% identity with murine nucleotide sequence and amino acid sequences [Itano 1996]; 22%, 54%, and 92% identity with S. pyogenes, X. laevis, and murine amino acid sequences [Shyjan 1996] - ubiquitous [Itano 1996]; high in ovary; moderate in spleen, thymus, prostate, testes, large intestine, and heart; weak in small intestine [Shyjan 1996] - isolated & expressed in CHO cells [Shyjan 1996] 3037: - cloned and expressed in human 293 and CHO cells [Watanabe 1996] - 55% amino acid identity with Xenopus DG42 and 52% identi 3038: - isolated partial ORF sequence [Spicer 1997] - 99% conservation between human (partial) and mouse amino
HAS2	3	Watanabe K, Yamaguchi Y	Molecular identification of a putative human hyaluronan synthase	J Biol Chem	1996	8798477	from [Varki 1999]: - GlcNAc and GlcA are copolymerized from UDP-GlcNAC and UDP-GlcA - synthesis occurs at plasma membrane; assembles from reducing end, causing its extrusion from cell surface - HA synthesizes can polymerize ~100 monosaccharides/sec in vitro from [Itano 2002]: - each HAS protein can independently synthesize HA - HAS2 synthesizes very large HA molecules; smaller molecules synthesized by HAS1, HAS3 - recombinant HAS1 has higher Km for UDP-GlcNAc and UDP-GlcA than HAS2 and HAS3 3036: - clone isolated from human fetal cDNA library [Itano 1996] - 84.4% and 96.0% identity with murine nucleotide sequence and amino acid sequences [Itano 1996]; 22%, 54%, and 92% identity with S. pyogenes, X. laevis, and murine amino acid sequences [Shyjan 1996] - ubiquitous [Itano 1996]; high in ovary; moderate in spleen, thymus, prostate, testes, large intestine, and heart; weak in small intestine [Shyjan 1996] - isolated & expressed in CHO cells [Shyjan 1996] 3037: - cloned and expressed in human 293 and CHO cells [Watanabe 1996] - 55% amino acid identity with Xenopus DG42 and 52% identi 3038: - isolated partial ORF sequence [Spicer 1997] - 99% conservation between human (partial) and mouse amino
HAS2	3	Shyjan AM, Heldin P, Butcher EC, Yoshino T, Briskin MJ	Functional cloning of the cDNA for a human hyaluronan synthase	J Biol Chem	1996	8798544	from [Varki 1999]: - GlcNAc and GlcA are copolymerized from UDP-GlcNAC and UDP-GlcA - synthesis occurs at plasma membrane; assembles from reducing end, causing its extrusion from cell surface - HA synthesizes can polymerize ~100 monosaccharides/sec in vitro from [Itano 2002]: - each HAS protein can independently synthesize HA - HAS2 synthesizes very large HA molecules; smaller molecules synthesized by HAS1, HAS3 - recombinant HAS1 has higher Km for UDP-GlcNAc and UDP-GlcA than HAS2 and HAS3 3036: - clone isolated from human fetal cDNA library [Itano 1996] - 84.4% and 96.0% identity with murine nucleotide sequence and amino acid sequences [Itano 1996]; 22%, 54%, and 92% identity with S. pyogenes, X. laevis, and murine amino acid sequences [Shyjan 1996] - ubiquitous [Itano 1996]; high in ovary; moderate in spleen, thymus, prostate, testes, large intestine, and heart; weak in small intestine [Shyjan 1996] - isolated & expressed in CHO cells [Shyjan 1996] 3037: - cloned and expressed in human 293 and CHO cells [Watanabe 1996] - 55% amino acid identity with Xenopus DG42 and 52% identi 3038: - isolated partial ORF sequence [Spicer 1997] - 99% conservation between human (partial) and mouse amino

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
HAS2	3	Spicer AP, Olson JS, McDonald JA	Molecular cloning and characterization of a cDNA encoding the third putative mammalian hyaluronan synthase	J Biol Chem	1997	9083017	from [Yaki 1999]: - GlcNAc and GlcA are copolymerized from UDP-GlcNAC and UDP-GlcA - synthesis occurs at plasma membrane; assembles from reducing end, causing its extrusion from cell surface - HA synthesizes can polymerize ~100 monosaccharides/sec in vitro from [Itano 2002]: - each HAS protein can independently synthesize HA - HAS2 synthesizes very large HA molecules; smaller molecules synthesized by HAS1, HAS3 - recombinant HAS1 has higher Km for UDP-GlcNAc and UDP-GlcA than HAS2 and HAS3 3036: - clone isolated from human fetal cDNA library [Itano 1996] - 84.4% and 96.0% identity with murine nucleotide sequence and amino acid sequences [Itano 1996]; 22%, 54%, and 92% identity with S. pyogenes, X. laevis, and murine amino acid sequences [Shyjan 1996] - ubiquitous [Itano 1996]; high in ovary; moderate in spleen, thymus, prostate, testes, large intestine, and heart; weak in small intestine [Shyjan 1996] - isolated & expressed in CHO cells [Shyjan 1996] 3037: - cloned and expressed in human 293 and CHO cells [Watanabe 1996] - 55% amino acid identity with Xenopus DG42 and 52% identi 3038: - isolated partial ORF sequence [Spicer 1997] - 99% conservation between human (partial) and mouse amino
HAS2	3	Itano N, Kimata K	Mammalian hyaluronan synthases	IUBMB Life	2002	12512858	from [Yaki 1999]: - GlcNAc and GlcA are copolymerized from UDP-GlcNAC and UDP-GlcA - synthesis occurs at plasma membrane; assembles from reducing end, causing its extrusion from cell surface - HA synthesizes can polymerize ~100 monosaccharides/sec in vitro from [Itano 2002]: - each HAS protein can independently synthesize HA - HAS2 synthesizes very large HA molecules; smaller molecules synthesized by HAS1, HAS3 - recombinant HAS1 has higher Km for UDP-GlcNAc and UDP-GlcA than HAS2 and HAS3 3036: - clone isolated from human fetal cDNA library [Itano 1996] - 84.4% and 96.0% identity with murine nucleotide sequence and amino acid sequences [Itano 1996]; 22%, 54%, and 92% identity with S. pyogenes, X. laevis, and murine amino acid sequences [Shyjan 1996] - ubiquitous [Itano 1996]; high in ovary; moderate in spleen, thymus, prostate, testes, large intestine, and heart; weak in small intestine [Shyjan 1996] - isolated & expressed in CHO cells [Shyjan 1996] 3037: - cloned and expressed in human 293 and CHO cells [Watanabe 1996] - 55% amino acid identity with Xenopus DG42 and 52% identi 3038: - isolated partial ORF sequence [Spicer 1997] - 99% conservation between human (partial) and mouse amino
HBZOPT10m	3	Kang DC, Takeshige K, Minakami S.	An intermediate of ubiquinone biosynthesis exists in the microsomal fraction of HepG2 cells.	J Biochem (Tokyo)	1990	1965190	I am not sure about reaction but sequence and physiological evidence for such a reaction it seems that the ubiquinone biosynthesis takes place in mitochondria altough direct evidence is missing but Jonassen and Clarke (2000) found a signal sequence typical for mitochondrial proteins in COQ3 gene sequence. IT
HBZOPT10m	3	Nambudiri AM, Ranganathan S, Rudney H	The role of 3-hydroxy-3-methylglutaryl coenzyme A reductase activity in the regulation of ubiquinone synthesis in human fibroblasts.	J Biol Chem	1980	7380842	I am not sure about reaction but sequence and physiological evidence for such a reaction it seems that the ubiquinone biosynthesis takes place in mitochondria altough direct evidence is missing but Jonassen and Clarke (2000) found a signal sequence typical for mitochondrial proteins in COQ3 gene sequence. IT
HBZOPT10m	3	Forsgren M, Attersand A, Lake S, Grunler J, Swiezewska E, Dallner G, Climent I.	Isolation and functional expression of human COQ2, a gene encoding a polyprenyl transferase involved in the synthesis of CoQ.	Biochem J	2004	15153069	I am not sure about reaction but sequence and physiological evidence for such a reaction it seems that the ubiquinone biosynthesis takes place in mitochondria altough direct evidence is missing but Jonassen and Clarke (2000) found a signal sequence typical for mitochondrial proteins in COQ3 gene sequence. IT

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HCO3_CLt	3	Parker MD, Ourmozdi EP, Tanner MJ.	Human BTR1, a new bicarbonate transporter superfamily member and human AE4 from kidney.		2001	11302728	Disease: defects in slc4a1 are a cause of hereditary elliptocytosis (he) [mim:109270, 166900, 130600]; also known as hereditary ovalocytosis (HO). It is a genetically heterogeneous, autosomal dominant disorder. It is characterized by variable hemolytic anemia and elliptical or oval red cell shape. Ovalocytosis/elliptocytosis due to SLC4A1 defects is rhesus- unliked (elliptocytosis 2 or EL2). It can also be referred to as elliptocytosis 4 (EL4). Disease: defects in slc4a1 are a cause of hereditary spherocytosis (hs) [mim:109270]. HS is a hematologic disorder leading to chronic hemolytic anemia and characterized by numerous abnormally shaped erythrocytes which are generally spheroidal. Disease: defects in slc4a1 are a cause of familial distal renal tubular acidosis (drta) [mim:179800]. This disease is characterized by reduced ability to acidify urine, variable hyperchloremic hypokalemic metabolic acidosis, nephrocalcinosis, and nephrolithiasis. Inheritance is generally autosomal dominant, but recessive forms have also been reported. MM
HCO3_CLt	3	Alper SL, Darman RB, Chernova MN, Dahl NK.	The AE gene family of Cl/HCO3- exchangers.		2003	12027221	Disease: defects in slc4a1 are a cause of hereditary elliptocytosis (he) [mim:109270, 166900, 130600]; also known as hereditary ovalocytosis (HO). It is a genetically heterogeneous, autosomal dominant disorder. It is characterized by variable hemolytic anemia and elliptical or oval red cell shape. Ovalocytosis/elliptocytosis due to SLC4A1 defects is rhesus- unliked (elliptocytosis 2 or EL2). It can also be referred to as elliptocytosis 4 (EL4). Disease: defects in slc4a1 are a cause of hereditary spherocytosis (hs) [mim:109270]. HS is a hematologic disorder leading to chronic hemolytic anemia and characterized by numerous abnormally shaped erythrocytes which are generally spheroidal. Disease: defects in slc4a1 are a cause of familial distal renal tubular acidosis (drta) [mim:179800]. This disease is characterized by reduced ability to acidify urine, variable hyperchloremic hypokalemic metabolic acidosis, nephrocalcinosis, and nephrolithiasis. Inheritance is generally autosomal dominant, but recessive forms have also been reported. MM
HCO3_CLt	3	Romero MF, Fulton CM, Boron WF.	The SLC4 family of HCO 3 - transporters.		2004	14722772	Disease: defects in slc4a1 are a cause of hereditary elliptocytosis (he) [mim:109270, 166900, 130600]; also known as hereditary ovalocytosis (HO). It is a genetically heterogeneous, autosomal dominant disorder. It is characterized by variable hemolytic anemia and elliptical or oval red cell shape. Ovalocytosis/elliptocytosis due to SLC4A1 defects is rhesus- unliked (elliptocytosis 2 or EL2). It can also be referred to as elliptocytosis 4 (EL4). Disease: defects in slc4a1 are a cause of hereditary spherocytosis (hs) [mim:109270]. HS is a hematologic disorder leading to chronic hemolytic anemia and characterized by numerous abnormally shaped erythrocytes which are generally spheroidal. Disease: defects in slc4a1 are a cause of familial distal renal tubular acidosis (drta) [mim:179800]. This disease is characterized by reduced ability to acidify urine, variable hyperchloremic hypokalemic metabolic acidosis, nephrocalcinosis, and nephrolithiasis. Inheritance is generally autosomal dominant, but recessive forms have also been reported. MM
HCO3_NAt	3	Wang CZ, Yano H, Nagashima K, Seino S.	The Na+-driven Cl-/HCO3- exchanger. Cloning, tissue distribution, and functional characterization.		2000	10993873	0
HDCAtr	2	Schaffer JE.	Fatty acid transport: the roads taken.	Am J Physiol Endocrinol Metab	2002	11788354	General mechanism - diffusional transport Fatty acids are transported as a result of multiple mechanisms - generally accepted that there are 2 main types of transporters: diffusional and protein mediated (e.g. FATP, OCTN, etc). Diffusional transport is concentration dependent (micromolar), whereease protein mediated transport can occur at lower concentrations (nanomolar). For both models transport is proposed to be bidirectional (the direction-determining step is FA activation once inside the cytosol. See Schaffer review (PMID: 11788354) and Pownall manuscript (PMID: 12864740) for further details. NJ

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
HDCAtr	2	Pownall HJ, Hamilton JA.	Energy translocation across cell membranes and membrane models.	Acta Physiol Scand	2003	12864740	General mechanism - diffusional transport Fatty acids are transported as a result of multiple mechanisms - generally accepted that there are 2 main types of transporters: diffusional and protein mediated (e.g. FATP, OCTN, etc). Diffusional transport is concentration dependent (micromolar), wherease protein mediated transport can occur at lower concentrations (nanomolar). For both models transport is proposed to be bidirectional (the direction-determining step is FA activation once inside the cytosol. See Schaffer review (PMID: 11788354) and Pownall manuscript (PMID: 12864740) for further details. NJ
HEX1	2	Orten JM, Neuhaus OW.	Human Biochemistry		1975		- irreversible under physiological conditions [Orten and Neuhaus, Human Biochem 1975] 2645: - outer mitochondrial membrane [RefSeq] - catalyzes initial step in utilization of glucose by the beta-cell and liver [UniProt] 3098: -Protein encoded by transcript 1 (NM_000188) is associated w/ outer mitochondrial membrane. Other transcripts enode cytosolic isoforms. [RefSeq] -Tissue localization from RefSeq, UniProt 3099: - predominant form found in skeletal muscle - outer membrane of mitochondria [RefSeq] 3101: - localized to lung, liver, placenta [Furuta et al, Genomics 1996] - soluble, cytosolic [HInv] 80201: - mito membrane [HInv]
HIBDm	3	Kedishvili NY, Popov KM, Jaskiewicz JA, Harris RA.	Coordinated expression of valine catabolic enzymes during adipogenesis: analysis of activity, mRNA, protein levels, and metabolic consequences.		1994	7527207	0
HIBDm	3	Sweetman, L, Williams, JC	Branched chain organic acidurias	The Metabolic and Molecular Bases of Inherited Disease, 8th ed (Scriver CR, et al, editors)	2001		0
HISD	3	Suchi M, Sano H, Mizuno H, Wada Y.	Molecular cloning and structural characterization of the human histidase gene (HAL).	Genomics	1995	8530107	Histidase (EC 4.3.1.3) is a cytosolic enzyme that catalyzes the nonoxidative deamination of histidine to urocanic acid. Histidinemia, resulting from reduced histidase activity as reported in Cambridge stock his/his mice and in humans, is the most frequent inborn metabolic error in Japan. 8530107
HISDC	3	LOVENBERG W, WEISSBACH H, UDENFRIEND S.	Aromatic L-amino acid decarboxylase.	J Biol Chem	1962	14466899	Textbook reaction.
HIStN1	3	Fei YJ, Sugawara M, Nakanishi T, Huang W, Wang H, Prasad PD, Leibach FH, Ganapathy V	Primary structure, genomic organization, and functional and electrogenic characteristics of human system N 1, a Na+- and H+-coupled glutamine transporter	J Biol Chem	2000	10823827	From first citation: "These data suggest that SN1 mediates the influx of two Na(+) and one amino acid substrate per transport cycle coupled to the efflux of one H(+), rendering the transport process electrogenic." At least somewhat resistant to lithium
HIStN1	3	Mackenzie B, Erickson JD	Sodium-coupled neutral amino acid (System N/A) transporters of the SLC38 gene family	Pflugers Arch	2004	12845534	From first citation: "These data suggest that SN1 mediates the influx of two Na(+) and one amino acid substrate per transport cycle coupled to the efflux of one H(+), rendering the transport process electrogenic." At least somewhat resistant to lithium
HKt	3	Maeda M, Oshiman K, Tamura S, Futai M.	Human gastric (H+ + K+)-ATPase gene. Similarity to (Na+ + K+)-ATPase genes in exon/intron organization but difference in control region.	J Biol Chem	1990	2160952	- Added by RS/TV There are primarily two types of H+/K+ ATPases: (1) Gastric - Gastric H+/K+ ATPase is located in a unique membrane system in parietal cells, is responsible for secretion of acid into the stomach lumen. (Stewart LA, van Driel IR, Toh BH, Gleeson PA. Glycobiology. 1999 Jun;9(6):601-6.) - Also located in cytoplasmic vesicles or apical plasma membranes of the secretory canaliculus (Maeda M, Oshiman K, Tamura S, Futai M. J Biol Chem. 1990 Jun 5;265(16):9027-32.) - Known to be expressed in the kidney as well (Callaghan JM, Tan SS, Khan MA, Curran KA, Campbell WG, Smolka AJ, Toh BH, Gleeson PA, Wingo CS, Cain BD, et al. Am J Physiol. 1995 Mar;268(3 Pt 2):F363-74.) - composed of an alpha and beta subunit [Entrez]

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HKt	3	Callaghan JM, Tan SS, Khan MA, Curran KA, Campbell WG, Smolka AJ, Toh BH, Gleeson PA, Wingo CS, Cain BD, et al.	Renal expression of the gene encoding the gastric H(+)-K(+)-ATPase beta-subunit.	Am J Physiol	1995	7900835	- Added by RS/TV There are primarily two types of H+/K+ ATPases: (1) Gastric - Gastric H+/K+ ATPase is located in a unique membrane system in parietal cells, is responsible for secretion of acid into the stomach lumen. (Stewart LA, van Driel IR, Toh BH, Gleeson PA. Glycobiology. 1999 Jun;9(6):601-6.) - Also located in cytoplasmic vesicles or apical plasma membranes of the secretory canaliculus (Maeda M, Oshiman K, Tamura S, Futai M. J Biol Chem. 1990 Jun 5;265(16):9027-32.) - Known to be expressed in the kidney as well (Callaghan JM, Tan SS, Khan MA, Curran KA, Campbell WG, Smolka AJ, Toh BH, Gleeson PA, Wingo CS, Cain BD, et al. Am J Physiol. 1995 Mar;268(3 Pt 2):F363-74.) - composed of an alpha and beta subunit [Entrez]
HKYNH	3	Toma S, Nakamura M, Tone S, Okuno E, Kido R, Breton J, Avanzi N, Cozzi L, Speciale C, Mostardini M, Gatti S, Benatti L	Cloning and recombinant expression of rat and human kynureninase	FEBS Lett	1997	9180257	Reaction and gene in intro of citation.
HMBS	2	Raich N, Romeo PH, Dubart A, Beaupain D, Cohen-Solal M, Goossens M.	Molecular cloning and complete primary sequence of human erythrocyte porphobilinogen deaminase.	Nucleic Acids Res	1986	2875434	- Added by RS/TV - Porphobilinogen deaminase (Hydroxymethylbilane synthase) (HMBS), catalyzes the head to tail condensation of four molecules of the monopyrrole porphobilinogen, to form the linear tetrapyrrole, hydroxymethylbilane. Catalytic activity also specified by Gene cards. (Raich N, Romeo PH, Dubart A, Beaupain D, Cohen-Solal M, Goossens M. Nucleic Acids Res. 1986 Aug 11;14(15):5955-68) - Although widely distributed in tissues, the enzymes of the heme biosynthesis pathways are particularly active in the liver.
HMGCOARr	3	Luskey KL, Stevens B.	Human 3-hydroxy-3-methylglutaryl coenzyme A reductase. Conserved domains responsible for catalytic activity and sterol-regulated degradation.	J Biol Chem	1985	2991281	ER version - see Olivier and Krisans 2000 ref see also PMID: 2991281 for localization and catalytic activity. NJ
HMGCOASim	3	Mascaro C, Buesa C, Ortiz JA, Haro D, Hegardt FG.	Molecular cloning and tissue expression of human mitochondrial 3-hydroxy-3-methylglutaryl-CoA synthase.	Arch Biochem Biophys	1995	7893153	mit - uniprot pathway for HMG-CoA synthesis for xol specificity: Liver and kidney NJ - Additional information by RS/TV 1) HMG-Coa catalyzes the condensation of acetyl-CoA and acetoacetyl-CoA, yielding Hmg-CoA and CoA. 2) Mitochondrial enzyme 3) Tissue Expression: Expressed highly in liver and colon; low in testis, kidney, heart, and skeletal muscles; very faint in pancreas. 1 through 3 according to Mascaro C, Buesa C, Ortiz JA, Haro D, Hegardt FG. Arch Biochem Biophys. 1995 Mar 10;317(2):385-90.
HMGCOAtm	2	Kovacs WJ, Krisans S.	Cholesterol biosynthesis and regulation: role of peroxisomes.	Adv Exp Med Biol	2003	14713247	Cholesterol biosynthetic pathway requires HMGCOA to be able to move between intracellular compartments (notably peroxisome <-> mitochondria). NJ
HMGCOAtm	2	Biardi L, Krisans K	Compartmentalization of cholesterol biosynthesis	Journal of Biological Chemistry	1996		Cholesterol biosynthetic pathway requires HMGCOA to be able to move between intracellular compartments (notably peroxisome <-> mitochondria). NJ
HPCLx	3	Foulon V, Antonenkov VD, Croes K, Waelkens E, Mannaerts GP, Van Veldhoven PP, Casteels M.	Purification, molecular cloning, and expression of 2-hydroxyphytanoyl-CoA lyase, a peroxisomal thiamine pyrophosphate-dependent enzyme that catalyzes the carbon-carbon bond cleavage during alpha-oxidation of 3-methyl-branched fatty acids.	Proc Natl Acad Sci U S A	1999	10468558	localization: peroxisome (uniprot) specificity: The compound pristanal is the fatty acid analog of pristanic acid. Pristanal is not in PubChem or KEGG, MW and charge inferred from reaction (lyase reaction producing formyl-CoA). Homotetramer catalyzing a carbon-carbon cleavage reaction; cleaves a 2-hydroxy-3-methylacyl-CoA into formyl-CoA and a 2-methyl-branched fatty aldehyde. Reaction specificty see PMID: 10468558 NJ
HPYRDC	2	Rofe AM, James HM, Bais R, Conyers RA	Hepatic oxalate production: the role of hydroxypyruvate	Biochem Med Metab Biol	1986	3778681	- reaction characterized in rat liver hepatocytes [Rofe, Biochem Med Metab Biol 1986] - activity found in mitochondrial and cytosolic fractions [Rofe, Biochem Med Metab Biol 1986]
HPYRR2x	2	Williams HE, Smith LH Jr.	L-glyceric aciduria. A new genetic variant of primary hyperoxaluria	N Eng J Med	1968	5635456	- catalyzed by lactate dehydrogenase [Cregeen, Human Mutat 2003], [Williams, N Eng J Med 1968]
HPYRR2x	2	Cregeen DP, Williams EL, Hulton S, Rumsby G	Molecular analysis of the glyoxylate reductase (GRHPR) gene and description of mutations underlying primary hyperoxaluria type 2	Hum Mutat	2003	14635115	- catalyzed by lactate dehydrogenase [Cregeen, Human Mutat 2003], [Williams, N Eng J Med 1968]

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HPYRtp	2	Johnson SA, Rumsby G, Cregeen D, Hulton SA	Primary hyperoxaluria type 2 in children.	Pediatr Nephrol	2002	12185464	- reaction shown on Mayo Clinic website at: http://mayoresearch.mayo.edu/mayo/research/nephrology/hyper oxaluria.cfm - also shown in Fig 1 of [Johnson 2002]
HS1ly	3	Scott HS, Blanch L, Guo XH, Freeman C, Orsborn A, Baker E, Sutherland GR, Morris CP, Hopwood JJ	Cloning of the sulphamidase gene and identification of mutations in Sanfilippo A syndrome	Nat Genet	1995	7493035	- O-sulfates have to be removed before sulfamidase desulfates amino group of GlcN [Winchester 1996] 6448: - lysosomal [UniProt] - cDNA was isolated and expressed [Scott, Nat Genet 1995] - recombinantly expressed in CHO cells, kinetic properties characterized [Bielicki, Biochem J 1998] - O-sulfates have to be removed before the desulfation of the amino group of glucosamine [Winchester 1996]
HS1ly	3	Bielicki J, Hopwood JJ, Melville EL, Anson DS	Recombinant human sulphamidase: expression, amplification, purification and characterization	Biochem J	1998	9405287	- O-sulfates have to be removed before sulfamidase desulfates amino group of GlcN [Winchester 1996] 6448: - lysosomal [UniProt] - cDNA was isolated and expressed [Scott, Nat Genet 1995] - recombinantly expressed in CHO cells, kinetic properties characterized [Bielicki, Biochem J 1998] - O-sulfates have to be removed before the desulfation of the amino group of glucosamine [Winchester 1996]
HSAT4ly	3	Ausseil J, Loredo-Osti JC, Verner A, Darmond-Zwaig C, Maire I, Poorthuis B, van Diggelen OP, Hudson TJ, Fujiwara TM, Morgan K, Pshezhetsky AV	Localisation of a gene for mucopolysaccharidosis IIIC to the pericentromeric region of chromosome 8	J Med Genet	2004	15591281	- the 2-amino group of GlcN has to be desulfated and reacetylated before hexosaminidic linkage is cleaved [Winchester 1996] - integral lysosomal membrane protein; transfers acetyl group from accoa in cytosol to desulfated 2-amino group of GlcN in the lumen of the lysosome [Winchester 1996] - gene has not been identified, although recent evidence suggests it resides in the pericentromeric region of chromosome 8 [Ausseil 2004] - Mucopolysaccharidosis type IIIC (MPS IIIC, or Sanfilippo syndrome C) is a hereditary disorder caused by deficiency of this enzyme [Ausseil 2004]
HSD11B2r	3	Albiston AL, Obeyesekere VR, Smith RE, Krozowski ZS.	Cloning and tissue distribution of the human 11 beta-hydroxysteroid dehydrogenase type 2 enzyme.	Mol Cell Endocrinol	1994	7859916	ER - uniprot Kidney specific - a/w HTN dz Found in placenta, kidney, pancreas, prostate, ovary, small intestine and colon. Defects in HSD11B2 are the cause of apparent mineralocorticoid excess (AME) [MIM:218030, 207765]. AME is a potentially fatal disease characterized by severe juvenile low-renin hypertension, sodium retention, hypokalemia and low levels of aldosterone. It often leads to nephrocalcinosis. Catalyzes the conversion of cortisol to the inactive metabolite cortisone. Modulates intracellular glucocorticoid levels, thus protecting the nonselective mineralocorticoid receptor from occupation by glucocorticoids. NJ
HSD11B2r	3	Yau JL, Seckl JR.	11beta-hydroxysteroid dehydrogenase type 1 in the brain; thickening the glucocorticoid soup.	Mol Psychiatry	2001	11673786	ER - uniprot Kidney specific - a/w HTN dz Found in placenta, kidney, pancreas, prostate, ovary, small intestine and colon. Defects in HSD11B2 are the cause of apparent mineralocorticoid excess (AME) [MIM:218030, 207765]. AME is a potentially fatal disease characterized by severe juvenile low-renin hypertension, sodium retention, hypokalemia and low levels of aldosterone. It often leads to nephrocalcinosis. Catalyzes the conversion of cortisol to the inactive metabolite cortisone. Modulates intracellular glucocorticoid levels, thus protecting the nonselective mineralocorticoid receptor from occupation by glucocorticoids. NJ

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HSD11B2r	3	Tomlinson JW, Walker EA, Bujalska IJ, Draper N, Lavery GG, Cooper MS, Hewison M, Stewart PM.	11beta-hydroxysteroid dehydrogenase type 1: a tissue specific regulator of glucocorticoid response.	Endocr Rev	2004	15466942	ER - uniprot Kidney specific - a/w HTN dz Found in placenta, kidney, pancreas, prostate, ovary, small intestine and colon. Defects in HSD11B2 are the cause of apparent mineralocorticoid excess (AME) [MIM:218030, 207765]. AME is a potentially fatal disease characterized by severe juvenile low-renin hypertension, sodium retention, hypokalemia and low levels of aldosterone. It often leads to nephrocalcinosis. Catalyzes the conversion of cortisol to the inactive metabolite cortisone. Modulates intracellular glucocorticoid levels, thus protecting the nonselective mineralocorticoid receptor from occupation by glucocorticoids. NJ
HSD17B1	3	Andersson S, Moghrabi N	Physiology and molecular genetics of 17beta-hydroxysteroid dehydrogenases	Steroids	1997		HSD 17beta dehyd: type I: cytosol type 2,3: microsomes type 4: peroxisomal type 7: inferred from Paynes paper: cytosolic or microsomal (set as microsomal for time being) - return for further searches HSD17B1 specificity: ovary, placenta, mammary glands HSD17B8: reverse direction: estradiol -> estrone. cytosol by default - no detailed localization info. NJ
HSD17B2r	3	Wu L, Einstein M, Geissler WM, Chan HK, Elliston KO, Andersson S.	Expression cloning and characterization of human 17 beta-hydroxysteroid dehydrogenase type 2, a microsomal enzyme possessing 20 alpha-hydroxysteroid dehydrogenase activity.	J Biol Chem	1993	8099587	localization: ER by analogy w/ HSD17B3 specificity: no details Cloning, sequence, biochem: PMID: 8099587 Capable of catalyzing the interconversion of testosterone and androstenedione, as well as estradiol and estrone. Also has 20-alpha-HSD activity. Uses NADH while EDH17B3 uses NADPH. NJ
HSD17B4x	3	Adamski J, Leenders F Carstensen JF, Kaumann M, Markus MM, Husen B, Tesdorpf JG, Seedor U, deLaunoit Y, Jakob F	Steroids, fatty acyl-CoA, and sterols are substrates of 80-kDa multifunctional protein	Steroids	1997		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Leenders F, Dolez V, Begue A, Moller G, Gloeckner JC, deLaunoit Y, Adamski J	Structure of the gene for the human 17-beta-hydroxysteroid dehydrogenase type IV	Mammalian Genome	1998		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Adamski J, Leenders F Carstensen JF, Kaumann M, Markus MM, Husen B, Tesdorpf JG, Seedor U, deLaunoit Y, Jakob F	Steroids, fatty acyl-CoA, and sterols are substrates of 80-kDa multifunctional protein	Steroids	1997		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
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HSD17B4x	3	Marijanovic Z, Laubner D, Moller G, Gege C, Husenn B, Adamski J, Breitling R	Closing the gap: Identification of human 3-ketosteroid reductase, the last unknown enzyme of mammalian cholesterol biosynthesis	Molecular Endocrinology	2005		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ

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HSD17B4x	3	Adamski J, Leenders F Carstensen JF, Kaumann M, Markus MM, Husen B, Tesdorpf JG, Seedor U, deLaunoit Y, Jakob F	Steroids, fatty acyl-CoA, and sterols are substrates of 80-kDa multifunctional protein	Steroids	1997		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Leenders F, Dolez V, Begue A, Moller G, Gloeckner JC, deLaunoit Y, Adamski J	Structure of the gene for the human 17-beta-hydroxysteroid dehydrogenase type IV	Mammalian Genome	1998		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Adamski J, Leenders F Carstensen JF, Kaumann M, Markus MM, Husen B, Tesdorpf JG, Seedor U, deLaunoit Y, Jakob F	Steroids, fatty acyl-CoA, and sterols are substrates of 80-kDa multifunctional protein	Steroids	1997		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Marijanovic Z, Laubner D, Moller G, Gege C, Husenn B, Adamski J, Breitling R	Closing the gap: Identification of human 3-ketosteroid reductase, the last unknown enzyme of mammalian cholesterol biosynthesis	Molecular Endocrinology	2005		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Leenders F, Dolez V, Begue A, Moller G, Gloeckner JC, deLaunoit Y, Adamski J	Structure of the gene for the human 17-beta-hydroxysteroid dehydrogenase type IV	Mammalian Genome	1998		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Adamski J, Leenders F Carstensen JF, Kaumann M, Markus MM, Husen B, Tesdorpf JG, Seedor U, deLaunoit Y, Jakob F	Steroids, fatty acyl-CoA, and sterols are substrates of 80-kDa multifunctional protein	Steroids	1997		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Marijanovic Z, Laubner D, Moller G, Gege C, Husenn B, Adamski J, Breitling R	Closing the gap: Identification of human 3-ketosteroid reductase, the last unknown enzyme of mammalian cholesterol biosynthesis	Molecular Endocrinology	2005		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Adamski J, Leenders F Carstensen JF, Kaumann M, Markus MM, Husen B, Tesdorpf JG, Seedor U, deLaunoit Y, Jakob F	Steroids, fatty acyl-CoA, and sterols are substrates of 80-kDa multifunctional protein	Steroids	1997		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD17B4x	3	Marijanovic Z, Laubner D, Moller G, Gege C, Husenn B, Adamski J, Breitling R	Closing the gap: Identification of human 3-ketosteroid reductase, the last unknown enzyme of mammalian cholesterol biosynthesis	Molecular Endocrinology	2005		peroxisome: uniprot specificity: Liver and placenta also involved in lanosterol -> cholesterol synthesis pathway NJ
HSD3A1r	3	Khanna M, Qin KN, Wang RW, Cheng KC.	Substrate specificity, gene structure, and tissue-specific distribution of multiple human 3 alpha-hydroxysteroid dehydrogenases.	J Biol Chem	1995	7650035	This gene encodes a member of the aldo/keto reductase superfamily, which consists of more than 40 known enzymes and proteins. These enzymes catalyze the conversion of aldehydes and ketones to their corresponding alcohols by utilizing NADH and/or NADPH as cofactors. The enzymes display overlapping but distinct substrate specificity. This enzyme catalyzes the bioreduction of chlordecone, a toxic organochlorine pesticide, to chlordecone alcohol in liver. This gene shares high sequence identity with three other gene members and is clustered with those three genes at chromosome 10p15-p14. see PMID: 7650035 NJ
HSD3B2r	3	Luu-The V.	Analysis and characteristics of multiple types of human 17beta-hydroxysteroid dehydrogenase.	J Steroid Biochem Mol Biol	2001	11384872	ER - uniprot, refs specificity: adrenal glands, testes, ovaries 3beta-HSD is a bifunctional enzyme, that catalyzes the oxidative conversion of delta(5)-ene-3-beta-hydroxy steroid, and the oxidative conversion of ketosteroids. The 3beta-HSD enzymatic system plays a crucial role in the biosynthesis of all classes of hormonal steroids. also PMID: 14643063, 11384872 NJ
HSPASEly	2	Burbach BJ, Friedl A, Mundhenke C, Rapraeger AC	Syndecan-1 accumulates in lysosomes of poorly differentiated breast carcinoma cells	Martix Biol	2003	12782143	- core protein proteolysis and initial GAG chain fragmentation occurs in endosomes, followed by the complete degradation of GAGs in lysosomes [Burbach, Matrix Biol 2003] - single heparan sulfate chains are broken down by a specific endoglucuronidase to oligosaccharides of approcimately 5kDA as they pass thru the endosomal system to lysosomes; NOTE: this functionality is not included in the model as the example heparan sulfate chain is already short - several cell types, including chondrocytes and liver endothelial cells, can bind and endocytose proteoglycans and deliver them to lysosomes for catabolism [Winchester 1996]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
HXPRT	3	Kim SH, Moores JC, David D, Respess JG, Jolly DJ, Friedmann T.	The organization of the human HPRT gene.	Nucleic Acids Res	1986	3008106	IT GeneCards: can act on both hxan and gua. homotetramer purine salvage
HXPRT	3	Wilson JM, Kobayashi R, Fox IH, Kelley WN.	Human hypoxanthine-guanine phosphoribosyltransferase.	J Biol Chem	1983	6853490	IT GeneCards: can act on both hxan and gua. homotetramer purine salvage
HYPOE	3	Jang YM, Kim DW, Kang TC, Won MH, Baek NI, Moon BJ, Choi SY, Kwon OS.	Human pyridoxal phosphatase. Molecular cloning, functional expression, and tissue distribution.	J Biol Chem	2003	14522954	IT homodimer. needs Mg2+ real name: Pyridoxamine-5-phosphate phosphatase
HYXNt	3	Yao SY, Ng AM, Vickers MF, Sundaram M, Cass CE, Baldwin SA, Young JD.	Functional and molecular characterization of nucleobase transport by recombinant human and rat equilibrative nucleoside transporters 1 and 2.	J Biol Chem	2002	12006583	IT
HYXNt	3	Baldwin SA, Beal PR, Yao SY, King AE, Cass CE, Young JD.	The equilibrative nucleoside transporter family, SLC29.	Pflugers Arch	2004	12838422	IT
ICDHyp	3	Geisbrecht BV, Gould SJ	The human PICD gene encodes a cytoplasmic and peroxisomal NADP(+)-dependent isocitrate dehydrogenase	J Biol Chem	1999	10521434	3417: - NADP(+)-dependent isocitrate dehydrogenase found in the cytoplasm and peroxisomes [RefSeq] - identified by homology search; 59% identity to yeast Idp3p [Geisbrecht 1999] - expression in yeast restored partial function to Idp3p knockou [Geisbrecht 1999] - found in peroxisomes and cytoplasm of human and rat liver cells, ~ 27% assoc w/ peroxisome [Geisbrecht 1999] - known gap; the source of intraperoxisomal isocitrate has not been determined [Geisbrecht 1999]
ICDHyrm	3	Sazanov LA, Jackson JB.	Proton-translocating transhydrogenase and NAD- and NADP-linked isocitrate dehydrogenases operate in a substrate cycle which contributes to fine regulation of the tricarboxylic acid cycle activity in mitochondria.	FEBS Lett	1994	8187868	3418: - NADP(+)-dependent isocitrate dehydrogenase found in the mitochondria [RefSeq] - mitochondria [UniProt] - reaction may be reversible [Orten, Human Biochem 1975], [Comte, Am J Physiol Heart Circ Physiol. 2002], [Sazanov, FEBS Lett 1994]
ICDHyrm	3	Comte B, Vincent G, Bouchard B, Benderdour M, Des Rosiers C.	Reverse flux through cardiac NADP(+)-isocitrate dehydrogenase under normoxia and ischemia.	Am J Physiol Heart Circ Physiol	2002	12234803	3418: - NADP(+)-dependent isocitrate dehydrogenase found in the mitochondria [RefSeq] - mitochondria [UniProt] - reaction may be reversible [Orten, Human Biochem 1975], [Comte, Am J Physiol Heart Circ Physiol. 2002], [Sazanov, FEBS Lett 1994]
IDHPOXOX3	3	Nakamura M, Yamazaki I, Kotani T, Ohtaki S	Thyroid peroxidase selects the mechanism of either 1 or 2-electron oxidation of phenols, depending on their substituents	J Biol Chem	1985	2997169	The citations are slightly inspecific, but the reaction mechanism from KEGG seems to be basically consistent, expecially when all four reactions IDHPOXOX(-4) are taken together. - identified [Kimura 1987] and cloned [Magnussen 1987]
IDHPOXOX3	3	Magnusson RP, Chazenbalk GD, Gestautas J, Seto P, Filetti S, DeGroot LJ, Rapoport B.	Molecular cloning of the complementary deoxyribonucleic acid for human thyroid peroxidase.	Mol Endocrinol	1987	3153466	The citations are slightly inspecific, but the reaction mechanism from KEGG seems to be basically consistent, expecially when all four reactions IDHPOXOX(-4) are taken together. - identified [Kimura 1987] and cloned [Magnussen 1987]
IDHPOXOX3	3	Kimura S, Kotani T, McBride OW, Umeki K, Hirai K, Nakayama T, Ohtaki S	Human thyroid peroxidase: complete cDNA and protein sequence, chromosome mapping, and identification of two alternately spliced mRNAs	Proc Natl Acad Sci U S A	1987	3475693	The citations are slightly inspecific, but the reaction mechanism from KEGG seems to be basically consistent, expecially when all four reactions IDHPOXOX(-4) are taken together. - identified [Kimura 1987] and cloned [Magnussen 1987]
IDOAASE1ly	3	Scott HS, Anson DS, Orsborn AM, Nelson PV, Clements PR, Morris CP, Hopwood JJ	Human alpha-L-iduronidase: cDNA isolation and expression	Proc Natl Acad Sci U S A	1991	1946389	3425: - hydrolyzes the teminal alpha-L-iduronic acid residues from heparan sulfate and dermatan sulfate [RefSeq] - isolated from human liver [Clements 1989] - cloned and expressed in CHO cells [Scott 1991]
IDOAASE1ly	3	Clements PR, Brooks DA, McCourt PA, Hopwood JJ	Immunopurification and characterization of human alpha-L-iduronidase with the use of monoclonal antibodies	Biochem J	1989	2470345	3425: - hydrolyzes the teminal alpha-L-iduronic acid residues from heparan sulfate and dermatan sulfate [RefSeq] - isolated from human liver [Clements 1989] - cloned and expressed in CHO cells [Scott 1991]
IDOURtly	3	Havelaar AC, Mancini GM, Beerens CE, Souren RM, Verheijen FW.	Purification of the lysosomal sialic acid transporter. Functional characteristics of a monocarboxylate transporter	J Biol Chem	1998	9852127	H symport with sialic acid (precursors) into lysososome. See PMID: 2768261for biochem characterization and PMID: 10581036 for discussion of particular SNPs with sialic acid storage diseases. Sialic acid storage disorders (due to transporter mutations) require import and export e.g. PMID: 2768266 - hence made reversible. NJ - iduronate was also been shown to be a substrate of purified rat liver enzyme [Havelaar 1998]
ILETA	3	Naylor SL, Shows TB.	Branched-chain aminotransferase deficiency in Chinese hamster cells complemented by two independent genes on human chromosomes 12 and 19.		1980	6933702	reversible reaction, substrate specificities- Somatic Cell Genet. 1980 Sep;6(5):641-52. cytosol - Biochim Biophys Acta. 1997 Apr 25;1339(1):9-13.

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
ILETA	3	Bledsoe RK, Dawson PA, Hutson SM.	Cloning of the rat and human mitochondrial branched chain aminotransferases (BCATm).		1997	9165094	reversible reaction, substrate specificities- Somatic Cell Genet. 1980 Sep;6(5):641-52. cytosol - Biochim Biophys Acta. 1997 Apr 25;1339(1):9-13.
IMPD	3	Natsumeda Y, Ohno S, Kawasaki H, Konno Y, Weber G, Suzuki K.	Two distinct cDNAs for human IMP dehydrogenase.	J Biol Chem	1990		IT
IMPD	3	Gu JJ, Kaiser-Rogers K, Rao K, Mitchell BS.	Assignment of the human type I IMP dehydrogenase gene (IMPDH1) to chromosome 7q31.3-q32).	Genomics	1994		IT
IMPD	3	Xiang B, Taylor JC, Markham GD.	Monovalent cation activation and kinetic mechanism of inosine 5'-monophosphate dehydrogenase.	J Biol Chem	1996		IT
INOSTO	3	Arner RJ, Prabhu KS, Reddy CC.	Molecular cloning, expression, and characterization of myo-inositol oxygenase from mouse, rat, and human kidney	Biochem Biophys Res Commun	2004	15504367	- cytoplasmic [UniProt] - myo-inositol oxygenase activity [UniProt] - kidney specific [UniProt] - cloning, expression, and characterization [Arner, Biochem Biophys Res Commun 2004]
INSt	3	Ward JL, Sherali A, Mo ZP, Tse CM.	Kinetic and pharmacological properties of cloned human equilibrative nucleoside transporters, ENT1 and ENT2, stably expressed in nucleoside transporter deficient PK15 cells.	J Biol Chem	2000	10722669	IT
INSTt2r	3	Uldry M, Ibberson M, Horisberger JD, Chatton JY, Riederer BM, Thorens B	Identification of a mammalian H(+)-myo-inositol symporter expressed predominantly in the brain	EMBO J	2001	11500374	114134: - cloned [Uldry, 2001] - transports only myo-inositol, no Glc [Uldry, 2004] - brain, white, brown, amd epididymal adipose, idney [Uldry, 2004]
INSTt4_2	3	Roll P, Massacrier A, Pereira S, Robaglia-Schlupp A, Cau P, Szepetowski P	New human sodium/glucose cotransporter gene (KST1): identification, characterization, and mutation analysis in ICCA (infantile convulsions and choreoathetosis) and BFIC (benign familial infantile convulsions) families	Gene	2002	12039040	- cloned [Roll 2002] and expressed [Coady 2002] - stoichiometry of 2 sodium for 1 MI molecule [Coady 2002] - heart, skeletal muscle, kidney, liver, placenta; weaker expression in brain, spleen, lung, WBC [Roll 2002] - also transports Glc, Xyl at moderate levels and a-methylGlc, Gal, Fuc, 3-O-methylGlc, 2-deoxyGlc at very low levels [Coady 2002]
INSTt4_2	3	Coady MJ, Wallendorff B, Gagnon DG, Lapointe JY	Identification of a novel Na+/myo-inositol cotransporter	J Biol Chem	2002	12133831	- cloned [Roll 2002] and expressed [Coady 2002] - stoichiometry of 2 sodium for 1 MI molecule [Coady 2002] - heart, skeletal muscle, kidney, liver, placenta; weaker expression in brain, spleen, lung, WBC [Roll 2002] - also transports Glc, Xyl at moderate levels and a-methylGlc, Gal, Fuc, 3-O-methylGlc, 2-deoxyGlc at very low levels [Coady 2002]
IPDDIx	3	Breitling R, Laubner D, Clizbe D, Adamski J, Krisans SK	Isopentenyl-diphosphate isomerases in human and mouse: evolutionary analysis of a mammalian gene duplication	J Mol Evol	2003		peroxisomal no tissue specificity IDI1 encodes a peroxisomally-localized enzyme that catalyzes the interconversion of isopentenyl diphosphate (IPP) to its highly electrophilic isomer, dimethylallyl diphosphate (DMAPP), which are the substrates for the successive reaction that results in the synthesis of farnesyl diphosphate and, ultimately, cholesterol. It has been shown in peroxisomal deficiency diseases such as Zellweger syndrome and neonatal adrenoleukodystrophy that there is reduction in IPP isomerase activity. See refs - IDI1 and IDI2 - evolutionarily distinct but appear to catalyze the same reaction! NJ
It	3	Rodriguez AM, Perron B, Lacroix L, Caillou B, Leblanc G, Schlumberger M, Bidart JM, Pourcher T	Identification and characterization of a putative human iodide transporter located at the apical membrane of thyrocytes	J Clin Endocrinol Metab	2002	1217270	160728: - cloned and expressed [Rodriguez 2002] - faciliated diffusion of I-, not cotransport [Rodriguez 2002] - apical pole of the thyroid cells facing the colloid lumen [Rodriguez 2002]
ITCOALm	1	WANG SF, ADLER J, LARDY HA	The pathway of itaconate metabolism by liver mitochondria	J Biol Chem	1961	13783048	- another function of EC 6.2.1.5 according to BRENDA - reaction known to occur w/ GTP as cofactor in rat liver mitochondria [Wang, J Biol Chem 1961] - Itaconate, citraconate, and mesaconate are probably metabolized in the dog, since they could be recovered in the urine to the extent of only 24, 28, and 64 per cent, respectively (see refs in [Adler 1957]) - In guinea pig liver, itaconate is oxidized as rapidly as most members of the tricarboxylic acid cycle, methyl succinate was oxidized 1/6 as fast and mesaconate 1/8 as fast as itaconate [Adler 1957]

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KCCt	3	Gillen CM, Brill S, Payne JA, Forbush B 3rd	Molecular cloning and functional expression of the K Cl cotransporter from rabbit, rat, and human. A new member of the cation-chloride cotransporter family	J Biol Chem	1996	8663127	- 1:1 stoichiometry of K+-Cl- transport [Hebert 2004] - RBC mediate K+-Cl- efflux, neurons mediate both efflux and influx [Hebert 2004] - KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6560: - cloned [Gillen 1996] - ubiquitous [Gillen 1996] - transport verified in HEK293 cells [Gillen 1996] 57468: - bi-directional transport [Entrez Gene], [Hebert 2004] - cloned [Song 2002] - brain [Song 2002]; also found in rat retina [Hebert 2004] - activity functionally verified in Xenopus laevis oocytes [Song 2002] 9990: - cloned [Hiki 1999], [Mount 1999], [Race 1999] - brain, heart, skeletal muscle, kidney [Hiki 1999]; most abundant in heart, kidney [Mount 1999]; high in kidney, heart, brain, lower in sk muscle, placenta, lung, liver, and pancreas [Race 1999] - 75-76% identical to human, pig, rat, and rabbit KCC1p, 7% identical to rat KCC2p [Race 1999] - activity functionally verified in HEK293 cells [Race 1999] 10723: - cloned [Mount 1999] - muscle, brain, lung, heart, kidney [Mount 1999] - basolateral membraine of type-A intercalated cells and proximal tubule (mouse) [Hebert 2004]
KCCt	3	Hiki K, D'Andrea RJ, Furze J, Crawford J, Woollatt E, Sutherland GR, Vadas MA, Gamble JR	Cloning, characterization, and chromosomal location of a novel human K+-Cl- cotransporter	J Biol Chem	1999	10187864	- 1:1 stoichiometry of K+-Cl- transport [Hebert 2004] - RBC mediate K+-Cl- efflux, neurons mediate both efflux and influx [Hebert 2004] - KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6560: - cloned [Gillen 1996] - ubiquitous [Gillen 1996] - transport verified in HEK293 cells [Gillen 1996] 57468: - bi-directional transport [Entrez Gene], [Hebert 2004] - cloned [Song 2002] - brain [Song 2002]; also found in rat retina [Hebert 2004] - activity functionally verified in Xenopus laevis oocytes [Song 2002] 9990: - cloned [Hiki 1999], [Mount 1999], [Race 1999] - brain, heart, skeletal muscle, kidney [Hiki 1999]; most abundant in heart, kidney [Mount 1999]; high in kidney, heart, brain, lower in sk muscle, placenta, lung, liver, and pancreas [Race 1999] - 75-76% identical to human, pig, rat, and rabbit KCC1p, 7% identical to rat KCC2p [Race 1999] - activity functionally verified in HEK293 cells [Race 1999] 10723: - cloned [Mount 1999] - muscle, brain, lung, heart, kidney [Mount 1999] - basolateral membraine of type-A intercalated cells and proximal tubule (mouse) [Hebert 2004]

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KCCt	3	Mount DB, Mercado A, Song L, Xu J, George AL Jr, Delpire E, Gamba G	Cloning and characterization of KCC3 and KCC4, new members of the cation-chloride cotransporter gene family	J Biol Chem	1999	10347194	- 1:1 stoichiometry of K+-Cl- transport [Hebert 2004] - RBC mediate K+-Cl- efflux, neurons mediate both efflux and influx [Hebert 2004] - KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6560: - cloned [Gillen 1996] - ubiquitous [Gillen 1996] - transport verified in HEK293 cells [Gillen 1996] 57468: - bi-directional transport [Entrez Gene], [Hebert 2004] - cloned [Song 2002] - brain [Song 2002]; also found in rat retina [Hebert 2004] - activity functionally verified in Xenopus laevis oocytes [Song 2002] 9990: - cloned [Hiki 1999], [Mount 1999], [Race 1999] - brain, heart, skeletal muscle, kidney [Hiki 1999]; most abundant in heart, kidney [Mount 1999]; high in kidney, heart, brain, lower in sk muscle, placenta, lung, liver, and pancreas [Race 1999] - 75-76% identical to human, pig, rat, and rabbit KCC1p, 7% identical to rat KCC2p [Race 1999] - activity functionally verified in HEK293 cells [Race 1999] 10723: - cloned [Mount 1999] - muscle, brain, lung, heart, kidney [Mount 1999] - basolateral membraine of type-A intercalated cells and proximal tubule (mouse) [Hebert 2004]
KCCt	3	Race JE, Makhlouf FN, Logue PJ, Wilson FH, Dunham PB, Holtzman EJ	Molecular cloning and functional characterization of KCC3, a new K-Cl cotransporter	Am J Physiol	1999	10600773	- 1:1 stoichiometry of K+-Cl- transport [Hebert 2004] - RBC mediate K+-Cl- efflux, neurons mediate both efflux and influx [Hebert 2004] - KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6560: - cloned [Gillen 1996] - ubiquitous [Gillen 1996] - transport verified in HEK293 cells [Gillen 1996] 57468: - bi-directional transport [Entrez Gene], [Hebert 2004] - cloned [Song 2002] - brain [Song 2002]; also found in rat retina [Hebert 2004] - activity functionally verified in Xenopus laevis oocytes [Song 2002] 9990: - cloned [Hiki 1999], [Mount 1999], [Race 1999] - brain, heart, skeletal muscle, kidney [Hiki 1999]; most abundant in heart, kidney [Mount 1999]; high in kidney, heart, brain, lower in sk muscle, placenta, lung, liver, and pancreas [Race 1999] - 75-76% identical to human, pig, rat, and rabbit KCC1p, 7% identical to rat KCC2p [Race 1999] - activity functionally verified in HEK293 cells [Race 1999] 10723: - cloned [Mount 1999] - muscle, brain, lung, heart, kidney [Mount 1999] - basolateral membraine of type-A intercalated cells and proximal tubule (mouse) [Hebert 2004]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
KCCt	3	Song L, Mercado A, Vazquez N, Xie Q, Desai R, George AL Jr, Gamba G, Mount DB	Molecular, functional, and genomic characterization of human KCC2, the neuronal K-Cl cotransporter	Brain Res Mol Brain Res	2002	121106695	- 1:1 stoichiometry of K+-Cl- transport [Hebert 2004] - RBC mediate K+-Cl- efflux, neurons mediate both efflux and influx [Hebert 2004] - KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6560: - cloned [Gillen 1996] - ubiquitous [Gillen 1996] - transport verified in HEK293 cells [Gillen 1996] 57468: - bi-directional transport [Entrez Gene], [Hebert 2004] - cloned [Song 2002] - brain [Song 2002]; also found in rat retina [Hebert 2004] - activity functionally verified in Xenopus laevis oocytes [Song 2002] 9990: - cloned [Hiki 1999], [Mount 1999], [Race 1999] - brain, heart, skeletal muscle, kidney [Hiki 1999]; most abundant in heart, kidney [Mount 1999]; high in kidney, heart, brain, lower in sk muscle, placenta, lung, liver, and pancreas [Race 1999] - 75-76% identical to human, pig, rat, and rabbit KCC1p, 7% identical to rat KCC2p [Race 1999] - activity functionally verified in HEK293 cells [Race 1999] 10723: - cloned [Mount 1999] - muscle, brain, lung, heart, kidney [Mount 1999] - basolateral membraine of type-A intercalated cells and proximal tubule (mouse) [Hebert 2004]
KHK2	3	Bais R, James HM, Rofe AM, Conyers RA.	The purification and properties of human liver ketohexokinase. A role for ketohexokinase and fructose-bisphosphate aldolase in the metabolic production of oxalate from xylitol	Biochem J	1985	2996495	- ketohexokinase and fructose-bisphosphate aldolase were purified from human liver [Bais 1985] - ketohexokinase and aldolase could catalyse a reaction sequence which forms glycolaldehyde from D-xylulose [Bais 1985], [Barngrover 1983], [James 1982] - this probably occurs mainly in the liver, to a lesser extent in the kidney, and very little in heart, brain and muscle based on the localization of the enzymes [Bais 1985] - ketohexokinase can phosphorylate D-xylulose as readily as D-fructose, except that higher concentrations of D-xylulose are required [Bais 1985]
KHK2	3	James HM, Bais R, Edwards JB, Rofe AM, Conyers AJ.	Models for the metabolic production of oxalate from xylitol in humans: a role for fructokinase and aldolase.	Aust J Exp Biol Med Sci	1982	6284103	- ketohexokinase and fructose-bisphosphate aldolase were purified from human liver [Bais 1985] - ketohexokinase and aldolase could catalyse a reaction sequence which forms glycolaldehyde from D-xylulose [Bais 1985], [Barngrover 1983], [James 1982] - this probably occurs mainly in the liver, to a lesser extent in the kidney, and very little in heart, brain and muscle based on the localization of the enzymes [Bais 1985] - ketohexokinase can phosphorylate D-xylulose as readily as D-fructose, except that higher concentrations of D-xylulose are required [Bais 1985]
KHK2	3	Barngrover DA, Dills WL Jr.	The involvement of liver fructokinase in the metabolism of D-xylulose and xylitol in isolated rat hepatocytes	J Nutr	1983	6298387	- ketohexokinase and fructose-bisphosphate aldolase were purified from human liver [Bais 1985] - ketohexokinase and aldolase could catalyse a reaction sequence which forms glycolaldehyde from D-xylulose [Bais 1985], [Barngrover 1983], [James 1982] - this probably occurs mainly in the liver, to a lesser extent in the kidney, and very little in heart, brain and muscle based on the localization of the enzymes [Bais 1985] - ketohexokinase can phosphorylate D-xylulose as readily as D-fructose, except that higher concentrations of D-xylulose are required [Bais 1985]
KYN3OX	3	Alberati-Giani D, Cesura AM, Broger C, Warren WD, Rover S, Malherbe P	Cloning and functional expression of human kynurenine 3-monooxygenase	FEBS Lett	1997	9237672	Gene found and enzyme characterized (at least for liver)
KYNAKGAT	3	Cooper AJ	The role of glutamine transaminase K (GTK) in sulfur and alpha-keto acid metabolism in the brain, and in the possible bioactivation of neurotoxicants	Neurochem Int	2004	15016471	First step of reaction described in citation. Also citation describes alternate splicing as the reason for mostly mitochondrial localization in brain and mostly cytosolic in kidney.
KYNATESYN	3	Hartai Z, Klivenyi P, Janaky T, Penke B, Dux L, Vecsei L	Kynurenine metabolism in multiple sclerosis	Acta Neurol Scand	2005	16008534	Second step of a reaction described in the first citation. Second citation (PMID 16008534): "To date, KYNA is the only known endogenous competitive antagonist of all three ionotrophic excitatory amino acid receptors."

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LALDO2	3	Bohren KM, Bullock B, Wermuth B, Gabbay KH	The aldo-keto reductase superfamily. cDNAs and deduced amino acid sequences of human aldehyde and aldose reductases	J Biol Chem	1989	2498333	- D-lactaldehyde is a minor product (5%) of methylglyoxal reduction by NADPH (alsoe reductase) [Vander Jagt 1992] 231: - cytosolic, NADPH [Bohren et al. J Biol Chem 264 (16): 9547, 1989] 10327: - NADP is cofactor [UniProt] 8574: - gene has been cloned, 78% identical with the Rattus norvegicus aflatoxin B1 aldehyde reductase (Afar) [Praml 1998] -rat homolog appears to be Golgi-associated [Kelly 2002] based on N-terminal sequence and immunohistochemistry, however I believe these results more strongly support localization in the outer Golgi membrane (cytosolic in our model) - human AKR7A2 also has Golgi signal sequence and [GO, UniProt] list as Golgi by similarity; however, I've assumed protein is cytosolic due to lack of specific info on location of functional domain and the fact that the only (weak) evidence thus far has come from the rat homolog - NADH or NADPH [UniProt]; specificity is based on mouse homology AFAR2 [Kelly 2002]
LCADi	2	Christopher MM, Eckfeldt JH, Eaton JW.	Propylene glycol ingestion causes D-lactic acidosis	Lab Invest	1990	2296157	- NAD-dependent aldehyde dehydrogenase can convert L or D-lactaldehyde to L or D-lactate, respectively (see Fig 3 of [Ewaschuk 2005]; this pathway is based on original work done in cats [Christopher 1990] suggesting that methylglyoxal metabolism can result in D-lactic acidosis under extreme conditions - aldehyde dehydrogenase from goat liver has been shown to oxidize lactaldehyde to lactic acid; lactaldehyde was found to be primarily oxidized by this enzyme; almost 90% of the total lactaldehyde-oxidizing activity is located in the cytosol; enzyme is also found in mitochondria [Ray 1984]
LCADi	2	Ray S, Ray M.	Oxidation of lactaldehyde by cytosolic aldehyde dehydrogenase and inhibition of cytosolic and mitochondrial aldehyde dehydrogenase by metabolites.	Biochim Biophys Acta	1984	6487654	- NAD-dependent aldehyde dehydrogenase can convert L or D-lactaldehyde to L or D-lactate, respectively (see Fig 3 of [Ewaschuk 2005]; this pathway is based on original work done in cats [Christopher 1990] suggesting that methylglyoxal metabolism can result in D-lactic acidosis under extreme conditions - aldehyde dehydrogenase from goat liver has been shown to oxidize lactaldehyde to lactic acid; lactaldehyde was found to be primarily oxidized by this enzyme; almost 90% of the total lactaldehyde-oxidizing activity is located in the cytosol; enzyme is also found in mitochondria [Ray 1984]
LCADi	2	Ewaschuk JB, Naylor JM, Zello GA	D-lactate in human and ruminant metabolism	J Nutr	2005	15987839	- NAD-dependent aldehyde dehydrogenase can convert L or D-lactaldehyde to L or D-lactate, respectively (see Fig 3 of [Ewaschuk 2005]; this pathway is based on original work done in cats [Christopher 1990] suggesting that methylglyoxal metabolism can result in D-lactic acidosis under extreme conditions - aldehyde dehydrogenase from goat liver has been shown to oxidize lactaldehyde to lactic acid; lactaldehyde was found to be primarily oxidized by this enzyme; almost 90% of the total lactaldehyde-oxidizing activity is located in the cytosol; enzyme is also found in mitochondria [Ray 1984]
LCAT1e	3	Krimbou L, Marcil M, Davignon J, Genest J Jr.	Interaction of lecithin:cholesterol acyltransferase (LCAT).alpha 2-macroglobulin complex with low density lipoprotein receptor-related protein (LRP). Evidence for an alpha 2-macroglobulin/LRP receptor-mediated system participating in LCAT clearance.	J Biol Chem	2001	11435418	localization: extracellular - uniprot This gene encodes the extracellular cholesterol esterifying enzyme, lecithin-cholesterol acyltransferase. The esterification of cholesterol is required for cholesterol transport. Mutations in this gene have been found to cause fish-eye disease as well as LCAT deficiency. NJ
LCAT1e	3	Wang K, Subbaiah PV.	Role of the interfacial binding domain in the oxidative susceptibility of lecithin:cholesterol acyltransferase.	Biochem J	2002	11966470	localization: extracellular - uniprot This gene encodes the extracellular cholesterol esterifying enzyme, lecithin-cholesterol acyltransferase. The esterification of cholesterol is required for cholesterol transport. Mutations in this gene have been found to cause fish-eye disease as well as LCAT deficiency. NJ
LCTStg	2	Alberts B, Johnson A, Lewis J, Raff M, Roberts K, Walter P	Molecular Biology of the Cell, 4th ed.		2002		- reaction would only be found in lactating mammary cells - lactose produced in the Golgi is transported out of the cell via a vesicle [Alberts 2002]
LCTStl	2	Gordon PB, Seglen PO.	Prelysosomal convergence of autophagic and endocytic pathways	Biochem Biophys Res Commun	1988	3126737	- cytosolic lactose can be autophagocytosed and taken into the lysosome (demonstrated in rat hepatocytes) [Gordon 1988]
LDH_D	2	Devlin, TM	Textbook of Biochemistry with Clinical Correlations		2001		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
LDH_D	2	Champe, P.C., Harvey, R.A., Ferrier D.R.	Biochemistry, 3rd edition		2005		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Devlin, TM	Textbook of Biochemistry with Clinical Correlations		2001		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Champe, P.C., Harvey, R.A., Ferrier D.R.	Biochemistry, 3rd edition		2005		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Devlin, TM	Textbook of Biochemistry with Clinical Correlations		2001		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Champe, P.C., Harvey, R.A., Ferrier D.R.	Biochemistry, 3rd edition		2005		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Devlin, TM	Textbook of Biochemistry with Clinical Correlations		2001		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Champe, P.C., Harvey, R.A., Ferrier D.R.	Biochemistry, 3rd edition		2005		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Devlin, TM	Textbook of Biochemistry with Clinical Correlations		2001		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Champe, P.C., Harvey, R.A., Ferrier D.R.	Biochemistry, 3rd edition		2005		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Devlin, TM	Textbook of Biochemistry with Clinical Correlations		2001		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Champe, P.C., Harvey, R.A., Ferrier D.R.	Biochemistry, 3rd edition		2005		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Devlin, TM	Textbook of Biochemistry with Clinical Correlations		2001		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Champe, P.C., Harvey, R.A., Ferrier D.R.	Biochemistry, 3rd edition		2005		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_D	2	Devlin, TM	Textbook of Biochemistry with Clinical Correlations		2001		- lactate formation is the major fate for pyruvate in red blood cells, lens and cornea of the eye, kidney medulla, testes, and leukocytes [Champe, Biochemistry 2005]
LDH_L	0	Millan JL, Driscoll CE, LeVan KM, Goldberg E.	Epitopes of human testis-specific lactate dehydrogenase deduced from a cDNA sequence.	Proc Natl Acad Sci U S A	1985	2440048	tissue localization: Ldh1 (HHHH): mycoardium, RBC Ldh2 (HHHM): myocardiam, RBC Ldh3 (HHMM): brain, kidney Ldh4 (HMMM) Ldh5 (MMMM): liver, sk muscle [Devlin, Textbook of Biochem, 2003] [Yu et al, Biochem Pharmacol. 2001 Jul 1;62(1):81-9] 3939: - predominantly expressed in muscle [RefSeq] -cytosolic [RefSeq] 3945: cytosolic [UniProt] 3948: - testis-specific [RefSeq] -cytosolic [UniProt] 92483: - testis specific [UniProt] - Additional information added by RS/TV: Cytosolic according to GeneCards Lactate dehydrogenase (LDH) catalyzes the NAD+-dependent conversion of lactate to pyruvate during anaerobic glycolysis according to Li X. Biochem Biophys Res Commun. 2004 Jul 30;320(3):625-34. LDH has three major isozymes each of which has a specific tissue localization: (1) Ldha.1 predominates in skeletal muscle and the liver (2) Ldhb.1 is highly expressed in the heart (3) Ldhc.1, Ldhc.2 are expressed only in testes and spermatoza. Tissue localization according to

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LDH_L	0	Yu Y, Deck JA, Hunsaker LA, Deck LM, Royer RE, Goldberg E, Vander Jagt DL	Selective active site inhibitors of human lactate dehydrogenases A4, B4, and C4.	Biochem Pharmacol	2001	11377399	tissue localization: Ldh1 (HHHH): mycoardium, RBC Ldh2 (HHHM): myocardiam, RBC Ldh3 (HHMM): brain, kidney Ldh4 (HMMM) Ldh5 (MMMM): liver, sk muscle [Devlin, Textbook of Biochem, 2003] [Yu et al, Biochem Pharmacol. 2001 Jul 1;62(1):81-9] 3939: - predominantly expressed in muscle [RefSeq] -cytosolic [RefSeq] 3945: cytosolic [UniProt] 3948: - testis-specific [RefSeq] -cytosolic [UniProt] 92483: - testis specific [UniProt] - Additional information added by RS/TV: Cytosolic according to GeneCards Lactate dehydrogenase (LDH) catalyzes the NAD+-dependent conversion of lactate to pyruvate during anaerobic glycolysis according to Li X. Biochem Biophys Res Commun. 2004 Jul 30;320(3):625-34. LDH has three major isozymes each of which has a specific tissue localization: (1) Ldha.1 predominates in skeletal muscle and the liver (2) Ldhb.1 is highly expressed in the heart (3) Ldhc.1, Ldhc.2 are expressed only in testes and spermatoza. Tissue localization according to
LDH_L	0	Li X, Qin C, Burghardt R, Safe S.	Hormonal regulation of lactate dehydrogenase-A through activation of protein kinase C pathways in MCF-7 breast cancer cells.	Biochem Biophys Res Commun	2004	15240094	tissue localization: Ldh1 (HHHH): mycoardium, RBC Ldh2 (HHHM): myocardiam, RBC Ldh3 (HHMM): brain, kidney Ldh4 (HMMM) Ldh5 (MMMM): liver, sk muscle [Devlin, Textbook of Biochem, 2003] [Yu et al, Biochem Pharmacol. 2001 Jul 1;62(1):81-9] 3939: - predominantly expressed in muscle [RefSeq] -cytosolic [RefSeq] 3945: cytosolic [UniProt] 3948: - testis-specific [RefSeq] -cytosolic [UniProt] 92483: - testis specific [UniProt] - Additional information added by RS/TV: Cytosolic according to GeneCards Lactate dehydrogenase (LDH) catalyzes the NAD+-dependent conversion of lactate to pyruvate during anaerobic glycolysis according to Li X. Biochem Biophys Res Commun. 2004 Jul 30;320(3):625-34. LDH has three major isozymes each of which has a specific tissue localization: (1) Ldha.1 predominates in skeletal muscle and the liver (2) Ldhb.1 is highly expressed in the heart (3) Ldhc.1, Ldhc.2 are expressed only in testes and spermatoza. Tissue localization according to
LDH_Lm	3	Brooks GA, Dubouchaud H, Brown M, Sicurello JP, Butz CE.	Role of mitochondrial lactate dehydrogenase and lactate oxidation in the intracellular lactate shuttle.	Proc Natl Acad Sci U S A	1999	9927705	- L-lactate is quickly metabolized to pyruvate in the liver [Ewaschuk 2005] - Additional information added by RS/TV: According to the paper listed below Ldh-1 and Ldh-5 are located in the mitochondria as well. According to Entrez gene database Ldh-1 is an alternative name for Ldha.1. Ldh-5 is not listed in Entrez gene database nor does there exist a 'Ldhe' protein. Tissue Localization: Ldh-1 (Ldha.1-m) is located primarily in the heart. All this according to the following paper:Brooks, G; "Role of mitochondrial lactate dehydrogenase and lactate oxidation in the intracellular lactate shuttle"; PNAS 1999.

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LEUKTRC4t	3	Abe T, Kakyo M, Tokui T, Nakagomi R, Nishio T, Nakai D, Nomura H, Unno M, Suzuki M, Naitoh T, Matsuno S, Yawo H.	Identification of a novel gene family encoding human liver-specific organic anion transporter LST-1.	J Biol Chem	1999	10358072	Tissue Specificity: SLCO2A1 - ubiquitous SLCO1A2 - brain, kidney, lung, testis, liver SLCO1B1 - liver SLCO1B3 - liver SLCO2B1 - liver, placenta, spleen, lung, kidney, heart, ovary SLCO3A1 - ubiquitously SLCO4A1 - ubiquitously SLCO1C1 - brain, testis SLCO1B1-Mediates the Na(+)-independent transport of organic anions such as pravastatin, taurocholate, methotrexate, dehydroepiandrosterone sulfate, 17-beta-glucuronosyl estradiol, estrone sulfate, prostaglandin E2, thromboxane B2, leukotriene C3, leukotriene E4, thyroxine and triiodothyronine. May play an important role in the clearance of bile acids and organic anions from the liver. NJ
LEUt4	3	Bertran J, Magagnin S, Werner A, Markovich D, Biber J, Testar X, Zorzano A, Kuhn LC, Palacin M, Murer H	Stimulation of system y(+)-like amino acid transport by the heavy chain of human 4F2 surface antigen in Xenopus laevis oocytes	Proc Natl Acad Sci U S A	1992	1376926	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, lysine, and ornithine across the plasma membrane (PMID 1348489) SLC3A2: PMID 10391915: 4F2hc alone induced, as previously reported (16, 18, 24-27), y+L amino acid transport activity (i.e. sodium-independent L-arginine transport and sodium-dependent L-leucine transport).
LEUt4	3	Pineda M, Fernandez E, Torrents D, Estevez R, Lopez C, Camps M, Lloberas J, Zorzano A, Palacin M	Identification of a membrane protein, LAT-2, that Co-expresses with 4F2 heavy chain, an L-type amino acid transport activity with broad specificity for small and large zwitterionic amino acids	J Biol Chem	1999	10391915	SLC38A4 is found predominantly in liver and transports both cationic and neutral amino acids. The transport of cationic amino acids by SLC38A4 is Na(+) and pH independent, while the transport of neutral amino acids is Na(+) and pH dependent (Hatanaka et al., 2001).[supplied by OMIM] Slc38a1: PMID 10891391 ATA2 mRNA is present in all tissues tested including the liver, kidney, colon, and small intestine as has been reported recently from our laboratory The transport of [14C]MeAIB is inhibited most markedly by alanine, serine, methionine, asparagine and glutamine. Moderate inhibition is observed with glycine, proline, threonine, leucine, and phenylalanine 1:1 Na: aa, no H transport reported SLC7A1 (and presumably 2,3): its role in normal cell metabolism is transport of the cationic amino acids, arginine, lysine, and ornithine across the plasma membrane (PMID 1348489) SLC3A2: PMID 10391915: 4F2hc alone induced, as previously reported (16, 18, 24-27), y+L amino acid transport activity (i.e. sodium-independent L-arginine transport and sodium-dependent L-leucine transport).

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LEUtec	3	Babu E, Kanai Y, Chairoungdua A, Kim do K, Iribe Y, Tangtrongsup S, Jutabha P, Li Y, Ahmed N, Sakamoto S, Anzai N, Nagamori S, Endou H	Identification of a novel system L amino acid transporter structurally distinct from heterodimeric amino acid transporters	J Biol Chem	2003	12930836	From PMID 12930836: Consistent with the results from the inhibition experiments, 14C-labeled L-leucine, L-isoleucine, L-valine, L-phenylalanine, and L-methionine (100 µM) were transported at relatively high rate by LAT3 (Fig. 2b). Among D-amino acids, D-leucine, for which a 14C-labeled compound was available, was confirmed to be transported by LAT3 (Fig. 2b). As observed for L-leucine uptake, the Eadie-Hofstee plots for the uptake of L-isoleucine, L-valine, and L-phenylalanine were curvilinear (data not shown). Kinetic parameters of these amino acid substrates are listed in Table I. From PMID 15659399: We next performed kinetic analysis for the induced transport activity of L-phenylalanine (Fig. 3 and Table I) and L-leucine. Interestingly, and similar to LAT3 (15), the expression of LAT4 in oocytes leads to the presence of a transport activity with two kinetic components. The low affinity component has a Km of 4694 ± 510 µM for L-phenylalanine and 3733 ± 1019 µM for L-leucine, and the high affinity component has a Km of 178 ± 29 µM for L-phenylalanine and 103 ± 62 µM for L-leucin To further characterize the activity induced by LAT4, we measu Other compounds transported by these genes as well.
LEUtec	3	Hediger MA, Romero MF, Peng JB, Rolfs A, Takanaga H, Bruford EA	The ABCs of solute carriers: physiological, pathological and therapeutic implications of human membrane transport proteinsIntroduction	Pflugers Arch	2004	14624363	From PMID 12930836: Consistent with the results from the inhibition experiments, 14C-labeled L-leucine, L-isoleucine, L-valine, L-phenylalanine, and L-methionine (100 µM) were transported at relatively high rate by LAT3 (Fig. 2b). Among D-amino acids, D-leucine, for which a 14C-labeled compound was available, was confirmed to be transported by LAT3 (Fig. 2b). As observed for L-leucine uptake, the Eadie-Hofstee plots for the uptake of L-isoleucine, L-valine, and L-phenylalanine were curvilinear (data not shown). Kinetic parameters of these amino acid substrates are listed in Table I. From PMID 15659399: We next performed kinetic analysis for the induced transport activity of L-phenylalanine (Fig. 3 and Table I) and L-leucine. Interestingly, and similar to LAT3 (15), the expression of LAT4 in oocytes leads to the presence of a transport activity with two kinetic components. The low affinity component has a Km of 4694 ± 510 µM for L-phenylalanine and 3733 ± 1019 µM for L-leucine, and the high affinity component has a Km of 178 ± 29 µM for L-phenylalanine and 103 ± 62 µM for L-leucin To further characterize the activity induced by LAT4, we measu Other compounds transported by these genes as well.
LEUtec	3	Bodoy S, Martin L, Zorzano A, Palacin M, Estevez R, Bertran J	Identification of LAT4, a novel amino acid transporter with system L activity	J Biol Chem	2005	15659399	From PMID 12930836: Consistent with the results from the inhibition experiments, 14C-labeled L-leucine, L-isoleucine, L-valine, L-phenylalanine, and L-methionine (100 µM) were transported at relatively high rate by LAT3 (Fig. 2b). Among D-amino acids, D-leucine, for which a 14C-labeled compound was available, was confirmed to be transported by LAT3 (Fig. 2b). As observed for L-leucine uptake, the Eadie-Hofstee plots for the uptake of L-isoleucine, L-valine, and L-phenylalanine were curvilinear (data not shown). Kinetic parameters of these amino acid substrates are listed in Table I. From PMID 15659399: We next performed kinetic analysis for the induced transport activity of L-phenylalanine (Fig. 3 and Table I) and L-leucine. Interestingly, and similar to LAT3 (15), the expression of LAT4 in oocytes leads to the presence of a transport activity with two kinetic components. The low affinity component has a Km of 4694 ± 510 µM for L-phenylalanine and 3733 ± 1019 µM for L-leucine, and the high affinity component has a Km of 178 ± 29 µM for L-phenylalanine and 103 ± 62 µM for L-leucin To further characterize the activity induced by LAT4, we measu Other compounds transported by these genes as well.
LGTHL	3	Kim NS, Umezawa Y, Ohmura S, Kato S	Human glyoxalase I. cDNA cloning, expression, and sequence similarity to glyoxalase I from Pseudomonas putida	J Biol Chem	1993	7684374	- Catalyzes the conversion of hemimercaptal, formed from methylglyoxal and glutathione, to S-lactoylglutathione [UniProt] - cloned and expressed [Ranganathan 1993], [Kim 1993] - 51% nucleotide homology and 42% amino acid homology with bacterial glyoxalase-I [Ranganathan 1993]; 57% identity with Pseudomonas putida glyoxalase I [Kim 1993]
LGTHL	3	Ranganathan S, Walsh ES, Godwin AK, Tew KD	Cloning and characterization of human colon glyoxalase-I	J Biol Chem	1993	8449929	- Catalyzes the conversion of hemimercaptal, formed from methylglyoxal and glutathione, to S-lactoylglutathione [UniProt] - cloned and expressed [Ranganathan 1993], [Kim 1993] - 51% nucleotide homology and 42% amino acid homology with bacterial glyoxalase-I [Ranganathan 1993]; 57% identity with Pseudomonas putida glyoxalase I [Kim 1993]

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L-LACt2r	3	Garcia CK, Li X, Luna J, Francke U	cDNA cloning of the human monocarboxylate transporter 1 and chromosomal localization of the SLC16A1 locus to 1p13.2-p12	Genomics	1994	7835905	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l
L-LACt2r	3	Poole RC, Halestrap AP	Transport of lactate and other monocarboxylates across mammalian plasma membranes	Am J Physiol	1993	8476015	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l
L-LACt2r	3	Price NT, Jackson VN, Halestrap AP	Cloning and sequencing of four new mammalian monocarboxylate transporter (MCT) homologues confirms the existence of a transporter family with an ancient past	Biochem J	1998	9425115	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l

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L-LACt2r	3	Ritzhaupt A, Ellis A, Hosie KB, Shirazi-Beechey SP	The characterization of butyrate transport across pig and human colonic luminal membrane	J Physiol	1998	9508842	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l
L-LACt2r	3	Ritzhaupt A, Wood IS, Ellis A, Hosie KB, Shirazi-Beechey SP	Identification of a monocarboxylate transporter isoform type 1 (MCT1) on the luminal membrane of human and pig colon	Biochem Soc Trans	1998	9649795	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l
L-LACt2r	3	Lin RY, Vera JC, Chaganti RS, Golde DW	Human monocarboxylate transporter 2 (MCT2) is a high affinity pyruvate transporter	J Biol Chem	1998	9786900	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l

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L-LACt2r	3	Ritzhaupt A, Wood IS, Ellis A, Hosie KB, Shirazi-Beechey SP	Identification and characterization of a monocarboxylate transporter (MCT1) in pig and human colon: its potential to transport L-lactate as well as butyrate	J Physiol	1998	9824713	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l
L-LACt2r	3	Yoon H, Donoso LA, Philp NJ	Cloning of the human monocarboxylate transporter MCT3 gene: localization to chromosome 22q12.3-q13.2.	Genomics	1999	10493836	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l
L-LACt2r	3	Halestrap AP, Price NT	The proton-linked monocarboxylate transporter (MCT) family: structure, function and regulation	Biochem J	1999	10510291	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l

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L-LACt2r	3	Manning Fox JE, Meredith D, Halestrap AP	Characterisation of human monocarboxylate transporter 4 substantiates its role in lactic acid efflux from skeletal muscle	J Physiol	2000	11101640	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l
L-LACt2r	3	Kim do K, Kanai Y, Matsuo H, Kim JY, Chairoungdua A, Kobayashi Y, Enomoto A, Cha SH, Goya T, Endou H	The human T-type amino acid transporter-1: characterization, gene organization, and chromosomal location	Genomics	2002	11827462	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l
L-LACt2r	3	Halestrap AP, Meredith D	The SLC16 gene family-from monocarboxylate transporters (MCTs) to aromatic amino acid transporters and beyond	Pflugers Arch	2004	12739169	- lactate transported into blood by exercising sk muscle and cells that lack mitochondria, such as red blood cells [Champe, Biochemistry 2005] 6566: - isolated [Garcia 1994] - 86% identtity to hamster protein [Garcia 1994] - expressed [Ritzhaupt, Ellis, J Physiol 1998], [Ritzhaupt, Wood, J Physiol et al 1998] - transports butyrate [Ritzhaupt, Ellis, J Physiol 1998] and lactate [Ritzhaupt, Wood, J Physiol et al 1998] - ubiquitous but predominant in colon [Ritzhaupt, Biochem Soc Trans 1998], [Price 1998], heart, red muscle [Halestrap 1999], [Price 1998] - L-Lactate transport in erythrocytes, cardiac myocytes, and hepatocytes due to proton-linked monocarboxylate transporter (see [Poole 1993] for review) - also transports pyruvate, b-hydroxybutyrate, acetoacetate [Halestrap 2004] - in most tissues MCT1 exports lactate to prevernt fall in cytosolic pH; in sk muscle and heart, MCT1 imports lactate to supply gluconeogenesis and lipogenesis; MCT1 or MCT2 imports lactate in liver, kidney and brain [Halestrap 2004] 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell l
LNS14DM	3	Bylund J, Finnstrom N, Oliw EH.	Gene expression of a novel cytochrome P450 of the CYP4F subfamily in human seminal vesicles.	Biochem Biophys Res Commun	1999	10405341	Need to resolve ER vs cytosol - membrane bound ER in uniprot (however some membrane bound enzymes catalyze reactions from the outer side --> substrates and products are cytosolic. NJ
LNS14DMr	3	Lepesheva GI, Waterman MR	CYP51 - the omnipotent P450	Molecular and Cell Endocrinology	2004		ER - see refs specificity: Ubiquitously expressed with highest levels in testis, ovary, adrenal, prostrate, liver, kidney, and lung. Catalyzes C14-demethylation of lanosterol; it transforms lanosterol into 4,4'-dimethyl cholesta-8,14,24-triene-3-beta-ol. NJ
LPASE	3	Perisic O, Fong S, Lynch DE, Bycroft M, Williams RL.	Crystal structure of a calcium-phospholipid binding domain from cytosolic phospholipase A2.	J Biol Chem	1998	9430701	cytoplasm - uniprot NJ

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LPCOXp	3	Ilst L, de Kromme I, Oostheim W, Wanders RJ.	Molecular cloning and expression of human L-pipecolate oxidase.		2000	10772957	0
LPS2	3	Emi M, Wilson DE, Iverius PH, Wu L, Hata A, Hegele R, Williams RR, Lalouel JM.	Missense mutation (Gly----Glu188) of human lipoprotein lipase imparting functional deficiency.	J Biol Chem	1990	1969408	cytosolic - uniprot See PMID: 1969408 for evidence of biochem, result of SNPs and associated dz. - actually degradation of tag, not synthesis NJ
LPS2e	2	Ben-Zeev O, Doolittle MH.	Maturation of hepatic lipase. Formation of functional enzyme in the endoplasmic reticulum is the rate-limiting step in its secretion.	J Biol Chem	2004	14630921	LIPC: location: secreted. specificity: liver (hepatic lipase). LIPC has the dual functions of triglyceride hydrolase and ligand/bridging factor for receptor-mediated lipoprotein uptake. Hepatic lipase has the capacity to catalyze hydrolysis of phospholipids, mono-, di-, and triglycerides, and acyl-CoA thioesters. It is an important enzyme in HDL metabolism. Hepatic lipase binds heparin. see PMID: 14630921 NJ
LPS3	3	Wall EM, Cao J, Chen N, Buller RM, Upton C.	A novel poxvirus gene and its human homolog are similar to an E. coli lysophospholipase.	Virus Res	1997	9495531	localization: cytosol (by default - no specificity) TISSUE SPECIFICITY: Detected in adipose tissue, lung, liver, kidney, brain and heart. Converts monoacylglycerides to free fatty acids and glycerol. Hydrolyzes 2-arachidonoylglycerol, a putative endocannabinoid By seq homology and needed functionality. See PMID: 9495531 NJ
LPS4e	3	Valentin E, Ghomashchi F, Gelb MH, Lazdunski M, Lambeau G.	Novel human secreted phospholipase A(2) with homology to the group III bee venom enzyme.	J Biol Chem	2000	10713052	location: secreted (uniprot) specificity: Expressed in kidney, heart, liver, and skeletal muscle. Also present in placenta and peripheral blood leukocytes. Not detected in brain, colon, thymus, spleen, small intestine and lung. PA2 catalyzes the calcium-dependent hydrolysis of the 2- acyl groups in 3-sn-phosphoglycerides. Shows an 11-fold preference for phosphatidylglycerol over phosphatidylcholine. NJ
LPSe	3	Giller T, Buchwald P, Blum-Kaelin D, Hunziker W.	Two novel human pancreatic lipase related proteins, hPLRP1 and hPLRP2. Differences in colipase dependence and in lipase activity.	J Biol Chem	1992	1379598	secreted - uniprot actually degradation of tag, not synthesis This gene is a member of the lipase gene family. It encodes a carboxyl esterase that hydrolyzes insoluble, emulsified triglycerides, and is essential for the efficient digestion of dietary fats. This gene is expressed specifically in the pancreas. LIPC: location: secreted. specificity: liver (hepatic lipase). LIPC has the dual functions of triglyceride hydrolase and ligand/bridging factor for receptor-mediated lipoprotein uptake. Hepatic lipase has the capacity to catalyze hydrolysis of phospholipids, mono-, di-, and triglycerides, and acyl-CoA thioesters. It is an important enzyme in HDL metabolism. Hepatic lipase binds heparin. see PMID: 14630921 PNLIPRP1 and PNLIPRP2: PMID: 1379598. Extracellular - secreted enzyme (uniport). TISSUE SPECIFICITY: Pancreas. LIPF: PMID: 3304425 and 2753032. SUBCELLULAR LOCATION: Secreted. Specificity: stomach (gastric lipase) LIPG: PMID: 10318835. Extracellular - secreted. TISSUE SPECIFICITY: High level of expression in the liver, placenta, lung, thyroid, kidney, testis and in the corpus luteum of the ova It is termed endothelial lipase due to the fact that it is synthesize NJ

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LPSe	3	Davis RC, Diep A, Hunziker W, Klisak I, Mohandas T, Schotz MC, Sparkes RS, Lusis AJ.	Assignment of human pancreatic lipase gene (PNLIP) to chromosome 10q24-q26.	Genomics	1991	1783385	secreted - uniprot actually degradation of tag, not synthesis This gene is a member of the lipase gene family. It encodes a carboxyl esterase that hydrolyzes insoluble, emulsified triglycerides, and is essential for the efficient digestion of dietary fats. This gene is expressed specifically in the pancreas. LIPC: location: secreted. specificity: liver (hepatic lipase). LIPC has the dual functions of triglyceride hydrolase and ligand/bridging factor for receptor-mediated lipoprotein uptake. Hepatic lipase has the capacity to catalyze hydrolysis of phospholipids, mono-, di-, and triglycerides, and acyl-CoA thioesters. It is an important enzyme in HDL metabolism. Hepatic lipase binds heparin. see PMID: 14630921 PNLIPRP1 and PNLIPRP2: PMID: 1379598. Extracellular - secreted enzyme (uniport). TISSUE SPECIFICITY: Pancreas. LIPF: PMID: 3304425 and 2753032. SUBCELLULAR LOCATION: Secreted. Specificity: stomach (gastric lipase) LIPG: PMID: 10318835. Extracellular - secreted. TISSUE SPECIFICITY: High level of expression in the liver, placenta, lung, thyroid, kidney, testis and in the corpus luteum of the ova It is termed endothelial lipase due to the fact that it is synthesize NJ
LPSe	3	Bernback S, Blackberg L.	Human gastric lipase. The N-terminal tetrapeptide is essential for lipid binding and lipase activity.	Eur J Biochem	1989	2753032	secreted - uniprot actually degradation of tag, not synthesis This gene is a member of the lipase gene family. It encodes a carboxyl esterase that hydrolyzes insoluble, emulsified triglycerides, and is essential for the efficient digestion of dietary fats. This gene is expressed specifically in the pancreas. LIPC: location: secreted. specificity: liver (hepatic lipase). LIPC has the dual functions of triglyceride hydrolase and ligand/bridging factor for receptor-mediated lipoprotein uptake. Hepatic lipase has the capacity to catalyze hydrolysis of phospholipids, mono-, di-, and triglycerides, and acyl-CoA thioesters. It is an important enzyme in HDL metabolism. Hepatic lipase binds heparin. see PMID: 14630921 PNLIPRP1 and PNLIPRP2: PMID: 1379598. Extracellular - secreted enzyme (uniport). TISSUE SPECIFICITY: Pancreas. LIPF: PMID: 3304425 and 2753032. SUBCELLULAR LOCATION: Secreted. Specificity: stomach (gastric lipase) LIPG: PMID: 10318835. Extracellular - secreted. TISSUE SPECIFICITY: High level of expression in the liver, placenta, lung, thyroid, kidney, testis and in the corpus luteum of the ova It is termed endothelial lipase due to the fact that it is synthesize NJ
LPSe	3	Bodmer MW, Angal S, Yarranton GT, Harris TJ, Lyons A, King DJ, Pieroni G, Riviere C, Verger R, Lowe PA.	Molecular cloning of a human gastric lipase and expression of the enzyme in yeast.	Biochim Biophys Acta	1987	3304425	secreted - uniprot actually degradation of tag, not synthesis This gene is a member of the lipase gene family. It encodes a carboxyl esterase that hydrolyzes insoluble, emulsified triglycerides, and is essential for the efficient digestion of dietary fats. This gene is expressed specifically in the pancreas. LIPC: location: secreted. specificity: liver (hepatic lipase). LIPC has the dual functions of triglyceride hydrolase and ligand/bridging factor for receptor-mediated lipoprotein uptake. Hepatic lipase has the capacity to catalyze hydrolysis of phospholipids, mono-, di-, and triglycerides, and acyl-CoA thioesters. It is an important enzyme in HDL metabolism. Hepatic lipase binds heparin. see PMID: 14630921 PNLIPRP1 and PNLIPRP2: PMID: 1379598. Extracellular - secreted enzyme (uniport). TISSUE SPECIFICITY: Pancreas. LIPF: PMID: 3304425 and 2753032. SUBCELLULAR LOCATION: Secreted. Specificity: stomach (gastric lipase) LIPG: PMID: 10318835. Extracellular - secreted. TISSUE SPECIFICITY: High level of expression in the liver, placenta, lung, thyroid, kidney, testis and in the corpus luteum of the ova It is termed endothelial lipase due to the fact that it is synthesize NJ

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LPSe	3	Hirata K, Dichek HL, Cioffi JA, Choi SY, Leeper NJ, Quintana L, Kronmal GS, Cooper AD, Quertermous T.	Cloning of a unique lipase from endothelial cells extends the lipase gene family.	J Biol Chem	1999	10318835	secreted - uniprot actually degradation of tag, not synthesis This gene is a member of the lipase gene family. It encodes a carboxyl esterase that hydrolyzes insoluble, emulsified triglycerides, and is essential for the efficient digestion of dietary fats. This gene is expressed specifically in the pancreas. LIPC: location: secreted. specificity: liver (hepatic lipase). LIPC has the dual functions of triglyceride hydrolase and ligand/bridging factor for receptor-mediated lipoprotein uptake. Hepatic lipase has the capacity to catalyze hydrolysis of phospholipids, mono-, di-, and triglycerides, and acyl-CoA thioesters. It is an important enzyme in HDL metabolism. Hepatic lipase binds heparin. see PMID: 14630921 PNLIPRP1 and PNLIPRP2: PMID: 1379598. Extracellular - secreted enzyme (uniport). TISSUE SPECIFICITY: Pancreas. LIPF: PMID: 3304425 and 2753032. SUBCELLULAR LOCATION: Secreted. Specificity: stomach (gastric lipase) LIPG: PMID: 10318835. Extracellular - secreted. TISSUE SPECIFICITY: High level of expression in the liver, placenta, lung, thyroid, kidney, testis and in the corpus luteum of the ova It is termed endothelial lipase due to the fact that it is synthesize NJ
LRAT	3	Jahng WJ, Xue L, Rando RR.	Lecithin retinol acyltransferase is a founder member of a novel family of enzymes.	Biochemistry	2003	14596594	IT
LRAT	3	Zolfaghari R, Ross AC.	Cloning, gene organization and identification of an alternative splicing process in lecithin:retinol acyltransferase cDNA from human liver.	Gene	2004	15474300	IT
LSTO1r	3	Taton M, Husselstein T, Benveniste P, Rahier A	Role of highly conserved residues in the reaction catalyzed by recombinant delta-sterol-C5(6)-desaturase studied by site-directed mutagensis	Biochemistry	2000		ER - see refs no tissue specificity Catalyzes a dehydrogenation to introduce C5-6 double bond into lathosterol. NJ
LTC4CP	2	Reddanna P, Prabhu KS, Whelan J, Reddy CC.	Carboxypeptidase A-catalyzed direct conversion of leukotriene C4 to leukotriene F4.	Arch Biochem Biophys	2003	12729612	Converts LTC4 directly to LTF4 cytoplasmic by default (no other specific information). Conversion performed by carboxypeptidase A (GPR not found yet). PMID: 12729612 NJ
LTD4DP	3	Hammarstrom S, Orning L, Bernstrom K.	Metabolism of leukotrienes.	Mol Cell Biochem	1985	3001504	Unknown GPR, known to occur biochemically. cytosolic by default NJ
LTDCL	3	Kitahama K, Ikemoto K, Jouvet A, Nagatsu I, Sakamoto N, Pearson J	Aromatic L-amino acid decarboxylase- and tyrosine hydroxylase-immunohistochemistry in the adult human hypothalamus	J Chem Neuroanat	1998	9924972	Citation suggests that this reaction produces tryptamine in minute quantities. The biochemical characterization is specific to the brain.
LYSOXp	2	Yung-Feng Chang	Lysine metabolism in the human and the monkey: Demonstration of pipecolic acid formation in the brain and other organs		1982	6811962	PMID 10772957: In higher eukaryotes L-lysine can be degraded via two distinct routes including the saccharopine pathway and the L-pipecolate pathway. since pipecolate oxidase (LPCOXp) occurs in the peroxisome, the three reactions (LYSOXp, PPD2CSPp, 1PPDCRp) preceding it are assumed to be peroxisomal as well MM
LYSOXp	2	Murthy SN, Janardanasarma MK.	Identification of L-amino acid/L-lysine alpha-amino oxidase in mouse brain.		1999	10485319	PMID 10772957: In higher eukaryotes L-lysine can be degraded via two distinct routes including the saccharopine pathway and the L-pipecolate pathway. since pipecolate oxidase (LPCOXp) occurs in the peroxisome, the three reactions (LYSOXp, PPD2CSPp, 1PPDCRp) preceding it are assumed to be peroxisomal as well MM
M1316Mg	0	Misago M, Liao YF, Kudo S, Eto S, Mattei MG, Moremen KW, Fukuda MN.	Molecular cloning and expression of cDNAs encoding human alpha-mannosidase II and a previously unrecognized alpha-mannosidase IIx isozyme.	Proc Natl Acad Sci U S A	1995	8524845	Man2a1p and Man2a2p ubiquitously expressed [Misago et al, PNAS 1995]
M13N2Tg	0	Yen CL, Farese RV Jr.	MGAT2, a monoacylglycerol acyltransferase expressed in the small intestine	J Biol Chem	2003	12621063	There are believed to be over 100 different glycosyltransferases involved in the synthesis of protein-bound and lipid-bound oligosaccharides. UDP-N-acetylglucosamine:alpha-3-D-mannoside beta-1,2-N-acetylglucosaminyltransferase I is a medial-Golgi enzyme essential for the synthesis of hybrid and complex N-glycans. The protein, encoded by a single exon, shows typical features of a type II transmembrane protein. The protein is believed to be essential for normal embryogenesis. [RefSeq] Mgat1p expressed in liver, kidney [Yen and Farese Jr, J Biol Chem 2003]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
M13N4Tg	0	Yoshida A, Minowa MT, Takamatsu S, Hara T, Oguri S, Ikenaga H, Takeuchi M.	Tissue specific expression and chromosomal mapping of a human UDP-N-acetylglucosamine: alpha1,3-d-mannoside beta1, 4-N-acetylglucosaminyltransferase.	Glycobiology	1999	10024668	Branching structures in complex N-glycans are synthesized on a commom core structure of Man3GlcNAc2Asn in the Golgi apparatus by the N-acetylglucosaminyltransferases. The mannosyl (alpha-1,3-)-glycoprotein beta-1,4-N-acetylglucosaminyltransferases, which include isoenzyme A (MGAT4A) and isoenzyme B (MGAT4B), are key N-acetylglucosaminyltransferases regulating formation of tri- and other multiantennary structures. MGAT4A and MGAT4B share 62% amino acid sequence identity. The huamn MGAT4A and bovine GnT-IV share 96% amino acid sequence identity. The expression levels of the MGAT4A mRNA are significantly different in various tissues and cell lines. Alternative splicing of MGAT4B results in two transcript variants encoding different isoforms. [RefSeq] Mgat4ap ubiquitously expressed [Yoshida et al, Glycobiology 1999] [Yoshida et al, Glycoconj J 1998] Mgat4b ubiquitously expressed [Yoshida et al, Glycoconj J 1998]
M13N4Tg	0	Yoshida A, Minowa MT, Takamatsu S, Hara T, Ikenaga H, Takeuchi M.	A novel second isoenzyme of the human UDP-N-acetylglucosamine:alpha1,3-D-mannoside beta1,4-N-acetylglucosaminyltransferase family: cDNA cloning, expression, and chromosomal assignment.	Glycoconj J	1998	10372966	Branching structures in complex N-glycans are synthesized on a commom core structure of Man3GlcNAc2Asn in the Golgi apparatus by the N-acetylglucosaminyltransferases. The mannosyl (alpha-1,3-)-glycoprotein beta-1,4-N-acetylglucosaminyltransferases, which include isoenzyme A (MGAT4A) and isoenzyme B (MGAT4B), are key N-acetylglucosaminyltransferases regulating formation of tri- and other multiantennary structures. MGAT4A and MGAT4B share 62% amino acid sequence identity. The huamn MGAT4A and bovine GnT-IV share 96% amino acid sequence identity. The expression levels of the MGAT4A mRNA are significantly different in various tissues and cell lines. Alternative splicing of MGAT4B results in two transcript variants encoding different isoforms. [RefSeq] Mgat4ap ubiquitously expressed [Yoshida et al, Glycobiology 1999] [Yoshida et al, Glycoconj J 1998] Mgat4b ubiquitously expressed [Yoshida et al, Glycoconj J 1998]
M4CET3er	3	Taron BW, Colussi PA, Wiedman JM, Orlean P, Taron CH	Human Smp3p adds a fourth mannose to yeast and human glycosylphosphatidylinositol precursors in vivo	J Biol Chem	2004	15208306	- M4A is more likely to be produced by the adding phosphoethanolamines to M4C rather than mannose addition to H8 [Taron, J Biol Chem 2004] 54872: - gene was cloned, function inferred from knockout [Shishioh, J Biol Chem 2005]
M8MASNterg	0	Spiro RG	Glucose residues as key determinants in the biosynthesis and quality control of glycoproteins with N-linked oligosaccharides.	J Biol Chem	2000	11007802	see Figure 2 in Spiro, J Biol Chem 275(46): 35657-35660 (2000).
MACACI	3	Fernandez-Canon JM, Hejna J, Reifsteck C, Olson S, Grompe M	Gene structure, chromosomal location, and expression pattern of maleylacetoacetate isomerase	Genomics	1999	10373324	This reaction appears to require glutathione as a cofactor, although it doesn't seem to appear in the reaction itself.
MACOXO	3	Zimatkin SM, Anichtchik OV	Alcohol-histamine interactions	Alcohol Alcohol	1999	10344773	Citation gives enzymes that catalyze reaction.
MALT	3	Martiniuk F, Hirschhorn R	Characterization of neutral isozymes of human alpha-glucosidase: differences in substrate specificity, molecular weight and electrophoretic mobility	Biochim Biophys Acta	1981	7018580	- alpha-glucosidase activity [UniProt], [Hirschorn, PNAS 2002] - maltose, maltotriose, and glycogen are substrates of Ganc (neutral alpha-glucosidase C); has similar catalytic properties as GAA [Martiniuk 1981]
MALT	3	Hirschhorn R, Huie ML, Kasper JS	Computer assisted cloning of human neutral alpha-glucosidase C (GANC): a new paralog in the glycosyl hydrolase gene family 31	Proc Natl Acad Sci U S A	2002	12370436	- alpha-glucosidase activity [UniProt], [Hirschorn, PNAS 2002] - maltose, maltotriose, and glycogen are substrates of Ganc (neutral alpha-glucosidase C); has similar catalytic properties as GAA [Martiniuk 1981]
MALTly	3	Hoefsloot LH, Hoogeveen-Westerveld M, Kroos MA, van Beeumen J, Reuser AJ, Oostra BA	Primary structure and processing of lysosomal alpha-glucosidase; homology with the intestinal sucrase-isomaltase complex	EMBO J	1988	3049072	- Essential for the degradation of glygogen to glucose in lysosomes [UniProt], [ReSeq] - lysosomal [RefSeq], [UniProt], [Hoefsloot, EMBO J, 1988] - alpha-glycosidase activity [Hoefsloot, EMBO J, 1988] Note: this rxn currently results in a modeling gap. This rxn is valid in vivo, malt usually arises from glycogen deg, but does not appear in our network for modeling reasons (only made representative structure for glycogen and malt doesn't happen to be one of its degradation products)

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MAN1_7Ber	0	Roth J, Zuber C, Guhl B, Fan JY, Ziak M.	The importance of trimming reactions on asparagine-linked oligosaccharides for protein quality control.	Histochem Cell Biol	2002	11935292	catalyzed by ER mannosidase I [Spiro, J Biol Chem (2000)] [Roth et al, Histochem Cell Biol 117: pp. 159-169 (2002)] Man1b1p ubiquitously expressed [Gonzalez et al, J Biol Chem 1999]
MAOX	2	Yu PH, Lai CT, Zuo DM	Formation of formaldehyde from adrenaline in vivo; a potential risk factor for stress-related angiopathy	Neurochem Res	1997	9131641	References state that when adrenaline is degraded, formaldehyde can be formed, and the amine oxidases are involved.
MAOX	2	Buffoni F, Ignesti G	The copper-containing amine oxidases: biochemical aspects and functional role	Mol Genet Metab	2000	11136547	References state that when adrenaline is degraded, formaldehyde can be formed, and the amine oxidases are involved.
MAOX	2	Conklin DJ, Cowley HR, Wiechmann RJ, Johnson GH, Trent MB, Boor PJ	Vasoactive effects of methylamine in isolated human blood vessels: role of semicarbazide-sensitive amine oxidase, formaldehyde, and hydrogen peroxide	Am J Physiol Heart Circ Physiol	2004	14715500	References state that when adrenaline is degraded, formaldehyde can be formed, and the amine oxidases are involved.
MCCCrm	3	Baumgartner MR, Almashanu S, Suormala T, Obie C, Cole RN, Packman S, Baumgartner ER, Valle D.	The molecular basis of human 3-methylcrotonyl-CoA carboxylase deficiency.		2001	11181649	see citation for all confidence evidence, reversibility, localization tissue - predominantly in kidney and liver MM
MCCCrm	3	Holzinger A, Roschinger W, Lagler F, Mayerhofer PU, Lichtner P, Kattenfeld T, Thuy LP, Nyhan WL, Koch HG, Muntau AC, Roscher AA.	Cloning of the human MCCA and MCCB genes and mutations therein reveal the molecular cause of 3-methylcrotonyl-CoA: carboxylase deficiency.		2001	11406611	see citation for all confidence evidence, reversibility, localization tissue - predominantly in kidney and liver MM
MCITS	2	Weidman SW, Drysdale GR.	he biosynthesis of methylcitrate.	Biochem J	1979	426765	- methylcitrate is known to be synthesized in humans [Weidman, Biochem J 1979] - methylcitrate was found to be a major product of proprionate metabolism in patients with proprionic acidemia and methylmalonic acidemia [Ando, PNAS 1972]
MCITS	2	Ando T, Rasmussen K, Nyhan WL, Hull D.	3-hydroxypropionate: significance of -oxidation of propionate in patients with propionic acidemia and methylmalonic acidemia	Proc Natl Acad Sci U S A	1972	4507604	- methylcitrate is known to be synthesized in humans [Weidman, Biochem J 1979] - methylcitrate was found to be a major product of proprionate metabolism in patients with proprionic acidemia and methylmalonic acidemia [Ando, PNAS 1972]
MCLACCYSR	2	Hannestad U, Martensson J, Sjodahl R, Sorbo B.	3-mercaptolactate cysteine disulfiduria: biochemical studies on affected and unaffected members of a family.		1981	6945862	-this is an inferred reaction based on below information: overproduction of mercaptolactate-cysteine disulfide is called 3 mercaptolactate cysteine disulfiduria, which is thought to occur when excess mercaptolactate, in the presence of cysteine, is converted to the mixed disulfide by oxidation (PMID:6945862) MM
MCLOR	3	Cooper AJ, Haber MT, Meister A.	On the chemistry and biochemistry of 3-mercaptopyruvic acid, the alpha-keto acid analog of cysteine		1982	7054184	lactate dehydrogenase biochemically shown to also use mercaptolactate/mercaptopyruvate as substrate - Cooper et. al. J Biol Chem. 1982 Jan 25;257(2):816-26.
MCOATAm	3	Zhang L, Joshi AK, Smith S.	Cloning, expression, characterization, and interaction of two components of a human mitochondrial fatty acid synthase. Malonyltransferase and acyl carrier protein.	J Biol Chem	2003	12882974	Mitochondrial by swiss prot See also: PMID: 12882974 The protein encoded by this gene is found exclusively in the mitochondrion, where it catalyzes the transfer of a malonyl group from malonyl-CoA to the mitochondrial acyl carrier protein. The encoded protein may be part of a fatty acid synthase complex that is more like the type II prokaryotic and plastid complexes rather than the type I human cytosolic complex. Two transcript variants encoding different isoforms have been found for this gene. Catalyzes the transfer of a malonyl moiety from malonyl- CoA to the free thiol group of the phosphopantetheine arm of the mitochondrial ACP protein (NDUFAB1). This suggests the existence of the biosynthesis of fatty acids in mitochondrias. NJ

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ME2	0	Loeber G, Infante AA, Maurer-Fogy I, Krystek E, Dworkin MB.	Human NAD(+)-dependent mitochondrial malic enzyme. cDNA cloning, primary structure, and expression in Escherichia coli.	J Biol Chem	1991	1993674	- cytosolic [RefSeq], [UniProt] - NADP is cofactor [RefSeq], [UniProt] - Additional information added by RS/TV: Malic enzyme catalyzes the oxidative decarboxylation of malate to pyruvate. Three different isoforms of malic enzyme have been found in mammalian tissues: (1) Me1.1: Cytosolic NADP+ dependent enzyme (2) Me3.1.-m: NADP+ dependent mitochondrial enzyme (3) Me2.1-m: NAD+/NADP+ dependent mitochondrial enzyme Each isoform has different tissue localizations: (1) Me1.1: Liver and adipose tissue (2) Me3.1-m: Brain, heart, skeletal muscle, and adrenals (3)Me2.1-m: Spleen, thymus, and the basal cells of the small intestinal mucosaAll of this according to Loeber G, Infante AA, Maurer-Fogy I, Krystek E, Dworkin MB. J Biol Chem. 1991 Feb 15;266(5):3016-21.
MELATN23DOX	2	Hirata F, Hayaishi O, Tokuyama T, Seno S	In vitro and in vivo formation of two new metabolites of melatonin	J Biol Chem	1974	4814344	Methods are old and not necessarily human experiments, so physiological evidence.
MELATNOX	3	Ma X, Idle JR, Krausz KW, Gonzalez FJ.	METABOLISM OF MELATONIN BY HUMAN CYTOCHROMES P450.	Drug Metab Dispos	2004	15616152	0
METAT	3	Horikawa,S., Tsukada,K.,	Molecular cloning and developmental expression of a human kidney S- adenosylmethionine synthetase.		1992	1426236	Entrez gene - catalyzes the formation of S-adenosylmethionine from methionine and ATP. Methionine adenosyltransferase deficiency is known to be caused by recessive as well as dominant mutations, the latter identified in autosomal dominant persistant hypermethioninemia. -in mammalian tissues, there are three distinct forms of adomet synthases designated as alpha, beta, and gamma. alpha and beta are expressed only in adult liver, while gamma is widely distributed in extrahepatic tissues. MM
METAT	3	Ubagai,T. , Lei,K.J. , Huang,S. , Mudd,S.H. , Levy,H.L. , Chou,J.Y.	Molecular mechanisms of an inborn error of methionine pathway. Methionine adenosyltransferase deficiency.		1995	7560086	Entrez gene - catalyzes the formation of S-adenosylmethionine from methionine and ATP. Methionine adenosyltransferase deficiency is known to be caused by recessive as well as dominant mutations, the latter identified in autosomal dominant persistant hypermethioninemia. -in mammalian tissues, there are three distinct forms of adomet synthases designated as alpha, beta, and gamma. alpha and beta are expressed only in adult liver, while gamma is widely distributed in extrahepatic tissues. MM
METAT	3	Alvarez,L., Corrales,F., Mato,J.M.,	Characterization of a full-length cDNA encoding human liver S- adenosylmethionine synthetase: tissue specific gene expression and mRNA levels in hepatopathies.		1993	8393662	Entrez gene - catalyzes the formation of S-adenosylmethionine from methionine and ATP. Methionine adenosyltransferase deficiency is known to be caused by recessive as well as dominant mutations, the latter identified in autosomal dominant persistant hypermethioninemia. -in mammalian tissues, there are three distinct forms of adomet synthases designated as alpha, beta, and gamma. alpha and beta are expressed only in adult liver, while gamma is widely distributed in extrahepatic tissues. MM
MGCHrm	3	Narisawa K, Gibson KM, Sweetman L, Nyhan WL, Duran M, Wadman SK.	Deficiency of 3-methylglutaconyl-coenzyme A hydratase in two siblings with 3-methylglutaconic aciduria.		1986	3082934	mitochondrial by similarity - UniProt AU-specific RNA-binding enoyl-CoA hydratase (AUH) protein binds to the AU-rich element (ARE), a common element found in the 3' UTR of rapidly decaying mRNA such as c-fos, c-myc and granulocyte/ macrophage colony stimulating factor. ARE elements are involved in directing RNA to rapid degradation and deadenylation. AUH is also homologous to enol-CoA hydratase, an enzyme involved in fatty acid degradation, and has been shown to have intrinsic hydratase enzymatic activity. AUH is thus a bifunctional chimera between RNA binding and metabolic enzyme activity. A possible subcellular localization in the mitochondria has been demonstrated for the mouse homolog of this protein which shares 92% identity with the human protein. It has been suggested that AUH may have a novel role as a mitochondrial located AU-binding protein. reversibility according to citations and Reactome

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MGCHrm	3	IJlst L, Loupatty FJ, Ruiter JP, Duran M, Lehnert W, Wanders RJ.	3-Methylglutaconic aciduria type I is caused by mutations in AUH.		2003	12434311	mitochondrial by similarity - UniProt AU-specific RNA-binding enoyl-CoA hydratase (AUH) protein binds to the AU-rich element (ARE), a common element found in the 3' UTR of rapidly decaying mRNA such as c-fos, c-myc and granulocyte/ macrophage colony stimulating factor. ARE elements are involved in directing RNA to rapid degradation and deadenylation. AUH is also homologous to enol-CoA hydratase, an enzyme involved in fatty acid degradation, and has been shown to have intrinsic hydratase enzymatic activity. AUH is thus a bifunctional chimera between RNA binding and metabolic enzyme activity. A possible subcellular localization in the mitochondria has been demonstrated for the mouse homolog of this protein which shares 92% identity with the human protein. It has been suggested that AUH may have a novel role as a mitochondrial located AU-binding protein. reversibility according to citations and Reactome
MGSA	3	Vander Jagt DL, Robinson B, Taylor KK, Hunsaker LA.	Reduction of trioses by NADPH-dependent aldo-keto reductases. Aldose reductase, methylglyoxal, and diabetic complications	J Biol Chem	1992	1537826	- methylglyoxylate can be produced from the nonenzymatic fragmentation of triose-phosphates [Vander Jagt 1992],[Thornalley 1996] - dhap and g3p are the primary triose-phosphates converted to methylglyoxylate [Beisswenger 2005] - this was the major pathway of methylglyoxalte formation in human red blood cells in vitro under normoglycaemic conditions [Thornalley 1996]
MGSA	3	Thornalley PJ	Pharmacology of methylglyoxal: formation, modification of proteins and nucleic acids, and enzymatic detoxification--a role in pathogenesis and antiproliferative chemotherapy.	Gen Pharmacol	1996	8853285	- methylglyoxylate can be produced from the nonenzymatic fragmentation of triose-phosphates [Vander Jagt 1992],[Thornalley 1996] - dhap and g3p are the primary triose-phosphates converted to methylglyoxylate [Beisswenger 2005] - this was the major pathway of methylglyoxalte formation in human red blood cells in vitro under normoglycaemic conditions [Thornalley 1996]
MGSA	3	Beisswenger BG, Delucia EM, Lapoint N, Sanford RJ, Beisswenger PJ.	Ketosis leads to increased methylglyoxal production on the atkins diet	Ann N Y Acad Sci	2005	16037240	- methylglyoxylate can be produced from the nonenzymatic fragmentation of triose-phosphates [Vander Jagt 1992],[Thornalley 1996] - dhap and g3p are the primary triose-phosphates converted to methylglyoxylate [Beisswenger 2005] - this was the major pathway of methylglyoxalte formation in human red blood cells in vitro under normoglycaemic conditions [Thornalley 1996]
MHISOR	3	Elmore BO, Bollinger JA, Dooley DM	Human kidney diamine oxidase: heterologous expression, purification, and characterization	J Biol Inorg Chem	2002	12072962	Fourth citation is specific for this reaction and gives some help regarding gene associations. MAO is not associated with this reaction because these other genes appeared more accurate.
MI1346PKn	3	Nalaskowski MM, Deschermeier C, Fanick W, Mayr GW	The human homologue of yeast ArgRIII protein is an inositol phosphate multikinase with predominantly nuclear localization	Biochem J	2002	12027805	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] - gene was cloned and has following activity [Nalaskowski, Biochem J 2002]: Ins(1,4,5)P3 -> Ins(1,4,5,6)P4 Ins(1,4,5)P3 -> Ins(1,3,4,5)P4 Ins(1,3,4,5)P4 -> Ins(1,3,4,5,6)P5 Ins(1,4,5,6(P4 -> Ins(1,3,4,5,6)P5 - protein is 83% identical to rat homolog and has following activity [Chang, J Biol Chem 2002]: Ins(1,3,4,6)P4 -> Ins(1,3,4,5,6)P5 - nucelar localization [Nalaskowski, Biochem J 2002]
MI1346PKn	3	Chang SC, Miller AL, Feng Y, Wente SR, Majerus PW.	The human homolog of the rat inositol phosphate multikinase is an inositol 1,3,4,6-tetrakisphosphate 5-kinase	J Biol Chem	2002	12223481	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] - gene was cloned and has following activity [Nalaskowski, Biochem J 2002]: Ins(1,4,5)P3 -> Ins(1,4,5,6)P4 Ins(1,4,5)P3 -> Ins(1,3,4,5)P4 Ins(1,3,4,5)P4 -> Ins(1,3,4,5,6)P5 Ins(1,4,5,6(P4 -> Ins(1,3,4,5,6)P5 - protein is 83% identical to rat homolog and has following activity [Chang, J Biol Chem 2002]: Ins(1,3,4,6)P4 -> Ins(1,3,4,5,6)P5 - nucelar localization [Nalaskowski, Biochem J 2002]
MI145PK	3	Takazawa K, Perret J, Dumont JE, Erneux C	Molecular cloning and expression of a new putative inositol 1,4,5-trisphosphate 3-kinase isoenzyme	Biochem J	1991	1654894	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3706, 3707: - inositol 1,4,5-trisphosphate 3-kinase activity [RefSeq] - gene was cloned and expressed [Takazawa, Biochem J 1991] 80271: - inositol 1,4,5-trisphosphate 3-kinase activity [RefSeq] - gene was cloned and expressed [Dewaste, Biochem J 2000]

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MI145PK	3	Dewaste V, Pouillon V, Moreau C, Shears S, Takazawa K, Erneux C	Cloning and expression of a cDNA encoding human inositol 1,4,5-trisphosphate 3-kinase C	Biochem J	2000	11085927	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3706, 3707: - inositol 1,4,5-trisphosphate 3-kinase activity [RefSeq] - gene was cloned and expressed [Takazawa, Biochem J 1991] 80271: - inositol 1,4,5-trisphosphate 3-kinase activity [RefSeq] - gene was cloned and expressed [Dewaste, Biochem J 2000]
MI145PP	3	Laxminarayan KM, Chan BK, Tetaz T, Bird PI, Mitchell CA	Characterization of a cDNA encoding the 43-kDa membrane-associated inositol-polyphosphate 5-phosphatase	J Biol Chem	1994	8006039	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3633: - Ins(1,3,4,5)P3 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - Inositol polyphosphate 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004], [Haffner, FEBS Lett 1997] 56623: - Inositol polyphophate 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Golgi stacks [UniProt] - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva, J Biol Chem 2000 3632: - inositol 1,4,5-trisphosphate 5-phosphatase [RefSeq] - membrane-associated [RefSeq], [UniProt] - Brain; high level in Purkinje cells [UniProt], [De Smedt, FEBS Lett 1994], [Laxminarayan, J Biol Chem 1994] - gene was cloned and expressed; 91% homology to dog homol 3635: - gene was cloned and expressed [Drayer, Biochem Biophys R 3636:
MI145PP	3	De Smedt F, Verjans B, Mailleux P, Erneux C	Cloning and expression of human brain type I inositol 1,4,5-trisphosphate 5-phosphatase. High levels of mRNA in cerebellar Purkinje cells	FEBS Lett	1994	8013665	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3633: - Ins(1,3,4,5)P3 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - Inositol polyphosphate 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004], [Haffner, FEBS Lett 1997] 56623: - Inositol polyphophate 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Golgi stacks [UniProt] - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva, J Biol Chem 2000 3632: - inositol 1,4,5-trisphosphate 5-phosphatase [RefSeq] - membrane-associated [RefSeq], [UniProt] - Brain; high level in Purkinje cells [UniProt], [De Smedt, FEBS Lett 1994], [Laxminarayan, J Biol Chem 1994] - gene was cloned and expressed; 91% homology to dog homol 3635: - gene was cloned and expressed [Drayer, Biochem Biophys R 3636:

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MI145PP	3	Hejna JA, Saito H, Merkens LS, Tittle TV, Jakobs PM, Whitney MA, Grompe M, Friedberg AS, Moses RE	Cloning and characterization of a human cDNA (INPPL1) sharing homology with inositol polyphosphate phosphatases	Genomics	1995	8530088	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3633: - Ins(1,3,4,5)P3 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - Inositol polyphosphate 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004], [Haffner, FEBS Lett 1997] 56623: - Inositol polyphophate 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Golgi stacks [UniProt] - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva, J Biol Chem 2000 3632: - inositol 1,4,5-trisphosphate 5-phosphatase [RefSeq] - membrane-associated [RefSeq], [UniProt] - Brain; high level in Purkinje cells [UniProt], [De Smedt, FEBS Lett 1994], [Laxminarayan, J Biol Chem 1994] - gene was cloned and expressed; 91% homology to dog homol 3635: - gene was cloned and expressed [Drayer, Biochem Biophys R 3636:
MI145PP	3	Drayer AL, Pesesse X, De Smedt F, Woscholski R, Parker P, Erneux C	Cloning and expression of a human placenta inositol 1,3,4,5-tetrakisphosphate and phosphatidylinositol 3,4,5-trisphosphate 5-phosphatase	Biochem Biophys Res Commun	1996	8769125	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3633: - Ins(1,3,4,5)P3 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - Inositol polyphosphate 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004], [Haffner, FEBS Lett 1997] 56623: - Inositol polyphophate 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Golgi stacks [UniProt] - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva, J Biol Chem 2000 3632: - inositol 1,4,5-trisphosphate 5-phosphatase [RefSeq] - membrane-associated [RefSeq], [UniProt] - Brain; high level in Purkinje cells [UniProt], [De Smedt, FEBS Lett 1994], [Laxminarayan, J Biol Chem 1994] - gene was cloned and expressed; 91% homology to dog homol 3635: - gene was cloned and expressed [Drayer, Biochem Biophys R 3636:

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MI145PP	3	Haffner C, Takei K, Chen H, Ringstad N, Hudson A, Butler MH, Salcini AE, Di Fiore PP, De Camilli P	Synaptojanin 1: localization on coated endocytic intermediates in nerve terminals and interaction of its 170 kDa isoform with Eps15	FEBS Lett	1997	9428629	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3633: - Ins(1,3,4,5)P3 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - Inositol polyphosphate 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004], [Haffner, FEBS Lett 1997] 56623: - Inositol polyphophate 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Golgi stacks [UniProt] - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva, J Biol Chem 2000 3632: - inositol 1,4,5-trisphosphate 5-phosphatase [RefSeq] - membrane-associated [RefSeq], [UniProt] - Brain; high level in Purkinje cells [UniProt], [De Smedt, FEBS Lett 1994], [Laxminarayan, J Biol Chem 1994] - gene was cloned and expressed; 91% homology to dog homol 3635: - gene was cloned and expressed [Drayer, Biochem Biophys R 3636:
MI145PP	3	Pesesse X, Moreau C, Drayer AL, Woscholski R, Parker P, Erneux C	The SH2 domain containing inositol 5-phosphatase SHIP2 displays phosphatidylinositol 3,4,5-trisphosphate and inositol 1,3,4,5-tetrakisphosphate 5-phosphatase activity	FEBS Lett	1998	9824312	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3633: - Ins(1,3,4,5)P3 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - Inositol polyphosphate 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004], [Haffner, FEBS Lett 1997] 56623: - Inositol polyphophate 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Golgi stacks [UniProt] - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva, J Biol Chem 2000 3632: - inositol 1,4,5-trisphosphate 5-phosphatase [RefSeq] - membrane-associated [RefSeq], [UniProt] - Brain; high level in Purkinje cells [UniProt], [De Smedt, FEBS Lett 1994], [Laxminarayan, J Biol Chem 1994] - gene was cloned and expressed; 91% homology to dog homol 3635: - gene was cloned and expressed [Drayer, Biochem Biophys R 3636:

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MI145PP	3	Mochizuki Y, Takenawa T	Novel inositol polyphosphate 5-phosphatase localizes at membrane ruffles	J Biol Chem	1999	10593988	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3633: - Ins(1,3,4,5)P3 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - Inositol polyphosphate 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004], [Haffner, FEBS Lett 1997] 56623: - Inositol polyphophate 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Golgi stacks [UniProt] - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva, J Biol Chem 2000 3632: - inositol 1,4,5-trisphosphate 5-phosphatase [RefSeq] - membrane-associated [RefSeq], [UniProt] - Brain; high level in Purkinje cells [UniProt], [De Smedt, FEBS Lett 1994], [Laxminarayan, J Biol Chem 1994] - gene was cloned and expressed; 91% homology to dog homol 3635: - gene was cloned and expressed [Drayer, Biochem Biophys R 3636:
MI145PP	3	Stopkova P, Vevera J, Paclt I, Zukov I, Lachman HM	Analysis of SYNJ1, a candidate gene for 21q22 linked bipolar disorder: a replication study	Psychiatry Res	2004	15261714	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3633: - Ins(1,3,4,5)P3 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - Inositol polyphosphate 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004], [Haffner, FEBS Lett 1997] 56623: - Inositol polyphophate 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Golgi stacks [UniProt] - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva, J Biol Chem 2000 3632: - inositol 1,4,5-trisphosphate 5-phosphatase [RefSeq] - membrane-associated [RefSeq], [UniProt] - Brain; high level in Purkinje cells [UniProt], [De Smedt, FEBS Lett 1994], [Laxminarayan, J Biol Chem 1994] - gene was cloned and expressed; 91% homology to dog homol 3635: - gene was cloned and expressed [Drayer, Biochem Biophys R 3636:
MI14PP	3	York JD, Veile RA, Donis-Keller H, Majerus PW	Cloning, heterologous expression, and chromosomal localization of human inositol polyphosphate 1-phosphatase	Proc Natl Acad Sci U S A	1993	8390685	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] - inositol polyphosphate-1-phosphatase activity [RefSeq] - Ubiquitously expressed, with highest levels in pancreas and kidney. [UniProt], [York, PNAS 1993] - gene was cloned and expressed; 84% identical to bovine homolog [York, PNAS 1993]
MI1PP	3	McAllister G, Whiting P, Hammond EA, Knowles MR, Atack JR, Bailey FJ, Maigetter R, Ragan CI.	cDNA cloning of human and rat brain myo-inositol monophosphatase. Expression and characterization of the human recombinant enzyme	Biochem J	1992	1377913	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3612: - cytosolic [UniProt] - gene was cloned and expressed; protein has 97% homology to rat, bovine homologs [McAllister, Biochem J 1992] 3613: - gene has been cloned and has homology to IMPA1 [Yoshikawa, Mol Psychiatry 1997]
MI1PP	3	Irvine RF, Moor RM, Pollock WK, Smith PM, Wreggett KA	Inositol phosphates: proliferation, metabolism and function	Philos Trans R Soc Lond B Biol Sci	1988	2906139	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3612: - cytosolic [UniProt] - gene was cloned and expressed; protein has 97% homology to rat, bovine homologs [McAllister, Biochem J 1992] 3613: - gene has been cloned and has homology to IMPA1 [Yoshikawa, Mol Psychiatry 1997]

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MI1PP	3	Yoshikawa T, Turner G, Esterling LE, Sanders AR, Detera-Wadleigh SD	A novel human myo-inositol monophosphatase gene, IMP.18p, maps to a susceptibility region for bipolar disorder	Mol Psychiatry	1997	9322233	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3612: - cytosolic [UniProt] - gene was cloned and expressed; protein has 97% homology to rat, bovine homologs [McAllister, Biochem J 1992] 3613: - gene has been cloned and has homology to IMPA1 [Yoshikawa, Mol Psychiatry 1997]
MI1PS	3	Guan G, Dai P, Shechter I.	cDNA cloning and gene expression analysis of human myo-inositol 1-phosphate synthase	Arch Biochem Biophys	2003	12941308	- gene has been cloned and expressed [Guan, Arch Biochem Biophys 2003] - highly expressed in human testis, ovary, heart, placenta, and pancreas; low expression in blood leukocyte, thymus, skeletal muscle, and colon [Guan, Arch Biochem Biophys 2003]
MI34PP	3	Norris FA, Auethavekiat V, Majerus PW	The isolation and characterization of cDNA encoding human and rat brain inositol polyphosphate 4-phosphatase	J Biol Chem	1995	7608176	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3631: - inositol polyphosphate 4-phosphatase activity [RefSeq] - gene was cloned; 97% identical to rat homolog [Norris, J Biol Chem 1995] - rat protein was highly expressed in brain, heart, and skeletal muscle [Norris, J Biol Chem 1995] 8821: - inositol polyphosphate 4-phosphatase activity [RefSeq] - gene has been cloned and its sequence has 37% similarity to INPP4A and 90% similarity to rat homolog [Norris, J Biol Chem 1997]
MI34PP	3	Norris FA, Atkins RC, Majerus PW	The cDNA cloning and characterization of inositol polyphosphate 4-phosphatase type II. Evidence for conserved alternative splicing in the 4-phosphatase family	J Biol Chem	1997	9295334	- reaction described in [Irvine, Philos Trans R Soc Lond B Biol Sci 1988] 3631: - inositol polyphosphate 4-phosphatase activity [RefSeq] - gene was cloned; 97% identical to rat homolog [Norris, J Biol Chem 1995] - rat protein was highly expressed in brain, heart, and skeletal muscle [Norris, J Biol Chem 1995] 8821: - inositol polyphosphate 4-phosphatase activity [RefSeq] - gene has been cloned and its sequence has 37% similarity to INPP4A and 90% similarity to rat homolog [Norris, J Biol Chem 1997]
MICITDr	2	Beach RL, Aogaichi T, Plaut GW.	Identification of D-threo-alpha-methylisocitrate as stereochemically specific substrate for bovine heart aconitase and inhibitor of TPN-linked isocitrate dehydrogenase.	J Biol Chem	1977	856801	-evidence from mitochondrial bovine heart aconitase suggests that it converts alpha-methyl-cis-aconitate to alpha-methylisocitrate but not alpha-methylcitrate; see [Lauble 1995] for structural modeling and [Beach 1977] for biochemical studies
MICITDr	2	Lauble H, Stout CD	Steric and conformational features of the aconitase mechanism.	Proteins	1995	7675781	-evidence from mitochondrial bovine heart aconitase suggests that it converts alpha-methyl-cis-aconitate to alpha-methylisocitrate but not alpha-methylcitrate; see [Lauble 1995] for structural modeling and [Beach 1977] for biochemical studies
MM8Ag	0	Tremblay LO, Herscovics A.	Characterization of a cDNA encoding a novel human Golgi alpha 1, 2-mannosidase (IC) involved in N-glycan biosynthesis	J Biol Chem	2000	10915796	pathway taken from Trembey and Hersovics. J Biol Chem 275(41):31655-60 (2000). MAN1A1 encodes a class I mammalian Golgi 1,2-mannosidase which is a type II transmembrane protein. This protein catalyzes the removal of 3 distinct mannose residues from peptide-bound Man(9)-GlcNAc(2) oligosaccharides and belongs to family 47 of glycosyl hydrolases. [RefSeq] Man1a1p and Man1a2p ubiquitously expressed [Tremblay et al, Glycobiology 1998] Man1c1p expressed in all tissue tested except lung, muscle, pancreas [Tremblay and Herscovic, J Biol Chem 2000]
MM8Ber	0	Tremblay LO, Campbell Dyke N, Herscovics A.	Molecular cloning, chromosomal mapping and tissue-specific expression of a novel human alpha1,2-mannosidase gene involved in N-glycan maturation.	Glycobiology	1998	9592125	cloning and expression of 11253 cleaves Man9GlcNAc2 to Man8GlcNAc2 isomer B localized to ER [Tremblay and Herscovics, Glycobiology 9(10): 1073-78 (1999)] Man1b1p ubiquitously expressed [Gonzalez et al, J Biol Chem 1999]
MM8Ber	0	Gonzalez DS, Karaveg K, Vandersall-Nairn AS, Lal A, Moremen KW.	Identification, expression, and characterization of a cDNA encoding human endoplasmic reticulum mannosidase I, the enzyme that catalyzes the first mannose trimming step in mammalian Asn-linked oligosaccharide biosynthesis.	J Biol Chem	1999	10409699	cloning and expression of 11253 cleaves Man9GlcNAc2 to Man8GlcNAc2 isomer B localized to ER [Tremblay and Herscovics, Glycobiology 9(10): 1073-78 (1999)] Man1b1p ubiquitously expressed [Gonzalez et al, J Biol Chem 1999]

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MMCD	1	Kerner J, Hoppel CL.	Radiochemical malonyl-CoA decarboxylase assay: activity and subcellular distribution in heart and skeletal muscle.	Anal Biochem	2002	12123667	- methylmalonate reaction added based on GO annotation - Additional information added by RS/TV: - Malonyl-CoA decarboxylase is the main route for the disposal of malonyl-CoA - MCD is reported to be predominantly localized in the heart, as well as skeletal muscle. - MCD has been found in the cytoplasm, mitochondria, and peroxisome. All this according to Kerner J, Hoppel CL. Anal Biochem. 2002 Jul 15;306(2):283-9 Radiochemical malonyl-CoA decarboxylase assay: activity and subcellular distribution in heart and skeletal muscle. summary of localization data (see refs in Wightman 2003]): - enzymatic activity: mito, cyto - Western blot: perox, cyto - seq analysis: mito, perox
MMCD	1	Wightman PJ, Santer R, Ribes A, Dougherty F, McGill N, Thorburn DR, FitzPatrick DR	MLYCD mutation analysis: evidence for protein mistargeting as a cause of MLYCD deficiency	Hum Mutat	2003	12955715	- methylmalonate reaction added based on GO annotation - Additional information added by RS/TV: - Malonyl-CoA decarboxylase is the main route for the disposal of malonyl-CoA - MCD is reported to be predominantly localized in the heart, as well as skeletal muscle. - MCD has been found in the cytoplasm, mitochondria, and peroxisome. All this according to Kerner J, Hoppel CL. Anal Biochem. 2002 Jul 15;306(2):283-9 Radiochemical malonyl-CoA decarboxylase assay: activity and subcellular distribution in heart and skeletal muscle. summary of localization data (see refs in Wightman 2003]): - enzymatic activity: mito, cyto - Western blot: perox, cyto - seq analysis: mito, perox
MMEm	3	Bobik TA, Rasche ME.	Identification of the human methylmalonyl-CoA racemase gene based on the analysis of prokaryotic gene arrangements. Implications for decoding the human genome.	J Biol Chem	2001	11481338	- mitochondrial (probable) [UniProt] - identification of gene by homology search and biochemical characterization of protein [Bobik, J Biol Chem 2001] - reaction described in Devlin p. 637, Orten p. 262
MMMm	3	Ledley FD, Rosenblatt DS.	Mutations in mut methylmalonic acidemia: clinical and enzymatic correlations.		1997	8990001	- reaction described in Devlin p. 637, Orten p. 262 MM
MMSAD1m	3	Kedishvili NY, Popov KM, Rougraff PM, Zhao Y, Crabb DW, Harris RA.	CoA-dependent methylmalonate-semialdehyde dehydrogenase, a unique member of the aldehyde dehydrogenase superfamily. cDNA cloning, evolutionary relationships, and tissue distribution.		1992	1527093	- reaction described in Devlin p. 812 - mitochondrial [RefSeq] - catalyzes the irreversible oxidative decarboxylation of methylmalonate semialdehydes to propionyl-CoA [RefSeq]
MMSAD1m	3	Chambliss,K.L. , Gray,R.G. , Rylance,G. , Pollitt,R.J. , Gibson,K.M.	Molecular characterization of methylmalonate semialdehyde dehydrogenase deficiency.		2000	10947204	- reaction described in Devlin p. 812 - mitochondrial [RefSeq] - catalyzes the irreversible oxidative decarboxylation of methylmalonate semialdehydes to propionyl-CoA [RefSeq]
MMSAD3m	3	Scholem RD, Brown GK	Metabolism of malonic semialdehyde in man	Biochem J	1983	6418146	- another function of EC 1.2.1.18, which was assumed to be present based on physiological data - malonic semialdehyde is directly converted into acetyl-CoA in man [Scholem, Biochem J 1983] - mitochondrial [RefSeq] - catalyzes the irreversible oxidative decarboxylation of malonate semialdehydes to acetyl-CoA [RefSeq]
MTAP	3	Della Ragione F, Carteni-Farina M, Gragnaniello V, Schettino MI, Zappia V	Purification and characterization of 5'-deoxy-5'-methylthioadenosine phosphorylase from human placenta	J Biol Chem	1986	3091600	This reaction is well characterized and degrades 5mta, a byproduct of sprm and spmd synthesis. The enzyme is deactivated in many cancers. It is possible that this reaction is reversible, depending on conditions of course. However, it is unlikely due to rapid removal of ade (see second citation).
MTAP	3	Evans GB, Furneaux RH, Lenz DH, Painter GF, Schramm VL, Singh V, Tyler PC	Second generation transition state analogue inhibitors of human 5'-methylthioadenosine phosphorylase	J Med Chem	2005	16000004	This reaction is well characterized and degrades 5mta, a byproduct of sprm and spmd synthesis. The enzyme is deactivated in many cancers. It is possible that this reaction is reversible, depending on conditions of course. However, it is unlikely due to rapid removal of ade (see second citation).

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MTHFDm	3	Yang XM, MacKenzie RE	NAD-dependent methylenetetrahydrofolate dehydrogenase-methenyltetrahydrofolate cyclohydrolase is the mammalian homolog of the mitochondrial enzyme encoded by the yeast MIS1 gene	Biochemistry	1993	8218174	25902: - encodes the mitochondrial isozyme of C1-tetrahydrofolate (THF) synthase, a trifunctional enzyme containing formyl-THF synthetase, methenyl-THF cyclohydrolase, and methylene-THF dehydrogenase activities [Prasannan, J Biol Chem 2003] 10797: - NAD is preferred cofactor, but can also use NADP at a lower Vmax, higher Km [Yang, Biochemistry 1993]
MTRI	2	Myers RW, Abeles RH	Conversion of 5-S-ethyl-5-thio-D-ribose to ethionine in Klebsiella pneumoniae. Basis for the selective toxicity of 5-S-ethyl-5-thio-D-ribose	J Biol Chem	1989	2543672	This reaction follows the scheme in the reference, checked for bacteria and given physiological data since the end product, met-L, is known for humans. A candidate gene has been determined based on homology with yeast: MGC3207 (84245). This association has not been included in the resonstruction because it is far from certain.
N3Tg	0	Ju T, Brewer K, D'Souza A, Cummings RD, Canfield WM.	Cloning and expression of human core 1 beta1,3-galactosyltransferase.	J Biol Chem	2002	11677243	C1galt1p expressed in kidney, heart, placenta, liver, brain, sk muscle [Ju et al, J Biol Chem 2002]
NACASPAH	3	Kaul R, Gao GP, Balamurugan K, Matalon R	Cloning of the human aspartoacylase cDNA and a common missense mutation in Canavan disease	Nat Genet	1993	8252036	This reaction, perhaps more correctly characterized under aspartate metabolism, is related to Canavan disease. KEGG also suggests an alternate form of this reaction involving N-Formyl-L-aspartate, but there is no particular evidence for that reaction and it doesn't not close a gap on its own.
NACHEX27ly	3	Franke I, Resch A, Dassler T, Maier T, Bock A.	YfiK from Escherichia coli promotes export of O-acetylserine and cysteine.	J Bacteriol	2003	12562784	- beta-N-acetylhexosaminidase needed for hyaluronan degradation [Varki, pg 275] 3073, 3074: -The subunits encoded by the genes HEXA and HEXB are synthesized as precursor proteins; processing and subunit assembly in the endoplasmatic reticulum yields three isoforms; beta-hexosaminidase A (alpha, beta), beta-hexosaminidase B (beta, beta) and beta-hexosaminidase S (alpha, alpha). The proteins are targeted to the lysosomes, where final processing produces the mature enzymes. [Maier et al, J Mol Biol. 328(3):669-81 (2003)] - lysosomal [Liu, Glycobiology 1999] - occurs in degradation of Asn-linked glycoproteins [Liu, Glycobiology 1999] - the active site of the beta -subunit hydrolyzes uncharged substrates, whereas the alpha -subunit, in addition, cleaves negatively charged substrate [Hepbildikler 2002] - Hexb -/- mice (expressing only HexS) showed no increased accumulation of glycosaminoglycans, indicating that Hex S involved in their catabolism [Hepbildikler 2002] - acts on N-acetylglucosides and N-acetylgalactosides [BRENDA]
NADK	3	Williams MB, Jones HP.	Calmodulin-dependent NAD kinase of human neutrophils.	Arch Biochem Biophys	1985	2982330	IT expressed in most tissues but not in skeletal muscle cells
NADK	3	Lerner F, Niere M, Ludwig A, Ziegler M.	Structural and functional characterization of human NAD kinase.	Biochem Biophys Res Commun	2001	11594753	IT expressed in most tissues but not in skeletal muscle cells
NADPN	2	Boyer CS, Moore GA, Moldeus P.	Submitochondrial localization of the NAD+ glycohydrolase. Implications for the role of pyridine nucleotide hydrolysis in mitochondrial calcium fluxes.	J Biol Chem	1993	8382685	IT Boyer et al reported NAD glycohydrolase location on outer membrane of rat mitochondria. In addition, Bender (book) mentioned degradation of NAD(P) in cell ADPribose moiety is normally transfered to protein which profoundly affect the target protein effector function. protein modification is reversed by hydrolases leading to liberation of ADPrib. the free ADPrib pool is tightly regulated in cell since it is a highly reactive molecule which causes non-enzymatic mom-ADP-ribosylation of proteins (from Rongvaux et alm BioEsssays 25, 683-690,2003; Yang et al, JBC, 275(12),8844-8853,2000;and Di Lisa, Ziegler, FEBS lett, 492, 2001,4-8)
NADPN	2	Bender DA	Nutritional Biochemistry of the Vitamins		2003		IT Boyer et al reported NAD glycohydrolase location on outer membrane of rat mitochondria. In addition, Bender (book) mentioned degradation of NAD(P) in cell ADPribose moiety is normally transfered to protein which profoundly affect the target protein effector function. protein modification is reversed by hydrolases leading to liberation of ADPrib. the free ADPrib pool is tightly regulated in cell since it is a highly reactive molecule which causes non-enzymatic mom-ADP-ribosylation of proteins (from Rongvaux et alm BioEsssays 25, 683-690,2003; Yang et al, JBC, 275(12),8844-8853,2000;and Di Lisa, Ziegler, FEBS lett, 492, 2001,4-8)
NADPNe	3	Kim UH, Han MK, Park BH, Kim HR, An NH.	Function of NAD glycohydrolase in ADP-ribose uptake from NAD by human erythrocytes.	Biochim Biophys Acta	1993	8394137	IT

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NADPNe	3	Kontani K, Nishina H, Ohoka Y, Takahashi K, Katada T.	NAD glycohydrolase specifically induced by retinoic acid in human leukemic HL-60 cells. Identification of the NAD glycohydrolase as leukocyte cell surface antigen CD38.	J Biol Chem	1993		IT
NADS2	3	Hara N, Yamada K, Terashima M, Osago H, Shimoyama M, Tsuchiya M.	Molecular identification of human glutamine- and ammonia-dependent NAD synthetases. Carbon-nitrogen hydrolase domain confers glutamine dependency.	J Biol Chem	2003	12547821	IT
NAGAly	3	Wang AM, Bishop DF, Desnick RJ.	Human alpha-N-acetylgalactosaminidase-molecular cloning, nucleotide sequence, and expression of a full-length cDNA. Homology with human alpha-galactosidase A suggests evolution from a common ancestral gene.	J Biol Chem	1990	2174888	lysosomal - uniprot, wang ref NAGA encodes the lysosomal enzyme alpha-N-acetylgalactosaminidase, which cleaves alpha-N-acetylgalactosaminyl moieties from glycoconjugates. Mutations in NAGA have been identified as the cause of Schindler disease types I and II (type II also known as Kanzaki disease). NJ 4668: - protein isolated from human placenta [Tsuji 1989] - purified from human lung, expressed in COS-1 cells [Wang 1990] - has "striking" homology to human alpha-galactosidase A and yeast alpha-galactosidase [Tsuji 1989]; 46.9-64.7% amino acid identity w/ alpha-Gal A exons 1 through 6, but exon 7 had only 15.8% homology with numerous gaps [Wang 1990]
NAGAly	3	Tsuji S, Yamauchi T, Hiraiwa M, Isobe T, Okuyama T, Sakimura K, Takahashi Y, Nishizawa M, Uda Y, Miyatake T.	Molecular cloning of a full-length cDNA for human alpha-N-acetylgalactosaminidase (alpha-galactosidase B)	Biochem Biophys Res Commun	1989	2551294	lysosomal - uniprot, wang ref NAGA encodes the lysosomal enzyme alpha-N-acetylgalactosaminidase, which cleaves alpha-N-acetylgalactosaminyl moieties from glycoconjugates. Mutations in NAGA have been identified as the cause of Schindler disease types I and II (type II also known as Kanzaki disease). NJ 4668: - protein isolated from human placenta [Tsuji 1989] - purified from human lung, expressed in COS-1 cells [Wang 1990] - has "striking" homology to human alpha-galactosidase A and yeast alpha-galactosidase [Tsuji 1989]; 46.9-64.7% amino acid identity w/ alpha-Gal A exons 1 through 6, but exon 7 had only 15.8% homology with numerous gaps [Wang 1990]
NaKt	3	Wang J, Schwinger RH, Frank K, Muller-Ehmsen J, Martin-Vasallo P, Pressley TA, Xiang A, Erdmann E, McDonough AA.	Regional expression of sodium pump subunits isoforms and Na+-Ca++ exchanger in the human heart.	J Clin Invest	1996	8833915	- Added by KSPV - The Na-K ATPase functions to maintain sodium and potassium gradients across membranes that subserve cellular activities such as volume regulation, action potentials, and secondary active transport. - This enzyme is composed of two subunits, a large catalytic subunit (alpha) and a smaller glycoprotein subunit (beta) [Entrez] - Expression for each subunit is as follows: 1) alpha1: ubiquitously 2) alpha2: brain, heart skeletal muscle 3) alpha3: brain and heart 4) alpha4: testis and skeletal muscle 5) beta1: ubiquitously 6) beta2: neural, heart 7) beta3: human placenta 8) beta4: skeletal muscle - Based on this expression data 11 different combinations of the two subunits were created: 1) Following combinations assumed to be expressed ubiquitously: alpha1/beta1, alpha1/beta2, alpha1/beta3, alpha1/beta4, alpha2/beta1, alpha3/beta1, alpha4/beta1 2) Following combination expresed in heart: alpha1/beta1, alpha2/beta1, alpha3/beta1 (according to Wang J, et al. J Clin Invest. 1996 Oct 1;98(7):1650-8.) 3) Following combinatons expressed in brain/neural tissues: alp 4) Following combination expressed in skeletal muscle: alpha4/ - Note: Protein name Atp1a3b4 indicates combination of alpha3

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NaKt	3	Stengelin MK, Hoffman JF.	Na,K-ATPase subunit isoforms in human reticulocytes: evidence from reverse transcription-PCR for the presence of alpha1, alpha3, beta2, beta3, and gamma.	Proc Natl Acad Sci U S A	1997	9159180	- Added by KS/TV - The Na-K ATPase functions to maintain sodium and potassium gradients across membranes that subserve cellular activities such as volume regulation, action potentials, and secondary active transport. - This enzyme is composed of two subunits, a large catalytic subunit (alpha) and a smaller glycoprotein subunit (beta) [Entrez] - Expression for each subunit is as follows: 1) alpha1: ubiquitously 2) alpha2: brain, heart skeletal muscle 3) alpha3: brain and heart 4) alpha4: testis and skeletal muscle 5) beta1: ubiquitously 6) beta2: neural, heart 7) beta3: human placenta 8) beta4: skeletal muscle - Based on this expression data 11 different combinations of the two subunits were created: 1) Following combinations assumed to be expressed ubiquitously: alpha1/beta1, alpha1/beta2, alpha1/beta3, alpha1/beta4, alpha2/beta1, alpha3/beta1, alpha4/beta1 2) Following combination expresed in heart: alpha1/beta1, alpha2/beta1, alpha3/beta1 (according to Wang J, et al. J Clin Invest. 1996 Oct 1;98(7):1650-8.) 3) Following combinatons expressed in brain/neural tissues: alp 4) Following combination expressed in skeletal muscle: alpha4/ - Note: Protein name Atp1a3b4 indicates combination of alpha3
NaKt	3	Pestov NB, Adams G, Shakhparonov MI, Modyanov NN.	Identification of a novel gene of the X,K-ATPase beta-subunit family that is predominantly expressed in skeletal and heart muscles	FEBS Lett	1999	10456317	- Added by KS/TV - The Na-K ATPase functions to maintain sodium and potassium gradients across membranes that subserve cellular activities such as volume regulation, action potentials, and secondary active transport. - This enzyme is composed of two subunits, a large catalytic subunit (alpha) and a smaller glycoprotein subunit (beta) [Entrez] - Expression for each subunit is as follows: 1) alpha1: ubiquitously 2) alpha2: brain, heart skeletal muscle 3) alpha3: brain and heart 4) alpha4: testis and skeletal muscle 5) beta1: ubiquitously 6) beta2: neural, heart 7) beta3: human placenta 8) beta4: skeletal muscle - Based on this expression data 11 different combinations of the two subunits were created: 1) Following combinations assumed to be expressed ubiquitously: alpha1/beta1, alpha1/beta2, alpha1/beta3, alpha1/beta4, alpha2/beta1, alpha3/beta1, alpha4/beta1 2) Following combination expresed in heart: alpha1/beta1, alpha2/beta1, alpha3/beta1 (according to Wang J, et al. J Clin Invest. 1996 Oct 1;98(7):1650-8.) 3) Following combinatons expressed in brain/neural tissues: alp 4) Following combination expressed in skeletal muscle: alpha4/ - Note: Protein name Atp1a3b4 indicates combination of alpha3
NAt	3	Kwon HM, Yamauchi A, Uchida S, Preston AS, Garcia-Perez A, Burg MB, Handler JS	Cloning of the cDNa for a Na+/myo-inositol cotransporter, a hypertonicity stress protein	J Biol Chem	1992	1372904	6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ in sugar cotransport [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6526: - cloned [Kwon 1992], [Berry 1995] - kidney, brain, placenta, pancreas, heart, skeletal muscle, lung [Berry 1995] - Na+/myo-inositol cotransport; also transports other sugars (incl glc) with low affinity [Hager 1995], [Kwon 1992] - plasma membrane; see [Wright 2004] for refs - transports Na+ in the absence of sugar [Wright, Physiology 2004] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandari, 1997] - primarily in thyroid gland [De La Vieja 2000], also expressed in breast, bladder, colon, endometrium, kidney, prostate, and pa - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs - transports Na+ in the absence of sugar [Wright, Physiology 20

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
NAt	3	Hager K, Hazama A, Kwon HM, Loo DD, Handler JS, Wright EM	Kinetics and specificity of the renal Na+/myo-inositol cotransporter expressed in Xenopus oocytes	J Membr Biol	1995	7537337	6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ in sugar cotransport [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6526: - cloned [Kwon 1992], [Berry 1995] - kidney, brain, placenta, pancreas, heart, skeletal muscle, lung [Berry 1995] - Na+/myo-inositol cotransport; also transports other sugars (incl glc) with low affinity [Hager 1995], [Kwon 1992] - plasma membrane; see [Wright 2004] for refs - transports Na+ in the absence of sugar [Wright, Physiology 2004] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandari, 1997] - primarily in thyroid gland [De La Vieja 2000], also expressed in breast, bladder, colon, endometrium, kidney, prostate, and pa - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs - transports Na+ in the absence of sugar [Wright, Physiology 20
NAt	3	Berry GT, Mallee JJ, Kwon HM, Rim JS, Mulla WR, Muenke M, Spinner NB	The human osmoregulatory Na+/myo-inositol cotransporter gene (SLC5A3): molecular cloning and localization to chromosome 21	Genomics	1995	7789985	6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ in sugar cotransport [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6526: - cloned [Kwon 1992], [Berry 1995] - kidney, brain, placenta, pancreas, heart, skeletal muscle, lung [Berry 1995] - Na+/myo-inositol cotransport; also transports other sugars (incl glc) with low affinity [Hager 1995], [Kwon 1992] - plasma membrane; see [Wright 2004] for refs - transports Na+ in the absence of sugar [Wright, Physiology 2004] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandari, 1997] - primarily in thyroid gland [De La Vieja 2000], also expressed in breast, bladder, colon, endometrium, kidney, prostate, and pa - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs - transports Na+ in the absence of sugar [Wright, Physiology 20
NAt	3	Wright EM, Loo DD, Hirayama BA, Turk E	Surprising versatility of Na+-glucose cotransporters: SLC5	Physiology (Bethesda)	2004	15546855	6523: -cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ in sugar cotransport [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6526: - cloned [Kwon 1992], [Berry 1995] - kidney, brain, placenta, pancreas, heart, skeletal muscle, lung [Berry 1995] - Na+/myo-inositol cotransport; also transports other sugars (incl glc) with low affinity [Hager 1995], [Kwon 1992] - plasma membrane; see [Wright 2004] for refs - transports Na+ in the absence of sugar [Wright, Physiology 2004] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandari, 1997] - primarily in thyroid gland [De La Vieja 2000], also expressed in breast, bladder, colon, endometrium, kidney, prostate, and pa - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs - transports Na+ in the absence of sugar [Wright, Physiology 20

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NAt3_1	3	Sardet C, Franchi A, Pouyssegur J	Molecular cloning, primary structure, and expression of the human growth factor-activatable Na+/H+ antiporter.	Cell	1989	2536298	6548: - cloned [Sardet 1989] - restored H+-activated Na+ influx in kockout [Sardet 1989] - ubiquitous [Orlowski 2004] - rabbit homolog localized to basolateral surface of plasma membrane (see ref in [Malakooti 1999]) - evidence for 1:1 stoichiometry reviewed in [Aronson 1985] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6549: - cloned [Ghishan 1995]*, [Malakooti 1999] * Ghishan cloned truncated sequence, Malakooti cloned full sequence - high in sk muscle, colon, and kidney and lower in testis, prostate, ovary, sm intestine [Malakooti 1999] - rabbit homolog localized in apical membrane (see refs in [Malakooti 1999]) - catalyzes Na+/H+ exchange activity in LAP1 cells lacking endogenous NHE activity [Malakooti 1999] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6550: - cloned [Brant 1995] - 89 and 88% amino acid identity with rat and rabbit NHE3 [Br - established Na+/H+ exchange in deficient PS120 cells [Brant kidney >> small intestine >> testes > ovary > colon = prostate > - rabbit homolog localized in apical membrane (see ref in [Bran - Na+/NH4+ exchange suggested in [Aronson 1985] for plasma - also exchanges Li+/H+ (not included in model) [Aronson 198
NAt3_1	3	Aronson PS	Kinetic properties of the plasma membrane Na+-H+ exchanger	Annu Rev Physiol	1985	2581505	6548: - cloned [Sardet 1989] - restored H+-activated Na+ influx in kockout [Sardet 1989] - ubiquitous [Orlowski 2004] - rabbit homolog localized to basolateral surface of plasma membrane (see ref in [Malakooti 1999]) - evidence for 1:1 stoichiometry reviewed in [Aronson 1985] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6549: - cloned [Ghishan 1995]*, [Malakooti 1999] * Ghishan cloned truncated sequence, Malakooti cloned full sequence - high in sk muscle, colon, and kidney and lower in testis, prostate, ovary, sm intestine [Malakooti 1999] - rabbit homolog localized in apical membrane (see refs in [Malakooti 1999]) - catalyzes Na+/H+ exchange activity in LAP1 cells lacking endogenous NHE activity [Malakooti 1999] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6550: - cloned [Brant 1995] - 89 and 88% amino acid identity with rat and rabbit NHE3 [Br - established Na+/H+ exchange in deficient PS120 cells [Brant kidney >> small intestine >> testes > ovary > colon = prostate > - rabbit homolog localized in apical membrane (see ref in [Bran - Na+/NH4+ exchange suggested in [Aronson 1985] for plasma - also exchanges Li+/H+ (not included in model) [Aronson 198
NAt3_1	3	Brant SR, Yun CH, Donowitz M, Tse CM	Cloning, tissue distribution, and functional analysis of the human Na+/N+ exchanger isoform, NHE3	Am J Physiol	1995	7631746	6548: - cloned [Sardet 1989] - restored H+-activated Na+ influx in kockout [Sardet 1989] - ubiquitous [Orlowski 2004] - rabbit homolog localized to basolateral surface of plasma membrane (see ref in [Malakooti 1999]) - evidence for 1:1 stoichiometry reviewed in [Aronson 1985] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6549: - cloned [Ghishan 1995]*, [Malakooti 1999] * Ghishan cloned truncated sequence, Malakooti cloned full sequence - high in sk muscle, colon, and kidney and lower in testis, prostate, ovary, sm intestine [Malakooti 1999] - rabbit homolog localized in apical membrane (see refs in [Malakooti 1999]) - catalyzes Na+/H+ exchange activity in LAP1 cells lacking endogenous NHE activity [Malakooti 1999] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6550: - cloned [Brant 1995] - 89 and 88% amino acid identity with rat and rabbit NHE3 [Br - established Na+/H+ exchange in deficient PS120 cells [Brant kidney >> small intestine >> testes > ovary > colon = prostate > - rabbit homolog localized in apical membrane (see ref in [Bran - Na+/NH4+ exchange suggested in [Aronson 1985] for plasma - also exchanges Li+/H+ (not included in model) [Aronson 198

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NAt3_1	3	Klanke CA, Su YR, Callen DF, Wang Z, Meneton P, Baird N, Kandasamy RA, Orlowski J, Otterud BE, Leppert M, et al	Molecular cloning and physical and genetic mapping of a novel human Na+/H+ exchanger (NHE5/SLC9A5) to chromosome 16q22.1	Genomics	1995	7759094	6548: - cloned [Sardet 1989] - restored H+-activated Na+ influx in kockout [Sardet 1989] - ubiquitous [Orlowski 2004] - rabbit homolog localized to basolateral surface of plasma membrane (see ref in [Malakooti 1999]) - evidence for 1:1 stoichiometry reviewed in [Aronson 1985] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6549: - cloned [Ghishan 1995]*, [Malakooti 1999] * Ghishan cloned truncated sequence, Malakooti cloned full sequence - high in sk muscle, colon, and kidney and lower in testis, prostate, ovary, sm intestine [Malakooti 1999] - rabbit homolog localized in apical membrane (see refs in [Malakooti 1999]) - catalyzes Na+/H+ exchange activity in LAP1 cells lacking endogenous NHE activity [Malakooti 1999] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6550: - cloned [Brant 1995] - 89 and 88% amino acid identity with rat and rabbit NHE3 [Br - established Na+/H+ exchange in deficient PS120 cells [Brant kidney >> small intestine >> testes > ovary > colon = prostate > - rabbit homolog localized in apical membrane (see ref in [Bran - Na+/NH4+ exchange suggested in [Aronson 1985] for plasma - also exchanges Li+/H+ (not included in model) [Aronson 198
NAt3_1	3	Szpirer C, Szpirer J, Riviere M, Levan G, Orlowski J	Chromosomal assignment of four genes encoding Na/H exchanger isoforms in human and rat.	Mamm Genome	1994	8199403	6548: - cloned [Sardet 1989] - restored H+-activated Na+ influx in kockout [Sardet 1989] - ubiquitous [Orlowski 2004] - rabbit homolog localized to basolateral surface of plasma membrane (see ref in [Malakooti 1999]) - evidence for 1:1 stoichiometry reviewed in [Aronson 1985] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6549: - cloned [Ghishan 1995]*, [Malakooti 1999] * Ghishan cloned truncated sequence, Malakooti cloned full sequence - high in sk muscle, colon, and kidney and lower in testis, prostate, ovary, sm intestine [Malakooti 1999] - rabbit homolog localized in apical membrane (see refs in [Malakooti 1999]) - catalyzes Na+/H+ exchange activity in LAP1 cells lacking endogenous NHE activity [Malakooti 1999] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6550: - cloned [Brant 1995] - 89 and 88% amino acid identity with rat and rabbit NHE3 [Br - established Na+/H+ exchange in deficient PS120 cells [Brant kidney >> small intestine >> testes > ovary > colon = prostate > - rabbit homolog localized in apical membrane (see ref in [Bran - Na+/NH4+ exchange suggested in [Aronson 1985] for plasma - also exchanges Li+/H+ (not included in model) [Aronson 198
NAt3_1	3	Ghishan FK, Knobel SM, Summar M	Molecular cloning, sequencing, chromosomal localization, and tissue distribution of the human Na+/H+ exchanger (SLC9A2)	Genomics	1995	8595899	6548: - cloned [Sardet 1989] - restored H+-activated Na+ influx in kockout [Sardet 1989] - ubiquitous [Orlowski 2004] - rabbit homolog localized to basolateral surface of plasma membrane (see ref in [Malakooti 1999]) - evidence for 1:1 stoichiometry reviewed in [Aronson 1985] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6549: - cloned [Ghishan 1995]*, [Malakooti 1999] * Ghishan cloned truncated sequence, Malakooti cloned full sequence - high in sk muscle, colon, and kidney and lower in testis, prostate, ovary, sm intestine [Malakooti 1999] - rabbit homolog localized in apical membrane (see refs in [Malakooti 1999]) - catalyzes Na+/H+ exchange activity in LAP1 cells lacking endogenous NHE activity [Malakooti 1999] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6550: - cloned [Brant 1995] - 89 and 88% amino acid identity with rat and rabbit NHE3 [Br - established Na+/H+ exchange in deficient PS120 cells [Brant kidney >> small intestine >> testes > ovary > colon = prostate > - rabbit homolog localized in apical membrane (see ref in [Bran - Na+/NH4+ exchange suggested in [Aronson 1985] for plasma - also exchanges Li+/H+ (not included in model) [Aronson 198

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NAt3_1	3	Baird NR, Orlowski J, Szabo EZ, Zaun HC, Schultheis PJ, Menon AG, Shull GE	Molecular cloning, genomic organization, and functional expression of Na+/H+ exchanger isoform 5 (NHE5) from human brain	J Biol Chem	1999	9933641	6548: - cloned [Sardet 1989] - restored H+-activated Na+ influx in kockout [Sardet 1989] - ubiquitous [Orlowski 2004] - rabbit homolog localized to basolateral surface of plasma membrane (see ref in [Malakooti 1999]) - evidence for 1:1 stoichiometry reviewed in [Aronson 1985] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6549: - cloned [Ghishan 1995]*, [Malakooti 1999] * Ghishan cloned truncated sequence, Malakooti cloned full sequence - high in sk muscle, colon, and kidney and lower in testis, prostate, ovary, sm intestine [Malakooti 1999] - rabbit homolog localized in apical membrane (see refs in [Malakooti 1999]) - catalyzes Na+/H+ exchange activity in LAP1 cells lacking endogenous NHE activity [Malakooti 1999] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6550: - cloned [Brant 1995] - 89 and 88% amino acid identity with rat and rabbit NHE3 [Br - established Na+/H+ exchange in deficient PS120 cells [Brant kidney >> small intestine >> testes > ovary > colon = prostate > - rabbit homolog localized in apical membrane (see ref in [Bran - Na+/NH4+ exchange suggested in [Aronson 1985] for plasma - also exchanges Li+/H+ (not included in model) [Aronson 198
NAt3_1	3	Malakooti J, Dahdal RY, Schmidt L, Layden TJ, Dudeja PK, Ramaswamy K	Molecular cloning, tissue distribution, and functional expression of the human Na(+)/H(+) exchanger NHE2	Am J Physiol	1999	10444453	6548: - cloned [Sardet 1989] - restored H+-activated Na+ influx in kockout [Sardet 1989] - ubiquitous [Orlowski 2004] - rabbit homolog localized to basolateral surface of plasma membrane (see ref in [Malakooti 1999]) - evidence for 1:1 stoichiometry reviewed in [Aronson 1985] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6549: - cloned [Ghishan 1995]*, [Malakooti 1999] * Ghishan cloned truncated sequence, Malakooti cloned full sequence - high in sk muscle, colon, and kidney and lower in testis, prostate, ovary, sm intestine [Malakooti 1999] - rabbit homolog localized in apical membrane (see refs in [Malakooti 1999]) - catalyzes Na+/H+ exchange activity in LAP1 cells lacking endogenous NHE activity [Malakooti 1999] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6550: - cloned [Brant 1995] - 89 and 88% amino acid identity with rat and rabbit NHE3 [Br - established Na+/H+ exchange in deficient PS120 cells [Brant kidney >> small intestine >> testes > ovary > colon = prostate > - rabbit homolog localized in apical membrane (see ref in [Bran - Na+/NH4+ exchange suggested in [Aronson 1985] for plasma - also exchanges Li+/H+ (not included in model) [Aronson 198
NAt3_1	3	Orlowski J, Grinstein S	Diversity of the mammalian sodium/proton exchange SLC9 gene family	Pflugers Arch	2004	12845533	6548: - cloned [Sardet 1989] - restored H+-activated Na+ influx in kockout [Sardet 1989] - ubiquitous [Orlowski 2004] - rabbit homolog localized to basolateral surface of plasma membrane (see ref in [Malakooti 1999]) - evidence for 1:1 stoichiometry reviewed in [Aronson 1985] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6549: - cloned [Ghishan 1995]*, [Malakooti 1999] * Ghishan cloned truncated sequence, Malakooti cloned full sequence - high in sk muscle, colon, and kidney and lower in testis, prostate, ovary, sm intestine [Malakooti 1999] - rabbit homolog localized in apical membrane (see refs in [Malakooti 1999]) - catalyzes Na+/H+ exchange activity in LAP1 cells lacking endogenous NHE activity [Malakooti 1999] - Na+/NH4+ exchange suggested in [Aronson 1985] for plasmalemmal NHE proteins [Orlowski 2004] - also exchanges Li+/H+ (not included in model) [Aronson 1985] 6550: - cloned [Brant 1995] - 89 and 88% amino acid identity with rat and rabbit NHE3 [Br - established Na+/H+ exchange in deficient PS120 cells [Brant kidney >> small intestine >> testes > ovary > colon = prostate > - rabbit homolog localized in apical membrane (see ref in [Bran - Na+/NH4+ exchange suggested in [Aronson 1985] for plasma - also exchanges Li+/H+ (not included in model) [Aronson 198

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NAt3_1g	3	Numata M, Orlowski J	Molecular cloning and characterization of a novel (Na+,K+)/H+ exchanger localized to the trans-Golgi network	J Biol Chem	2001	11279194	84679: - cloned [Numata 2001] - ubiquitous [Numata 2001] - localizes predominantly to trans-Golgi network [Numata 2001], [Nakamura 2005] - catalyzes Na+/H+ and K+/H+ exchange [Numata 2001] 23315: - cloned [Nakamura 2005] - localizes to mid-to trans-Golgi [Nakamura 2005] - ubiquitous; most highly expressed in sk muscle, kidney [Nakamura 2005] - catalyzes Na+/H+ and K+/H+ exchange [Nakamura 2005]
NAt3_1g	3	Nakamura N, Tanaka S, Teko Y, Mitsui K, Kanazawa H.	Four Na+/H+ exchanger isoforms are distributed to Golgi and post-Golgi compartments and are involved in organelle pH regulation.	J Biol Chem	2005	15522866	84679: - cloned [Numata 2001] - ubiquitous [Numata 2001] - localizes predominantly to trans-Golgi network [Numata 2001], [Nakamura 2005] - catalyzes Na+/H+ and K+/H+ exchange [Numata 2001] 23315: - cloned [Nakamura 2005] - localizes to mid-to trans-Golgi [Nakamura 2005] - ubiquitous; most highly expressed in sk muscle, kidney [Nakamura 2005] - catalyzes Na+/H+ and K+/H+ exchange [Nakamura 2005]
NCAMUP	2	Sofue M, Yoshimura Y, Nishida M, Kawada J.	Possible multifunction of glucose transporter. Transport of nicotinamide by reconstituted liposomes.	Biochem J	1992	1463467	IT it is taken up from blood in one way or another - it does not seem to be GLUT1 as reported formerly Ball (book) mentioned that it is thought that ncam anf nac can diffuse through membrane
NCAMUP	2	Reyes AM, Bustamante F, Rivas CI, Ortega M, Donnet C, Rossi JP, Fischbarg J, Vera JC.	Nicotinamide is not a substrate of the facilitative hexose transporter GLUT1.	Biochemistry	2002	12069599	IT it is taken up from blood in one way or another - it does not seem to be GLUT1 as reported formerly Ball (book) mentioned that it is thought that ncam anf nac can diffuse through membrane
NCCt	3	Simon DB, Nelson-Williams C, Bia MJ, Ellison D, Karet FE, Molina AM, Vaara I, Iwata F, Cushner HM, Koolen M, Gainza FJ, Gitleman HJ, Lifton RP	Gitelman's variant of Bartter's syndrome, inherited hypokalaemic alkalosis, is caused by mutations in the thiazide-sensitive Na-Cl cotransporter	Nat Genet	1996	8528245	6559: - major salt reabsorption pathway in apical membrane of distal convoluted tubule [Hebert 2004] - kidney [Mastroianni 1996], [Chang 1996], lower in sm intestine, placenta, prostate, colon, spleen [Chang 1996] - cloned [Simon 1996], [Mastroianni 1996], [Chang 1996] - predominantly expressed in kidney [Hebert 2004] - 1:1 stoiciometry of Na+-Cl- transport [Hebert 2001]
NCCt	3	Chang H, Tashiro K, Hirai M, Ikeda K, Kurokawa K, Fujita T	Identification of a cDNA encoding a thiazide-sensitive sodium-chloride cotransporter from the human and its mRNA expression in various tissues	Biochem Biophys Res Commun	1996	8670281	6559: - major salt reabsorption pathway in apical membrane of distal convoluted tubule [Hebert 2004] - kidney [Mastroianni 1996], [Chang 1996], lower in sm intestine, placenta, prostate, colon, spleen [Chang 1996] - cloned [Simon 1996], [Mastroianni 1996], [Chang 1996] - predominantly expressed in kidney [Hebert 2004] - 1:1 stoiciometry of Na+-Cl- transport [Hebert 2001]
NCCt	3	Mastroianni N, De Fusco M, Zollo M, Arrigo G, Zuffardi O, Bettinelli A, Ballabio A, Casari G	Molecular cloning, expression pattern, and chromosomal localization of the human Na-Cl thiazide-sensitive cotransporter (SLC12A3).	Genomics	1996	8812482	6559: - major salt reabsorption pathway in apical membrane of distal convoluted tubule [Hebert 2004] - kidney [Mastroianni 1996], [Chang 1996], lower in sm intestine, placenta, prostate, colon, spleen [Chang 1996] - cloned [Simon 1996], [Mastroianni 1996], [Chang 1996] - predominantly expressed in kidney [Hebert 2004] - 1:1 stoiciometry of Na+-Cl- transport [Hebert 2001]

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NCKt	3	Tucker JE, Winkfein RJ, Cooper CB, Schnetkamp PP	cDNA cloning of the human retinal rod Na-Ca + K exchanger: comparison with a revised bovine sequence	Invest Ophthalmol Vis Sci	1998	9478004	9187: - cloned [Tucker 1998] - 64.3% identity with bovine protein [Tucker 1998] - only found in retinal rod photoreceptors and platelets [Kimura 1999] NCKX1, NCKX2 have following characterisitics: - exchanges in 4Na+:1Ca2+:1K+ ratio* - bidirectional transporter* - selectivity for Na+ is absolute; Ca2+ can be replaced by Sr2+; K+ can be replaced by Rb+, Nh4+* * see refs in [Schnetkamp 2004] for refs 25769: - cloned [Prinsen 2000] - detected K-dependent Na-Ca exchange activity when recombinantly expressed in insect cells [Prinsen 2000] - only found in brain, retinal ganglion cels, cone photoreceptors (chicken transcript Northern blot) [Prinsen 2000] - see notes for 9187 about function 57419: - cloned [Kraev 2001] - displayed K+-dependent Na+/Ca2+ exchanger activity [Kraev 2001] but stoichiometry has not been established (assumed the same as NCKX1 & NCKX2) - most abundant in brain, lower levels in aorta, uterus, and intestine [Kraev 2001] 123041: - cloned [Li 2002] - demonstrated K+-dependent Na+/Ca2+ exchanger activity [Li 2002] but stoichiometry has not been established (assumed - abundantly expressed in brain , aorta, lung, and thymus, lower
NCKt	3	Kimura M, Jeanclos EM, Donnelly RJ, Lytton J, Reeves JP, Aviv A.	Physiological and molecular characterization of the Na+/Ca2+ exchanger in human platelets	Am J Physiol	1999	10484410	9187: - cloned [Tucker 1998] - 64.3% identity with bovine protein [Tucker 1998] - only found in retinal rod photoreceptors and platelets [Kimura 1999] NCKX1, NCKX2 have following characterisitics: - exchanges in 4Na+:1Ca2+:1K+ ratio* - bidirectional transporter* - selectivity for Na+ is absolute; Ca2+ can be replaced by Sr2+; K+ can be replaced by Rb+, Nh4+* * see refs in [Schnetkamp 2004] for refs 25769: - cloned [Prinsen 2000] - detected K-dependent Na-Ca exchange activity when recombinantly expressed in insect cells [Prinsen 2000] - only found in brain, retinal ganglion cels, cone photoreceptors (chicken transcript Northern blot) [Prinsen 2000] - see notes for 9187 about function 57419: - cloned [Kraev 2001] - displayed K+-dependent Na+/Ca2+ exchanger activity [Kraev 2001] but stoichiometry has not been established (assumed the same as NCKX1 & NCKX2) - most abundant in brain, lower levels in aorta, uterus, and intestine [Kraev 2001] 123041: - cloned [Li 2002] - demonstrated K+-dependent Na+/Ca2+ exchanger activity [Li 2002] but stoichiometry has not been established (assumed - abundantly expressed in brain , aorta, lung, and thymus, lower

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
NCKt	3	Prinsen CF, Szerencsei RT, Schnetkamp PP	Molecular cloning and functional expression of the potassium-dependent sodium-calcium exchanger from human and chicken retinal cone photoreceptors	J Neurosci	2000	10662833	9187: - cloned [Tucker 1998] - 64.3% identity with bovine protein [Tucker 1998] - only found in retinal rod photoreceptors and platelets [Kimura 1999] NCKX1, NCKX2 have following characterisitics: - exchanges in 4Na+:1Ca2+:1K+ ratio* - bidirectional transporter* - selectivity for Na+ is absolute; Ca2+ can be replaced by Sr2+; K+ can be replaced by Rb+, Nh4+* * see refs in [Schnetkamp 2004] for refs 25769: - cloned [Prinsen 2000] - detected K-dependent Na-Ca exchange activity when recombinantly expressed in insect cells [Prinsen 2000] - only found in brain, retinal ganglion cels, cone photoreceptors (chicken transcript Northern blot) [Prinsen 2000] - see notes for 9187 about function 57419: - cloned [Kraev 2001] - displayed K+-dependent Na+/Ca2+ exchanger activity [Kraev 2001] but stoichiometry has not been established (assumed the same as NCKX1 & NCKX2) - most abundant in brain, lower levels in aorta, uterus, and intestine [Kraev 2001] 123041: - cloned [Li 2002] - demonstrated K+-dependent Na+/Ca2+ exchanger activity [Li 2002] but stoichiometry has not been established (assumed - abundantly expressed in brain , aorta, lung, and thymus, lower
NCKt	3	Kraev A, Quednau BD, Leach S, Li XF, Dong H, Winkfein R, Perizzolo M, Cai X, Yang R, Philipson KD, Lytton J	Molecular cloning of a third member of the potassium-dependent sodium-calcium exchanger gene family, NCKX3	J Biol Chem	2001	11294880	9187: - cloned [Tucker 1998] - 64.3% identity with bovine protein [Tucker 1998] - only found in retinal rod photoreceptors and platelets [Kimura 1999] NCKX1, NCKX2 have following characterisitics: - exchanges in 4Na+:1Ca2+:1K+ ratio* - bidirectional transporter* - selectivity for Na+ is absolute; Ca2+ can be replaced by Sr2+; K+ can be replaced by Rb+, Nh4+* * see refs in [Schnetkamp 2004] for refs 25769: - cloned [Prinsen 2000] - detected K-dependent Na-Ca exchange activity when recombinantly expressed in insect cells [Prinsen 2000] - only found in brain, retinal ganglion cels, cone photoreceptors (chicken transcript Northern blot) [Prinsen 2000] - see notes for 9187 about function 57419: - cloned [Kraev 2001] - displayed K+-dependent Na+/Ca2+ exchanger activity [Kraev 2001] but stoichiometry has not been established (assumed the same as NCKX1 & NCKX2) - most abundant in brain, lower levels in aorta, uterus, and intestine [Kraev 2001] 123041: - cloned [Li 2002] - demonstrated K+-dependent Na+/Ca2+ exchanger activity [Li 2002] but stoichiometry has not been established (assumed - abundantly expressed in brain , aorta, lung, and thymus, lower

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NCKt	3	Li XF, Kraev AS, Lytton J	Molecular cloning of a fourth member of the potassium-dependent sodium-calcium exchanger gene family, NCKX4	J Biol Chem	2002	12379639	9187: - cloned [Tucker 1998] - 64.3% identity with bovine protein [Tucker 1998] - only found in retinal rod photoreceptors and platelets [Kimura 1999] NCKX1, NCKX2 have following characterisitics: - exchanges in 4Na+:1Ca2+:1K+ ratio* - bidirectional transporter* - selectivity for Na+ is absolute; Ca2+ can be replaced by Sr2+; K+ can be replaced by Rb+, Nh4+* * see refs in [Schnetkamp 2004] for refs 25769: - cloned [Prinsen 2000] - detected K-dependent Na-Ca exchange activity when recombinantly expressed in insect cells [Prinsen 2000] - only found in brain, retinal ganglion cels, cone photoreceptors (chicken transcript Northern blot) [Prinsen 2000] - see notes for 9187 about function 57419: - cloned [Kraev 2001] - displayed K+-dependent Na+/Ca2+ exchanger activity [Kraev 2001] but stoichiometry has not been established (assumed the same as NCKX1 & NCKX2) - most abundant in brain, lower levels in aorta, uterus, and intestine [Kraev 2001] 123041: - cloned [Li 2002] - demonstrated K+-dependent Na+/Ca2+ exchanger activity [Li 2002] but stoichiometry has not been established (assumed - abundantly expressed in brain , aorta, lung, and thymus, lower
NCKt	3	Schnetkamp PP	The SLC24 Na+/Ca2+-K+ exchanger family: vision and beyond.	Pflugers Arch	2004	14770312	9187: - cloned [Tucker 1998] - 64.3% identity with bovine protein [Tucker 1998] - only found in retinal rod photoreceptors and platelets [Kimura 1999] NCKX1, NCKX2 have following characterisitics: - exchanges in 4Na+:1Ca2+:1K+ ratio* - bidirectional transporter* - selectivity for Na+ is absolute; Ca2+ can be replaced by Sr2+; K+ can be replaced by Rb+, Nh4+* * see refs in [Schnetkamp 2004] for refs 25769: - cloned [Prinsen 2000] - detected K-dependent Na-Ca exchange activity when recombinantly expressed in insect cells [Prinsen 2000] - only found in brain, retinal ganglion cels, cone photoreceptors (chicken transcript Northern blot) [Prinsen 2000] - see notes for 9187 about function 57419: - cloned [Kraev 2001] - displayed K+-dependent Na+/Ca2+ exchanger activity [Kraev 2001] but stoichiometry has not been established (assumed the same as NCKX1 & NCKX2) - most abundant in brain, lower levels in aorta, uterus, and intestine [Kraev 2001] 123041: - cloned [Li 2002] - demonstrated K+-dependent Na+/Ca2+ exchanger activity [Li 2002] but stoichiometry has not been established (assumed - abundantly expressed in brain , aorta, lung, and thymus, lower
NDP7g	3	Wang TF, Guidotti G	Golgi localization and functional expression of human uridine diphosphatase	J Biol Chem	1998	9556635	9583: - gene is alternatively spliced into 2 transcripts which have different localizations and enzymatic activities: LALP70 is identical to the human Golgi UDPase with the exception of additional 24 bp in the central region of the LALP70 cDNA [Biederbick 2000] - UDPase cleaves CTP most efficiently followed by CDP, UDP, and GTP; LALP70 has highest enzyme activity on UTP and TTP [Biederbick 2000] UDPase (from [Wang 1998]): - identified by homology search & cloned - heart, brain, placenta, lung, liver, skeletal muscle, kidney, pancreas; - increased disphosphatase activity detected in transfected COS-7 cells; highest w/ UDP as substrate, lower activity w/ GDP, CDP, TDP - immunofluorescence staining suggests Golgi lumen localization LALP70 (from [Biederbick 1999]): - cloned and expressed - ubiquitous - lysosomal/autophagic vacuole membrane protein NOTE: only included reaction for UDP

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
NDP7g	3	Biederbick A, Rose S, Elsasser HP	A human intracellular apyrase-like protein, LALP70, localizes to lysosomal/autophagic vacuoles	J Cell Sci	1999	10393803	9583: - gene is alternatively spliced into 2 transcripts which have different localizations and enzymatic activities: LALP70 is identical to the human Golgi UDPase with the exception of additional 24 bp in the central region of the LALP70 cDNA [Biederbick 2000] - UDPase cleaves CTP most efficiently followed by CDP, UDP, and GTP; LALP70 has highest enzyme activity on UTP and TTP [Biederbick 2000] UDPase (from [Wang 1998]): - identified by homology search & cloned - heart, brain, placenta, lung, liver, skeletal muscle, kidney, pancreas; - increased disphosphatase activity detected in transfected COS-7 cells; highest w/ UDP as substrate, lower activity w/ GDP, CDP, TDP - immunofluorescence staining suggests Golgi lumen localization LALP70 (from [Biederbick 1999]): - cloned and expressed - ubiquitous - lysosomal/autophagic vacuole membrane protein NOTE: only included reaction for UDP
NDP7g	3	Biederbick A, Kosan C, Kunz J, Elsasser HP	First apyrase splice variants have different enzymatic properties.	J Biol Chem	2000	10858452	9583: - gene is alternatively spliced into 2 transcripts which have different localizations and enzymatic activities: LALP70 is identical to the human Golgi UDPase with the exception of additional 24 bp in the central region of the LALP70 cDNA [Biederbick 2000] - UDPase cleaves CTP most efficiently followed by CDP, UDP, and GTP; LALP70 has highest enzyme activity on UTP and TTP [Biederbick 2000] UDPase (from [Wang 1998]): - identified by homology search & cloned - heart, brain, placenta, lung, liver, skeletal muscle, kidney, pancreas; - increased disphosphatase activity detected in transfected COS-7 cells; highest w/ UDP as substrate, lower activity w/ GDP, CDP, TDP - immunofluorescence staining suggests Golgi lumen localization LALP70 (from [Biederbick 1999]): - cloned and expressed - ubiquitous - lysosomal/autophagic vacuole membrane protein NOTE: only included reaction for UDP
NDPK2m	2	Milon L, Rousseau-Merck MF, Munier A, Erent M, Lascu I, Capeau J, Lacombe ML.	nm23-H4, a new member of the family of human nm23/nucleoside diphosphate kinase genes localised on chromosome 16p13.	Hum Genet	1997	9099850	0
NH4t3r	3	Ridgwell K, Spurr NK, Laguda B, MacGeoch C, Avent ND, Tanner MJ	Isolation of cDNA clones for a 50 kDa glycoprotein of the human erythrocyte membrane associated with Rh (rhesus) blood-group antigen expression	Biochem J	1992	1417776	6005: - cloned [Ridgwell 1992] - NH4+/H+ antiport [Westhoff 2002] - found in RBC as part of a multi-subunit complex with the Rh polypeptides [Nakhoul 2004] 57127: - cloned [Liu 2001] - liver, kidney, and skin [Liu 2001] - NH4+/H+ electroneutral exchange [Ludewig 2004]
NH4t3r	3	Liu Z, Peng J, Mo R, Hui C, Huang CH	Rh type B glycoprotein is a new member of the Rh superfamily and a putative ammonia transporter in mammals	J Biol Chem	2001	11024028	6005: - cloned [Ridgwell 1992] - NH4+/H+ antiport [Westhoff 2002] - found in RBC as part of a multi-subunit complex with the Rh polypeptides [Nakhoul 2004] 57127: - cloned [Liu 2001] - liver, kidney, and skin [Liu 2001] - NH4+/H+ electroneutral exchange [Ludewig 2004]
NH4t3r	3	Nakhoul NL, Hamm LL	Non-erythroid Rh glycoproteins: a putative new family of mammalian ammonium transporters	Pflugers Arch	2004	12920597	6005: - cloned [Ridgwell 1992] - NH4+/H+ antiport [Westhoff 2002] - found in RBC as part of a multi-subunit complex with the Rh polypeptides [Nakhoul 2004] 57127: - cloned [Liu 2001] - liver, kidney, and skin [Liu 2001] - NH4+/H+ electroneutral exchange [Ludewig 2004]
NH4t3r	3	Westhoff CM, Siegel DL, Burd CG, Foskett JK	Mechanism of genetic complementation of ammonium transport in yeast by human erythrocyte Rh-associated glycoprotein	J Biol Chem	2004	14966114	6005: - cloned [Ridgwell 1992] - NH4+/H+ antiport [Westhoff 2002] - found in RBC as part of a multi-subunit complex with the Rh polypeptides [Nakhoul 2004] 57127: - cloned [Liu 2001] - liver, kidney, and skin [Liu 2001] - NH4+/H+ electroneutral exchange [Ludewig 2004]

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NH4t3r	3	Ludewig U	Electroneutral ammonium transport by basolateral rhesus B glycoprotein	J Physiol	2004	15284342	6005: - cloned [Ridgwell 1992] - NH4+/H+ antiport [Westhoff 2002] - found in RBC as part of a multi-subunit complex with the Rh polypeptides [Nakhoul 2004] 57127: - cloned [Liu 2001] - liver, kidney, and skin [Liu 2001] - NH4+/H+ electroneutral exchange [Ludewig 2004]
NKCCt	3	Payne JA, Xu JC, Haas M, Lytle CY, Ward D, Forbush B 3rd	Primary structure, functional expression, and chromosomal localization of the bumetanide-sensitive Na-K-Cl cotransporter in human colon	J Biol Chem	1995	7629105	- NKCC1, NKCC2, KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6557: - kidney-specific; apical membrane of the thick ascending limb of Henle's loop and the macula densa [Entrez Gene], [Hebert 2004] - accounts for most of the NaCl resorption [Entrez Gene], [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Entrez Gene], [Hebert 2004] - cloned [Simon 1996] 6558: - ubiquitous; basolateral membrane of epithelial cells, also found in non-epithelial cells [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Russell 2000] - cloned [Payne 1995]
NKCCt	3	Simon DB, Karet FE, Hamdan JM, DiPietro A, Sanjad SA, Lifton RP	Bartter's syndrome, hypokalaemic alkalosis with hypercalciuria, is caused by mutations in the Na-K-2Cl cotransporter NKCC2	Nat Genet	1996	8640224	- NKCC1, NKCC2, KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6557: - kidney-specific; apical membrane of the thick ascending limb of Henle's loop and the macula densa [Entrez Gene], [Hebert 2004] - accounts for most of the NaCl resorption [Entrez Gene], [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Entrez Gene], [Hebert 2004] - cloned [Simon 1996] 6558: - ubiquitous; basolateral membrane of epithelial cells, also found in non-epithelial cells [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Russell 2000] - cloned [Payne 1995]
NKCCt	3	Russell JM	Sodium-potassium-chloride cotransport	Physiol Rev	2000	10617769	- NKCC1, NKCC2, KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6557: - kidney-specific; apical membrane of the thick ascending limb of Henle's loop and the macula densa [Entrez Gene], [Hebert 2004] - accounts for most of the NaCl resorption [Entrez Gene], [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Entrez Gene], [Hebert 2004] - cloned [Simon 1996] 6558: - ubiquitous; basolateral membrane of epithelial cells, also found in non-epithelial cells [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Russell 2000] - cloned [Payne 1995]
NKCCt	3	Bergeron MJ, Gagnon E, Wallendorff B, Lapointe JY, Isenring P.	Ammonium transport and pH regulation by K(+)-Cl(-) cotransporters	Am J Physiol Renal Physiol	2003	12657561	- NKCC1, NKCC2, KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6557: - kidney-specific; apical membrane of the thick ascending limb of Henle's loop and the macula densa [Entrez Gene], [Hebert 2004] - accounts for most of the NaCl resorption [Entrez Gene], [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Entrez Gene], [Hebert 2004] - cloned [Simon 1996] 6558: - ubiquitous; basolateral membrane of epithelial cells, also found in non-epithelial cells [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Russell 2000] - cloned [Payne 1995]

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NKCCt	3	Hebert SC, Mount DB, Gamba G	Molecular physiology of cation-coupled Cl- cotransport: the SLC12 family	Pflugers Arch	2004	12739168	- NKCC1, NKCC2, KCC1, KCC3, KCC4 shown to transport NH4+ in place of K+ [Bergeron 2003] 6557: - kidney-specific; apical membrane of the thick ascending limb of Henle's loop and the macula densa [Entrez Gene], [Hebert 2004] - accounts for most of the NaCl resorption [Entrez Gene], [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Entrez Gene], [Hebert 2004] - cloned [Simon 1996] 6558: - ubiquitous; basolateral membrane of epithelial cells, also found in non-epithelial cells [Hebert 2004] - stoichiometry of 1Na:1K:2Cl [Russell 2000] - cloned [Payne 1995]
NMNATn	3	Schweiger M, Hennig K, Lerner F, Niere M, Hirsch-Kauffmann M, Specht T, Weise C, Oei SL, Ziegler M.	Characterization of recombinant human nicotinamide mononucleotide adenylyl transferase (NMNAT), a nuclear enzyme essential for NAD synthesis.	FEBS Lett	2001	11248244	IT
NMNATr	3	Raffaelli N, Sorci L, Amici A, Emanuelli M, Mazzola F, Magni G.	Identification of a novel human nicotinamide mononucleotide adenylyltransferase.	Biochem Biophys Res Commun	2003	12359228	IT
NMPTRCOX	2	Frydman J, Ruiz O, Robetto E, Dellacha JM, Frydman RB	Modulation of insulin induced ornithine decarboxylase by putrescine and methylputrescines in H-35 hepatoma cells	Mol Cell Biochem	1991	2051998	these compounds are found in mamallian cells according to the reference--so physiological evidence diamine oxidase from pig apparantly does NOT do this reaction, but may do others (PMID 3111885)
NMPTRCOX	2	Frydman RB, Ruiz O, Kreisel M, Bachrach U	Oxidation of N-alkyl and C-alkylputrescines by diamine oxidases	FEBS Lett	1987	3111885	these compounds are found in mamallian cells according to the reference--so physiological evidence diamine oxidase from pig apparantly does NOT do this reaction, but may do others (PMID 3111885)
NNAT	3	Yalowitz JA, Xiao S, Biju MP, Antony AC, Cummings OW, Deeg MA, Jayaram HN.	Characterization of human brain nicotinamide 5'-mononucleotide adenylyltransferase-2 and expression in human pancreas.	Biochem J	2004	14516279	IT
NNAT	3	Zhou T, Kurnasov O, Tomchick DR, Binns DD, Grishin NV, Marquez VE, Osterman AL, Zhang H.	Structure of human nicotinamide/nicotinic acid mononucleotide adenylyltransferase. Basis for the dual substrate specificity and activation of the oncolytic agent tiazofurin	J Biol Chem	2002		IT
NNATm	3	Zhang X, Kurnasov OV, Karthikeyan S, Grishin NV, Osterman AL, Zhang H.	Structural characterization of a human cytosolic NMN/NaMN adenylyltransferase and implication in human NAD biosynthesis.	J Biol Chem	2003	12574164	IT
NNATn	3	Emanuelli M, Carnevali F, Saccucci F, Pierella F, Amici A, Raffaelli N, Magni G.	Molecular cloning, chromosomal localization, tissue mRNA levels, bacterial expression, and enzymatic properties of human NMN adenylyltransferase.	J Biol Chem	2001	11027696	IT
NNDPR	2	Okuno E, White RJ, Schwarcz R.	Quinolinic acid phosphoribosyltransferase: purification and partial characterization from human liver and brain.	J Biochem (Tokyo)	1988	3139649	IT
NNDPR	2	Fukuoka SI, Nyaruhucha CM, Shibata K.	Characterization and functional expression of the cDNA encoding human brain quinolinate phosphoribosyltransferase.	Biochim Biophys Acta	1998	9473669	IT
NNMT	3	Aksoy S, Szumlanski CL, Weinshilboum RM.	Human liver nicotinamide N-methyltransferase. cDNA cloning, expression, and biochemical characterization.	J Biol Chem	1994	8182091	IT can be further oxidzied to n-methyl-pyridone-2-carbamide and n-methyl-pyridone-4-carbamide (but rxn in kegg seems to be wrong --> not included yet)
NOS1	3	Brenman JE, Chao DS, Xia H, Aldape K, Bredt DS	Nitric oxide synthase complexed with dystrophin and absent from skeletal muscle sarcolemma in Duchenne muscular dystrophy	Cell	1995	7545544	split form based on references
NOS1	3	Bloch KD, Wolfram JR, Brown DM, Roberts JD Jr, Zapol DG, Lepore JJ, Filippov G, Thomas JE, Jacob HJ, Bloch DB.	Three members of the nitric oxide synthase II gene family (NOS2A, NOS2B, and NOS2C) colocalize to human chromosome 17.	Genomics	1995	7558036	split form based on references
NOS1	3	Geoffrey M. Cooper	The Cell A Molecular Approach		2000		split form based on references
NS26T2g	0	Ikehara Y, Kojima N, Kurosawa N, Kudo T, Kono M, Nishihara S, Issiki S, Morozumi K, Itzkowitz S, Tsuda T, Nishimura SI, Tsuji S, Narimatsu H	Cloning and expression of a human gene encoding an N-acetylgalactosamine-alpha2,6-sialyltransferase (ST6GalNAc I): a candidate for synthesis of cancer-associated sialyl-Tn antigens	Glycobiology	1999	10536037	Siat7ap expressed in intestine [Ikehara et al, Glycobiology 1999]
NTD1	3	Rampazzo C, Gallinaro L, Milanesi E, Frigimelica E, Reichard P, Bianchi V	A deoxyribonucleotidase in mitochondria: involvement in regulation of dNTP pools and possible link to genetic disease.	Proc Natl Acad Sci U S A	2000	10899995	IT 30833.1: highest activity on 5'dUMP (100%), follwoed by 5'dIMP (96%): rampazzo et al, 2000; acts also on 3'dTMP
NTD1	3	Oka J, Matsumoto A, Hosokawa Y, Inoue S.	Molecular cloning of human cytosolic purine 5'-nucleotidase.	Biochem Biophys Res Commun	1994		IT 30833.1: highest activity on 5'dUMP (100%), follwoed by 5'dIMP (96%): rampazzo et al, 2000; acts also on 3'dTMP
NTD2	3	Amici A, Magni G.	Human erythrocyte pyrimidine 5'-nucleotidase, PN-I.	Arch Biochem Biophys	2002	11795870	IT
NTD3	3	Hunsucker SA, Spychala J, Mitchell BS.	Human cytosolic 5'-nucleotidase I: characterization and role in nucleoside analog resistance.	J Biol Chem	2001	11133996	IT 93034: highest affinity with dCMP, but acts also on AMP and IMP. ADP was necessary for max activity: Hunsucker et al 2001

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NTD7e	3	Hashikawa T, Takedachi M, Terakura M, Saho T, Yamada S, Thompson LF, Shimabukuro Y, Murakami S.	Involvement of CD73 (ecto-5'-nucleotidase) in adenosine generation by human gingival fibroblasts.	J Dent Res	2003	14578500	IT
NTP3e	3	Smith TM, Hicks-Berger CA, Kim S, Kirley TL.	Cloning, expression, and characterization of a soluble calcium-activated nucleotidase, a human enzyme belonging to a new family of extracellular nucleotidases.	Arch Biochem Biophys	2002	12234496	IT preferred substrates depends on publication: (Smith, 2002): UDP>GDP> UTP>GTP>ADP>ATP (since activity with ADP and ATP were really weak I did not included these reactions) (Murphy , 2003): GDP>UDP>IDP calcium dependent activation
NTP3e	3	Murphy DM, Ivanenkov VV, Kirley TL.	Bacterial expression and characterization of a novel, soluble, calcium-binding, and calcium-activated human nucleotidase.	Biochemistry	2003	12600208	IT preferred substrates depends on publication: (Smith, 2002): UDP>GDP> UTP>GTP>ADP>ATP (since activity with ADP and ATP were really weak I did not included these reactions) (Murphy , 2003): GDP>UDP>IDP calcium dependent activation
NTPP9	3	Lin S, McLennan AG, Ying K, Wang Z, Gu S, Jin H, Wu C, Liu W, Yuan Y, Tang R, Xie Y, Mao Y.	Cloning, expression, and characterization of a human inosine triphosphate pyrophosphatase encoded by the itpa gene.	J Biol Chem	2001	11278832	enzyme can also act on ATP, dATp, CTP, dCTP,UTP, dTTP, GTP, dGTP. However, the activity for those compounds were almost 100 time smaller than for XTP, ITP, dITP. I would only consider these compounds for hte enzyme if there is no other enzyme that uses these compounds better. IT
O2Stx	2	Archibald F.	Oxygen toxicity and the health and survival of eukaryote cells: a new piece is added to the puzzle.	Proc Natl Acad Sci U S A	2003	12939409	- inferred that superoxide can diffuse across cell membranes; according to [Archibald 2003], superoxide dismutase can "catalyze the dismutation of two superoxide anions to O2 + H2O2 at superoxide diffusion-limited rates, making them the fastest enzymes known"
O2tm	1	Koyama T, Kinjo M, Araiso T.	Oxygen diffusion through mitochondrial membranes.	Adv Exp Med Biol	1989	2551143	- Additional info by RS/TV - No genes found. - Some data found for the existence of a diffusion mechanism for oxygen across the mitochondrial membrane. Oxygen gradients were recorded in the following paper: Koyama T, Kinjo M, Araiso T. Adv Exp Med Biol. 1989;248:763-7. - oxygen assumed to be transported freely into all compartments
OCBTm	3	Horwich AL, Fenton WA, Williams KR, Kalousek F, Kraus JP, Doolittle RF, Konigsberg W, Rosenberg LE	Structure and expression of a complementary DNA for the nuclear coded precursor of human mitochondrial ornithine transcarbamylase	Science	1984	6372096	SAB reviewed - Additional information added by RS/TV: - Mitochondrial according to Entrez Gene database - Expressed solely in the liver and small intestine (Dekaney CM, Wu G, Jaeger LA. Pediatr Res. 2003 Feb;53(2):274-80.)
OCBTm	3	Dekaney CM, Wu G, Jaeger LA.	Gene expression and activity of enzymes in the arginine biosynthetic pathway in porcine fetal small intestine.	Pediatr Res	2003	12538786	SAB reviewed - Additional information added by RS/TV: - Mitochondrial according to Entrez Gene database - Expressed solely in the liver and small intestine (Dekaney CM, Wu G, Jaeger LA. Pediatr Res. 2003 Feb;53(2):274-80.)
OCOAT1m	3	Fukao T, Mitchell GA, Song XQ, Nakamura H, Kassovska-Bratinova S, Orii KE, Wraith JE, Besley G, Wanders RJ, Niezen-Koning KE, Berry GT, Palmieri M, Kondo N.	Succinyl-CoA:3-ketoacid CoA transferase (SCOT): cloning of the human SCOT gene, tertiary structural modeling of the human SCOT monomer, and characterization of three pathogenic mutations.		2000	10964512	tissue - 5019 (abundant in heart, followed in order by kidney, brain, and muscle, whereas in liver it is undetectable; also detectable in leukocytes and fibroblasts.); 64064 (testis specific) mitochondrial - Harvester, UniProt, GeneCards MM - Checked over by RS/TV
OCOAT1m	3	Tanaka H, Kohroki J, Iguchi N, Onishi M, Nishimune Y.	Cloning and characterization of a human orthologue of testis-specific succinyl CoA: 3-oxo acid CoA transferase (Scot-t) cDNA.		2002	11756565	tissue - 5019 (abundant in heart, followed in order by kidney, brain, and muscle, whereas in liver it is undetectable; also detectable in leukocytes and fibroblasts.); 64064 (testis specific) mitochondrial - Harvester, UniProt, GeneCards MM - Checked over by RS/TV

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OIVD1m	3	Lau KS, Herring WJ, Chuang JL, McKean M, Danner DJ, Cox RP, Chuang DT.	Structure of the gene encoding dihydrolipoyl transacylase (E2) component of human branched chain alpha-keto acid dehydrogenase complex and characterization of an E2 pseudogene.		1992	1429740	This process, carried out by the mitochondrial branched-chain alpha-ketoacid dehydrogenase complex, consists of five distinct reactions. First, alpha-ketoisocaproate is oxidatively decarboxylated, catalyzed by the E1 component of the complex. Lipoamide associated with E1 is reduced at the same time. Next, the isovaleryl group derived from alpha-ketoisocaproate is transferred to coenzyme A in two steps catalyzed by the E2 component of the complex (dihydrolipoyl transacetylase). Finally, the oxidized form of lipoamide is regenerated and electrons are transferred to NAD+ in two steps catalyzed by the E3 component of the complex (dihydrolipoyl dehydrogenase). [Reactome]
OIVD1m	3	Fisher CW, Chuang JL, Griffin TA, Lau KS, Cox RP, Chuang DT.	Molecular phenotypes in cultured maple syrup urine disease cells. Complete E1 alpha cDNA sequence and mRNA and subunit contents of the human branched chain alpha-keto acid dehydrogenase complex.		1989	2914958	This process, carried out by the mitochondrial branched-chain alpha-ketoacid dehydrogenase complex, consists of five distinct reactions. First, alpha-ketoisocaproate is oxidatively decarboxylated, catalyzed by the E1 component of the complex. Lipoamide associated with E1 is reduced at the same time. Next, the isovaleryl group derived from alpha-ketoisocaproate is transferred to coenzyme A in two steps catalyzed by the E2 component of the complex (dihydrolipoyl transacetylase). Finally, the oxidized form of lipoamide is regenerated and electrons are transferred to NAD+ in two steps catalyzed by the E3 component of the complex (dihydrolipoyl dehydrogenase). [Reactome]
OMPDC	3	Suttle DP, Bugg BY, Winkler JK, Kanalas JJ.	Molecular cloning and nucleotide sequence for the complete coding region of human UMP synthase.	Proc Natl Acad Sci U S A	1988	3279416	IT no infos about localization in cell
OMPDC	3	Patterson D, Jones C, Morse H, Rumsby P, Miller Y, Davis R.	Structural gene coding for multifunctional protein carrying orotate phosphoribosyltransferase and OMP decarboxylase activity is located on long arm of human chromosome 3.	Somatic Cell Genet	1983	6574608	IT no infos about localization in cell
OMPDC	3	McClard RW, Black MJ, Livingstone LR, Jones ME.	Isolation and initial characterization of the single polypeptide that synthesizes uridine 5'-monophosphate from orotate in Ehrlich ascites carcinoma. Purification by tandem affinity chromatography of uridine-5'-monophosphate synthase.	Biochemistry	1980	6893554	IT no infos about localization in cell
OMPDC	3	Suchi M, Mizuno H, Kawai Y, Tsuboi T, Sumi S, Okajima K, Hodgson ME, Ogawa H, Wada Y.	Molecular cloning of the human UMP synthase gene and characterization of point mutations in two hereditary orotic aciduria families.	Am J Hum Genet	1997	9042911	IT no infos about localization in cell
ORNDC	3	Hsieh JT, Denning MF, Heidel SM, Verma AK.	Expression of human chromosome 2 ornithine decarboxylase gene in ornithine decarboxylase-deficient Chinese hamster ovary cells	Cancer Res	1990	2317811	Gene and enzyme characterized
ORNDC	3	Zhu MY, Iyo A, Piletz JE, Regunathan S	Expression of human arginine decarboxylase, the biosynthetic enzyme for agmatine	Biochim Biophys Acta	2004		Gene and enzyme characterized
ORNTArm	3	Shen BW, Hennig M, Hohenester E, Jansonius JN, Schirmer T	Crystal structure of human recombinant ornithine aminotransferase	J Mol Biol	1998	9514741	Enzyme and reaction characterized
P45011A1m	3	Sakaki T, Inouye K.	Practical application of mammalian cytochrome P450.	J Biosci Bioeng	2000	16232916	mit- see refs specificity: adrenal cortex, ovary, testis, placenta, giant trophoblast Catalyzes the side-chain cleavage reaction of cholesterol to pregnenolone Further details of 4mptnl not known - degraded to isocaproic acid and isocapryl alcohol (see PMID: 8645003) - details and downstream metabolism not known. NJ
P45017A2r	3	Payne AH, Hales DB	Overview of steroidogenic enzymes in the pathway from cholesterol to active steroid hormones	Endocrine Reviews	2004		ER - by refs - also noted to be microsomal, need to return and review lit again specificity: leydig cells (testis), adrenal cortex, thecal cells (ovary) Conversion of pregnenolone and progesterone to their 17- alpha hydroxylated products and subsequently to dehydroepiandrosterone (DHEA) and androstenedione. Catalyzes both the 17-alpha-hydroxylation and the 17,20-lyase reaction. Involved in sexual development during fetal life and a puberty. NJ
P45017A2r	3	Korzekwa KR, Trager WF, Mancewicz J, Osawa Y	Studies on the mechanism of aromatase and other cytochrome P450 mediated deformylation reactions	J Steroid Biochem Mol Biol	1993		ER - by refs - also noted to be microsomal, need to return and review lit again specificity: leydig cells (testis), adrenal cortex, thecal cells (ovary) Conversion of pregnenolone and progesterone to their 17- alpha hydroxylated products and subsequently to dehydroepiandrosterone (DHEA) and androstenedione. Catalyzes both the 17-alpha-hydroxylation and the 17,20-lyase reaction. Involved in sexual development during fetal life and a puberty. NJ

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P45017A2r	3	Korzekwa KR, Trager WF, Mancewicz J, Osawa Y	Studies on the mechanism of aromatase and other cytochrome P450 mediated deformylation reactions	J Steroid Biochem Mol Biol	1993		ER - by refs - also noted to be microsomal, need to return and review lit again specificity: leydig cells (testis), adrenal cortex, thecal cells (ovary) Conversion of pregnenolone and progesterone to their 17- alpha hydroxylated products and subsequently to dehydroepiandrosterone (DHEA) and androstenedione. Catalyzes both the 17-alpha-hydroxylation and the 17,20-lyase reaction. Involved in sexual development during fetal life and a puberty. NJ
P4501B1r	3	Tang YM, Wo YY, Stewart J, Hawkins AL, Griffin CA, Sutter TR, Greenlee WF.	Isolation and characterization of the human cytochrome P450 CYP1B1 gene.	J Biol Chem	1996	8910454	ER - uniprot The enzyme encoded by this gene localizes to the endoplasmic reticulum and metabolizes procarcinogens such as polycyclic aromatic hydrocarbons and 17beta-estradiol. Mutations in this gene have been associated with primary congenital glaucoma; therefore it is thought that the enzyme also metabolizes a signaling molecule involved in eye development, possibly a steroid. biochem and gene exp - Tang ref NJ
P4502A6	3	Oscarson M, Gullsten H, Rautio A, Bernal ML, Sinues B, Dahl ML, Stengard JH, Pelkonen O, Raunio H, Ingelman-Sundberg M.	Genotyping of human cytochrome P450 2A6 (CYP2A6), a nicotine C-oxidase.	FEBS Let	1998	9827545	ER - placed on cytsolic side because of xenobiotic rxn. CYP2A6, encodes a member of the cytochrome P450 superfamily of enzymes. The cytochrome P450 proteins are monooxygenases which catalyze many reactions involved in drug metabolism and synthesis of cholesterol, steroids and other lipids. This protein localizes to the endoplasmic reticulum and its expression is induced by phenobarbital. The enzyme is known to hydroxylate coumarin, and also metabolizes nicotine, aflatoxin B1, nitrosamines, and some pharmaceuticals. Individuals with certain allelic variants are said to have a poor metabolizer phenotype, meaning they do not efficiently metabolize coumarin or nicotine. This gene is part of a large cluster of cytochrome P450 genes from the CYP2A, CYP2B and CYP2F subfamilies on chromosome 19q. The gene was formerly referred to as CYP2A3; however, it has been renamed CYP2A6. Oscarson - 2A6 NJ NJ
P4502C18	2	Goldstein JA, Faletto MB, Romkes-Sparks M, Sullivan T, Kitareewan S, Raucy JL, Lasker JM, Ghanayem BI.	Evidence that CYP2C19 is the major (S)-mephenytoin 4'-hydroxylase in humans.	Biochemistry	1994	8110777	ER - placed on cytsolic side because of xenobiotic rxn. NJ
P4502C19	3	Meier UT, Meyer UA.	Genetic polymorphism of human cytochrome P-450 (S)-mephenytoin 4-hydroxylase. Studies with human autoantibodies suggest a functionally altered cytochrome P-450 isozyme as cause of the genetic deficiency.	Biochemistry	1987	3442670	ER - placed on cytsolic side because of xenobiotic rxn. This protein localizes to the endoplasmic reticulum and is known to metabolize many xenobiotics, including the anticonvulsive drug mephenytoin, omeprazole, diazepam and some barbiturates. Polymorphism within this gene is associated with variable ability to metabolize mephenytoin, known as the poor metabolizer and extensive metabolizer phenotypes. NJ

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
P4502C9	3	Umbenhauer DR, Martin MV, Lloyd RS, Guengerich FP.	Cloning and sequence determination of a complementary DNA related to human liver microsomal cytochrome P-450 S-mephenytoin 4-hydroxylase.	Biochemistry	1987		ER - placed on cytsolic side because of xenobiotic rxn. This protein localizes to the endoplasmic reticulum and its expression is induced by rifampin. The enzyme is known to metabolize many xenobiotics, including phenytoin, tolbutamide, ibuprofen and S-warfarin. Studies identifying individuals who are poor metabolizers of phenytoin and tolbutamide suggest that this gene is polymorphic. NJ
P4502C9	3	Meehan RR, Gosden JR, Rout D, Hastie ND, Friedberg T, Adesnik M, Buckland R, van Heyningen V, Fletcher J, Spurr NK, et al.	Human cytochrome P-450 PB-1: a multigene family involved in mephenytoin and steroid oxidations that maps to chromosome 10.	Am J Hum Genet	1988	2827463	ER - placed on cytsolic side because of xenobiotic rxn. This protein localizes to the endoplasmic reticulum and its expression is induced by rifampin. The enzyme is known to metabolize many xenobiotics, including phenytoin, tolbutamide, ibuprofen and S-warfarin. Studies identifying individuals who are poor metabolizers of phenytoin and tolbutamide suggest that this gene is polymorphic. NJ
P4502C92	3	Miyazawa M, Shindo M, Shimada T.	Metabolism of (+)- and (-)-limonenes to respective carveols and perillyl alcohols by CYP2C9 and CYP2C19 in human liver microsomes.	Drug Metab Dispos	2002	11950794	ER bound enzyme - assumed to take place on outer membrane (no evidence to support 1 direction vs another). specificity: liver (possibly intestines) see Miyazawa ref PMID 11950794 for rxn specificity NJ
P4502E1	3	Umeno M, McBride OW, Yang CS, Gelboin HV, Gonzalez FJ.	Human ethanol-inducible P450IIE1: complete gene sequence, promoter characterization, chromosome mapping, and cDNA-directed expression.	Biochemistry	1988	3233219	ER - placed on cytsolic side because of xenobiotic rxn. This protein localizes to the endoplasmic reticulum and is induced by ethanol, the diabetic state, and starvation. The enzyme metabolizes both endogenous substrates, such as ethanol, acetone, and acetal, as well as exogenous substrates including benzene, carbon tetrachloride, ethylene glycol, and nitrosamines which are premutagens found in cigarette smoke. Due to its many substrates, this enzyme may be involved in such varied processes as gluconeogenesis, hepatic cirrhosis, diabetes, and cancer. NJ
P4502F1	3	Nhamburo PT, Kimura S, McBride OW, Kozak CA, Gelboin HV, Gonzalez FJ.	The human CYP2F gene subfamily: identification of a cDNA encoding a new cytochrome P450, cDNA-directed expression, and chromosome mapping.	Biochemistry	1990	1974816	ER - placed on cytsolic side because of xenobiotic rxn. This protein localizes to the endoplasmic reticulum and is known to dehydrogenate 3-methylindole, an endogenous toxin derived from the fermentation of tryptophan, as well as xenobiotic substrates such as naphthalene and ethoxycoumarin. This gene is part of a large cluster of cytochrome P450 genes from the CYP2A, CYP2B and CYP2F subfamilies on chromosome 19q. NJ
P4503A4	3	Molowa DT, Schuetz EG, Wrighton SA, Watkins PB, Kremers P, Mendez-Picon G, Parker GA, Guzelian PS.	Complete cDNA sequence of a cytochrome P-450 inducible by glucocorticoids in human liver.	Proc Natl Acad Sci U S A	1986	3460094	ER -does not specifiy inner vs outer - assumed outer membrane since aflatoxin is an exogenous metabolite This protein localizes to the endoplasmic reticulum and its expression is induced by glucocorticoids and some pharmacological agents. This enzyme is involved in the metabolism of approximately half the drugs which are are used today, including acetaminophen, codeine, cyclosporin A, diazepam and erythromycin. The enzyme also metabolizes some steroids and carcinogens. This gene is part of a cluster of cytochrome P450 genes on chromosome 7q21.1. Previously another CYP3A gene, CYP3A3, was thought to exist; however, it is now thought that this sequence represents a transcript variant of CYP3A4. NJ
P4503A4	3	Finta C, Zaphiropoulos PG.	Intergenic mRNA molecules resulting from trans-splicing.	J Biol Chem	2002	11726664	ER -does not specifiy inner vs outer - assumed outer membrane since aflatoxin is an exogenous metabolite This protein localizes to the endoplasmic reticulum and its expression is induced by glucocorticoids and some pharmacological agents. This enzyme is involved in the metabolism of approximately half the drugs which are are used today, including acetaminophen, codeine, cyclosporin A, diazepam and erythromycin. The enzyme also metabolizes some steroids and carcinogens. This gene is part of a cluster of cytochrome P450 genes on chromosome 7q21.1. Previously another CYP3A gene, CYP3A3, was thought to exist; however, it is now thought that this sequence represents a transcript variant of CYP3A4. NJ

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P4503A7r	3	Komori M, Nishio K, Ohi H, Kitada M, Kamataki T.	Molecular cloning and sequence analysis of cDNA containing the entire coding region for human fetal liver cytochrome P-450.	J Biochem (Tokyo)	1989	2722762	ER - uniprot biochem act, seq see Komori ref This enzyme hydroxylates testosterone and dehydroepiandrosterone 3-sulphate, which is involved in the formation of estriol during pregnancy. The enzyme also metabolizes some drugs such as aflatoxin B1. This gene is part of a cluster of cytochrome P450 genes on chromosome 7q21.1. Transcript variants have been described, but it is not known whether these transcripts are normally produced. NJ
P4504B1r	3	Nhamburo PT, Gonzalez FJ, McBride OW, Gelboin HV, Kimura S.	Identification of a new P450 expressed in human lung: complete cDNA sequence, cDNA-directed expression, and chromosome mapping.	Biochemistry	1989		Membrane-bound. Endoplasmic reticulum. - uniprot P450 can be induced to high levels in liver and other tissues by various foreign compounds, including drugs, pesticides, and carcinogens. Rxn specificity - from Lewis ref (which refers to rendic ref). NJ
P4504B1r	3	Rendic S.	Summary of information on human CYP enzymes: human P450 metabolism data.	Drug Metab Rev	2002	11996015	Membrane-bound. Endoplasmic reticulum. - uniprot P450 can be induced to high levels in liver and other tissues by various foreign compounds, including drugs, pesticides, and carcinogens. Rxn specificity - from Lewis ref (which refers to rendic ref). NJ
P4504F121r	3	Lewis DF.	57 varieties: the human cytochromes P450.	Pharmacogenomics	2004	15579107	ER - uniprot - inner vs outer membrane not specified, assumed inner specific hydroxylation product not defined, general omega hydroxy arachidonic acid used When expressed in yeast the enzyme is capable of oxdizing arachidonic acid; however, its physiological function has not been determined. This gene is part of a cluster of cytochrome P450 genes on chromosome 19.
P4504F122r	3	Kikuta Y, Kusunose E, Kondo T, Yamamoto S, Kinoshita H, Kusunose M.	Cloning and expression of a novel form of leukotriene B4 omega-hydroxylase from human liver.	FEBS Lett	1994	8026587	ER - uniprot - inner vs outer membrane not specified, assumed inner kikuta (1993) ref for exp evidence leukotriene B4 specificty not as good as cyp4f2 - see lewis ref Cyp4f2 see Kikuta 1994 ref For possible additional, alternative substrates see PMID: 15145985. This protein localizes to the endoplasmic reticulum. The enzyme starts the process of inactivating and degrading leukotriene B4, a potent mediator of inflammation. This gene is part of a cluster of cytochrome P450 genes on chromosome 19. NJ
P4504F122r	3	Kikuta Y, Kusunose E, Endo K, Yamamoto S, Sogawa K, Fujii-Kuriyama Y, Kusunose M.	A novel form of cytochrome P-450 family 4 in human polymorphonuclear leukocytes. cDNA cloning and expression of leukotriene B4 omega-hydroxylase.	J Biol Chem	1993	8486631	ER - uniprot - inner vs outer membrane not specified, assumed inner kikuta (1993) ref for exp evidence leukotriene B4 specificty not as good as cyp4f2 - see lewis ref Cyp4f2 see Kikuta 1994 ref For possible additional, alternative substrates see PMID: 15145985. This protein localizes to the endoplasmic reticulum. The enzyme starts the process of inactivating and degrading leukotriene B4, a potent mediator of inflammation. This gene is part of a cluster of cytochrome P450 genes on chromosome 19. NJ

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P4504F122r	3	Le Quere V, Plee-Gautier E, Potin P, Madec S, Salaun JP.	Human CYP4F3s are the main catalysts in the oxidation of fatty acid epoxides.	J Lipid Res	2004	15145985	ER - uniprot - inner vs outer membrane not specified, assumed inner kikuta (1993) ref for exp evidence leukotriene B4 specificty not as good as cyp4f2 - see lewis ref Cyp4f2 see Kikuta 1994 ref For possible additional, alternative substrates see PMID: 15145985. This protein localizes to the endoplasmic reticulum. The enzyme starts the process of inactivating and degrading leukotriene B4, a potent mediator of inflammation. This gene is part of a cluster of cytochrome P450 genes on chromosome 19. NJ
P4507A1r	3	Russell DW	The enzymes, regulation, and genetics of bile acid synthesis	Annu Rev Biochem	2003		ER: uniprot NJ
P4508B11r	3	Gafvels M, Olin M, Chowdhary BP, Raudsepp T, Andersson U, Persson B, Jansson M, Bjorkhem I, Eggertsen G	Structure and chromosomal assignment of the sterol 12-alpha-hydroxylase gene (cyp8b1) in human and mouse	Genomics	1999		ER - uniprot NADH/NADPH can be used as cofactors specificity: liver (also kidney to a smaller degree) Involved in bile acid synthesis and is responsible for the conversion of 7 alpha-hydroxy-4-cholesten-3-one into 7 alpha, 12 alpha-dihydroxy-4-cholesten-3-one. Responsible for the balance between formation of cholic acid and chenodeoxycholic acid. Has a rather broad substrate specificity including a number of 7-alpha- hydroxylated C27 steroids. NJ
P450LTB4r	2	Soberman RJ.	Cytochrome P-450LTB and inactivation of leukotriene B4.	Methods Enzymol	1990	2172735	ER localization inferred by similarity w/ preceding rxn (P450F122r). See refs for supporting physiological data (PMID: 2172735, etc). No GPR identified yet. NJ
P5CDm	3	Hu CA, Lin WW, Valle D.	Cloning, characterization, and expression of cDNAs encoding human delta 1-pyrroline-5-carboxylate dehydrogenase.		1996	8621661	Mitochondrial matrix; preferred cofactor NAD - Hu et al. J Biol Chem. 1996 Apr 19;271(16):9795-800. Genetic data - Geraghty et. al. Hum Mol Genet. 1998 Sep;7(9):1411-5. Reversible according to Reactome database predominantly in liver
P5CDm	3	Geraghty MT, Vaughn D, Nicholson AJ, Lin WW, Jimenez-Sanchez G, Obie C, Flynn MP, Valle D, Hu CA.	Mutations in the Delta1-pyrroline 5-carboxylate dehydrogenase gene cause type II hyperprolinemia.		1998	9700195	Mitochondrial matrix; preferred cofactor NAD - Hu et al. J Biol Chem. 1996 Apr 19;271(16):9795-800. Genetic data - Geraghty et. al. Hum Mol Genet. 1998 Sep;7(9):1411-5. Reversible according to Reactome database predominantly in liver
P5CR	3	Merrill MJ, Yeh GC, Phang JM.	Purified human erythrocyte pyrroline-5-carboxylate reductase. Preferential oxidation of NADPH.	J Biol Chem	1989	2722838	prefers NADPH according to ref
PAFH	3	Hattori M, Adachi H, Tsujimoto M, Arai H, Inoue K.	Miller-Dieker lissencephaly gene encodes a subunit of brain platelet-activating factor acetylhydrolase [corrected]	Nature	1994		cytoplasmic - uniprot and refs TISSUE SPECIFICITY: In the adult, expressed in brain, skeletal muscle, kidney, thymus, spleen, colon, testis, ovary and peripheral blood leukocytes. In the fetus, highest expression occurs in brain. Specificity for isoform 2: high in B and T lymphocytes. In brain, expression is restricted to amygdala and frontal cortex. Isoform 2: cloning and expression ref: Hattori (1996) Cytosolic PAF-AH IB is formed of three subunits of 45 kDa (alpha), 30 kDa (beta) and 29 kDa (gamma). The catalytic activity of the enzyme resides in the beta and gamma subunits, whereas the alpha subunit has regulatory activity. Trimer formation is not essential for the catalytic activity. PAFAH1B1 was identified as encoding a gene that when mutated or lost caused the lissencephaly associated with Miller-Dieker lissencephaly syndrome. PAFAH1B1 encodes the non-catalytic alpha subunit of the intracellular Ib isoform of platelet-activating factor acteylhydrolase, a heterotrimeric enzyme that specifically catalyzes the removal of the acetyl group at the SN-2 position of platelet-activating factor (identified as 1-O-alkyl-2 NJ

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PAFH	3	Adachi H, Tsujimoto M, Hattori M, Arai H, Inoue K.	cDNA cloning of human cytosolic platelet-activating factor acetylhydrolase gamma-subunit and its mRNA expression in human tissues.	Biochem Biophys Res Commun	1995	7669037	cytoplasmic - uniprot and refs TISSUE SPECIFICITY: In the adult, expressed in brain, skeletal muscle, kidney, thymus, spleen, colon, testis, ovary and peripheral blood leukocytes. In the fetus, highest expression occurs in brain. Specificity for isoform 2: high in B and T lymphocytes. In brain, expression is restricted to amygdala and frontal cortex. Isoform 2: cloning and expression ref: Hattori (1996) Cytosolic PAF-AH IB is formed of three subunits of 45 kDa (alpha), 30 kDa (beta) and 29 kDa (gamma). The catalytic activity of the enzyme resides in the beta and gamma subunits, whereas the alpha subunit has regulatory activity. Trimer formation is not essential for the catalytic activity. PAFAH1B1 was identified as encoding a gene that when mutated or lost caused the lissencephaly associated with Miller-Dieker lissencephaly syndrome. PAFAH1B1 encodes the non-catalytic alpha subunit of the intracellular Ib isoform of platelet activating factor acteylhydrolase, a heterotrimeric enzyme that specifically catalyzes the removal of the acetyl group at the SN-2 position of platelet-activating factor (identified as 1-O-alkyl-2 NJ
PAFH	3	Hattori K, Adachi H, Matsuzawa A, Yamamoto K, Tsujimoto M, Aoki J, Hattori M, Arai H, Inoue K.	cDNA cloning and expression of intracellular platelet activating factor (PAF) acetylhydrolase II. Its homology with plasma PAF acetylhydrolase.	J Biol Chem	1996	8955149	cytoplasmic - uniprot and refs TISSUE SPECIFICITY: In the adult, expressed in brain, skeletal muscle, kidney, thymus, spleen, colon, testis, ovary and peripheral blood leukocytes. In the fetus, highest expression occurs in brain. Specificity for isoform 2: high in B and T lymphocytes. In brain, expression is restricted to amygdala and frontal cortex. Isoform 2: cloning and expression ref: Hattori (1996) Cytosolic PAF-AH IB is formed of three subunits of 45 kDa (alpha), 30 kDa (beta) and 29 kDa (gamma). The catalytic activity of the enzyme resides in the beta and gamma subunits, whereas the alpha subunit has regulatory activity. Trimer formation is not essential for the catalytic activity. PAFAH1B1 was identified as encoding a gene that when mutated or lost caused the lissencephaly associated with Miller-Dieker lissencephaly syndrome. PAFAH1B1 encodes the non-catalytic alpha subunit of the intracellular Ib isoform of platelet activating factor acteylhydrolase, a heterotrimeric enzyme that specifically catalyzes the removal of the acetyl group at the SN-2 position of platelet-activating factor (identified as 1-O-alkyl-2 NJ
PAFHe	3	Stafforini DM, Satoh K, Atkinson DL, Tjoelker LW, Eberhardt C, Yoshida H, Imaizumi T, Takamatsu S, Zimmerman GA, McIntyre TM, Gray PW, Prescott SM.	Platelet-activating factor acetylhydrolase deficiency. A missense mutation near the active site of an anti-inflammatory phospholipase.	J Clin Invest	2005	8675689	localization: plasma/extracellular (uniprot) specificity: plasma Modulates the action of platelet-activating factor (PAF) by hydrolyzing the sn-2 ester bond to yield the biologically inactive lyso-PAF. Has a specificity for substrates with a short residue at the sn-2 position. It is inactive against long-chain phospholipids. See PMID: 8675689 NJ
PAPStg	3	Kamiyama S, Suda T, Ueda R, Suzuki M, Okubo R, Kikuchi N, Chiba Y, Goto S, Toyoda H, Saigo K, Watanabe M, Narimatsu H, Jigami Y, Nishihara S	Molecular cloning and identification of 3'-phosphoadenosine 5'-phosphosulfate transporter	J Biol Chem	2003	12716889	- 48.1% identity w/ Drosophila protein [Kamiyama 2003] - Golgi [Kamiyama 2003] - PAPS transport [Kamiyama 2003] - high in placenta and pancreas, low in colon and heart [Kamiyama 2003]
PCHOLP_hs	3	Colley WC, Sung TC, Roll R, Jenco J, Hammond SM, Altshuller Y, Bar-Sagi D, Morris AJ, Frohman MA.	Phospholipase D2, a distinct phospholipase D isoform with novel regulatory properties that provokes cytoskeletal reorganization.	Curr Biol	1997	9395408	cytosol - uniprot - Colley ref - PMID 9395408 NJ
PCHOLP_hs	3	Lopez I, Arnold RS, Lambeth JD.	Cloning and initial characterization of a human phospholipase D2 (hPLD2). ADP-ribosylation factor regulates hPLD2.	J Biol Chem	1998	9582313	cytosol - uniprot - Colley ref - PMID 9395408 NJ
PCHOLPm_hs	3	Hammond SM, Altshuller YM, Sung TC, Rudge SA, Rose K, Engebrecht J, Morris AJ, Frohman MA.	Human ADP-ribosylation factor-activated phosphatidylcholine-specific phospholipase D defines a new and highly conserved gene family.	J Biol Chem	1995	8530346	perinuclear regions for PLD1 - need to doublecheck for mit specificity - localization: Colley PMID 9395408 Tissue specificity: Expressed abundantly in the pancreas and heart and at high levels in brain, placenta, spleen, uterus and small intestine. Implicated as a critical step in numerous cellular pathways, including signal transduction, membrane trafficking, and the regulation of mitosis. May be involved in the regulation of perinuclear intravesicular membrane traffic (By similarity). NJ

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PCHOLPm_hs	3	Cases S, Stone SJ, Zhou P, Yen E, Tow B, Lardizabal KD, Voelker T, Farese RV Jr.	Cloning of DGAT2, a second mammalian diacylglycerol acyltransferase, and related family members.	J Biol Chem	2001	11481335	perinuclear regions for PLD1 - need to doublecheck for mit specificity - localization: Colley PMID 9395408 Tissue specificity: Expressed abundantly in the pancreas and heart and at high levels in brain, placenta, spleen, uterus and small intestine. Implicated as a critical step in numerous cellular pathways, including signal transduction, membrane trafficking, and the regulation of mitosis. May be involved in the regulation of perinuclear intravesicular membrane traffic (By similarity). NJ
PCLAD	3	Fukuoka S, Ishiguro K, Yanagihara K, Tanabe A, Egashira Y, Sanada H, Shibata K	Identification and expression of a cDNA encoding human alpha-amino-beta-carboxymuconate-epsilon-semialdehyde decarboxylase (ACMSD). A key enzyme for the tryptophan-niacine pathway and "quinolinate hypothesis"	J Biol Chem	2002	12140278	Only known conversion of cmusa to avoid quln formation.
PCm	3	Freytag SO, Collier KJ.	Molecular cloning of a cDNA for human pyruvate carboxylase. Structural relationship to other biotin-containing carboxylases and regulation of mRNA content in differentiating preadipocytes.	J Biol Chem	1984	6548474	mitochondrial matrix [RefSeq], [UniProt] - found in liver, kidney, and intestine, NOT sk muscle, heart, or brain [Orten, Human Biochem 1975] - Additional information added by RS/TV: Two transcriptional variants according to Entrez Gene database Pyruvate carboxylase catalyzes the formation of oxaloacetate from pyruvate and HCO3-. Pyruvate carboxylase is located solely in the mitochondrial matrix. Pyruvate carboxylate is present in a variety of tissues for various reasons: (1) Gluconeogenic tissues such as the liver and kideny (2) Lipogenic tissues such as the liver, adipose, lactating mammary gland, and adrenal gland. (3) Other tissues where it has an anapleurotic role. All this according to Freytag SO, Collier KJ. J Biol Chem. 1984 Oct 25;259(20):12831-7.
PCRNtc	3	Jakobs BS, Wanders RJ.	Fatty acid beta-oxidation in peroxisomes and mitochondria: the first, unequivocal evidence for the involvement of carnitine in shuttling propionyl-CoA from peroxisomes to mitochondria.	Biochem Biophys Res Commun	1995	7654220	Peroxisomes don't have general carnitine transport shuttles, an exception is ppcoa/pcrn. PMID: 7654220 NJ
PCt	3	Ruetz S, Gros P.	Phosphatidylcholine translocase: a physiological role for the mdr2 gene.	Cell	1994	7912658	ABC transporter for phosphatidyl choline transport into bile. Localized on basolateral membrane of hepatocytes and cholangiocytes (liver and bile ducts). Alternative splicing of this gene results in several products of undetermined function. See PMID: 7912658 for characterization and function. Also Meier and Steiger review. PMID: 7893760 for promoter characterization PMID: 7912658 - PC translocase NJ
PCt	3	Meier PJ, Stieger B.	Bile salt transporters.	Annu Rev Physiol	2002	11826283	ABC transporter for phosphatidyl choline transport into bile. Localized on basolateral membrane of hepatocytes and cholangiocytes (liver and bile ducts). Alternative splicing of this gene results in several products of undetermined function. See PMID: 7912658 for characterization and function. Also Meier and Steiger review. PMID: 7893760 for promoter characterization PMID: 7912658 - PC translocase NJ

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PDE1	3	Miki T, Taira M, Hockman S, Shimada F, Lieman J, Napolitano M, Ward D, Taira M, Makino H, Manganiello VC.	Characterization of the cDNA and gene encoding human PDE3B, the cGIP1 isoform of the human cyclic GMP-inhibited cyclic nucleotide phosphodiesterase family.	Genomics	1996	8884271	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Rosman GJ, Martins TJ, Sonnenburg WK, Beavo JA, Ferguson K, Loughney K.	Isolation and characterization of human cDNAs encoding a cGMP-stimulated 3',5'-cyclic nucleotide phosphodiesterase.	Gene	1997	9210593	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Yu J, Wolda SL, Frazier AL, Florio VA, Martins TJ, Snyder PB, Harris EA, McCaw KN, Farrell CA, Steiner B, Bentley JK, Beavo JA, Ferguson K, Gelinas R.	Identification and characterisation of a human calmodulin-stimulated phosphodiesterase PDE1B1.	Cell Signal	1997	9419816	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT

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PDE1	3	Fawcett L, Baxendale R, Stacey P, McGrouther C, Harrow I, Soderling S, Hetman J, Beavo JA, Phillips SC.	Molecular cloning and characterization of a distinct human phosphodiesterase gene family: PDE11A.	Proc Natl Acad Sci U S A	2000	10725373	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Gardner C, Robas N, Cawkill D, Fidock M.	Cloning and characterization of the human and mouse PDE7B, a novel cAMP-specific cyclic nucleotide phosphodiesterase.	Biochem Biophys Res Commun	2000	10872825	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Yuasa K, Kotera J, Fujishige K, Michibata H, Sasaki T, Omori K.	Isolation and characterization of two novel phosphodiesterase PDE11A variants showing unique structure and tissue-specific expression.	J Biol Chem	2000	10906126	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT

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PDE1	3	Shakur Y, Takeda K, Kenan Y, Yu ZX, Rena G, Brandt D, Houslay MD, Degerman E, Ferrans VJ, Manganiello VC.	Membrane localization of cyclic nucleotide phosphodiesterase 3 (PDE3). Two N-terminal domains are required for the efficient targeting to, and association of, PDE3 with endoplasmic reticulum.	J Biol Chem	2000	10952971	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Hetman JM, Robas N, Baxendale R, Fidock M, Phillips SC, Soderling SH, Beavo JA.	Cloning and characterization of two splice variants of human phosphodiesterase 11A.	Proc Natl Acad Sci U S A	2000	11050148	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Secchiero P, Zella D, Curreli S, Mirandola P, Capitani S, Gallo RC, Zauli G.	Pivotal role of cyclic nucleoside phosphodiesterase 4 in Tat-mediated CD4+ T cell hyperactivation and HIV type 1 replication.	Proc Natl Acad Sci U S A	2000	11114167	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT

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PDE1	3	Manns JM, Brenna KJ, Colman RW, Sheth SB.	Differential regulation of human platelet responses by cGMP inhibited and stimulated cAMP phosphodiesterases.	Thromb Haemost	2002	12038792	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Lefievre L, de Lamirande E, Gagnon C.	Presence of cyclic nucleotide phosphodiesterases PDE1A, existing as a stable complex with calmodulin, and PDE3A in human spermatozoa.	Biol Reprod	2002	12135876	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Wechsler J, Choi YH, Krall J, Ahmad F, Manganiello VC, Movsesian MA.	Isoforms of cyclic nucleotide phosphodiesterase PDE3A in cardiac myocytes.	J Biol Chem	2002	12154085	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT

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PDE1	3	Smith SJ, Brookes-Fazakerley S, Donnelly LE, Barnes PJ, Barnette MS, Giembycz MA.	Ubiquitous expression of phosphodiesterase 7A in human proinflammatory and immune cells	Am J Physiol Lung Cell Mol Physiol	2003	12388353	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Gamanuma M, Yuasa K, Sasaki T, Sakurai N, Kotera J, Omori K.	Comparison of enzymatic characterization and gene organization of cyclic nucleotide phosphodiesterase 8 family in humans.	Cell Signal	2003	12681444	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Pryzwansky KB, Madden VJ	Type 4A cAMP-specific phosphodiesterase is stored in granules of human neutrophils and eosinophils.	Cell Tissue Res	2003	12764607	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT

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PDE1	3	Kotera J, Sasaki T, Kobayashi T, Fujishige K, Yamashita Y, Omori K.	Subcellular localization of cyclic nucleotide phosphodiesterase type 10A variants, and alteration of the localization by cAMP-dependent protein kinase-dependent phosphorylation.	J Biol Chem	2004	14604994	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Scapin G, Patel SB, Chung C, Varnerin JP, Edmondson SD, Mastracchio A, Parmee ER, Singh SB, Becker JW, Van der Ploeg LH, Tota MR.	Crystal structure of human phosphodiesterase 3B: atomic basis for substrate and inhibitor specificity.	Biochemistry	2004	15147193	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE1	3	Goraya TA, Masada N, Ciruela A, Cooper DM.	Sustained entry of Ca2+ is required to activate Ca2+-calmodulin-dependent phosphodiesterase 1A.	J Biol Chem	2004	15272012	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT

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PDE1	3	White JB, Thompson WJ, Pittler SJ.	Characterization of 3',5' cyclic nucleotide phosphodiesterase activity in Y79 retinoblastoma cells: absence of functional PDE6.	Mol Vis	2004	15480303	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDH4: White et al, fund activity in Y79 retinoblastoma cells; PDHa: are stored in granules of neutrophiles and eosinophiles PDH4: Lefievre et al, Spermatozoa. PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind;can used cGMP and cAMP as substrate but they compete for binding site PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, placenta, lung, skeletal muscle, kidney and pancreas PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodulin-dependent; in spermatozoa, in Y79 retinoblastoma cells PDE1B: has preference for cGMP, calmodulin-dependent PDE7B: specific for cAMP, expressed in brain, heart, liver PDE7A:Messenger RNA transcripts were detected by RT-PCR PDE11A: no infos where located. Different groups identified in IT
PDE4	3	Shimizu-Matsumoto A, Itoh K, Inazawa J, Nishida K, Matsumoto Y, Kinoshita S, Matsubara K, Okubo K.	Isolation and chromosomal localization of the human cone cGMP phosphodiesterase gamma cDNA (PDE6H).	Genomics	1996	8786098	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDE6: I am not sure if I understood the GPR in GeneCards right. --> therefore Conf level 0 PDE6 mRNA expression is very high in retina, and some groups found it only there expressed, however, White et al, 2004, Mol. Vis., 10,738-749, measured no PDE6 activity in Y70 retinoblastoma cells but the mRNA expression. There might be postranslational modification for a active stable PDE6 that do not occurs in retinoblasoma cells ... PDE1, PDE4: White et al, 2004, Mol. Vis., 10,738-749, measured activity in Y70 retinoblastoma cells b PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind; can used cGMP and cAMP as substrate but the PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, pla PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodu PDE1B: has preference for cGMP, calmodulin-dependent, high
PDE4	3	Fisher DA, Smith JF, Pillar JS, St Denis SH, Cheng JB.	Isolation and characterization of PDE9A, a novel human cGMP-specific phosphodiesterase.	J Biol Chem	1998	9624146	IMPORTANT: IT is thought that the PDE's contribute to a compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDE6: I am not sure if I understood the GPR in GeneCards right. --> therefore Conf level 0 PDE6 mRNA expression is very high in retina, and some groups found it only there expressed, however, White et al, 2004, Mol. Vis., 10,738-749, measured no PDE6 activity in Y70 retinoblastoma cells but the mRNA expression. There might be postranslational modification for a active stable PDE6 that do not occurs in retinoblasoma cells ... PDE1, PDE4: White et al, 2004, Mol. Vis., 10,738-749, measured activity in Y70 retinoblastoma cells b PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind; can used cGMP and cAMP as substrate but the PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, pla PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodu PDE1B: has preference for cGMP, calmodulin-dependent, high

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PDE4	3	Yanaka N, Kotera J, Ohtsuka A, Akatsuka H, Imai Y, Michibata H, Fujishige K, Kawai E, Takebayashi S, Okumura K, Omori K.	Expression, structure and chromosomal localization of the human cGMP-binding cGMP-specific phosphodiesterase PDE5A gene.	Eur J Biochem	1998	9716380	compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDE6: I am not sure if I understood the GPR in GeneCards right. --> therefore Conf level 0 PDE6 mRNA expression is very high in retina, and some groups found it only there expressed, however, White et al, 2004, Mol. Vis., 10,738-749, measured no PDE6 activity in Y70 retinoblastoma cells but the mRNA expression. There might be postranslational modification for a active stable PDE6 that do not occurs in retinoblasoma cells ... PDE1, PDE4: White et al, 2004, Mol. Vis., 10,738-749, measured activity in Y70 retinoblastoma cells b PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind; can used cGMP and cAMP as substrate but the PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, pla PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodu PDE1B: has preference for cGMP, calmodulin-dependent, high
PDE4	3	Identification and distribution of different mRNA variants produced by differential splicing in the human phosphodiesterase 9A gene.	Identification and distribution of different mRNA variants produced by differential splicing in the human phosphodiesterase 9A gene.	Biochem Biophys Res Commun	2003	12565835	compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDE6: I am not sure if I understood the GPR in GeneCards right. --> therefore Conf level 0 PDE6 mRNA expression is very high in retina, and some groups found it only there expressed, however, White et al, 2004, Mol. Vis., 10,738-749, measured no PDE6 activity in Y70 retinoblastoma cells but the mRNA expression. There might be postranslational modification for a active stable PDE6 that do not occurs in retinoblasoma cells ... PDE1, PDE4: White et al, 2004, Mol. Vis., 10,738-749, measured activity in Y70 retinoblastoma cells b PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind; can used cGMP and cAMP as substrate but the PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, pla PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodu PDE1B: has preference for cGMP, calmodulin-dependent, high
PDE4	3	Wang P, Wu P, Egan RW, Billah MM.	Identification and characterization of a new human type 9 cGMP-specific phosphodiesterase splice variant (PDE9A5). Differential tissue distribution and subcellular localization of PDE9A variants.	Gene	2003	14527714	compartimentalization of cell signaling. Although I found ER-membrane associated PDE's, there is no experimental evidence that they act in ER but in cytoplasm (by inhibition) PDE6: I am not sure if I understood the GPR in GeneCards right. --> therefore Conf level 0 PDE6 mRNA expression is very high in retina, and some groups found it only there expressed, however, White et al, 2004, Mol. Vis., 10,738-749, measured no PDE6 activity in Y70 retinoblastoma cells but the mRNA expression. There might be postranslational modification for a active stable PDE6 that do not occurs in retinoblasoma cells ... PDE1, PDE4: White et al, 2004, Mol. Vis., 10,738-749, measured activity in Y70 retinoblastoma cells b PDE3A: is ER-membrane associated. However, if inhibited, intracellular cAMP concentration increase. expressed in cardiomycytes, paletes, spermatozoa, vascular smooth muscle cells. Wechsler et al, found 3 isoforms of PDE3A, produced by 2 transcripts -- > in LocusLink & SimPheny only 1 transcript -- > keep in mind; can used cGMP and cAMP as substrate but the PDE3B: adipocytes and hepatocytes PDE2A: is expressed in brain and to a lesser extent in heart, pla PDE1A: has a higher affinitiy to cGMP than to cAMP; calmodu PDE1B: has preference for cGMP, calmodulin-dependent, high
PDX5PO	3	Kang JH, Hong ML, Kim DW, Park J, Kang TC, Won MH, Baek NI, Moon BJ, Choi SY, Kwon OS.	Genomic organization, tissue distribution and deletion mutation of human pyridoxine 5'-phosphate oxidase.	Eur J Biochem	2004	15182361	mainly expressed in liver, skeletal muscle and kidney. Erys also have PDX5PO activity (see Review) IT
PDX5PO	3	Mehansho H, Henderson LM.	Transport and accumulation of pyridoxine and pyridoxal by erythrocytes.	J Biol Chem	1980	J Biol Chem.	mainly expressed in liver, skeletal muscle and kidney. Erys also have PDX5PO activity (see Review) IT
PE_HSter	2	Daleke DL, Lyles JV.	Identification and purification of aminophospholipid flippases.	Biochim Biophys Acta	2000	10856717	- evidence for scramblase that translocates phosphatidylethanolamine, phosphatidylcholine, and phosphatidylserine in rat liver ER membranes [Chang 2004] NJ: additional comments: Energy independent, so rev, scramblase activity.

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PE_HSter	2	Chang QL, Gummadi SN, Menon AK	Chemical modification identifies two populations of glycerophospholipid flippase in rat liver ER	Biochemistry	2004	15311932	- evidence for scramblase that translocates phosphatidylethanolamine, phosphatidylcholine, and phosphatidylserine in rat liver ER membranes [Chang 2004] NJ: additional comments: Energy independent, so rev, scramblase activity.
PEAMNO	2	Lyles GA	Mammalian plasma and tissue-bound semicarbazide-sensitive amine oxidases: biochemical, pharmacological and toxicological aspects	Int J Biochem Cell Biol	1996	8920635	4th citation gives evidence that MAOB does this more than MAOA, but the journal is slightly suspect and the data may be from rats, so physiological evidence.
PEAMNO	2	Smith DJ, Salmi M, Bono P, Hellman J, Leu T, Jalkanen S.	Cloning of vascular adhesion protein 1 reveals a novel multifunctional adhesion molecule	J Exp Med	1998	9653080	4th citation gives evidence that MAOB does this more than MAOA, but the journal is slightly suspect and the data may be from rats, so physiological evidence.
PEAMNO	2	Shih JC, Chen K, Ridd MJ	Role of MAO A and B in neurotransmitter metabolism and behavior	Pol J Pharmacol	1999	10389141	4th citation gives evidence that MAOB does this more than MAOA, but the journal is slightly suspect and the data may be from rats, so physiological evidence.
PEAMNO	2	Buffoni F, Ignesti G	Biochemical aspects and functional role of the copper containing amine oxidases	Inflammopharmacology	2003	15035803	4th citation gives evidence that MAOB does this more than MAOA, but the journal is slightly suspect and the data may be from rats, so physiological evidence.
PEFLIP	3	Mouro I, Halleck MS, Schlegel RA, Mattei MG, Williamson P, Zachowski A, Devaux P, Cartron JP, Colin Y.	Cloning, expression, and chromosomal mapping of a human ATPase II gene, member of the third subfamily of P-type ATPases and orthologous to the presumed bovine and murine aminophospholipid translocase.	Biochem Biophys Res Commun	1999	10198212	transport out of mitochondria - flip and flip (not to be confused w/ flip-flop) mechanisms are ATP dependent, whereas scramblase transport of lipids is not - PMID: 10856717 Largely inferred transport at this time (sequence homology characterizes it as a phosphlipid, ATP dependent transporter). ATP10A: PMID: 11353404 - The protein encoded by this gene belongs to the family of P-type cation transport ATPases, and to the subfamily of aminophospholipid-transporting ATPases. The aminophospholipid translocases transport phosphatidylserine and phosphatidylethanolamine from one side of a bilayer to another. This gene is maternally expressed. It maps within the most common interval of deletion responsible for Angelman syndrome, also known as 'happy puppet syndrome'. ATP8A1: PMID: 10198212 for cloning and expression. The P-type adenosinetriphosphatases (P-type ATPases) are a family of proteins which use the free energy of ATP hydrolysis to drive uphill transport of ions across membranes. Several subfamilies of P-type ATPases have been identified. One subfamily catalyzes transport of heavy metal ions. Another subfamily tran NJ
PEFLIP	3	Herzing LB, Kim SJ, Cook EH Jr, Ledbetter DH.	The human aminophospholipid-transporting ATPase gene ATP10C maps adjacent to UBE3A and exhibits similar imprinted expression.	Am J Hum Genet	2001	11353404	transport out of mitochondria - flip and flip (not to be confused w/ flip-flop) mechanisms are ATP dependent, whereas scramblase transport of lipids is not - PMID: 10856717 Largely inferred transport at this time (sequence homology characterizes it as a phosphlipid, ATP dependent transporter). ATP10A: PMID: 11353404 - The protein encoded by this gene belongs to the family of P-type cation transport ATPases, and to the subfamily of aminophospholipid-transporting ATPases. The aminophospholipid translocases transport phosphatidylserine and phosphatidylethanolamine from one side of a bilayer to another. This gene is maternally expressed. It maps within the most common interval of deletion responsible for Angelman syndrome, also known as 'happy puppet syndrome'. ATP8A1: PMID: 10198212 for cloning and expression. The P-type adenosinetriphosphatases (P-type ATPases) are a family of proteins which use the free energy of ATP hydrolysis to drive uphill transport of ions across membranes. Several subfamilies of P-type ATPases have been identified. One subfamily catalyzes transport of heavy metal ions. Another subfamily tran NJ
PEPCKm	0	Modaressi S, Christ B, Bratke J, Zahn S, Heise T, Jungermann K.	Molecular cloning, sequencing and expression of the cDNA of the mitochondrial form of phosphoenolpyruvate carboxykinase from human liver.	Biochem J	1996	8645161	mitochondrial [UniProt] - found in liver, kidney, and intestine, NOT sk muscle, heart, or brain [Orten, Human Biochem 1975] - Additional information by RS/TV: Phosphoenolpyruvate carboykinase (PCK) (EC 4.1.1.32) catalyses the GTP-driven conversion of oxaloacetate to phosphoenolpyruvate. Tissue localization: Mainly in the liver and the kidney Subcellular Localization: Exists as two isozymes, Pck1.1 is in the cytosol. Pck2.1-m is in the mitochondria. All according to Modaressi S, Christ B, Bratke J, Zahn S, Heise T, Jungermann K. Biochem J. 1996 May 1;315 (Pt 3):807-14.

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PEROXx	3	Adamski J, Normand T, Leenders F, Monte D, Begue A, Stehelin D, Jungblut PW, de Launoit Y.	Molecular cloning of a novel widely expressed human 80 kDa 17 beta-hydroxysteroid dehydrogenase IV.	Biochem J	1995	7487879	to dmnoncoa which is transported out of the peroxisome, into the mitochondria and degraded 1x -> a heptanoyl-CoA - however "the metabolic fate of 2,6 dimethylheptanoyl-CoA IS NOT KNOWN" see PMID: 11591435 (Verhoeven review article). See also Mukherji (PMID:12814641) ACOX3: PMID: 15599942, Zha et al ACOX3, which is expressed at extremely low level in other human organs studied including the liver, might contribute significantly to peroxisomal branched chain fatty acid beta-oxidation in human prostate tissue and some prostate cancer cell lines. Acyl-Coenzyme A oxidase 3 also know as pristanoyl CoA oxidase (ACOX3)is involved in the desaturation of 2-methyl branched fatty acids in peroxisomes. Unlike the rat homolog, the human gene is expressed in very low amounts in liver such that its mRNA was undetectable by routine Northern-blot analysis or its product by immunoblotting or by enzyme activity measurements. However the human cDNA encoding a 700 amino acid protein with a peroxisomal targeting C-terminal tripeptide S-K-L was isolated and is thought to be expressed un HSD17B4: PMID: 7487879 ACAA1: PMID: 1679347, Bout Acetyl-Coenzyme A acyltransferase (ACAA1) is an enzyme op
PEROXx	3	Zha S, Ferdinandusse S, Hicks JL, Denis S, Dunn TA, Wanders RJ, Luo J, De Marzo AM, Isaacs WB.	Peroxisomal branched chain fatty acid beta-oxidation pathway is upregulated in prostate cancer.	Prostate	2005	15599942	to dmnoncoa which is transported out of the peroxisome, into the mitochondria and degraded 1x -> a heptanoyl-CoA - however "the metabolic fate of 2,6 dimethylheptanoyl-CoA IS NOT KNOWN" see PMID: 11591435 (Verhoeven review article). See also Mukherji (PMID:12814641) ACOX3: PMID: 15599942, Zha et al ACOX3, which is expressed at extremely low level in other human organs studied including the liver, might contribute significantly to peroxisomal branched chain fatty acid beta-oxidation in human prostate tissue and some prostate cancer cell lines. Acyl-Coenzyme A oxidase 3 also know as pristanoyl CoA oxidase (ACOX3)is involved in the desaturation of 2-methyl branched fatty acids in peroxisomes. Unlike the rat homolog, the human gene is expressed in very low amounts in liver such that its mRNA was undetectable by routine Northern-blot analysis or its product by immunoblotting or by enzyme activity measurements. However the human cDNA encoding a 700 amino acid protein with a peroxisomal targeting C-terminal tripeptide S-K-L was isolated and is thought to be expressed un HSD17B4: PMID: 7487879 ACAA1: PMID: 1679347, Bout Acetyl-Coenzyme A acyltransferase (ACAA1) is an enzyme op
PETHCT	3	Nakashima A, Hosaka K, Nikawa J	Cloning of a human cDNA for CTP-Phosphoethanolamine cytidylyltransferase by complementation in vivo of a yeast mutant	Journal of Biological Chemistry	1997		cytoplasmic - unitprot NJ
PETOHMm_hs	2	Walkey CJ, Shields DJ, Vance DE.	Identification of three novel cDNAs for human phosphatidylethanolamine N-methyltransferase and localization of the human gene on chromosome 17p11.2.	Biochim Biophys Acta	1999	9989271	Original version added to subset of amino acid metabolism and created an unknown compound. Literature appears to support the conversion of pe-> pchol in humans. localization: ER and mit according to Swiss-Prot * Function: Catalyzes three sequential methylation of phosphatidylethanolamine (PE) by AdoMet, thus producing phosphatidylcholine (PC). * Catalytic activity: S-adenosyl-L-methionine + phosphatidylethanolamine = S-adenosyl-L-homocysteine + phosphatidyl-N-methylethanolamine. * Enzyme regulation: The first methylation is rate-limiting. NJ
PFK	2	Eto K, Sakura H, Yasuda K, Hayakawa T, Kawasaki E, Moriuchi R, Nagataki S, Yazaki Y, Kadowaki T	Cloning of a complete protein-coding sequence of human platelet-type phosphofructokinase isozyme from pancreatic islet	Biochem Biophys Res Commun	1994	8117307	-muscle only contains homotetratmer of M subunits - liver only contains homotetramer of L subunits - platelets, white blood cells contain all 5 L,M heterotetramer combinations (4L, 3LM, 2L2M, LM3, M4) [Eto et al, Biochem Biophys Res Comm 1994] -tetramer of randomly associated isozymes available in individual tissues [Eto et al, Biochem Biophys Res Comm 1994] - higher proportion of P subunits found in brain, platelet, fibroids [UniProt] - physiologically irreversible [Orten, Human Biochm 1975]
PGCD	3	Cho HM, Jun DY, Bae MA, Ahn JD, Kim YH.	Nucleotide sequence and differential expression of the human 3-phosphoglycerate dehydrogenase gene.		2000	10713460	irreversible according to Lehninger (pg. 844, 4th ed.) cytosolic based on mouse localisation (Kazuyuki et. al, 2004 PMID:14645240)

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PGDIr	3	Yamashima T, Sakuda K, Tohma Y, Yamashita J, Oda H, Irikura D, Eguchi N, Beuckmann CT, Kanaoka Y, Urade Y, Hayaishi O	Prostaglandin D synthase (beta-trace) in human arachnoid and meningioma cells: roles as a cell marker or in cerebrospinal fluid absorption, tumorigenesis, and calcification process.	J Neurosci	1997		location: found in cytosol + ER membrane specificity: different transcripts have different specificities - see refs, e.g. PTGDS has brain (oligodendrocyte specificity) and PGDS has hematopoietic cell specificity. The protein encoded by this gene is a glutathione-independent prostaglandin D synthase that catalyzes the conversion of prostaglandin H2 (PGH2) to postaglandin D2 (PGD2). PGD2 functions as a neuromodulator as well as a trophic factor in the central nervous system. PGD2 is also involved in smooth muscle contraction/relaxation and is a potent inhibitor of platelet aggregation. This gene is preferentially expressed in brain. Studies with transgenic mice overexpressing this gene suggest that this gene may be also involved in the regulation of non-rapid eye movement sleep. NJ
PGDIr	3	Mahmud I, Ueda N, Yamaguchi H, Yamashita R, Yamamoto S, Kanaoka Y, Urade Y, Hayaishi O.	Prostaglandin D synthase in human megakaryoblastic cells.	J Biol Chem	1997	9353279	location: found in cytosol + ER membrane specificity: different transcripts have different specificities - see refs, e.g. PTGDS has brain (oligodendrocyte specificity) and PGDS has hematopoietic cell specificity. The protein encoded by this gene is a glutathione-independent prostaglandin D synthase that catalyzes the conversion of prostaglandin H2 (PGH2) to postaglandin D2 (PGD2). PGD2 functions as a neuromodulator as well as a trophic factor in the central nervous system. PGD2 is also involved in smooth muscle contraction/relaxation and is a potent inhibitor of platelet aggregation. This gene is preferentially expressed in brain. Studies with transgenic mice overexpressing this gene suggest that this gene may be also involved in the regulation of non-rapid eye movement sleep. NJ
PGDIr	3		Goodman & Gilman's the pharmacological basis of therapeutics		2001		location: found in cytosol + ER membrane specificity: different transcripts have different specificities - see refs, e.g. PTGDS has brain (oligodendrocyte specificity) and PGDS has hematopoietic cell specificity. The protein encoded by this gene is a glutathione-independent prostaglandin D synthase that catalyzes the conversion of prostaglandin H2 (PGH2) to postaglandin D2 (PGD2). PGD2 functions as a neuromodulator as well as a trophic factor in the central nervous system. PGD2 is also involved in smooth muscle contraction/relaxation and is a potent inhibitor of platelet aggregation. This gene is preferentially expressed in brain. Studies with transgenic mice overexpressing this gene suggest that this gene may be also involved in the regulation of non-rapid eye movement sleep. NJ
PGESr	3	Han R, Tsui S, Smith TJ.	Up-regulation of prostaglandin E2 synthesis by interleukin-1beta in human orbital fibroblasts involves coordinate induction of prostaglandin-endoperoxide H synthase-2 and glutathione-dependent prostaglandin E2 synthase expression.	J Biol Chem	2002	11847219	ER: uniprot specificity: none The protein encoded by this gene is a glutathione-dependent prostaglandin E synthase. The expression of this gene has been shown to be induced by proinflammatory cytokine interleukin 1 beta (IL1B). Its expression can also be induced by tumor suppressor protein TP53, and may be involved in TP53 induced apoptosis. Knockout studies in mice suggest that this gene may contribute to the pathogenesis of collagen-induced arthritis and mediate acute pain during inflammatory responses. Alternatively spliced transcript variants encoding distinct isoforms have been observed. For PTGES2: The protein encoded by this gene is a membrane-associated prostaglandin E synthase, which catalyzes the conversion of prostaglandin H2 to prostaglandin E2. This protein also has been shown to activate the transcription regulated by a gamma-interferon-activated transcription element (GATE). Four alternatively spliced transcript variants encoding three distinct isoforms have been observed. NJ
PGLYCP	3	Mulley JC, Barton N, Callen DF	Localisation of human PGP and HAGH genes to 16p13.3	Cytogenet Cell Genet	1990	2164460	- existence of PGP in human RBC is well established; has been purified and biochemically characterized [Zecher 1982] - has been detected in all human tissues [- chr location of gene has been identified [Mulley 1990]
PGLYCP	3	Zecher R, Schwulera U, Wolf HU.	Purification, isolation and characterization of a phosphoglycolate phosphatase isoenzyme from human erythrocytes.	Int J Biochem	1982	6290284	- existence of PGP in human RBC is well established; has been purified and biochemically characterized [Zecher 1982] - has been detected in all human tissues [- chr location of gene has been identified [Mulley 1990]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
PGPPT	2	Schlame M, Rua D, Greenberg ML.	The biosynthesis and functional role of cardiolipin.	Prog Lipid Res	2000	10799718	cytoplasm - uniprot NJ
PGS	3	Funk CD, Funk LB, Kennedy ME, Pong AS, Fitzgerald GA.	Human platelet/erythroleukemia cell prostaglandin G/H synthase: cDNA cloning, expression, and gene chromosomal assignment.	FASEB J	1991	1907252	ER membrane - uniprot, cytoplasmic: see ref by Maihofner Cloned version came from platelets - PMID: 1907252 Specific cofactor determined based on consistency w/ other parts of pathway (biosynthetic, O2 dependent). HemeB is a cofactor, but no evidence that it is a metabolized cofactor. aka COX1 and COX2 Prostaglandin-endoperoxide synthase (PTGS), also known as cyclooxygenase, is the key enzyme in prostaglandin biosynthesis, and acts both as a dioxygenase and as a peroxidase. There are two isozymes of PTGS: a constitutive PTGS1 and an inducible PTGS2, which differ in their regulation of expression and tissue distribution. This gene encodes PTGS2, which shows 86% - 89% amino acid sequence identity with mouse, rat, sheep, bovine, horse and rabit PTGS2 proteins, respectively. Human PTGS2 is expressed in a limited number of cell types and regulated by specific stimulatory events, suggesting that it is responsible for the prostanoid biosynthesis involved in inflammation and mitogenesis. The expression of this gene is deregulated in epithelial tumors. NJ
PGS	3	Maihofner C, Probst-Cousin S, Bergmann M, Neuhuber W, Neundorfer B, Heuss D.	Expression and localization of cyclooxygenase-1 and 2 in human sporadic amyotrophic lateral sclerosis.	Eur J Neurosci	2003	14511332	ER membrane - uniprot, cytoplasmic: see ref by Maihofner Cloned version came from platelets - PMID: 1907252 Specific cofactor determined based on consistency w/ other parts of pathway (biosynthetic, O2 dependent). HemeB is a cofactor, but no evidence that it is a metabolized cofactor. aka COX1 and COX2 Prostaglandin-endoperoxide synthase (PTGS), also known as cyclooxygenase, is the key enzyme in prostaglandin biosynthesis, and acts both as a dioxygenase and as a peroxidase. There are two isozymes of PTGS: a constitutive PTGS1 and an inducible PTGS2, which differ in their regulation of expression and tissue distribution. This gene encodes PTGS2, which shows 86% - 89% amino acid sequence identity with mouse, rat, sheep, bovine, horse and rabit PTGS2 proteins, respectively. Human PTGS2 is expressed in a limited number of cell types and regulated by specific stimulatory events, suggesting that it is responsible for the prostanoid biosynthesis involved in inflammation and mitogenesis. The expression of this gene is deregulated in epithelial tumors. NJ
PHCDm	2	Valle D, Goodman SI, Harris SC, Phang JM.	Genetic evidence for a common enzyme catalyzing the second step in the degradation of proline and hydroxyproline.	J Clin Invest	1979	500817	based on KEGG and citation - ALDH4A1 catalyzes the oxidation of both delta 1-pyrroline-5-carboxylate and delta 1-pyrroline-3-hydroxy-5-carboxylate (the second steps in the degradation of proline and hydroxyproline, respectively) [Valle 1979] - isolated mouse liver mitochondria produced glyoxylate from hydroxyproline [Knight 2005]
PHCDm	2	Takayama T, Fujita K, Suzuki K, Sakaguchi M, Fujie M, Nagai E, Watanabe S, Ichiyama A, Ogawa Y	Control of oxalate formation from L-hydroxyproline in liver mitochondria	J Am Soc Nephrol	2003	12660328	based on KEGG and citation - ALDH4A1 catalyzes the oxidation of both delta 1-pyrroline-5-carboxylate and delta 1-pyrroline-3-hydroxy-5-carboxylate (the second steps in the degradation of proline and hydroxyproline, respectively) [Valle 1979] - isolated mouse liver mitochondria produced glyoxylate from hydroxyproline [Knight 2005]
PHEACGLNt	0	Vanholder RC, Glorieux G, De Smet R, De Deyn PP	Low water-soluble uremic toxins	Adv Ren Replace Ther	2003	14681857	Compound (or one with a very similar name) is said to be present in the urine, so some transport is necessary. Gene and mechanism unknown. SAB
PHEMEt	3	Shayeghi M, Latunde-Dada GO, Oakhill JS, Laftah AH, Takeuchi K, Halliday N, Khan Y, Warley A, McCann FE, Hider RC, Frazer DM, Anderson GJ, Vulpe CD, Simpson RJ, McKie AT	Identification of an intestinal heme transporter	Cell	2005	16143108	- mouse protein was isolated from duodenum; shown to mediate heme transport in a temperature-dependent and saturable manner [Shayeghi 2005] - sequence of mouse gene was aligned with human, rabbit, rat, and zebrafish orthologs [Shayeghi 2005] - organic heme is known to be absorbed from the diet (breakdown of hemoglobin and myoglobin contained in red meat) [Shayeghi 2005] - in mammals, duodenal enterocytes and hepatocytes are major sites of heme transport [Shayeghi 2005] - heme has been shown to diffuse across model lipid membranes; also, studies in isolated hepatocytes and intestine-like cell line Caco-2 have demonstrated that heme uiptake takes place by a staurable carrier mediated process [Shayeghi 2005]
PHETA1	2	Jeremy M. Berg, John L. Tymoczko, Lubert Stryer, Neil D. Clarke	Biochemistry		2002		Well-established that this reaction, or one with alternate cofactors, takes place excessively in individuals with phenylketonuria.

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PHETHPTOX2	3	Waters PJ, Parniak MA, Nowacki P, Scriver CR	In vitro expression analysis of mutations in phenylalanine hydroxylase: linking genotype to phenotype and structure to function	Hum Mutat	1998	9450897	Gene and reaction characterized. A cause of PKU.
PHYHx	3	Jansen GA, Mihalik SJ, Watkins PA, Moser HW, Jakobs C, Denis S, Wanders RJ.	Phytanoyl-CoA hydroxylase is present in human liver, located in peroxisomes, and deficient in Zellweger syndrome: direct, unequivocal evidence for the new, revised pathway of phytanic acid alpha-oxidation in humans.	Biochem Biophys Res Commun	1996	8954107	localization: peroxisome (uniprot) specificity: Expressed in liver, kidney, and T-cells, but not in spleen, brain, heart, lung and skeletal muscle. The protein encoded by this gene is a peroxisomal enzyme. It catalyzes the initial alpha-oxidation step in the degradation of phytanic acid and converts phytanoyl-CoA to 2-hydroxyphytanoyl-CoA. It interacts specifically with the immunophilin FKBP52. Refsum disease, an autosomal recessive neurologic disorder, is caused by the deficiency of this encoded protein.
PHYHx	3	Jansen GA, Ofman R, Ferdinandusse S, Ijlst L, Muijsers AO, Skjeldal OH, Stokke O, Jakobs C, Besley GT, Wraith JE, Wanders RJ.	Refsum disease is caused by mutations in the phytanoyl-CoA hydroxylase gene.	Nat Genet	1997	9326940	localization: peroxisome (uniprot) specificity: Expressed in liver, kidney, and T-cells, but not in spleen, brain, heart, lung and skeletal muscle. The protein encoded by this gene is a peroxisomal enzyme. It catalyzes the initial alpha-oxidation step in the degradation of phytanic acid and converts phytanoyl-CoA to 2-hydroxyphytanoyl-CoA. It interacts specifically with the immunophilin FKBP52. Refsum disease, an autosomal recessive neurologic disorder, is caused by the deficiency of this encoded protein.
PHYQt	3	David A. Bender	Vitamin K		2003		uptake in proximal small intestin and incorporation in chylomicrons extrahepatic tissues take up phyQ from chylomicrons and synthesize menaquinone-4 which is hte principal vitamer in tissues other than the liver. some menaquinone-4 is also absorbed into the portal system from the colon - vitamin K1, comes from plants - estrogens increase PhyQ absorption - menaquinones (vit k2) are mainly absorbed from terminal ileum (where bile salts are present) into hepatic portal vein - about 90% of total liver content of vit K is menaquinone 7 to 13 and the hepatic pool of phyQ turns over considerably faster than that of menaquinone --> little storage of vit k liver makes catabolism of Vit k - however I could not find out how! IT
PI345P3P	3	Ono H, Katagiri H, Funaki M, Anai M, Inukai K, Fukushima Y, Sakoda H, Ogihara T, Onishi Y, Fujishiro M, Kikuchi M, Oka Y, Asano T	Regulation of phosphoinositide metabolism, Akt phosphorylation, and glucose transport by PTEN (phosphatase and tensin homolog deleted on chromosome 10) in 3T3-L1 adipocytes	Mol Endocrinol	2001	11463863	- has PtdIns(3,4,5)P3 phosphatase activity [RefSeq], [UniProt], [Ono, Mol Endocrinol 2001]
PI34P5K	3	Cunningham TW, Majerus PW	Pathway for the formation of D-3 phosphate containing inositol phospholipids in PDGF stimulated NIH 3T3 fibroblasts	Biochem Biophys Res Commun	1991	1850246	- reaction described in [Tolias, Chem Phys Lipids 1999] - whether this rxn occurs significantly in vivo is contraversial [Cunningham, J Biol Chem 1990], [Cunningham, Biophys Biochem Res Commun 1991], [Stephens, Nature 1991], [Carter, Biochem J 1994]
PI34P5K	3	Stephens LR, Hughes KT, Irvine RF	Pathway of phosphatidylinositol(3,4,5)-trisphosphate synthesis in activated neutrophils	Nature	1991	1851250	- reaction described in [Tolias, Chem Phys Lipids 1999] - whether this rxn occurs significantly in vivo is contraversial [Cunningham, J Biol Chem 1990], [Cunningham, Biophys Biochem Res Commun 1991], [Stephens, Nature 1991], [Carter, Biochem J 1994]
PI34P5K	3	Cunningham TW, Lips DL, Bansal VS, Caldwell KK, Mitchell CA, Majerus PW	Pathway for the formation of D-3 phosphate containing inositol phospholipids in intact human platelets	J Biol Chem	1990	2174884	- reaction described in [Tolias, Chem Phys Lipids 1999] - whether this rxn occurs significantly in vivo is contraversial [Cunningham, J Biol Chem 1990], [Cunningham, Biophys Biochem Res Commun 1991], [Stephens, Nature 1991], [Carter, Biochem J 1994]
PI34P5K	3	Carter AN, Huang R, Sorisky A, Downes CP, Rittenhouse SE	Phosphatidylinositol 3,4,5-trisphosphate is formed from phosphatidylinositol 4,5-bisphosphate in thrombin-stimulated platelets	Biochem J	1994	8042983	- reaction described in [Tolias, Chem Phys Lipids 1999] - whether this rxn occurs significantly in vivo is contraversial [Cunningham, J Biol Chem 1990], [Cunningham, Biophys Biochem Res Commun 1991], [Stephens, Nature 1991], [Carter, Biochem J 1994]
PI35P3P	3	Walker DM, Urbe S, Dove SK, Tenza D, Raposo G, Clague MJ	Characterization of MTMR3. an inositol lipid 3-phosphatase with novel substrate specificity	Curr Biol	2001	11676921	- isolated from HeLa cells; was shown to hydrolyze PtdIns3P and PtdIns(3,5)P2 to PtdIns and PtdIns5P, respeectively, in vitro and when heterologously expressed in S. cerevisiae [Walker 2001]

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PI45P3K	3	Carpenter CL, Duckworth BC, Auger KR, Cohen B, Schaffhausen BS, Cantley LC	Purification and characterization of phosphoinositide 3-kinase from rat liver	J Biol Chem	1990	2174051	- rat protein is heterodimer of an 85 kd regulatory subunit that mediates binding to phosphorylated proteins and a 110 kd catalytic subunit [Carpenter, J Biol Chem 1990] - 3 known class IA catalytic subunits: PIK3CA (p110a), PIK3CB (p110b), PIK3CD (p110c) which form complexes with 85 kDa regulatory subunits: PIK3R1 (p85a, p55a, p50a), PIK3R2 (p85b), PIK3R3 (p55c) [Wymann, Curr Opin Cell Biol 2005] - PIK3CG is class IB member; interacts with PIK3R5 (p101) and PIK adapter protein (p87). [Wymann, Curr Opin Cell Biol 2005] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Vanhaesebroeck, PNAS 19997] - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - Pancreas, skeletal muscle, liver and heart [UniProt]
PI45P3K	3	Stoyanov B, Volinia S, Hanck T, Rubio I, Loubtchenkov M, Malek D, Stoyanova S, Vanhaesebroeck B, Dhand R, Nurnberg B, et al	Cloning and characterization of a G protein-activated human phosphoinositide-3 kinase	Science	1995	7624799	- rat protein is heterodimer of an 85 kd regulatory subunit that mediates binding to phosphorylated proteins and a 110 kd catalytic subunit [Carpenter, J Biol Chem 1990] - 3 known class IA catalytic subunits: PIK3CA (p110a), PIK3CB (p110b), PIK3CD (p110c) which form complexes with 85 kDa regulatory subunits: PIK3R1 (p85a, p55a, p50a), PIK3R2 (p85b), PIK3R3 (p55c) [Wymann, Curr Opin Cell Biol 2005] - PIK3CG is class IB member; interacts with PIK3R5 (p101) and PIK adapter protein (p87). [Wymann, Curr Opin Cell Biol 2005] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Vanhaesebroeck, PNAS 19997] - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - Pancreas, skeletal muscle, liver and heart [UniProt]
PI45P3K	3	Volinia S, Hiles I, Ormondroyd E, Nizetic D, Antonacci R, Rocchi M, Waterfield MD	Molecular cloning, cDNA sequence, and chromosomal localization of the human phosphatidylinositol 3-kinase p110 alpha (PIK3CA) gene	Genomics	1994	7713498	- rat protein is heterodimer of an 85 kd regulatory subunit that mediates binding to phosphorylated proteins and a 110 kd catalytic subunit [Carpenter, J Biol Chem 1990] - 3 known class IA catalytic subunits: PIK3CA (p110a), PIK3CB (p110b), PIK3CD (p110c) which form complexes with 85 kDa regulatory subunits: PIK3R1 (p85a, p55a, p50a), PIK3R2 (p85b), PIK3R3 (p55c) [Wymann, Curr Opin Cell Biol 2005] - PIK3CG is class IB member; interacts with PIK3R5 (p101) and PIK adapter protein (p87). [Wymann, Curr Opin Cell Biol 2005] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Vanhaesebroeck, PNAS 19997] - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - Pancreas, skeletal muscle, liver and heart [UniProt]

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PI45P3K	3	Hu P, Mondino A, Skolnik EY, Schlessinger J	Cloning of a novel, ubiquitously expressed human phosphatidylinositol 3-kinase and identification of its binding site on p85	Mol Cell Biol	1993	8246984	- fat protein is heterodimer of an 85 kd regulatory subunit that mediates binding to phosphorylated proteins and a 110 kd catalytic subunit [Carpenter, J Biol Chem 1990] - 3 known class IA catalytic subunits: PIK3CA (p110a), PIK3CB (p110b), PIK3CD (p110c) which form complexes with 85 kDa regulatory subunits: PIK3R1 (p85a, p55a, p50a), PIK3R2 (p85b), PIK3R3 (p55c) [Wymann, Curr Opin Cell Biol 2005] - PIK3CG is class IB member; interacts with PIK3R5 (p101) and PIK adapter protein (p87). [Wymann, Curr Opin Cell Biol 2005] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Vanhaesebroeck, PNAS 19997] - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - Pancreas, skeletal muscle, liver and heart [UniProt]
PI45P3K	3	Vanhaesebroeck B, Welham MJ, Kotani K, Stein R, Warne PH, Zvelebil MJ, Higashi K, Volinia S, Downward J, Waterfield MD	P110delta, a novel phosphoinositide 3-kinase in leukocytes	Proc Natl Acad Sci U S A	1997	9113989	- fat protein is heterodimer of an 85 kd regulatory subunit that mediates binding to phosphorylated proteins and a 110 kd catalytic subunit [Carpenter, J Biol Chem 1990] - 3 known class IA catalytic subunits: PIK3CA (p110a), PIK3CB (p110b), PIK3CD (p110c) which form complexes with 85 kDa regulatory subunits: PIK3R1 (p85a, p55a, p50a), PIK3R2 (p85b), PIK3R3 (p55c) [Wymann, Curr Opin Cell Biol 2005] - PIK3CG is class IB member; interacts with PIK3R5 (p101) and PIK adapter protein (p87). [Wymann, Curr Opin Cell Biol 2005] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Vanhaesebroeck, PNAS 19997] - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - Pancreas, skeletal muscle, liver and heart [UniProt]
PI45P3K	3	Brock C, Schaefer M, Reusch HP, Czupalla C, Michalke M, Spicher K, Schultz G, Nurnberg B	Roles of G beta gamma in membrane recruitment and activation of p110 gamma/p101 phosphoinositide 3-kinase gamma.	J Cell Biol	2003	12507995	- fat protein is heterodimer of an 85 kd regulatory subunit that mediates binding to phosphorylated proteins and a 110 kd catalytic subunit [Carpenter, J Biol Chem 1990] - 3 known class IA catalytic subunits: PIK3CA (p110a), PIK3CB (p110b), PIK3CD (p110c) which form complexes with 85 kDa regulatory subunits: PIK3R1 (p85a, p55a, p50a), PIK3R2 (p85b), PIK3R3 (p55c) [Wymann, Curr Opin Cell Biol 2005] - PIK3CG is class IB member; interacts with PIK3R5 (p101) and PIK adapter protein (p87). [Wymann, Curr Opin Cell Biol 2005] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Vanhaesebroeck, PNAS 19997] - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - Pancreas, skeletal muscle, liver and heart [UniProt]

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PI45P3K	3	Wymann MP, Marone R	Phosphoinositide 3-kinase in disease: timing, location, and scaffolding	Curr Opin Cell Biol	2005	15780590	- fat protein is heterodimer of an 85 kd regulatory subunit that mediates binding to phosphorylated proteins and a 110 kd catalytic subunit [Carpenter, J Biol Chem 1990] - 3 known class IA catalytic subunits: PIK3CA (p110a), PIK3CB (p110b), PIK3CD (p110c) which form complexes with 85 kDa regulatory subunits: PIK3R1 (p85a, p55a, p50a), PIK3R2 (p85b), PIK3R3 (p55c) [Wymann, Curr Opin Cell Biol 2005] - PIK3CG is class IB member; interacts with PIK3R5 (p101) and PIK adapter protein (p87). [Wymann, Curr Opin Cell Biol 2005] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned: 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Vanhaesebroeck, PNAS 19997] - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - Pancreas, skeletal muscle, liver and heart [UniProt]
PI45P5P	3	Jefferson AB, Majerus PW	Properties of type II inositol polyphosphate 5-phosphatase	J Biol Chem	1995	7721860	8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - PtdIns4P 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004] 4952: - phospatidylinositol polyphosphate 5-phosphatase activity [RefSeq], [UniProt], [Zhang, PNAS 1995] - golgi cisternae [RefSeq] - Brain, skeletal muscle, heart, kidney, lung, placenta, and fibroblasts [UniProt] - associates w/ Rac GTPase in trans-Golgi (assume this occurs at cytosolic surface of outer membrane) [Faucherre, Hum Mol Genet 2003] 8871: - PtdIns4P 5-phosphatase activity [UniProt], [Spaenij-Dekking, Leukemia 2003] - cytosolic and synaptic nerve termini [Malecz, Curr Biol 2000] 3633: - PtdIns4P 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 56623: - PtdIns4P 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Gc - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva,

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PI45P5P	3	Zhang X, Jefferson AB, Auethavekiat V, Majerus PW	The protein deficient in Lowe syndrome is a phosphatidylinositol-4,5-bisphosphate 5-phosphatase	Proc Natl Acad Sci U S A	1995	7761412	8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - PtdIns4P 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004] 4952: - phospatidylinositol polyphosphate 5-phosphatase activity [RefSeq], [UniProt], [Zhang, PNAS 1995] - golgi cisternae [RefSeq] - Brain, skeletal muscle, heart, kidney, lung, placenta, and fibroblasts [UniProt] - associates w/ Rac GTPase in trans-Golgi (assume this occurs at cytosolic surface of outer membrane) [Faucherre, Hum Mol Genet 2003] 8871: - PtdIns4P 5-phosphatase activity [UniProt], [Spaenij-Dekking, Leukemia 2003] - cytosolic and synaptic nerve termini [Malecz, Curr Biol 2000] 3633: - PtdIns4P 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 56623: - PtdIns4P 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Gc - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva,
PI45P5P	3	Kisseleva MV, Wilson MP, Majerus PW	The isolation and characterization of a cDNA encoding phospholipid-specific inositol polyphosphate 5-phosphatase	J Biol Chem	2000	10764818	8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - PtdIns4P 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004] 4952: - phospatidylinositol polyphosphate 5-phosphatase activity [RefSeq], [UniProt], [Zhang, PNAS 1995] - golgi cisternae [RefSeq] - Brain, skeletal muscle, heart, kidney, lung, placenta, and fibroblasts [UniProt] - associates w/ Rac GTPase in trans-Golgi (assume this occurs at cytosolic surface of outer membrane) [Faucherre, Hum Mol Genet 2003] 8871: - PtdIns4P 5-phosphatase activity [UniProt], [Spaenij-Dekking, Leukemia 2003] - cytosolic and synaptic nerve termini [Malecz, Curr Biol 2000] 3633: - PtdIns4P 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 56623: - PtdIns4P 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Gc - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva,

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PI45P5P	3	Faucherre A, Desbois P, Satre V, Lunardi J, Dorseuil O, Gacon G	Lowe syndrome protein OCRL1 interacts with Rac GTPase in the trans-Golgi network.	Hum Mol Genet	2003	12915445	8867: - localizes to clathrin-coated invaginations of synaptic nerve terminals [Haffner, FEBS Lett 1997], [UniProt] - Isoform 1 is more enriched than isoform 2 in developing brain as well as non-neuronal cells. Isoform 2 is very abundant in nerve terminals [UniProt] - PtdIns4P 5-phosphatase activity [UniProt], [Stopkova, Psychiatry Res 2004] 4952: - phospatidylinositol polyphosphate 5-phosphatase activity [RefSeq], [UniProt], [Zhang, PNAS 1995] - golgi cisternae [RefSeq] - Brain, skeletal muscle, heart, kidney, lung, placenta, and fibroblasts [UniProt] - associates w/ Rac GTPase in trans-Golgi (assume this occurs at cytosolic surface of outer membrane) [Faucherre, Hum Mol Genet 2003] 8871: - PtdIns4P 5-phosphatase activity [UniProt], [Spaenij-Dekking, Leukemia 2003] - cytosolic and synaptic nerve termini [Malecz, Curr Biol 2000] 3633: - PtdIns4P 5-phosphatase activity [UniProt], [Jefferson, J Biol Chem 1995] - found in platelets [UniProt] 56623: - PtdIns4P 5-phosphatase activity [UniProt], [Kisseleva, J Biol Chem 200] - Cytoplasmic; peripheral membrane protein associated with Gc - brain, heart, pancreas, testis and spleen [UniProt], [Kisseleva,
PI45PLC	3	Park D, Jhon DY, Kriz R, Knopf J, Rhee SG	Cloning, sequencing, expression, and Gq-independent activation of phospholipase C-beta 2	J Biol Chem	1992	1644792	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Burgess WH, Dionne CA, Kaplow J, Mudd R, Friesel R, Zilberstein A, Schlessinger J, Jaye M	Characterization and cDNA cloning of phospholipase C-gamma, a major substrate for heparin-binding growth factor 1 (acidic fibroblast growth factor)-activated tyrosine kinase	Mol Cell Biol	1990	2167438	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Lagercrantz J, Carson E, Phelan C, Grimmond S, Rosen A, Dare E, Nordenskjold M, Hayward NK, Larsson C, Weber G	Genomic organization and complete cDNA sequence of the human phosphoinositide-specific phospholipase C beta 3 gene (PLCB3)	Genomics	1995	7607669	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Mazuruk K, Schoen TJ, Chader GJ, Rodriguez IR	Structural organization and expression of the human phosphatidylinositol-specific phospholipase C beta-3 gene	Biochem Biophys Res Commun	1995	7612006	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Kohno T, Otsuka T, Takano H, Yamamoto T, Hamaguchi M, Terada M, Yokota J	Identification of a novel phospholipase C family gene at chromosome 2q33 that is homozygously deleted in human small cell lung carcinoma	Hum Mol Genet	1995	7633416	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Sinke RJ, Geurts van Kessel AG	Localization of the human phosphatidylinositol-specific phospholipase c beta 3 gene (PLCB3) within chromosome band 11q13	Genoics	1995	7789993	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Hernandez D, Egan SE, Yulug IG, Fisher EM	Mapping the gene that encodes phosphatidylinositol-specific phospholipase C-gamma 2 in the human and the mouse	Genomics	1994	7835906	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Weber G, Friedman E, Grimmond S, Hayward NK, Phelan C, Skogseid B, Gobl A, Zedenius J, Sandelin K, Teh BT, et al.	The phospholipase C beta 3 gene located in the MEN1 region shows loss of expression in endocrine tumours	Hum Mol Genet	1994	7849701	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Cheng HF, Jiang MJ, Chen CL, Liu SM, Wong LP, Lomasney JW, King K	Cloning and identification of amino acid residues of human phospholipase C delta 1 essential for catalysis	J Biol Chem	1995	7890667	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Alvarez RA, Ghalayini AJ, Xu P, Hardcastle A, Bhattacharya S, Rao PN, Pettenati MJ, Anderson RE, Baehr W.	cDNA sequence and gene locus of the human retinal phosphoinositide-specific phospholipase-C beta 4 (PLCB4)	Genomics	1995	8530101	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Ishikawa S, Takahashi T, Ogawa M, Nakamura Y	Genomic structure of the human PLCD1 (phospholipase C delta 1) locus on 3p22-->p21.3	Cytogenet Cell Genet	1997	9345909	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Kim H, Suh PG, Ryu SH, Park SH	Assignment of the human PLC delta3 gene (PLCD3) to human chromosome band 17q21 by fluorescence in situ hybridization	Cytogenet Cell Genet	1999	10702670	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Kim H, Suh PG, Ryu SH, Park SH	Assignment of the human PLC delta4 gene (PLCD4) to human chromosome band 2q35 by fluorescence in situ hybridization	Cytogenet Cell Genet	1999	10702683	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Peruzzi D, Calabrese G, Faenza I, Manzoli L, Matteucci A, Gianfrancesco F, Billi AM, Stuppia L, Palka G, Cocco L	Identification and chromosomal localisation by fluorescence in situ hybridisation of human gene of phosphoinositide-specific phospholipase C beta(1)	Biochim Biophy Acta	2000	10760467	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Lopez I, Mak EC, Ding J, Hamm HE, Lomasney JW	A novel bifunctional phospholipase c that is regulated by Galpha 12 and stimulates the Ras/mitogen-activated protein kinase pathway	J Biol Chem	2001	11022047	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Song C, Hu CD, Masago M, Kariyai K, Yamawaki-Kataoka Y, Shibatohge M, Wu D, Satoh T, Kataoka T	Regulation of a novel human phospholipase C, PLCepsilon, through membrane targeting by Ras	J Biol Chem	2001	11022048	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Caricasole A, Sala C, Roncarati R, Formenti E, Terstappen GC	Cloning and characterization of the human phosphoinositide-specific phospholipase C-beta 1 (PLC beta 1)	Biochim Biophys Acta	2000	11118617	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
PI45PLC	3	Saunders CM, Larman MG, Parrington J, Cox LJ, Royse J, Blayney LM, Swann K, Lai FA	PLC zeta: a sperm-specific trigger of Ca(2+) oscillations in eggs and embryo development	Development	2002	12117804	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002] - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Peruzzi D, Aluigi M, Manzoli L, Billi AM, Di Giorgio FP, Morleo M, Martelli AM, Cocco L	Molecular characterization of the human PLC beta1 gene	Biochim Biophys Acta	2002	12213492	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002] - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Cox LJ, Larman MG, Saunders CM, Hashimoto K, Swann K, Lai FA	Sperm phospholipase Czeta from humans and cynomolgus monkeys triggers Ca2+ oscillations, activation and development of mouse oocytes	Reproduction	2002	12416999	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLC	3	Carozzi AJ, Kriz RW, Webster C, Parker PJ	Identification, purification and characterization of a novel phosphatidylinositol-specific phospholipase C, a third member of the beta subfamily	Eur J Biochem	1992	13333955	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]

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PI45PLC	3	Leung DW, Tompkins C, Brewer J, Ball A, Coon M, Morris V, Waggoner D, Singer JW	Phospholipase C delta-4 overexpression upregulates ErbB1/2 expression, Erk signaling pathway, and proliferation in MCF-7 cells	Mol Cancer	2004	15140260	substrates [Leung, Mol Cancer 2004] - PLC family is composed of 11 subtypes: ß1, 2, 3 and 4, ¿ 1 and 2, d1, 3 and 4, e, and ¿ [Leung, Mol Cancer 2004] 84812: - gene was cloned [Kim, Cytogenet Cell Genet 1999] 5333: - gene was cloned [Ishikawa, Cytogenet Cell Genet 1997] - expression and biochemical characterization [Cheng, J Biol Chem 1995] 23007: - function inferred from electronic annotation [GO] 51196: - gene was cloned and characterized [Song, J Biol Chem 2001], [Lopez, J Biol Chem 2001] 5336: - gene was mapped [Hernandez, Genomics 1994] 5332: - predominantly expressed in retina [UniProt], [Alvarez, Genomics 1995] - gene was cloned and compared to mammalian homologs [Alvarez, Genomics 1995] 26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] 89869: - gene was cloned and expressed [Saunders, Development 2002 - sperm-specific [Saunders, Development 2002]
PI45PLCn	3	Irvine RF	Nuclear lipid signalling	Nat Rev Mol Cell Biol	2003	12728269	26236: - gene was identified, cloned and expressed [Peruzzi, Biochim Biophys Acta 2002], [Caricasole, Biochim Biophys Acta 2000], [Peruzzi, Biochim Biophys Acta 2000] - nuclear [Peruzzi, Biochim Biophys Acta 2000] and cytosolic [Caricasole, Biochim Biophys Acta 2000] - see fig1 of [Irvine 2003]
PI4P5K	3	Burriss Garrett RJ, Redman CM	Localization of enzymes involved in polyphosphoinositids metabolism on the cytoplasmic surface of the human erythrocyte membrane	Biochim Biophys Acta	1975	164238	cytoplasm - by default Particularly abundant in platelets and in brain. Present in most tissues, except notably skeletal muscle and small intestine. Catalyzes the phosphorylation of phosphatidylinositol-4-phosphate on the fifth hydroxyl of the myo-inositol ring, to form phosphatidylinositol-4,5-biphosphate. NJ - present on the cytosolic surface of human erythrocyte membranes [Burriss Garrett, Biochim Biophys Acta 1975] 5305: - catalyzes PtdIns4P 5-kinase rxn [RefSeq], [UniProt] - expressed ubiquitously, with high levels in the brain [UniProt] - homolgy to S. cerevisiae proteins Fab1p and Mss4p [Boronenkov, J Biol Chem 1995] - protein expressed in E. coli [Boronenkov, J Biol Chem 1995] 8394: - gene has been cloned, expressed, and characterized [Loijens, J Biol Chem 1996] 8395: - gene has been cloned and shown to have PtdIns4P 5-kinase activity [Carvajal, Nat Genet 1996] 8396: - catalyzes PtdIns4P 5-kinase rxn [RefSeq], [UniProt] - Cytoplasmic. Peripheral membrane protein associated with the plasma membrane and the endoplasmic reticulum (By similarity). [UniProt] - Highly expressed in brain, heart, pancreas, skeletal muscle and

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
PI4P5K	3	Boronenkov IV, Anderson RA	The sequence of phosphatidylinositol-4-phosphate 5-kinase defines a novel family of lipid kinases	J Biol Chem	1995	7852364	cytoplasm - by default Particularly abundant in platelets and in brain. Present in most tissues, except notably skeletal muscle and small intestine. Catalyzes the phosphorylation of phosphatidylinositol-4-phosphate on the fifth hydroxyl of the myo-inositol ring, to form phosphatidylinositol-4,5-biphosphate. NJ - present on the cytosolic surface of human erythrocyte membranes [Burriss Garrett, Biochim Biophys Acta 1975] 5305: - catalyzes PtdIns4P 5-kinase rxn [RefSeq], [UniProt] - expressed ubiquitously, with high levels in the brain [UniProt] - homolgy to S. cerevisiae proteins Fab1p and Mss4p [Boronenkov, J Biol Chem 1995] - protein expressed in E. coli [Boronenkov, J Biol Chem 1995] 8394: - gene has been cloned, expressed, and characterized [Loijens, J Biol Chem 1996] 8395: - gene has been cloned and shown to have PtdIns4P 5-kinase activity [Carvajal, Nat Genet 1996] 8396: - catalyzes PtdIns4P 5-kinase rxn [RefSeq], [UniProt] - Cytoplasmic. Peripheral membrane protein associated with the plasma membrane and the endoplasmic reticulum (By similarity). [UniProt] - Highly expressed in brain, heart, pancreas, skeletal muscle and
PI4P5K	3	Carvajal JJ, Pook MA, dos Santos M, Doudney K, Hillermann R, Minogue S, Williamson R, Hsuan JJ, Chamberlain S	The Friedreich's ataxia gene encodes a novel phosphatidylinositol-4- phosphate 5-kinase	Nat Genet	1996	8841185	cytoplasm - by default Particularly abundant in platelets and in brain. Present in most tissues, except notably skeletal muscle and small intestine. Catalyzes the phosphorylation of phosphatidylinositol-4-phosphate on the fifth hydroxyl of the myo-inositol ring, to form phosphatidylinositol-4,5-biphosphate. NJ - present on the cytosolic surface of human erythrocyte membranes [Burriss Garrett, Biochim Biophys Acta 1975] 5305: - catalyzes PtdIns4P 5-kinase rxn [RefSeq], [UniProt] - expressed ubiquitously, with high levels in the brain [UniProt] - homolgy to S. cerevisiae proteins Fab1p and Mss4p [Boronenkov, J Biol Chem 1995] - protein expressed in E. coli [Boronenkov, J Biol Chem 1995] 8394: - gene has been cloned, expressed, and characterized [Loijens, J Biol Chem 1996] 8395: - gene has been cloned and shown to have PtdIns4P 5-kinase activity [Carvajal, Nat Genet 1996] 8396: - catalyzes PtdIns4P 5-kinase rxn [RefSeq], [UniProt] - Cytoplasmic. Peripheral membrane protein associated with the plasma membrane and the endoplasmic reticulum (By similarity). [UniProt] - Highly expressed in brain, heart, pancreas, skeletal muscle and

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PI4P5K	3	Loijens JC, Anderson RA	Type I phosphatidylinositol-4-phosphate 5-kinases are distinct members of this novel lipid kinase family	J Biol Chem	1996	8955136	cytoplasm - by default Particularly abundant in platelets and in brain. Present in most tissues, except notably skeletal muscle and small intestine. Catalyzes the phosphorylation of phosphatidylinositol-4-phosphate on the fifth hydroxyl of the myo-inositol ring, to form phosphatidylinositol-4,5-biphosphate. NJ - present on the cytosolic surface of human erythrocyte membranes [Burriss Garrett, Biochim Biophys Acta 1975] 5305: - catalyzes PtdIns4P 5-kinase rxn [RefSeq], [UniProt] - expressed ubiquitously, with high levels in the brain [UniProt] - homolgy to S. cerevisiae proteins Fab1p and Mss4p [Boronenkov, J Biol Chem 1995] - protein expressed in E. coli [Boronenkov, J Biol Chem 1995] 8394: - gene has been cloned, expressed, and characterized [Loijens, J Biol Chem 1996] 8395: - gene has been cloned and shown to have PtdIns4P 5-kinase activity [Carvajal, Nat Genet 1996] 8396: - catalyzes PtdIns4P 5-kinase rxn [RefSeq], [UniProt] - Cytoplasmic. Peripheral membrane protein associated with the plasma membrane and the endoplasmic reticulum (By similarity). [UniProt] - Highly expressed in brain, heart, pancreas, skeletal muscle and
PI4PP	2	Holub BJ	Metabolism and function of myo-inositol and inositol phospholipids	Annu Rev Nutr	1986	2425833	- reaction described in [Holub, Ann Rev Nutr 1986]
PI5P4K	3	Zhang X, Loijens JC, Boronenkov IV, Parker GJ, Norris FA, Chen J, Thum O, Prestwich GD, Majerus PW, Anderson RA	Phosphatidylinositol-4-phosphate 5-kinase isozymes catalyze the synthesis of 3-phosphate-containing phosphatidylinositol signaling molecules	J Biol Chem	1997	9211928	- type II a and b PIP4Ks have much higher 4-kinase activity for or PtdIns5P than PtdIns3P in vitro [Rameh, Nature 1997], [Zhang, J Biol Chem 1997] - reaction described in [Tolias, Chem Phys Lipids 1999]
PI5P4K	3	Rameh LE, Tolias KF, Duckworth BC, Cantley LC	A new pathway for synthesis of phosphatidylinositol-4,5-bisphosphate	Nature	1997	9367159	- type II a and b PIP4Ks have much higher 4-kinase activity for or PtdIns5P than PtdIns3P in vitro [Rameh, Nature 1997], [Zhang, J Biol Chem 1997] - reaction described in [Tolias, Chem Phys Lipids 1999]
PI5P4K	3	Tolias KF, Cantley LC	Pathways for phosphoinositide synthesis	Chem Phys Lipids	1999	10358929	- type II a and b PIP4Ks have much higher 4-kinase activity for or PtdIns5P than PtdIns3P in vitro [Rameh, Nature 1997], [Zhang, J Biol Chem 1997] - reaction described in [Tolias, Chem Phys Lipids 1999]
PIACGT	3	Miyata T, Takeda J, Iida Y, Yamada N, Inoue N, Takahashi M, Maeda K, Kitani T, Kinoshita T	The cloning of PIG-A, a component in the early step of GPI-anchor biosynthesis	Science	1993	7680492	9322 (PIGF): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):

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PIACGT	3	Iida Y, Takeda J, Miyata T, Inoue N, Nishimura J, Kitani T, Maeda K, Kinoshita T	Characterization of genomic PIG-A gene: a gene for glycosylphosphatidylinositol-anchor biosynthesis and paroxysmal nocturnal hemoglobinuria	Blood	1994	8193350	51227 (PIGP): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):
PIACGT	3	Kamitani T, Chang HM, Rollins C, Waneck GL, Yeh ET.	Correction of the class H defect in glycosylphosphatidylinositol anchor biosynthesis in Ltk- cells by a human cDNA clone	J Biol Chem	1993	8407896	51227 (PIGP): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):
PIACGT	3	Inoue N, Watanabe R, Takeda J, Kinoshita T	PIG-C, one of the three human genes involved in the first step of glycosylphosphatidylinositol biosynthesis is a homologue of Saccharomyces cerevisiae GPI2	Biochem Biophys Res Commun	1996	8806613	51227 (PIGP): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):

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PIACGT	3	Hong Y, Ohishi K, Inoue N, Endo Y, Fujita T, Takeda J, Kinoshita T	Structures and chromosomal localizations of the glycosylphosphatidylinositol synthesis gene PIGC and its pseudogene PIGCP1	Genomics	1997	9325057	51227 (PIGF): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):
PIACGT	3	Watanabe R, Inoue N, Westfall B, Taron CH, Orlean P, Takeda J, Kinoshita T	The first step of glycosylphosphatidylinositol biosynthesis is mediated by a complex of PIG-A, PIG-H, PIG-C and GPI1	EMBO J	1998	9463366	51227 (PIGF): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):
PIACGT	3	Shibuya K, Kudoh J, Minoshima S, Kawasaki K, Asakawa S, Shimizu N	Isolation of two novel genes, DSCR5 and DSCR6, from Down syndrome critical region on human chromosome 21q22.2	Biochem Biophys Res Commun	2000	10814524	51227 (PIGF): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):

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PIACGT	3	Togashi T, Choi DK, Taylor TD, Suzuki Y, Sugano S, Hattori M, Sakaki Y	A novel gene, DSCR5, from the distal Down syndrome critical region on chromosome 21q22.2	DNA Res	2000	10907851	51227 (PIGP): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):
PIACGT	3	Choi DK, Suzuki Y, Yoshimura S, Togashi T, Hida M, Taylor TD, Wang Y, Sugano S, Hattori M, Sakaki Y	Molecular cloning and characterization of a gene expressed in mouse developing tongue, mDscr5 gene, a homolog of human DSCR5 (Down syndrome Critical Region gene 5)	Mamm Genome	2001	11331941	51227 (PIGP): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):
PIACGT	3	Tiede A, Daniels RJ, Higgs DR, Mehrein Y, Schmidt RE, Schubert J	The human GPI1 gene is required for efficient glycosylphosphatidylinositol biosynthesis	Gene	2001	11418246	51227 (PIGP): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):

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PIACGT	3	Eisenhaber B, Maurer-Stroh S, Novatchkova M, Schneider G, Eisenhaber F	Enzymes and auxiliary factors for GPI lipid anchor biosynthesis and post-translational transfer to proteins	Bioesssays	2003	12655644	5122 (PIGP): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - ubiquitous [UniProt], [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - gene was cloned [Shibuya, Biochem Biophys Res Commun 2000], [Togashi, DNA Res 2000] - mouse homolog has been cloned and characterized [Choi, Mamm Genome 2001] 5277 (PIGA): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene was cloned [Miyata, Science 1993] and characterized [Iida, Blood 1994] 5279 (PIGC): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - gene has 20% similarity w/ yeast homolog [Inoue, Biochem Biophys Res Commun 1996] - gene was isolated [Hong, Genomics 1997] 5283 (PIGH): - component of phosphatidylinositol N-acetylglucosaminyltransferase complex [RefSeq], [UniProt], [Eisenhaber, Bioessays 2003] - cloning and expression [Kamitami, J Biol Chem 1993] 9091 (PIGQ):
PIK3	3	Volinia S, Dhand R, Vanhaesebroeck B, MacDougall LK, Stein R, Zvelebil MJ, Domin J, Panaretou C, Waterfield MD	A human phosphatidylinositol 3-kinase complex related to the yeast Vps34p-Vps15p protein sorting system	EMBO J	1995	7628435	5286: - phosphoinositide 3-kinase activity [RefSeq] - acts on PtdIns and PtdIns4P [Domin, Biochem J 1997] - gene has been cloned [Domin, Biochem J 1997] 5287: - Found mostly in the microsome, but also in the plasma membrane and cytosol [UniProt] - gene was cloned and expressed [Arcaro, J Biol Chem 1998] - acts on PtdIns and PtdIns4P [Arcaro, J Biol Chem 1998] 5288: - acts on PtdIns and PtdIns4P [UniProt], [Rozycka, Genomics 1998] - gene has been cloned [Rozycka, Genomics 1998] - Highly expressed in liver, prostate and testis. Lower levels in small intestine, kidney and pancreas.[UniProt] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
PIK3	3	Domin J, Pages F, Volinia S, Rittenhouse SE, Zvelebil MJ, Stein RC, Waterfield MD	Cloning of a human phosphoinositide 3-kinase with a C2 domain that displays reduced sensitivity to the inhibitor wortmannin	Biochem J	1997	9337861	5286: - phosphoinositide 3-kinase activity [RefSeq] - acts on PtdIns and PtdIns4P [Domin, Biochem J 1997] - gene has been cloned [Domin, Biochem J 1997] 5287: - Found mostly in the microsome, but also in the plasma membrane and cytosol [UniProt] - gene was cloned and expressed [Arcaro, J Biol Chem 1998] - acts on PtdIns and PtdIns4P [Arcaro, J Biol Chem 1998] 5288: - acts on PtdIns and PtdIns4P [UniProt], [Rozycka, Genomics 1998] - gene has been cloned [Rozycka, Genomics 1998] - Highly expressed in liver, prostate and testis. Lower levels in small intestine, kidney and pancreas.[UniProt] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt
PIK3	3	Arcaro A, Volinia S, Zvelebil MJ, Stein R, Watton SJ, Layton MJ, Gout I, Ahmadi K, Downward J, Waterfield MD	Human phosphoinositide 3-kinase C2beta, the role of calcium and the C2 domain in enzyme activity	J Biol Chem	1998	9830063	5286: - phosphoinositide 3-kinase activity [RefSeq] - acts on PtdIns and PtdIns4P [Domin, Biochem J 1997] - gene has been cloned [Domin, Biochem J 1997] 5287: - Found mostly in the microsome, but also in the plasma membrane and cytosol [UniProt] - gene was cloned and expressed [Arcaro, J Biol Chem 1998] - acts on PtdIns and PtdIns4P [Arcaro, J Biol Chem 1998] 5288: - acts on PtdIns and PtdIns4P [UniProt], [Rozycka, Genomics 1998] - gene has been cloned [Rozycka, Genomics 1998] - Highly expressed in liver, prostate and testis. Lower levels in small intestine, kidney and pancreas.[UniProt] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt

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PIK3	3	Rozycka M, Lu YJ, Brown RA, Lau MR, Shipley JM, Fry MJ	cDNA cloning of a third human C2-domain-containing class II phosphoinositide 3-kinase, PI3K-C2gamma, and chromosomal assignment of this gene (PIK3C2G) to 12p12	Genomics	1998	9878262	5286: - phosphoinositide 3-kinase activity [RefSeq] - acts on PtdIns and PtdIns4P [Domin, Biochem J 1997] - gene has been cloned [Domin, Biochem J 1997] 5287: - Found mostly in the microsome, but also in the plasma membrane and cytosol [UniProt] - gene was cloned and expressed [Arcaro, J Biol Chem 1998] - acts on PtdIns and PtdIns4P [Arcaro, J Biol Chem 1998] 5288: - acts on PtdIns and PtdIns4P [UniProt], [Rozycka, Genomics 1998] - gene has been cloned [Rozycka, Genomics 1998] - Highly expressed in liver, prostate and testis. Lower levels in small intestine, kidney and pancreas.[UniProt] 5290: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [RefSeq], [UniProt] - gene has been cloned; 99% identical to bovine protein [Volinia, Genomics 1994] 5291: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, PtdIns(4,5)P2 [UniProt], [Hu, Mol Cell Biol 1993] - expressed ubiquitously [UniProt], [Hu, Mol Cell Biol 1993] - 42% identical to bovine PI 3-kinase and 28% identical to S. cerevisiae Vps34 [Hu, Mol Cell Biol 1993] 5293: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt - leukocytes [UniProt], [Vanhaesebroeck, PNAS 19997] - gene has been cloned, characterized [Vanhaesebroeck, PNAS 5294: - catalytic subunit of PIK3, phosphorylates PtdIns, PtdIns4P, Pt
PIK4	3	Nakagawa T, Goto K, Kondo H	Cloning, expression, and localization of 230-kDa phosphatidylinositol 4-kinase	J Biol Chem	1996	8662589	cytoplasmic - uniprot Expressed ubiquitously. Highest levels in placenta and brain. Little or no expression in lung, liver, pancreas, testis or leukocytes. PI4KII, PI4K2B: no separte reaction explicitly associated w/ it -> assumed to have same PIK4 NJ 5297: - rat protein was found to be closely assoc w/ Golgi vesicles and vacuoles [Nakagawa, J Biol Chem 1996] - present on the cytosolic surface of human erythrocyte membranes [Burriss Garrett, Biochim Biophys Acta 1975] 5298: - enzyme is primarily cytosolic; associates peripherally with plasma membranes, endoplasmic reticulum, and Golgi [Wei, J Biol Chem 2002]
PIK4	3	Wei YJ, Sun HQ, Yamamoto M, Wlodarski P, Kunii K, Martinez M, Barylko B, Albanesi JP, Yin HL.	ype II phosphatidylinositol 4-kinase beta is a cytosolic and peripheral membrane protein that is recruited to the plasma membrane and activated by Rac-GTP	J Biol Chem	2002	12324459	cytoplasmic - uniprot Expressed ubiquitously. Highest levels in placenta and brain. Little or no expression in lung, liver, pancreas, testis or leukocytes. PI4KII, PI4K2B: no separte reaction explicitly associated w/ it -> assumed to have same PIK4 NJ 5297: - rat protein was found to be closely assoc w/ Golgi vesicles and vacuoles [Nakagawa, J Biol Chem 1996] - present on the cytosolic surface of human erythrocyte membranes [Burriss Garrett, Biochim Biophys Acta 1975] 5298: - enzyme is primarily cytosolic; associates peripherally with plasma membranes, endoplasmic reticulum, and Golgi [Wei, J Biol Chem 2002]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
PIK4	3	Minogue S, Anderson JS, Waugh MG, dosSantos M, Corless S, Cramer R, Hsuan JJ	Cloning of a human type II phosphatidylinositol 4-kinase reveals a novel lipid kinase family	Journal of Biological Chemistry	2001		cytoplasmic - uniprot Expressed ubiquitously. Highest levels in placenta and brain. Little or no expression in lung, liver, pancreas, testis or leukocytes. PI4KII, PI4K2B: no separte reaction explicitly associated w/ it -> assumed to have same PIK4 NJ 5297: - rat protein was found to be closely assoc w/ Golgi vesicles and vacuoles [Nakagawa, J Biol Chem 1996] - present on the cytosolic surface of human erythrocyte membranes [Burriss Garrett, Biochim Biophys Acta 1975] 5298: - enzyme is primarily cytosolic; associates peripherally with plasma membranes, endoplasmic reticulum, and Golgi [Wei, J Biol Chem 2002]
PIK4	3	Wong K, Cantley LC	Cloning and characterization of a human phosphatidylinositol 4-kinase	Journal of Biological Chemsitry	1994		cytoplasmic - uniprot Expressed ubiquitously. Highest levels in placenta and brain. Little or no expression in lung, liver, pancreas, testis or leukocytes. PI4KII, PI4K2B: no separte reaction explicitly associated w/ it -> assumed to have same PIK4 NJ 5297: - rat protein was found to be closely assoc w/ Golgi vesicles and vacuoles [Nakagawa, J Biol Chem 1996] - present on the cytosolic surface of human erythrocyte membranes [Burriss Garrett, Biochim Biophys Acta 1975] 5298: - enzyme is primarily cytosolic; associates peripherally with plasma membranes, endoplasmic reticulum, and Golgi [Wei, J Biol Chem 2002]
PIK5	3	Tolias KF, Rameh LE, Ishihara H, Shibasaki Y, Chen J, Prestwich GD, Cantley LC, Carpenter CL	Type I phosphatidylinositol-4-phosphate 5-kinases synthesize the novel lipids phosphatidylinositol 3,5-bisphosphate and phosphatidylinositol 5-phosphate	J Biol Chem	1998	9660759	- pathway for synthesis of PtdIns5P is unclear, but cpd has been detected in mammalian fibroblasts [Tolias, Chem Phys Lipids 1999] - 8394 & 8395 have been shown to catalyze PtdIns -> PtdIns5P rxn in vitro [Tolias, J Biol Chem 1998] - 23396 has not been fully characterized but is inferred to catalyze reaction as well
PIt2m	3	Palmieri, F.	The mitochondrial transporter family (SLC25): physiological and pathological implications.	Pflugers Archive	2004	14598172	- Added by RS/TV Mitochondrial according to Entrez Gene Database Four transcript variants according to RefSeq found on Entrez Gene Database 1) Substrate specificity: Phosphate 2) Transport mechanism: Proton antiport 3) Tissue Localization: Variant 1(isoform a: heart, muscle, skeletal muscle, diaphragm), Variant 2&3(isoform b: liver, kidney, brain, thymus, lung, heart, skeletal muscle, diaphragm), Variant 4 (isoform c: unknown) 4) GPR association as shown 1 through 4 according to Table 1 in Palmieri, F. The mitochondrial transporter family (SLC25): physiological and pathological implications. Pflugers Arch. 2004 Feb. (PMID: 14598172)
PIt7	3	Murer H, Forster I, Biber J.	The sodium phosphate cotransporter family SLC34.		2004	12750889	- A1 and A2 transporters are electrogenic w/ probably 3:1 Na:Pi cotransport MM
PIt7ir	2	Murer H, Biber J	Molecular mechanisms of renal apical Na/phosphate cotransport	Annu Rev Physiol	1996	8815811	The 3:1 ratio Na to Pi is from tentative data gathered for the type II transporter, which is NOT SLC17A1!! The ratio for the type I transporter (SLC17A1-4) is not known. From PMID 8815811: The calculated Hill coefficients of these interactions suggest a 3:l coupling ratio of Na vs Pi. The discrepancy of the coupling ratio of 2 1, as derived from Pi transport studies in brush-border membrane vesicles (40), may be explained by factors such as heterogeneity and electrical properties of the vesicle population. Three other proteins closely related to NPT1 have been identified through genomic analysis, and designated NPT3 (SLC17A2), NPT4 (SLC17A3) and Na+/PO4 cotransporter homologue (SLC17A4) [36, 38]. As of yet, there has been no functional characterization of SLC17A2 4.

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
Plt7ir	2	Reimer RJ, Edwards RH	Organic anion transport is the primary function of the SLC17/type I phosphate transporter family	Pflugers Arch	2004	12811560	The 3:1 ratio Na to Pi is from tentative data gathered for the type II transporter, which is NOT SLC17A1!! The ratio for the type I transporter (SLC17A1-4) is not known. From PMID 8815811: The calculated Hill coefficients of these interactions suggest a 3:l coupling ratio of Na vs Pi. The discrepancy of the coupling ratio of 2 1, as derived from Pi transport studies in brush-border membrane vesicles (40), may be explained by factors such as heterogeneity and electrical properties of the vesicle population Three other proteins closely related to NPT1 have been identified through genomic analysis, and designated NPT3 (SLC17A2), NPT4 (SLC17A3) and Na+/PO4 cotransporter homologue (SLC17A4) [36, 38]. As of yet, there has been no functional characterization of SLC17A2 4.
PIt8	3	Collins JF, Bai L, Ghishan FK.	The SLC20 family of proteins: dual functions as sodium-phosphate cotransporters and viral receptors.		2003	12759754	-paper states that stoichiometry is >1 Na per Pi cotransported, but no other papers specified the actual number. 1.5:1 was used as an estimated average value between 1 and 2 MM
PLA2_2	3	Nimmrich I, Friedl W, Kruse R, Pietsch S, Hentsch S, Deuter R, Winde G, Muller O.	Loss of the PLA2G2A gene in a sporadic colorectal tumor of a patient with a PLA2G2A germline mutation and absence of PLA2G2A germline alterations in patients with FAP.	Hum Genet	1997	9272153	cytoplasm - for group VI by uniport, other variants: by default The protein encoded by this gene is an A2 phospholipase, a class of enzyme that catalyzes the release of fatty acids from phospholipids. The encoded protein may play a role in phospholipid remodelling, arachidonic acid release, leukotriene and prostaglandin synthesis, fas-mediated apoptosis, and transmembrane ion flux in glucose-stimulated B-cells. Several transcript variants encoding multiple isoforms have been described, but the full-length nature of only two of them have been determined to date. group VI variant has been sequenced and described, others have not been identified yet (genetically) PLA2G2A: see PMID: 9272153. Membrane associated. Group II phospholipase A2 is found in many cells and also extracellularly. The membrane-bound and secreted forms are identical and are encoded by a single gene. NJ
PLA2_2	3	Larsson PK, Claesson HE, Kennedy BP.	Multiple splice variants of the human calcium-independent phospholipase A2 and their effect on enzyme activity.	J Biol Chem	1998	9417066	cytoplasm - for group VI by uniport, other variants: by default The protein encoded by this gene is an A2 phospholipase, a class of enzyme that catalyzes the release of fatty acids from phospholipids. The encoded protein may play a role in phospholipid remodelling, arachidonic acid release, leukotriene and prostaglandin synthesis, fas-mediated apoptosis, and transmembrane ion flux in glucose-stimulated B-cells. Several transcript variants encoding multiple isoforms have been described, but the full-length nature of only two of them have been determined to date. group VI variant has been sequenced and described, others have not been identified yet (genetically) PLA2G2A: see PMID: 9272153. Membrane associated. Group II phospholipase A2 is found in many cells and also extracellularly. The membrane-bound and secreted forms are identical and are encoded by a single gene. NJ

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PLA2_2e	3	Nardelli B, Tiffany HL, Bong GW, Yourey PA, Morahan DK, Li Y, Murphy PM, Alderson RF.	Characterization of the signal transduction pathway activated in human monocytes and dendritic cells by MPIF-1, a specific ligand for CC chemokine receptor 1.	J Immunol	1999		secreted enzyme -> extracellular designation: uniprot PA2 catalyzes the calcium-dependent hydrolysis of the 2- acyl groups in 3-sn-phosphoglycerides. Pla2g1b: specificity: pancreas role in signaling pathways, etc (Nardelli ref) Pla2g12a: Abundantly expressed in heart, skeletal muscle, kidney, liver and pancreas. (see Gelb et al ref) Pla2g2e: specificity: Restricted to the brain, heart, lung, and placenta. Promotes stimulus-induced arachidonic acid release and prostaglandin (PG) production similar to those elicited by HSPG-dependent sPLA(2)s, suggesting that this enzyme plays a role in the inflammatory process. see PMID: 10681567. PLA2G2A: see PMID: 9272153. Membrane associated. Group II phospholipase A2 is found in many cells and also extracellularly. The membrane-bound and secreted forms are identical and are encoded by a single gene. PLA2G5: PMID: 8300559. Localization: extracellular because enzyme is secreted (uniprot). Specificity: Heart, placenta and less abundantly, in lung. May be involved in the production of lung surfactant, the remodeling or regulation of cardiac muscle. PLA2G2F: PMID: 11112443. Localization: extracellular - secre PLA2G10: PMID: 9188469 Localization: extracellular - secrete PLA2G2D: PMID: 10455175. Localization: extracellular - secr
PLA2_2e	3	Cupillard L, Koumanov K, Mattei MG, Lazdunski M, Lambeau G.	Cloning, chromosomal mapping, and expression of a novel human secretory phospholipase A2.	J Biol Chem	1997	9188469	secreted enzyme -> extracellular designation: uniprot PA2 catalyzes the calcium-dependent hydrolysis of the 2- acyl groups in 3-sn-phosphoglycerides. Pla2g1b: specificity: pancreas role in signaling pathways, etc (Nardelli ref) Pla2g12a: Abundantly expressed in heart, skeletal muscle, kidney, liver and pancreas. (see Gelb et al ref) Pla2g2e: specificity: Restricted to the brain, heart, lung, and placenta. Promotes stimulus-induced arachidonic acid release and prostaglandin (PG) production similar to those elicited by HSPG-dependent sPLA(2)s, suggesting that this enzyme plays a role in the inflammatory process. see PMID: 10681567. PLA2G2A: see PMID: 9272153. Membrane associated. Group II phospholipase A2 is found in many cells and also extracellularly. The membrane-bound and secreted forms are identical and are encoded by a single gene. PLA2G5: PMID: 8300559. Localization: extracellular because enzyme is secreted (uniprot). Specificity: Heart, placenta and less abundantly, in lung. May be involved in the production of lung surfactant, the remodeling or regulation of cardiac muscle. PLA2G2F: PMID: 11112443. Localization: extracellular - secre PLA2G10: PMID: 9188469 Localization: extracellular - secrete PLA2G2D: PMID: 10455175. Localization: extracellular - secr

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PLA2_2e	3	Ishizaki J, Suzuki N, Higashino K, Yokota Y, Ono T, Kawamoto K, Fujii N, Arita H, Hanasaki K.	Cloning and characterization of novel mouse and human secretory phospholipase A(2)s.	J Biol Chem	1999	10455175	secreted enzyme -> extracellular designation: uniprot PA2 catalyzes the calcium-dependent hydrolysis of the 2- acyl groups in 3-sn-phosphoglycerides. Pla2g1b: specificity: pancreas role in signaling pathways, etc (Nardelli ref) Pla2g12a: Abundantly expressed in heart, skeletal muscle, kidney, liver and pancreas. (see Gelb et al ref) Pla2g2e: specificity: Restricted to the brain, heart, lung, and placenta. Promotes stimulus-induced arachidonic acid release and prostaglandin (PG) production similar to those elicited by HSPG-dependent sPLA(2)s, suggesting that this enzyme plays a role in the inflammatory process. see PMID: 10681567. PLA2G2A: see PMID: 9272153. Membrane associated. Group II phospholipase A2 is found in many cells and also extracellularly. The membrane-bound and secreted forms are identical and are encoded by a single gene. PLA2G5: PMID: 8300559. Localization: extracellular because enzyme is secreted (uniprot). Specificity: Heart, placenta and less abundantly, in lung. May be involved in the production of lung surfactant, the remodeling or regulation of cardiac muscle. PLA2G2F: PMID: 11112443. Localization: extracellular - secre PLA2G10: PMID: 9188469 Localization: extracellular - secrete PLA2G2D: PMID: 10455175. Localization: extracellular - secr
PLA2_2e	3	Suzuki N, Ishizaki J, Yokota Y, Higashino K, Ono T, Ikeda M, Fujii N, Kawamoto K, Hanasaki K.	Structures, enzymatic properties, and expression of novel human and mouse secretory phospholipase A(2)s.	J Biol Chem	2000	10681567	secreted enzyme -> extracellular designation: uniprot PA2 catalyzes the calcium-dependent hydrolysis of the 2- acyl groups in 3-sn-phosphoglycerides. Pla2g1b: specificity: pancreas role in signaling pathways, etc (Nardelli ref) Pla2g12a: Abundantly expressed in heart, skeletal muscle, kidney, liver and pancreas. (see Gelb et al ref) Pla2g2e: specificity: Restricted to the brain, heart, lung, and placenta. Promotes stimulus-induced arachidonic acid release and prostaglandin (PG) production similar to those elicited by HSPG-dependent sPLA(2)s, suggesting that this enzyme plays a role in the inflammatory process. see PMID: 10681567. PLA2G2A: see PMID: 9272153. Membrane associated. Group II phospholipase A2 is found in many cells and also extracellularly. The membrane-bound and secreted forms are identical and are encoded by a single gene. PLA2G5: PMID: 8300559. Localization: extracellular because enzyme is secreted (uniprot). Specificity: Heart, placenta and less abundantly, in lung. May be involved in the production of lung surfactant, the remodeling or regulation of cardiac muscle. PLA2G2F: PMID: 11112443. Localization: extracellular - secre PLA2G10: PMID: 9188469 Localization: extracellular - secrete PLA2G2D: PMID: 10455175. Localization: extracellular - secr

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PLA2_2e	3	Gelb MH, Valentin E, Ghomashchi F, Lazdunski M, Lambeau G.	Cloning and recombinant expression of a structurally novel human secreted phospholipase A2.	J Biol Chem	2000	11031251	secreted enzyme -> extracellular designation: uniprot PA2 catalyzes the calcium-dependent hydrolysis of the 2- acyl groups in 3-sn-phosphoglycerides. Pla2g1b: specificity: pancreas role in signaling pathways, etc (Nardelli ref) Pla2g12a: Abundantly expressed in heart, skeletal muscle, kidney, liver and pancreas. (see Gelb et al ref) Pla2g2e: specificity: Restricted to the brain, heart, lung, and placenta. Promotes stimulus-induced arachidonic acid release and prostaglandin (PG) production similar to those elicited by HSPG-dependent sPLA(2)s, suggesting that this enzyme plays a role in the inflammatory process. see PMID: 10681567. PLA2G2A: see PMID: 9272153. Membrane associated. Group II phospholipase A2 is found in many cells and also extracellularly. The membrane-bound and secreted forms are identical and are encoded by a single gene. PLA2G5: PMID: 8300559. Localization: extracellular because enzyme is secreted (uniprot). Specificity: Heart, placenta and less abundantly, in lung. May be involved in the production of lung surfactant, the remodeling or regulation of cardiac muscle. PLA2G2F: PMID: 11112443. Localization: extracellular - secre PLA2G10: PMID: 9188469 Localization: extracellular - secrete PLA2G2D: PMID: 10455175. Localization: extracellular - secr
PLA2_2e	3	Valentin E, Singer AG, Ghomashchi F, Lazdunski M, Gelb MH, Lambeau G.	Cloning and recombinant expression of human group IIF-secreted phospholipase A(2).	Biochem Biophys Res Commun	2000	11112443	secreted enzyme -> extracellular designation: uniprot PA2 catalyzes the calcium-dependent hydrolysis of the 2- acyl groups in 3-sn-phosphoglycerides. Pla2g1b: specificity: pancreas role in signaling pathways, etc (Nardelli ref) Pla2g12a: Abundantly expressed in heart, skeletal muscle, kidney, liver and pancreas. (see Gelb et al ref) Pla2g2e: specificity: Restricted to the brain, heart, lung, and placenta. Promotes stimulus-induced arachidonic acid release and prostaglandin (PG) production similar to those elicited by HSPG-dependent sPLA(2)s, suggesting that this enzyme plays a role in the inflammatory process. see PMID: 10681567. PLA2G2A: see PMID: 9272153. Membrane associated. Group II phospholipase A2 is found in many cells and also extracellularly. The membrane-bound and secreted forms are identical and are encoded by a single gene. PLA2G5: PMID: 8300559. Localization: extracellular because enzyme is secreted (uniprot). Specificity: Heart, placenta and less abundantly, in lung. May be involved in the production of lung surfactant, the remodeling or regulation of cardiac muscle. PLA2G2F: PMID: 11112443. Localization: extracellular - secre PLA2G10: PMID: 9188469 Localization: extracellular - secrete PLA2G2D: PMID: 10455175. Localization: extracellular - secr

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PLA2_2e	3	Chen J, Engle SJ, Seilhamer JJ, Tischfield JA.	Cloning and recombinant expression of a novel human low molecular weight Ca(2+)-dependent phospholipase A2.	J Biol Chem	1994	8300559 [	secreted enzyme -> extracellular designation: uniprot PA2 catalyzes the calcium-dependent hydrolysis of the 2- acyl groups in 3-sn-phosphoglycerides. Pla2g1b: specificity: pancreas role in signaling pathways, etc (Nardelli ref) Pla2g12a: Abundantly expressed in heart, skeletal muscle, kidney, liver and pancreas. (see Gelb et al ref) Pla2g2e: specificity: Restricted to the brain, heart, lung, and placenta. Promotes stimulus-induced arachidonic acid release and prostaglandin (PG) production similar to those elicited by HSPG-dependent sPLA(2)s, suggesting that this enzyme plays a role in the inflammatory process. see PMID: 10681567. PLA2G2A: see PMID: 9272153. Membrane associated. Group II phospholipase A2 is found in many cells and also extracellularly. The membrane-bound and secreted forms are identical and are encoded by a single gene. PLA2G5: PMID: 8300559. Localization: extracellular because enzyme is secreted (uniprot). Specificity: Heart, placenta and less abundantly, in lung. May be involved in the production of lung surfactant, the remodeling or regulation of cardiac muscle. PLA2G2F: PMID: 11112443. Localization: extracellular - secre PLA2G10: PMID: 9188469 Localization: extracellular - secrete PLA2G2D: PMID: 10455175. Localization: extracellular - secr
PMI12346PH	3	Saiardi A, Nagata E, Luo HR, Snowman AM, Snyder SH	Identification and characterization of a novel inositol hexakisphosphate kinase	J Biol Chem	2001	11502751	has the activity [RefSeq, [Saiardi, J Biol Chem 2001]]: InsP6 -> PP-Ins(1,2,3,4,6)P5 Ins(1,3,4,5,6)P5 -> PP-Ins(1,3,4,6)P4 - nuclear (predominant) and cytoplasmic [UniProt], [Saiardi, J Biol Chem 2001] - gene was cloned; 50, 45% identity to isozymes [Saiardi, J Biol Chem 2001]
PNP	3	Wielgus-Kutrowska B, Kulikowska E, Wierzchowski J, Bzowska A, Shugar D.	Nicotinamide riboside, an unusual, non-typical, substrate of purified purine-nucleoside phosphorylases.	Eur J Biochem	1997	9030766	IT
PNP	3	Magni G, Amici A, Emanuelli M, Orsomando G, Raffaelli N, Ruggieri S.	Enzymology of NAD+ homeostasis in man.	Cell Mol Life Sci	2004	14704851	IT
PNTEH	3	Maras B, Barra D, Dupre S, Pitari G.	Is pantetheinase the actual identity of mouse and human vanin-1 proteins?	FEBS Lett	1999	10567687	IT I am not 100 % sure if all three genes are necessary for pantetheinase activity.
PNTEH	3	Martin F, Malergue F, Pitari G, Philippe JM, Philips S, Chabret C, Granjeaud S, Mattei MG, Mungall AJ, Naquet P, Galland F.	Vanin genes are clustered (human 6q22-24 and mouse 10A2B1) and encode isoforms of pantetheinase ectoenzymes.	Immunogenetics	2001	11491533	IT I am not 100 % sure if all three genes are necessary for pantetheinase activity.
PNTK	3	Zhou B, Westaway SK, Levinson B, Johnson MA, Gitschier J, Hayflick SJ.	A novel pantothenate kinase gene (PANK2) is defective in Hallervorden-Spatz syndrome.	Nat Genet	2001	11479594	This gene is defective in Hallervorden-Spatz syndrome (PanK2) 01-24-05 IT Expression: PanK1: heart, liver, kidney PanK2: ubiquitously, inluding retina and infant basal ganglia PanK3: mostly liver PanK4: most abundant in muscle, but expressed in all tissues There are 2 other reactions of this enzyme - I did not included them yet
PNTK	3	Daugherty M, Polanuyer B, Farrell M, Scholle M, Lykidis A, de Crecy-Lagard V, Osterman A.	Complete reconstitution of the human coenzyme A biosynthetic pathway via comparative genomics.	J Biol Chem	2002	11923312	This gene is defective in Hallervorden-Spatz syndrome (PanK2) 01-24-05 IT Expression: PanK1: heart, liver, kidney PanK2: ubiquitously, inluding retina and infant basal ganglia PanK3: mostly liver PanK4: most abundant in muscle, but expressed in all tissues There are 2 other reactions of this enzyme - I did not included them yet

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PNTK	3	Ching KH, Westaway SK, Gitschier J, Higgins JJ, Hayflick SJ.	HARP syndrome is allelic with pantothenate kinase-associated neurodegeneration.	Neurology	2002	Neurology	This gene is defective in Hallervorden-Spatz syndrome (PanK2) 01-24-05 IT Expression: PanK1: heart, liver, kidney PanK2: ubiquitously, inluding retina and infant basal ganglia PanK3: mostly liver PanK4: most abundant in muscle, but expressed in all tissues There are 2 other reactions of this enzyme - I did not included them yet
PNTKm	3	Hortnagel K, Prokisch H, Meitinger T.	An isoform of hPANK2, deficient in pantothenate kinase-associated neurodegeneration, localizes to mitochondria.	Hum Mol Genet	2003	12554685	01-24-05 I am not quite sure if this is a new isoform or not, however the isoforms listed in Locuslink do not cover the same CDS (7..1711). In Hoertnagel et al, 2003, Human Mol. genetics, 12(3), 321-327, it seems as they identified this as new transcript. IT They also proposed that a complete intramitochondrial pathway for de novo synthesis of CoA exists
PPA	3	Fairchild TA, Patejunas G.	Cloning and expression profile of human inorganic pyrophosphatase.	Biochim Biophys Acta	1999	10542310	- Added by RS/TV Catalytic Activity: Inorganic phosphatase catalyzes the hydrolysis of pyrophosphate to form orthophosphate. There are two groups of Ppases (type I and type 2). Both catalyzes the same aforementioned reaction, however vary in their kinetic information, co-factor usuage. More differences described Fabrichniy IP, Lehtio L, Salminen A, Zyryanov AB, Baykov AA, Lahti R, Goldman A.Biochemistry. 2004 Nov 16;43(45):14403-11. There are four transcriptional variants according to Entrez for PPA2 Subcellular Localization: Ppa2.1-4: Located in mitochondria according to GeneCards Pp.1: Located in cytosol according to Gene Cards Tissue Localization: Both are distributed fairly ubquitiously throughout the body. There is a slightly greater concentration in the heart and brain. Catalytic activity and tissue localization according to Fairchild TA, Patejunas G.iochim Biophys Acta. 1999 Oct 28;1447(2-3):133-6.
PPA	3	Fabrichniy IP, Lehtio L, Salminen A, Zyryanov AB, Baykov AA, Lahti R, Goldman A.	Structural studies of metal ions in family II pyrophosphatases: the requirement for a Janus ion.	Biochemistry	2004	15533045	- Added by RS/TV Catalytic Activity: Inorganic phosphatase catalyzes the hydrolysis of pyrophosphate to form orthophosphate. There are two groups of Ppases (type I and type 2). Both catalyzes the same aforementioned reaction, however vary in their kinetic information, co-factor usuage. More differences described Fabrichniy IP, Lehtio L, Salminen A, Zyryanov AB, Baykov AA, Lahti R, Goldman A.Biochemistry. 2004 Nov 16;43(45):14403-11. There are four transcriptional variants according to Entrez for PPA2 Subcellular Localization: Ppa2.1-4: Located in mitochondria according to GeneCards Pp.1: Located in cytosol according to Gene Cards Tissue Localization: Both are distributed fairly ubquitiously throughout the body. There is a slightly greater concentration in the heart and brain. Catalytic activity and tissue localization according to Fairchild TA, Patejunas G.iochim Biophys Acta. 1999 Oct 28;1447(2-3):133-6.
PPAP	3	Coleman RA, Lee DP	Enzymes of triacylglycerol synthesis and their regulation	Prog in Lipid Research	2004		ER - external ER surface (see refs) multiple substrate specificities NJ
PPAP	3	Roberts R, Sciorra VA, Morris AJ	Human type 2 phosphatidic acid phosphohydrolases	The Journal of Biological Chemistry	1998		ER - external ER surface (see refs) multiple substrate specificities NJ
PPAP	3	Coleman RA, Lee DP	Enzymes of triacylglycerol synthesis and their regulation	Prog in Lipid Research	2004		ER - external ER surface (see refs) multiple substrate specificities NJ
PPAt	2	Tassani V, Cattapan F, Magnanimi L, Peschechera A.	Anaplerotic effect of propionyl carnitine in rat heart mitochondria.	Biochem Biophys Res Commun	1994	8135845	Tassani, 1994 Anaplerotic effect of propionyl carnintine in rat heart mitochondria - reference and description taken from Thuy's mitochondrial model ("Heart Mito Isotopomer"); ND

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
PPBNGS	3	Ishida N, Fujita H, Fukuda Y, Noguchi T, Doss M, Kappas A, Sassa S.	Cloning and expression of the defective genes from a patient with delta-aminolevulinate dehydratase porphyria.	J Clin Invest	1992		- Aminolevulinate dehydratase (ALAD), the second enzyme in the heme biosyntheic pathway, catalyzes the asymmetric condensation of two molecules of 5-aminolevulic acid to form the monopyrrole, porphobilinogen. (Ishida N, Fujita H, Fukuda Y, Noguchi T, Doss M, Kappas A, Sassa S. J Clin Invest. 1992 May;89(5):1431-7.) Added by RS/TV
PPCOACm	3	Lamhonwah AM, Barankiewicz TJ, Willard HF, Mahuran DJ, Quan F, Gravel RA	Isolation of cDNA clones coding for the alpha and beta chains of human propionyl-CoA carboxylase: chromosomal assignments and DNA polymorphisms associated with PCCA and PCCB genes	Proc Natl Acad Sci U S A	1986	3460076	- enzyme is heterodimer of PCCA & PCCB [RefSeq], [UniProt], [Lamhonwah, PNAS 1986] - mitochondrial [UniProt], [RefSeq], [Lamhonwah, Genomics 1994] - reaction described in Devlin p. 637, Orten p. 262
PPCOACm	3	Lamhonwah AM, Leclerc D, Loyer M, Clarizio R, Gravel RA	Correction of the metabolic defect in propionic acidemia fibroblasts by microinjection of a full-length cDNA or RNA transcript encoding the propionyl-CoA carboxylase beta subunit	Genomics	1994	8188292	- enzyme is heterodimer of PCCA & PCCB [RefSeq], [UniProt], [Lamhonwah, PNAS 1986] - mitochondrial [UniProt], [RefSeq], [Lamhonwah, Genomics 1994] - reaction described in Devlin p. 637, Orten p. 262
PPCOAOm	3	Rozen R, Vockley J, Zhou L, Milos R, Willard J, Fu K, Vicanek C, Low-Nang L, Torban E, Fournier B	Isolation and expression of a cDNA encoding the precursor for a novel member (ACADSB) of the acyl-CoA dehydrogenase gene family	Genomics	1994	7698750	chains can also act on propinoyl-CoA 34: - mitochondrial [RefSeq], [UniProt] - functions on C4 to C12 fatty acyl-CoA chains [RefSeq] - functions on C4 to C16 fatty acyl-CoA chains [UniProt] - 88% sequence identity w/ porcine gene [Kelly, PNAS 1987] 35: - mitochondrial [RefSeq], [UniProt] - specificity inferred from mouse protein [Kelly, Genomics 1993] - identification of cDNA [Naito, J Clin Invest 1985] 36: - mitochondrial [RefSeq], [UniProt] - greatest activity towards (S)-2-methylbutyryl-CoA, but also reacts significantly with other 2-methyl branched chain substrates and with short straight chain acyl-CoAs [RefSeq], [Rozen, Genomics 1994], [UniProt] -ubiquitous [UniProt] 27034: - has activity with isobutyryl-CoA, (S) 2-methylbutyryl-CoA, and n-propionyl-CoA [Ngyuen, Mol Genet Metab 2002] - mitochondrial [UniProt] - Detected at comparable levels in all tissues examined (heart, lung, brain, skeletal muscle, pancreas and placenta). Weakly expressed in liver and kidney. [UniProt] 28976: - primarily active on palmitoyl-CoA (C16) and stearoyl-CoA (C - mitochondrial (probable) [UniProt] - Ubiquitously expressed in most normal human tissues and can 80724:
PPCOAOm	3	Kelly CL, Hinsdale ME, Wood PA	Cloning and characterization of the mouse short-chain acyl-CoA dehydrogenase cDNA	Genomics	1993	8276399	chains can also act on propinoyl-CoA 34: - mitochondrial [RefSeq], [UniProt] - functions on C4 to C12 fatty acyl-CoA chains [RefSeq] - functions on C4 to C16 fatty acyl-CoA chains [UniProt] - 88% sequence identity w/ porcine gene [Kelly, PNAS 1987] 35: - mitochondrial [RefSeq], [UniProt] - specificity inferred from mouse protein [Kelly, Genomics 1993] - identification of cDNA [Naito, J Clin Invest 1985] 36: - mitochondrial [RefSeq], [UniProt] - greatest activity towards (S)-2-methylbutyryl-CoA, but also reacts significantly with other 2-methyl branched chain substrates and with short straight chain acyl-CoAs [RefSeq], [Rozen, Genomics 1994], [UniProt] -ubiquitous [UniProt] 27034: - has activity with isobutyryl-CoA, (S) 2-methylbutyryl-CoA, and n-propionyl-CoA [Ngyuen, Mol Genet Metab 2002] - mitochondrial [UniProt] - Detected at comparable levels in all tissues examined (heart, lung, brain, skeletal muscle, pancreas and placenta). Weakly expressed in liver and kidney. [UniProt] 28976: - primarily active on palmitoyl-CoA (C16) and stearoyl-CoA (C - mitochondrial (probable) [UniProt] - Ubiquitously expressed in most normal human tissues and can 80724:

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PPCOAOm	3	Nguyen TV, Andresen BS, Corydon TJ, Ghisla S, Abd-El Razik N, Mohsen AW, Cederbaum SD, Roe DS, Roe CR, Lench NJ, Vockley J	Identification of isobutyryl-CoA dehydrogenase and its deficiency in humans	Mol Genet Metab	2002	12359132	chains can also act on propinoyl-CoA 34: - mitochondrial [RefSeq], [UniProt] - functions on C4 to C12 fatty acyl-CoA chains [RefSeq] - functions on C4 to C16 fatty acyl-CoA chains [UniProt] - 88% sequence identity w/ porcine gene [Kelly, PNAS 1987] 35: - mitochondrial [RefSeq], [UniProt] - specificity inferred from mouse protein [Kelly, Genomics 1993] - identification of cDNA [Naito, J Clin Invest 1985] 36: - mitochondrial [RefSeq], [UniProt] - greatest activity towards (S)-2-methylbutyryl-CoA, but also reacts significantly with other 2-methyl branched chain substrates and with short straight chain acyl-CoAs [RefSeq], [Rozen, Genomics 1994], [UniProt] -ubiquitous [UniProt] 27034: - has activity with isobutyryl-CoA, (S) 2-methylbutyryl-CoA, and n-propionyl-CoA [Ngyuen, Mol Genet Metab 2002] - mitochondrial [UniProt] - Detected at comparable levels in all tissues examined (heart, lung, brain, skeletal muscle, pancreas and placenta). Weakly expressed in liver and kidney. [UniProt] 28976: - primarily active on palmitoyl-CoA (C16) and stearoyl-CoA (C - mitochondrial (probable) [UniProt] - Ubiquitously expressed in most normal human tissues and can 80724:
PPCOAOm	3	Zhang J, Zhang W, Zou D, Chen G, Wan T, Zhang M, Cao X	Cloning and functional characterization of ACAD-9, a novel member of human acyl-CoA dehydrogenase family	Biochem Biophys Res Commun	2002	12359260	chains can also act on propinoyl-CoA 34: - mitochondrial [RefSeq], [UniProt] - functions on C4 to C12 fatty acyl-CoA chains [RefSeq] - functions on C4 to C16 fatty acyl-CoA chains [UniProt] - 88% sequence identity w/ porcine gene [Kelly, PNAS 1987] 35: - mitochondrial [RefSeq], [UniProt] - specificity inferred from mouse protein [Kelly, Genomics 1993] - identification of cDNA [Naito, J Clin Invest 1985] 36: - mitochondrial [RefSeq], [UniProt] - greatest activity towards (S)-2-methylbutyryl-CoA, but also reacts significantly with other 2-methyl branched chain substrates and with short straight chain acyl-CoAs [RefSeq], [Rozen, Genomics 1994], [UniProt] -ubiquitous [UniProt] 27034: - has activity with isobutyryl-CoA, (S) 2-methylbutyryl-CoA, and n-propionyl-CoA [Ngyuen, Mol Genet Metab 2002] - mitochondrial [UniProt] - Detected at comparable levels in all tissues examined (heart, lung, brain, skeletal muscle, pancreas and placenta). Weakly expressed in liver and kidney. [UniProt] 28976: - primarily active on palmitoyl-CoA (C16) and stearoyl-CoA (C - mitochondrial (probable) [UniProt] - Ubiquitously expressed in most normal human tissues and can 80724:

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PPCOAOm	3	Ye X, Ji C, Zhou C, Zeng L, Gu S, Ying K, Xie Y, Mao Y	Cloning and characterization of a human cDNA ACAD10 mapped to chromosome 12q24.1	Mol Biol Rep	2004	15560374	chains can also act on propinoyl-CoA 34: - mitochondrial [RefSeq], [UniProt] - functions on C4 to C12 fatty acyl-CoA chains [RefSeq] - functions on C4 to C16 fatty acyl-CoA chains [UniProt] - 88% sequence identity w/ porcine gene [Kelly, PNAS 1987] 35: - mitochondrial [RefSeq], [UniProt] - specificity inferred from mouse protein [Kelly, Genomics 1993] - identification of cDNA [Naito, J Clin Invest 1985] 36: - mitochondrial [RefSeq], [UniProt] - greatest activity towards (S)-2-methylbutyryl-CoA, but also reacts significantly with other 2-methyl branched chain substrates and with short straight chain acyl-CoAs [RefSeq], [Rozen, Genomics 1994], [UniProt] -ubiquitous [UniProt] 27034: - has activity with isobutyryl-CoA, (S) 2-methylbutyryl-CoA, and n-propionyl-CoA [Ngyuen, Mol Genet Metab 2002] - mitochondrial [UniProt] - Detected at comparable levels in all tissues examined (heart, lung, brain, skeletal muscle, pancreas and placenta). Weakly expressed in liver and kidney. [UniProt] 28976: - primarily active on palmitoyl-CoA (C16) and stearoyl-CoA (C - mitochondrial (probable) [UniProt] - Ubiquitously expressed in most normal human tissues and can 80724:
PPNCL3	3	Manoj N, Strauss E, Begley TP, Ealick SE.	Structure of human phosphopantothenoylcysteine synthetase at 2.3 A resolution.	Structure (Camb)	2003	12906824	01-24-05 IT The functional enzyme is a homodimer
PPPGOm	2	Dailey TA, Dailey HA.	Human protoporphyrinogen oxidase: expression, purification, and characterization of the cloned enzyme.	Protein Sci	1996	8771201	- Added by RS/TV Proteome - Mitochondrial according to Entrez Gene database. - Protoporphyrinogen oxidase catalyzes the oxygen-dependent oxidation of protoporphyrinogen IX to protoporphyrin IX. Northern blot analysis of of eight different human tissues show evidence for only a single transcript in all tissue types. (Dailey TA, Dailey HA. Protein Sci. 1996 Jan;5(1):98-105.)
PRAGSr	3	Schild D, Brake AJ, Kiefer MC, Young D, Barr PJ.	Cloning of three human multifunctional de novo purine biosynthetic genes by functional complementation of yeast mutations.	Proc Natl Acad Sci U S A	1990	2183217	IT no infos about compartiment
PRAGSr	3	Brodsky G, Barnes T, Bleskan J, Becker L, Cox M, Patterson D.	The human GARS-AIRS-GART gene encodes two proteins which are differentially expressed during human brain development and temporally overexpressed in cerebellum of individuals with Down syndrome.	Hum Mol Genet	1997	9328467	IT no infos about compartiment
PRAGSr	3	Poch MT, Qin W, Caperelli CA.	The human trifunctional enzyme of de novo purine biosynthesis: heterologous expression, purification, and preliminary characterization.	Protein Expr Purif	1998	9473452	IT no infos about compartiment
PRAGSr	3	Zhang Y, Desharnais J, Greasley SE, Beardsley GP, Boger DL, Wilson IA.	Crystal structures of human GAR Tfase at low and high pH and with substrate beta-GAR.	Biochemistry	2002	12450384	IT no infos about compartiment
PRDX	0	Wu W, Chen Y, Hazen SL.	Eosinophil peroxidase nitrates protein tyrosyl residues. Implications for oxidative damage by nitrating intermediates in eosinophilic inflammatory disorders.	J Biol Chem	1999	10464338	- RS/TV (6/3/2005) - cytosolic accoding to GeneCards - EPX belongs to the peroxidase family. It has the ability to reduce hydrogen peroxide while simultaneously using a co-substrate which is consequently oxidized. Studies thus far have shown that EPX is able to use bromide, SCN salts; Furthermore it is also shown that EPX readily uses NO2(1-) as substrate to generate a reactive intermediate that nitrates protein tyrosyl residues in high yield. (Wu W, Chen Y, Hazen SL. J Biol Chem. 1999 Sep 3;274(36):25933-44.) - Based on this observation as well as GO annotation this GPR association has been made. - the catalase-peroxidase system is used to oxidize methanol in non-primates whereas in primates the alcohol dehydrogenase system is the primary mechanism; see: http://antizol.info/mpoisono.htm

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PRDXl	0	Burner U, Furtmuller PG, Kettle AJ, Koppenol WH, Obinger C.	Mechanism of reaction of myeloperoxidase with nitrite	J Biol Chem	2000	10777476	- RS/TV (6/3/2005) - Lysosomal according to GeneCards - MPO is a part of the peroxidase family. Generally, ferric or native myeloperoxidase reacts with hydrogen peroxide forming a redox intermediate. This redox intermediate is known to oxidize halides via a single two-electron reaction to produce the respective hypohalous acids and regenerate the native enzyme. However, halides are not the only co-substrates that MPO works with. These co-substrates include tyrosine, tryptophan, sulfhydryls, phenol and indole derivatives, nitrite, hydrogen peroxide, xenobiotics, and others. According to 1) Arnhold J.Biochemistry (Mosc). 2004 Jan;69(1):4-9. Review. 2) Burner U, Furtmuller PG, Kettle AJ, Koppenol WH, Obinger C.J Biol Chem. 2000 Jul 7;275(27):20597-601. - Based on GO annotation and known catalytic activity, this GPR association has been made. - the catalase-peroxidase system is used to oxidize methanol in non-primates whereas in primates the alcohol dehydrogenase system is the primary mechanism; see: http://antizol.info/mpoisono.htm
PRDXl	0	Arnhold J.	Properties, functions, and secretion of human myeloperoxidase	Biochemistry (Mosc)	2004	14972011	- RS/TV (6/3/2005) - Lysosomal according to GeneCards - MPO is a part of the peroxidase family. Generally, ferric or native myeloperoxidase reacts with hydrogen peroxide forming a redox intermediate. This redox intermediate is known to oxidize halides via a single two-electron reaction to produce the respective hypohalous acids and regenerate the native enzyme. However, halides are not the only co-substrates that MPO works with. These co-substrates include tyrosine, tryptophan, sulfhydryls, phenol and indole derivatives, nitrite, hydrogen peroxide, xenobiotics, and others. According to 1) Arnhold J.Biochemistry (Mosc). 2004 Jan;69(1):4-9. Review. 2) Burner U, Furtmuller PG, Kettle AJ, Koppenol WH, Obinger C.J Biol Chem. 2000 Jul 7;275(27):20597-601. - Based on GO annotation and known catalytic activity, this GPR association has been made. - the catalase-peroxidase system is used to oxidize methanol in non-primates whereas in primates the alcohol dehydrogenase system is the primary mechanism; see: http://antizol.info/mpoisono.htm
PRFGS	3	Patterson D, Bleskan J, Gardiner K, Bowersox J.	Human phosphoribosylformylglycineamide amidotransferase (FGARAT): regional mapping, complete coding sequence, isolation of a functional genomic clone, and DNA sequence analysis.	Gene	1999	10548741	cytoplasm (GeneCards) IT
PROAKGOX1r	3	Helaakoski T, Vuori K, Myllyla R, Kivirikko KI, Pihlajaniemi T.	Molecular cloning of the alpha-subunit of human prolyl 4-hydroxylase: the complete cDNA-derived amino acid sequence and evidence for alternative splicing of RNA transcripts.	Proc Natl Acad Sci U S A	1989	2543975	this is done with enzyme complex vs subunits and one enzyme because "The beta subunit (P4HB; MIM 176790) is an unusual multifunctional polypeptide identical to protein disulfide isomerase (EC 5.3.4.1). P4HA2 is one of at least 2 alpha subunit isoforms (Helaakoski et al., 1995 [PubMed 7753822])" This reaction represents proline hydroxylation in peptide likages and NOT free proline [Helaakoski et al., 1995] Collagen prolyl 4-hydroxylases (P4Hs, EC 1.14.11.2) are located within the lumen of the endoplasmic reticulum and catalyze the formation of 4-hydroxyproline by the hydroxylation of prolines in -X-Pro-Gly- sequences in collagens and more than 15 other proteins that have collagen-like domains [Myllyharju 2003] The 4hpro-L in in proline could eventually be broken down and used to form glyoxylate.

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PROAKGOX1r	3	Pihlajaniemi T, Helaakoski T, Tasanen K, Myllyla R, Huhtala ML, Koivu J, Kivirikko KI.	Molecular cloning of the beta-subunit of human prolyl 4-hydroxylase. This subunit and protein disulphide isomerase are products of the same gene.	EMBO J	1987	3034602	this is done with enzyme complex vs subunits and one enzyme because "The beta subunit (P4HB; MIM 176790) is an unusual multifunctional polypeptide identical to protein disulfide isomerase (EC 5.3.4.1). P4HA2 is one of at least 2 alpha subunit isoforms (Helaakoski et al., 1995 [PubMed 7753822])" This reaction represents proline hydroxylation in peptide likages and NOT free proline [Helaakoski et al., 1995] Collagen prolyl 4-hydroxylases (P4Hs, EC 1.14.11.2) are located within the lumen of the endoplasmic reticulum and catalyze the formation of 4-hydroxyproline by the hydroxylation of prolines in -X-Pro-Gly- sequences in collagens and more than 15 other proteins that have collagen-like domains [Myllyharju 2003] The 4hpro-L in in proline could eventually be broken down and used to form glyoxylate.
PROAKGOX1r	3	Myllyharju J	Prolyl 4-hydroxylases, the key enzymes of collagen biosynthesis	Matrix Biol	2003	12714038	this is done with enzyme complex vs subunits and one enzyme because "The beta subunit (P4HB; MIM 176790) is an unusual multifunctional polypeptide identical to protein disulfide isomerase (EC 5.3.4.1). P4HA2 is one of at least 2 alpha subunit isoforms (Helaakoski et al., 1995 [PubMed 7753822])" This reaction represents proline hydroxylation in peptide likages and NOT free proline [Helaakoski et al., 1995] Collagen prolyl 4-hydroxylases (P4Hs, EC 1.14.11.2) are located within the lumen of the endoplasmic reticulum and catalyze the formation of 4-hydroxyproline by the hydroxylation of prolines in -X-Pro-Gly- sequences in collagens and more than 15 other proteins that have collagen-like domains [Myllyharju 2003] The 4hpro-L in in proline could eventually be broken down and used to form glyoxylate.
PROD2	2	Gogos JA, Santha M, Takacs Z, Beck KD, Luine V, Lucas LR, Nadler JV, Karayiorgou M.	The gene encoding proline dehydrogenase modulates sensorimotor gating in mice.	Nat Genet	1999	10192398	unclear which acceptors are used for this reaction (NADPH specifically excluded at this point because it would cause a loop that could turn NADH into NADPH) (right now a loop can turn NADH into FADH2 which should be okay)
PROSTGD2t	3	Lu R, Kanai N, Bao Y, Schuster VL.	Cloning, in vitro expression, and tissue distribution of a human prostaglandin transporter cDNA(hPGT).	J Clin Invest	1996	8787677	Tissue Specificity: SLCO2A1 - ubiquitous SLCO1A2 - brain, kidney, lung, testis, liver SLCO1B1 - liver SLCO1B3 - liver SLCO2B1 - liver, placenta, spleen, lung, kidney, heart, ovary SLCO3A1 - ubiquitously SLCO4A1 - ubiquitously SLCO1C1 - brain, testis May mediate the release of newly synthesized prostaglandins from cells, the transepithelial transport of prostaglandins, and the clearance of prostaglandins from the circulation. NJ
PROSTGD2t	3	Satlin LM, Amin V, Wolkoff AW.	Organic anion transporting polypeptide mediates organic anion/HCO3- exchange.	J Biol Chem	1997	9334206	Tissue Specificity: SLCO2A1 - ubiquitous SLCO1A2 - brain, kidney, lung, testis, liver SLCO1B1 - liver SLCO1B3 - liver SLCO2B1 - liver, placenta, spleen, lung, kidney, heart, ovary SLCO3A1 - ubiquitously SLCO4A1 - ubiquitously SLCO1C1 - brain, testis May mediate the release of newly synthesized prostaglandins from cells, the transepithelial transport of prostaglandins, and the clearance of prostaglandins from the circulation. NJ

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PROSTGD2t	3	Hagenbuch B, Meier PJ.	The superfamily of organic anion transporting polypeptides.	Biochim Biophys Acta	2003	12507753	Tissue Specificity: SLCO2A1 - ubiquitous SLCO1A2 - brain, kidney, lung, testis, liver SLCO1B1 - liver SLCO1B3 - liver SLCO2B1 - liver, placenta, spleen, lung, kidney, heart, ovary SLCO3A1 - ubiquitously SLCO4A1 - ubiquitously SLCO1C1 - brain, testis May mediate the release of newly synthesized prostaglandins from cells, the transepithelial transport of prostaglandins, and the clearance of prostaglandins from the circulation. NJ
PROSTGE2t2	3	Zhang L, Dresser MJ, Gray AT, Yost SC, Terashita S, Giacomini KM.	Cloning and functional expression of a human liver organic cation transporter.	Mol Pharmacol	1997	9187257	Tissue specificity: kidney (basolat membrane of prox tub), brain Km and Ki listed in Koepsell 2003 (in addition to detailed info about localization and other members of organic cation transporters) PMID: 12827517 cloning in PMID 9187257 Polyspecific organic cation transporters in the liver, kidney, intestine, and other organs are critical for elimination of many endogenous small organic cations as well as a wide array of drugs and environmental toxins. This gene is one of three similar cation transporter genes located in a cluster on chromosome 6. The encoded protein contains twelve putative transmembrane domains and is a plasma integral membrane protein. Two transcript variants encoding two different isoforms have been found for this gene, but only the longer variant encodes a functional transporter. NJ
PRPNCOAHYDx	3	Hoefler G, Forstner M, McGuinness MC, Hulla W, Hiden M, Krisper P, Kenner L, Ried T, Lengauer C, Zechner R, et al.	DNA cloning of the human peroxisomal enoyl-CoA hydratase: 3-hydroxyacyl-CoA dehydrogenase bifunctional enzyme and localization to chromosome 3q26.3-3q28: a free left Alu Arm is inserted in the 3' noncoding region.	Genomics	1994	8188243	- peroxisomal [Hoefler, Genomics 1994], [UniProt] - highest expression in liver and kidney [Hoefler, Genomics 1994] lower amounts also in brain [UniProt]
PRPPS	0	Taira M, Iizasa T, Shimada H, Kudoh J, Shimizu N, Tatibana M.	A human testis-specific mRNA for phosphoribosylpyrophosphate synthetase that initiates from a non-AUG codon.	J Biol Chem	1990	2168892	221823: - testis [RefSeq, UniProt]
PRPPS	0	Becker MA, Kim M	Regulation of purine synthesis de novo in human fibroblasts by purine nucleotides and phosphoribosylpyrophosphate.	J Biol Chem	1987	2444588	221823: - testis [RefSeq, UniProt]
PSDm_hs	3	Voelker DR.	Phosphatidylserine decarboxylase.	Biochim Biophys Acta	1997	9370338	from TV model - needs to be updated w/ refs, etc Localized to inner mit membrane (PMID: 9370338). This enzyme is actually believed to be a minor contributor to the production of pe - PMID: 15052331 (hatch rev) comment on data suggesting that the pe production pathway may involve the beta ox pathway. NJ
PSDm_hs	3	Hatch GM.	Cell biology of cardiac mitochondrial phospholipids.	Biochem Cell Biol	2004	15052331	from TV model - needs to be updated w/ refs, etc Localized to inner mit membrane (PMID: 9370338). This enzyme is actually believed to be a minor contributor to the production of pe - PMID: 15052331 (hatch rev) comment on data suggesting that the pe production pathway may involve the beta ox pathway. NJ
PSERT	3	Baek,J.Y. , Jun,d. o Y , Taub,D. , Kim,Y.H.	Characterization of human phosphoserine aminotransferase involved in the phosphorylated pathway of L-serine biosynthesis		2003	12633500	irreversible according to Lehninger (pg. 844, 4th ed.)
PSP_L	3	Collet JF, Gerin I, Rider MH, Veiga-da-Cunha M, Van Schaftingen E.	Human L-3-phosphoserine phosphatase: sequence, expression and evidence for a phosphoenzyme intermediate.		1997	9188776	irreversibility based on Lehninger (4th ed.)
PSP_L	3	Planitzer SA, Machl AW, Rueckels M, Kubbies M.	Identification of a novel c-DNA overexpressed in Fanconi's anemia fibroblasts partially homologous to a putative L-3-phosphoserine-phosphatase.		1998	9573387	irreversibility based on Lehninger (4th ed.)

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PTE2x	3	Jones JM, Nau K, Geraghty MT, Erdmann R, Gould SJ.	Identification of peroxisomal acyl-CoA thioesterases in yeast and humans.	J Biol Chem	1999	10092594	localization: peroxisomal (uniprot) specificity: ubiquitous Acyl-CoA thioesterases are a group of enzymes that catalyze the hydrolysis of acyl-CoAs to the free fatty acid and coenzyme A (CoASH), providing the potential to regulate intracellular levels of acyl-CoAs, free fatty acids and CoASH. May mediate Nef-induced down-regulation of CD4. Major thioesterase in peroxisomes. Competes with BAAT (Bile acid CoA: amino acid N-acyltransferase) for bile acid-CoA substrate (such as chenodeoxycholoyl-CoA). Shows a preference for medium-length fatty acyl-CoAs (By similarity). May be involved in the metabolic regulation of peroxisome proliferation. see also PMID: 10944470 for support for Zap128 NJ
PTE3x	3	Jones JM, Gould SJ.	Identification of PTE2, a human peroxisomal long-chain acyl-CoA thioesterase.	Biochem Biophys Res Commun	2000	10944470	localization: peroxisome (uniprot) specificity: none-noted Descriptions only noted medium and long chain fatty acids, in the future re-check literature for possible addition of other FA, particularly bile acid precursors (PTE1x, PTE6x, ...) Acyl-CoA thioesterases are a group of enzymes that catalyze the hydrolysis of acyl-CoAs to the free fatty acid and coenzyme A (CoASH), providing the potential to regulate intracellular levels of acyl-CoAs, free fatty acids and CoASH. Displays high levels of activity on medium- and long chain acyl CoAs. See PMID: 10944470 for thioesterase activity and localization. NJ
PTPAT	3	Aghajanian S, Worrall DM.	Identification and characterization of the gene encoding the human phosphopantetheine adenylyltransferase and dephospho-CoA kinase bifunctional enzyme (CoA synthase).	Biochem J	2002	11994049	01-24-05 IT
PTRCAT1	3	Chen Y, Vujcic S, Liang P, Diegelman P, Kramer DL, Porter CW.	Genomic identification and biochemical characterization of a second spermidine/spermine N1-acetyltransferase	Biochem J	2003	12803540	preference is for other substrates, but citation says ptrc is acceptable
PTRCOX1	2	Imamura Y, Kubota R, Wang Y, Asakawa S, Kudoh J, Mashima Y, Oguchi Y, Shimizu N	Human retina-specific amine oxidase (RAO): cDNA cloning, tissue expression, and chromosomal mapping	Genomics	1997	9119395	physiological data based on at least one of these genes associated with ptrc degradation in some way
PUNP1	3	Williams SR, Goddard JM, Martin DW Jr.	Human purine nucleoside phosphorylase cDNA sequence and genomic clone characterization.	Nucleic Acids Res	1984	6087295	IT
PUNP1	3	Erion MD, Stoeckler JD, Guida WC, Walter RL, Ealick SE.	Purine nucleoside phosphorylase. 2. Catalytic mechanism.	Biochemistry	1997	9305963	IT
PUNP5	3	Canduri F, dos Santos DM, Silva RG, Mendes MA, Basso LA, Palma MS, de Azevedo WF, Santos DS.	Structures of human purine nucleoside phosphorylase complexed with inosine and ddI.	Biochem Biophys Res Commun	2004	14706628	IT
PYDX5Ptm	2	Lui A, Lumeng L, Li TK.	Metabolism of vitamin B6 in rat liver mitochondria.	J Biol Chem	1981	6263901	IT studies on rat hepatocytes suggests that the mitochondrial transport occurs via diffusion. A number of enzymes located in the mitochondria are using Vit B6 (either pydx5p or pyam5p) as cofactor.
PYDX5Ptm	2	Lui A, Lumeng L, Li TK.	Transport of pyridoxine and pyridoxal 5'-phosphate in isolated rat liver mitochondria.	J Biol Chem	1982	7174673	IT studies on rat hepatocytes suggests that the mitochondrial transport occurs via diffusion. A number of enzymes located in the mitochondria are using Vit B6 (either pydx5p or pyam5p) as cofactor.
PYDXK	3	Hanna MC, Turner AJ, Kirkness EF.	Human pyridoxal kinase. cDNA cloning, expression, and modulation by ligands of the benzodiazepine receptor.	J Biol Chem	1997	9099727	IT
PYDXNK	3	Chern CJ, Beutler E.	Biochemical and electrophoretic studies of erythrocyte pyridoxine kinase in white and black Americans.	Am J Hum Genet	1976	2009	IT
PYDXNK	3	Ink SL, Henderson LM.	Vitamin B6 metabolism.	Annu Rev Nutr	1984	6380540	IT
PYNP2r	3	Watanabe S, Uchida T.	Cloning and expression of human uridine phosphorylase.	Biochem Biophys Res Commun	1995	7488099	IT
PYNP2r	3	Russell RL, Cao D, Zhang D, Handschumacher RE, Pizzorno G.	Uridine phosphorylase association with vimentin. Intracellular distribution and localization.	J Biol Chem	2001	11278417	IT
PYNP2r	3	Pizzorno G, Cao D, Leffert JJ, Russell RL, Zhang D, Handschumacher RE.	Homeostatic control of uridine and the role of uridine phosphorylase: a biological and clinical update.	Biochim Biophys Acta	2002	12084455	IT
PYNP2r	3	Johansson M.	Identification of a novel human uridine phosphorylase.	Biochem Biophys Res Commun	2003	12849978	IT

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PYRt2m	0	Halestrap AP, Scott RD, Thomas AP	Mitochondrial pyruvate transport and its hormonal regulation	Int J Biochem	1980	6987111	Halestrap et al, Int J Biochem 1980 - Additional Info by RS/TV: Ferdinando Palmieri, The mitochondrial transporter family (SLC25): physiological and pathological implications. "not yet identified at the molecular level"
PYRt2p	3	McClelland GB, Khanna S, Gonzalez GF, Butz CE, Brooks GA	Peroxisomal membrane monocarboxylate transporters: evidence for a redox shuttle system?	Biochem Biophys Res Commun	2003	12705896	Mechanism based on references and yeast The associated genes probably function in the transport into/out of various compartments The first citation also has evidence that pyruvate is converted into lactate inside peroxisomes 9194: - cloned [Lin 1998] - high affinity for pyruvate [Lin 1998] - restricted expression in normal tissues but high in cancer cell lines [Lin 1998] - also transports lactate, hydroxybutrayte [Lin 1998], ketone bodies[Halestrap 2004]
RAI3	2	Gough WH, VanOoteghem S, Sint T, Kedishvili NY.	cDNA cloning and characterization of a new human microsomal NAD+-dependent dehydrogenase that oxidizes all-trans-retinol and 3alpha-hydroxysteroids.	J Biol Chem	1998	9677409	IT
RAI3	2	Chen H, Juchau MR.	Recombinant human glutathione S-transferases catalyse enzymic isomerization of 13-cis-retinoic acid to all-trans-retinoic acid in vitro.	Biochem J	1998	9806904	IT
RBFK	3	Karthikeyan S, Zhou Q, Mseeh F, Grishin NV, Osterman AL, Zhang H.	Crystal structure of human riboflavin kinase reveals a beta barrel fold and a novel active site arch.	Structure (Camb)	2003	12623014	GeneCards located enzyme in cytoplasm based on sequence. IT
RBFK	3	Karthikeyan S, Zhou Q, Osterman AL, Zhang H.	Ligand binding-induced conformational changes in riboflavin kinase: structural basis for the ordered mechanism.	Biochemistry	2003	14580199	GeneCards located enzyme in cytoplasm based on sequence. IT
RBK	2	Agranoff BW, Brady RO	Purification and properties of calf liver ribokinase	J Biol Chem	1956	13295274	- exogenous ribose enters the PPP and is converted to R-5-P by ribokinase [Segal 1958] - enzyme has been purified from calf liver [Agranoff 1956]
RBK	2	Segal S, Foley J	The metabolism of D-ribose in man.	J Clin Invest	1958	13539215	- exogenous ribose enters the PPP and is converted to R-5-P by ribokinase [Segal 1958] - enzyme has been purified from calf liver [Agranoff 1956]
RBK_D	2	Huck JH, Roos B, Jakobs C, van der Knaap MS, Verhoeven NM	Evaluation of pentitol metabolism in mammalian tissues provides new insight into disorders of human sugar metabolism	Mol Genet Metab	2004	15234337	- ribulokinase has been identified in mammals [Huck, Mol Genet Metab 2004]
RDH1	3	Jurukovski V, Markova NG, Karaman-Jurukovska N, Randolph RK, Su J, Napoli JL, Simon M.	Cloning and characterization of retinol dehydrogenase transcripts expressed in human epidermal keratinocytes.	Mol Genet Metab	1999	10329026	IT RDH5 (5959): substrate spec: 11-cis-reinal = 13-cis-retinal > 9-cis-retinal (pro-S) (not all-trans-retinal), pro-S NADH
RDH1	3	Haeseleer F, Jang GF, Imanishi Y, Driessen CA, Matsumura M, Nelson PS, Palczewski K.	Dual-substrate specificity short chain retinol dehydrogenases from the vertebrate retina.	J Biol Chem	2002	12226107	IT RDH5 (5959): substrate spec: 11-cis-reinal = 13-cis-retinal > 9-cis-retinal (pro-S) (not all-trans-retinal), pro-S NADH
RDH1	3	Lapshina EA, Belyaeva OV, Chumakova OV, Kedishvili NY.	Differential recognition of the free versus bound retinol by human microsomal retinol/sterol dehydrogenases: characterization of the holo-CRBP dehydrogenase activity of RoDH-4.	Biochemistry	2003	12534290	IT RDH5 (5959): substrate spec: 11-cis-reinal = 13-cis-retinal > 9-cis-retinal (pro-S) (not all-trans-retinal), pro-S NADH
RDH1a	3	Matsuzaka Y, Okamoto K, Tsuji H, Mabuchi T, Ozawa A, Tamiya G, Inoko H.	Identification of the hRDH-E2 gene, a novel member of the SDR family, and its increased expression in psoriatic lesion.	Biochem Biophys Res Commun	2002	12372410	all-trans-retinol is predominant form, with 100% Viatmin A activity. 13-cis-retinol has 75% relative activity. 11-cis-retinaldehyde(retinal) is the chromophore in the retina of the eye, while all-trans and 9-cis retinoic acid are active metabolitess of retinol found in most if not in all tissues. changes in the molecular state of oxidation and cis/trans isomerization are of physiological importance in modifying the biological activity of retinoids. (Ball, Vitamins, book, 2004, 1st Ed) RDH12 (145226.1): Substrate spec: 9-cis-retinal> 11-cis-retinal>all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002) RDH14 (57665.1): Substrate spec: : 9-cis-retinal, 11-cis-retinal,all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002), pancreas RDH11 (51109.1): 9-cis-retinal>all-trans-retinal>11-cis-retinal,pro-S NADPH, eye (Haeseleer, 2002), prostate RDH13 (112724):NADPH (Haeseleer, 2002) RDH8(50700.1): pro-S NADPH, pro-R all-trans=retinol>9-cis-retinal; retina (Haeseleer,2002) RHD10 (157506.1): NADPH (Wu, 2002), retinal pigment epithelium IT

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RDH1a	3	Wu BX, Chen Y, Chen Y, Fan J, Rohrer B, Crouch RK, Ma JX.	Cloning and characterization of a novel all-trans retinol short-chain dehydrogenase/reductase from the RPE.	Invest Ophthalmol Vis Sci	2002	12407145	all-trans-retinol is predominant form, with 100% Viatmin A activity. 13-cis-retinol has 75% relative activity. 11-cis-retinaldehyde(retinal) is the chromophore in the retina of the eye, while all-trans and 9-cis retinoic acid are active metabolitess of retinol found in most if not in all tissues. changes in the molecular state of oxidation and cis/trans isomerization are of physiological importance in modifying the biological activity of retinoids. (Ball, Vitamins, book, 2004, 1st Ed) RDH12 (145226.1): Substrate spec: 9-cis-retinal> 11-cis-retinal>all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002) RDH14 (57665.1): Substrate spec: : 9-cis-retinal, 11-cis-retinal,all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002), pancreas RDH11 (51109.1): 9-cis-retinal>all-trans-retinal>11-cis-retinal,pro-S NADPH, eye (Haeseleer, 2002), prostate RDH13 (112724):NADPH (Haeseleer, 2002) RDH8(50700.1): pro-S NADPH, pro-R all-trans=retinol>9-cis-retinal; retina (Haeseleer,2002) RHD10 (157506.1): NADPH (Wu, 2002), retinal pigment epithelium IT
RDH1a	3	Janecke AR, Thompson DA, Utermann G, Becker C, Hubner CA, Schmid E, McHenry CL, Nair AR, Ruschendorf F, Heckenlively J, Wissinger B, Nurnberg P, Gal	Mutations in RDH12 encoding a photoreceptor cell retinol dehydrogenase cause childhood-onset severe retinal dystrophy.	Nat Genet	2004	15258582	all-trans-retinol is predominant form, with 100% Viatmin A activity. 13-cis-retinol has 75% relative activity. 11-cis-retinaldehyde(retinal) is the chromophore in the retina of the eye, while all-trans and 9-cis retinoic acid are active metabolitess of retinol found in most if not in all tissues. changes in the molecular state of oxidation and cis/trans isomerization are of physiological importance in modifying the biological activity of retinoids. (Ball, Vitamins, book, 2004, 1st Ed) RDH12 (145226.1): Substrate spec: 9-cis-retinal> 11-cis-retinal>all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002) RDH14 (57665.1): Substrate spec: : 9-cis-retinal, 11-cis-retinal,all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002), pancreas RDH11 (51109.1): 9-cis-retinal>all-trans-retinal>11-cis-retinal,pro-S NADPH, eye (Haeseleer, 2002), prostate RDH13 (112724):NADPH (Haeseleer, 2002) RDH8(50700.1): pro-S NADPH, pro-R all-trans=retinol>9-cis-retinal; retina (Haeseleer,2002) RHD10 (157506.1): NADPH (Wu, 2002), retinal pigment epithelium IT
RDH1a	3	Perrault I, Hanein S, Gerber S, Barbet F, Ducroq D, Dollfus H, Hamel C, Dufier JL, Munnich A, Kaplan J, Rozet JM.	Retinal dehydrogenase 12 (RDH12) mutations in leber congenital amaurosis.	Am J Hum Genet	2004	15322982	all-trans-retinol is predominant form, with 100% Viatmin A activity. 13-cis-retinol has 75% relative activity. 11-cis-retinaldehyde(retinal) is the chromophore in the retina of the eye, while all-trans and 9-cis retinoic acid are active metabolitess of retinol found in most if not in all tissues. changes in the molecular state of oxidation and cis/trans isomerization are of physiological importance in modifying the biological activity of retinoids. (Ball, Vitamins, book, 2004, 1st Ed) RDH12 (145226.1): Substrate spec: 9-cis-retinal> 11-cis-retinal>all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002) RDH14 (57665.1): Substrate spec: : 9-cis-retinal, 11-cis-retinal,all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002), pancreas RDH11 (51109.1): 9-cis-retinal>all-trans-retinal>11-cis-retinal,pro-S NADPH, eye (Haeseleer, 2002), prostate RDH13 (112724):NADPH (Haeseleer, 2002) RDH8(50700.1): pro-S NADPH, pro-R all-trans=retinol>9-cis-retinal; retina (Haeseleer,2002) RHD10 (157506.1): NADPH (Wu, 2002), retinal pigment epithelium IT
RDH2	3	Simon A, Hellman U, Wernstedt C, Eriksson U.	The retinal pigment epithelial-specific 11-cis retinol dehydrogenase belongs to the family of short chain alcohol dehydrogenases.	J Biol Chem	1995	7836368	IT RDH5 (5959): substrate spec: 11-cis-reinal = 13-cis-retinal > 9-cis-retinal (pro-S) (not all-trans-retinal), pro-S NADH
RDH2	3	Mertz JR, Shang E, Piantedosi R, Wei S, Wolgemuth DJ, Blaner WS.	Identification and characterization of a stereospecific human enzyme that catalyzes 9-cis-retinol oxidation. A possible role in 9-cis-retinoic acid formation.	J Biol Chem	1997	9115228	IT RDH5 (5959): substrate spec: 11-cis-reinal = 13-cis-retinal > 9-cis-retinal (pro-S) (not all-trans-retinal), pro-S NADH

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RDH2a	3	Belyaeva OV, Kedishvili NY.	Human pancreas protein 2 (PAN2) has a retinal reductase activity and is ubiquitously expressed in human tissues.	FEBS Lett	2002	12435598	RDH12 (145226.1): Substrate spec: 9-cis-retinal> 11-cis-retinal>all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002) RDH14 (57665.1): Substrate spec: : 9-cis-retinal, 11-cis-retinal,all-trans-retinal, NADPH, eye, other tissue? (Haeseleer, 2002), pancreas RDH11 (51109.1): 9-cis-retinal>all-trans-retinal>11-cis-retinal,pro-S NADPH, eye (Haeseleer, 2002), prostate RDH13 (112724):NADPH (Haeseleer, 2002) RDH8(50700.1): pro-S NADPH, pro-R all-trans=retinol>9-cis-retinal; retina (Haeseleer,2002) IT
RETH	3	Gao J, Simon M.	Identification of a novel keratinocyte retinyl ester hydrolase as a transacylase and lipase.	J Invest Dermatol	2005	15955102	IT main R group = palminate
RETI3	3	Blaner WS.	Cellular metabolism and actions of 13-cis-retinoic acid.	J Am Acad Dermatol	2001	11606944	it
RETNGLCt2	2	Tang GW, Russell RM.	13-cis-retinoic acid is an endogenous compound in human serum.	J Lipid Res	1990	2324641	mechanism unknown. produced in ER. found in blood. IT
RETNt	2	Hodam JR, Creek KE.	Uptake and metabolism of [3H]retinoic acid delivered to human foreskin keratinocytes either bound to serum albumin or added directly to the culture medium.	Biochim Biophys Acta	1996	8630327	IT
RETt	2	Blomhoff R, Green MH, Norum KR.	Vitamin A: physiological and biochemical processing.	Annu Rev Nutr	1992	1503811	IT at pharmacological concentration levels retinol can be absorbed by passive diffusion retinol is transported (absorbed) by passive diffusion from intestinal lumen to enterocytes. there it is enclosed by chylomicron and released. In this from it can get to extrahepatic target cells. However, the most part of chylomicron with retinol is taken up by liver cells (parenchymal cells), retinol is released there and binds directly to RBP (retinol binding protein) and then either stored or transported to other extrahepatic cells via blood
RIBFLVt3	3	Said HM, Ma TY.	Mechanism of riboflavine uptake by Caco-2 human intestinal epithelial cells.	Am J Physiol	2003	8304455	IT
RNDR1	2	Shao J, Zhou B, Zhu L, Qiu W, Yuan YC, Xi B, Yen Y.	In vitro characterization of enzymatic properties and inhibition of the p53R2 subunit of human ribonucleotide reductase.	Cancer Res	2004	14729598	IT needs ATP, iron -protein (kegg note)
RNMK	3	Bieganowski P, Brenner C.	Discoveries of nicotinamide riboside as a nutrient and conserved NRK genes establish a Preiss-Handler independent route to NAD+ in fungi and humans.	Cell	2004	15137942	IT There are two human genes for this reaction however i could not identify the second gene in Entrz-Gene
S23T2g	3	Sasaki K, Watanabe E, Kawashima K, Sekine S, Dohi T, Oshima M, Hanai N, Nishi T, Hasegawa M	Expression cloning of a novel Gal beta (1-3/1-4) GlcNAc alpha 2,3-sialyltransferase using lectin resistance selection	J Biol Chem	1993	7901202	- reaction described in Varki, pg 235 6482: - most activity due to ST3Gal-1, low levels due to ST3Gal-2, -4 in vitro [Varki] - gene was cloned and expressed [Shang, Eur J Biochem 1999] - found in Golgi [RefSeq], also inferred from author statement in [Shang, Eur J Biochem 1999] - Siat4ap expressed in placenta, liver, sk muscle [Kitagawa and Paulson, JBC 1994] 6483: - most activity due to ST3Gal-1, low levels due to ST3Gal-2, -4 in vitro [Varki] - gene was cloned and expressed [Kim, Biochem, Biophys Res Commun 1996] - Golgi, see [Kolter, Brain Pathol 1998] 6484: - most activity due to ST3Gal-1, low levels due to ST3Gal-2, -4 in vitro [Varki] - gene was cloned [Sasaki, J Biol Chem 1993], [Kitagawa, J Biol Chem 1994]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
S23T2g	3	Kitagawa H, Paulson JC	Cloning of a novel alpha 2,3-sialyltransferase that sialylates glycoprotein and glycolipid carbohydrate groups	J Biol Chem	1994	8288606	- reaction described in Varki, pg 235 6482: - most activity due to ST3Gal-1, low levels due to ST3Gal-2, -4 in vitro [Varki] - gene was cloned and expressed [Shang, Eur J Biochem 1999] - found in Golgi [RefSeq], also inferred from author statement in [Shang, Eur J Biochem 1999] - Siat4ap expressed in placenta, liver, sk muscle [Kitagawa and Paulson, JBC 1994] 6483: - most activity due to ST3Gal-1, low levels due to ST3Gal-2, -4 in vitro [Varki] - gene was cloned and expressed [Kim, Biochem, Biophys Res Commun 1996] - Golgi, see [Kolter, Brain Pathol 1998] 6484: - most activity due to ST3Gal-1, low levels due to ST3Gal-2, -4 in vitro [Varki] - gene was cloned [Sasaki, J Biol Chem 1993], [Kitagawa, J Biol Chem 1994]
S23T2g	3	Kim YJ, Kim KS, Kim SH, Kim CH, Ko JH, Choe IS, Tsuji S, Lee YC	Molecular cloning and expression of human Gal beta 1,3GalNAc alpha 2,3-sialytransferase (hST3Gal II)	Biochem Biophys Res Commun	1996	8920913	- reaction described in Varki, pg 235 6482: - most activity due to ST3Gal-1, low levels due to ST3Gal-2, -4 in vitro [Varki] - gene was cloned and expressed [Shang, Eur J Biochem 1999] - found in Golgi [RefSeq], also inferred from author statement in [Shang, Eur J Biochem 1999] - Siat4ap expressed in placenta, liver, sk muscle [Kitagawa and Paulson, JBC 1994] 6483: - most activity due to ST3Gal-1, low levels due to ST3Gal-2, -4 in vitro [Varki] - gene was cloned and expressed [Kim, Biochem, Biophys Res Commun 1996] - Golgi, see [Kolter, Brain Pathol 1998] 6484: - most activity due to ST3Gal-1, low levels due to ST3Gal-2, -4 in vitro [Varki] - gene was cloned [Sasaki, J Biol Chem 1993], [Kitagawa, J Biol Chem 1994]
S23T3g	3	Kitagawa H, Paulson JC	Cloning and expression of human Gal beta 1,3(4)GlcNAc alpha 2,3-sialyltransferase	Biochem Biophys Res Commun	1993	8333853	- normally found in Golgi [RefSeq] - gene was cloned and expressed [Kitagawa, Biochem Biophys Res Commun 1993] - 91% similiarity w/ rat cDNA [Kitagawa, Biochem Biophys Res Commun 1993] - abundant in sk muscle & fetal tissues, low exp in placenta [Kitagawa, Biochem Biophys Res Commun 1993]
S23Tg	3	Shang J, Qiu R, Wang J, Liu J, Zhou R, Ding H, Yang S, Zhang S, Jin C	Molecular cloning and expression of Galbeta1,3GalNAc alpha2, 3-sialyltransferase from human fetal liver	Eur J Biochem	1999	10504389	- reaction described in Varki, pg 235 - gene was cloned and expressed [Shang, Eur J Biochem 1999] - found in Golgi [RefSeq], also inferred from author statement in [Shang, Eur J Biochem 1999] - Siat4ap expressed in placenta, liver, sk muscle [Kitagawa and Paulson, JBC 1994]
S26Tg	0	Kitagawa H, Paulson JC	Differential expression of five sialyltransferase genes in human tissues	J Biol Chem	1994	8027041	The protein encoded by SIAT1 is a type II membrane protein that catalyzes the transfer of sialic acid from CMP-sialic acid to galactose-containing substrates. The encoded protein, which is normally found in the Golgi but which can be proteolytically processed to a soluble form, is involved in the generation of the cell-surface carbohydrate determinants and differentiation antigens HB-6, CDw75, and CD76. This protein is a member of glycosyltransferase family 29. Three transcript variants encoding two different isoforms have been found for this gene. [RefSeq] Siat1p expressed in sk muscle, liver, placenta [Kitagawa and Paulson, JBC 1994]
S2T1g	3	Kobayashi M, Sugumaran G, Liu J, Shworak NW, Silbert JE, Rosenberg RD	Molecular cloning and characterization of a human uronyl 2-sulfotransferase that sulfates iduronyl and glucuronyl residues in dermatan/chondroitin sulfate	J Biol Chem	1999	10187838	- Golgi localization [Silbert, IUBMB LIfe 2002] - identified based on sequence homology to heparan sulfate IdA 6-sulfotransferase [Kobayashi, J Biol Chem 1999] - gene was cloned and expressed [Kobayashi, J Biol Chem 1999] - ubiquitously expressed [Kobayashi, J Biol Chem 1999] - substantial sulfotransferase activity w/ dermatan sulfate, small degree of activity w/ chondroitin sulfate [Kobayashi, J Biol Chem 1999]
S2T4g	3	Seki N, Ohira M, Nagase T, Ishikawa K, Miyajima N, Nakajima D, Nomura N, Ohara O	Characterization of cDNA clones in size-fractionated cDNA libraries from human brain.	DNA Res	1997	9455484	- 2-sulfation of IdoA and GlcA in heparan sulfate is catalyzed by the same enzyme [Rong, Biochem J 2000], but has higher affinity for transfer to IdoA [Sugahara, IUBMB Life 2002] - distinct from uronyl-2-O-sulfotransferase for synthesis of chondroitin and dermatan sulfate [Kobayashi, J Biol Chem 1999] - gene was identified via high-throughput study [Seki, DNA Res 1997]

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S2T4g	3	Rong J, Habuchi H, Kimata K, Lindahl U, Kusche-Gullberg M	Expression of heparan sulphate L-iduronyl 2-O-sulphotransferase in human kidney 293 cells results in increased D-glucuronyl 2-O-sulphation	Biochem J	2000	10677367	- 2-sulfation of IdoA and GlcA in heparan sulfate is catalyzed by the same enzyme [Rong, Biochem J 2000], but has higher affinity for transfer to IdoA [Sugahara, IUBMB Life 2002] - distinct from uronyl-2-O-sulfotransferase for synthesis of chondroitin and dermatan sulfate [Kobayashi, J Biol Chem 1999] - gene was identified via high-throughput study [Seki, DNA Res 1997]
S2TASE1ly	3	Wilson PJ, Morris CP, Anson DS, Occhiodoro T, Bielicki J, Clements PR, Hopwood JJ	Hunter syndrome: isolation of an iduronate-2-sulfatase cDNA clone and analysis of patient DNA	Proc Natl Acad Sci U S A	1990	2122463	- hexuronic acids have to be desulfated before the hexuronidic linkage is hydrolyzed [Winchester 1996] 3423: - cloned isolated [Wilson 1990] - genomic sequence identified [Wilson 1993] - enzyme has been isolated from human placenta, serum, urine, liver (see refs in [Malmgren 1985]) - alternative transcript identified [Malmgren 1985])
S2TASE1ly	3	Wilson PJ, Meaney CA, Hopwood JJ, Morris CP	Sequence of the human iduronate 2-sulfatase (IDS) gene	Genomics	1993	8244397	- hexuronic acids have to be desulfated before the hexuronidic linkage is hydrolyzed [Winchester 1996] 3423: - cloned isolated [Wilson 1990] - genomic sequence identified [Wilson 1993] - enzyme has been isolated from human placenta, serum, urine, liver (see refs in [Malmgren 1985]) - alternative transcript identified [Malmgren 1985])
S2TASE1ly	3	Malmgren H, Carlberg BM, Pettersson U, Bondeson ML	dentification of an alternative transcript from the human iduronate-2-sulfatase (IDS) gene	Genomics	1995	8530090	- hexuronic acids have to be desulfated before the hexuronidic linkage is hydrolyzed [Winchester 1996] 3423: - cloned isolated [Wilson 1990] - genomic sequence identified [Wilson 1993] - enzyme has been isolated from human placenta, serum, urine, liver (see refs in [Malmgren 1985]) - alternative transcript identified [Malmgren 1985])
S2TASE4ly	3	Freeman C, Hopwood JJ	Human liver glucuronate 2-sulphatase. Purification, characterization and catalytic properties	Biochem J	1989	2497731	- hexuronic acids have to be desulfated before the hexuronidic linkage is hydrolyzed [Winchester 1996] - cultured human skin fibroblasts were shown to have glucuronate 2-sulfatase activity [Shaklee 1985] - glucuronate 2-sulfatase was purified from human liver, but it was unstable w/o the addition of BSA [Freeman 1989]
S2TASE4ly	3	Shaklee PN, Glaser JH, Conrad HE	A sulfatase specific for glucuronic acid 2-sulfate residues in glycosaminoglycans	J Biol Chem	1985	4019466	- hexuronic acids have to be desulfated before the hexuronidic linkage is hydrolyzed [Winchester 1996] - cultured human skin fibroblasts were shown to have glucuronate 2-sulfatase activity [Shaklee 1985] - glucuronate 2-sulfatase was purified from human liver, but it was unstable w/o the addition of BSA [Freeman 1989]
S3T1g	3	Shworak NW, Liu J, Fritze LM, Schwartz JJ, Zhang L, Logeart D, Rosenberg RD	Molecular cloning and expression of mouse and human cDNAs encoding heparan sulfate D-glucosaminyl 3-O-sulfotransferase	J Biol Chem	1997	9346953	9957: - Golgi [RefSeq], [Shworak, J Biol Chem 1997] - cDNA was cloned; protein has 93% similarity to mouse homolog [Shworak, J Biol Chem 1997] - expressed highly in kidney and brain, intermediately in heart and lung, and lowly in other tissues [Shworak, J Biol Chem 1999], [Sugahara, IUBMB Life 2002] - transfers sulfate to C3 position of NSGlcNAc and NSGLcNAc(6S) adjacent to the reducing side of GlcA [Sugahara, IUBMB Life 2002] 222537: - cDNA was isolated and expressed [Xia, J Biol Chem 2002] - major products of recombinantly expressed protein were: DeltaHexA-GlcN(NS,3S,6S), DeltaHexA(2S)-GlcN(NS,3S), and DeltaHexA(2S)-GlcN(NS,3S,6S) [Mochizuki, J Biol Chem 2003], [Sugahara, IUBMB Life 2002] - highly expressed in fetal brain, adult brain and spinal cord, low or undetectable in other tissues [Mochizuki, J Biol Chem 2003] 9951: - partial length clone identified (incomplete coding sequence) [Shworak, J Biol Chem 1999] - exclusively expressed in brain [Shworak, J Biol Chem 1999]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
S3T1g	3	Shworak NW, Liu J, Petros LM, Zhang L, Kobayashi M, Copeland NG, Jenkins NA, Rosenberg RD	Multiple isoforms of heparan sulfate D-glucosaminyl 3-O-sulfotransferase. Isolation, characterization, and expression of human cdnas and identification of distinct genomic loci	J Biol Chem	1999	9988767	9957: - Golgi [RefSeq], [Shworak, J Biol Chem 1997] - cDNA was cloned; protein has 93% similarity to mouse homolog [Shworak, J Biol Chem 1997] - expressed highly in kidney and brain, intermediately in heart and lung, and lowly in other tissues [Shworak, J Biol Chem 1999], [Sugahara, IUBMB Life 2002] - transfers sulfate to C3 position of NSGlcNAc and NSGLcNAc(6S) adjacent to the reducing side of GlcA [Sugahara, IUBMB Life 2002] 222537: - cDNA was isolated and expressed [Xia, J Biol Chem 2002] - major products of recombinantly expressed protein were: DeltaHexA-GlcN(NS,3S,6S), DeltaHexA(2S)-GlcN(NS,3S), and DeltaHexA(2S)-GlcN(NS,3S,6S) [Mochizuki, J Biol Chem 2003], [Sugahara, IUBMB Life 2002] - highly expressed in fetal brain, adult brain and spinal cord, low or undetectable in other tissues [Mochizuki, J Biol Chem 2003] 9951: - partial length clone identified (incomplete coding sequence) [Shworak, J Biol Chem 1999] - exclusively expressed in brain [Shworak, J Biol Chem 1999]
S3T1g	3	Xia G, Chen J, Tiwari V, Ju W, Li JP, Malmstrom A, Shukla D, Liu J	Heparan sulfate 3-O-sulfotransferase isoform 5 generates both an antithrombin-binding site and an entry receptor for herpes simplex virus, type 1	J Biol Chem	2002	12138164	9957: - Golgi [RefSeq], [Shworak, J Biol Chem 1997] - cDNA was cloned; protein has 93% similarity to mouse homolog [Shworak, J Biol Chem 1997] - expressed highly in kidney and brain, intermediately in heart and lung, and lowly in other tissues [Shworak, J Biol Chem 1999], [Sugahara, IUBMB Life 2002] - transfers sulfate to C3 position of NSGlcNAc and NSGLcNAc(6S) adjacent to the reducing side of GlcA [Sugahara, IUBMB Life 2002] 222537: - cDNA was isolated and expressed [Xia, J Biol Chem 2002] - major products of recombinantly expressed protein were: DeltaHexA-GlcN(NS,3S,6S), DeltaHexA(2S)-GlcN(NS,3S), and DeltaHexA(2S)-GlcN(NS,3S,6S) [Mochizuki, J Biol Chem 2003], [Sugahara, IUBMB Life 2002] - highly expressed in fetal brain, adult brain and spinal cord, low or undetectable in other tissues [Mochizuki, J Biol Chem 2003] 9951: - partial length clone identified (incomplete coding sequence) [Shworak, J Biol Chem 1999] - exclusively expressed in brain [Shworak, J Biol Chem 1999]
S3T1g	3	Mochizuki H, Yoshida K, Gotoh M, Sugioka S, Kikuchi N, Kwon YD, Tawada A, Maeyama K, Inaba N, Hiruma T, Kimata K, Narimatsu H	Characterization of a heparan sulfate 3-O-sulfotransferase-5, an enzyme synthesizing a tetrasulfated disaccharide	J Biol Chem	2003	12740361	9957: - Golgi [RefSeq], [Shworak, J Biol Chem 1997] - cDNA was cloned; protein has 93% similarity to mouse homolog [Shworak, J Biol Chem 1997] - expressed highly in kidney and brain, intermediately in heart and lung, and lowly in other tissues [Shworak, J Biol Chem 1999], [Sugahara, IUBMB Life 2002] - transfers sulfate to C3 position of NSGlcNAc and NSGLcNAc(6S) adjacent to the reducing side of GlcA [Sugahara, IUBMB Life 2002] 222537: - cDNA was isolated and expressed [Xia, J Biol Chem 2002] - major products of recombinantly expressed protein were: DeltaHexA-GlcN(NS,3S,6S), DeltaHexA(2S)-GlcN(NS,3S), and DeltaHexA(2S)-GlcN(NS,3S,6S) [Mochizuki, J Biol Chem 2003], [Sugahara, IUBMB Life 2002] - highly expressed in fetal brain, adult brain and spinal cord, low or undetectable in other tissues [Mochizuki, J Biol Chem 2003] 9951: - partial length clone identified (incomplete coding sequence) [Shworak, J Biol Chem 1999] - exclusively expressed in brain [Shworak, J Biol Chem 1999]

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
S3T2g	3	Liu J, Shworak NW, Sinay P, Schwartz JJ, Zhang L, Fritze LM, Rosenberg RD	Expression of heparan sulfate D-glucosaminyl 3-O-sulfotransferase isoforms reveals novel substrate specificities	J Biol Chem	1999	9988768	222537: - cDNA was isolated and expressed [Xia, J Biol Chem 2002] - major products of recombinantly expressed protein were: DeltaHexA-GlcN(NS,3S,6S), DeltaHexA(2S)-GlcN(NS,3S), and DeltaHexA(2S)-GlcN(NS,3S,6S) [Mochizuki, J Biol Chem 2003], [Sugahara, IUBMB Life 2002] - highly expressed in fetal brain, adult brain and spinal cord, low or undetectable in other tissues [Mochizuki, J Biol Chem 2003] 9956: - cDNA was isolated, characterized, and expressed [Shworak, J Biol Chem 1999] - predominantly expressed in the brain, low expression in heart and placenta [Shworak, J Biol Chem 1999] - transfers sulfate to C3 position of GlcA2S-GlcNS and IdoA2S GlcNS [Liu, J Biol Chem 1999], [Sugahara, IUBMB Life 2002] 9951: - partial length clone identified (incomplete coding sequence) [Shworak, J Biol Chem 1999] - exclusively expressed in brain [Shworak, J Biol Chem 1999]
S3T3g	3	Liu J, Shriver Z, Blaiklock P, Yoshida K, Sasisekharan R, Rosenberg RD	Heparan sulfate D-glucosaminyl 3-O-sulfotransferase 3A sulfates N-unsubstituted glucosamine residues	J Biol Chem	1999	10608887	9953: - cDNA was isolated, characterized, and expressed [Shworak, J Biol Chem 1999] - highly expressed in heart and placenta, moderately expressed in liver and kidney, lowly expressed in lung and pancreas [Shworak, J Biol Chem 1999] - transfers sulfate to C3 position of IdoA2S-GlcNH2 [Liu, J Biol Chem 1999], [Liu, J Biol Chem 1999], [Sugahara, J Biol Chem 2002] 9954: - cDNA was isolated and expressed [Shworak, J Biol Chem 1999] - functionality remains to be established [Shworak, J Biol Chem 1999]; assumed to have same functionality as HS3ST3A1 9953: - cDNA was isolated, characterized, and expressed [Shworak, J Biol Chem 1999] - highly expressed in placenta and liver, moderately expressed in heart, kidney, and pancreas, lowly expressed in brain, lung, skeletal muscle [Shworak, J Biol Chem 1999] - transfers sulfate to C3 position of IdoA2S-GlcNH2 [Sugahara, IUBMB Life 2002] 9952: - cDNA was isolated and expressed [Shworak, J Biol Chem 1999] - functionality remains to be established [Shworak, J Biol Chem 1999]; assumed to have same functionality as HS3ST3B1 64711:
S3T3g	3	Xu D, Tiwari V, Xia G, Clement C, Shukla D, Liu J	Characterization of heparan sulphate 3-O-sulphotransferase isoform 6 and its role in assisting the entry of herpes simplex virus type 1	Biochem J	2005	15303968	9953: - cDNA was isolated, characterized, and expressed [Shworak, J Biol Chem 1999] - highly expressed in heart and placenta, moderately expressed in liver and kidney, lowly expressed in lung and pancreas [Shworak, J Biol Chem 1999] - transfers sulfate to C3 position of IdoA2S-GlcNH2 [Liu, J Biol Chem 1999], [Liu, J Biol Chem 1999], [Sugahara, J Biol Chem 2002] 9954: - cDNA was isolated and expressed [Shworak, J Biol Chem 1999] - functionality remains to be established [Shworak, J Biol Chem 1999]; assumed to have same functionality as HS3ST3A1 9953: - cDNA was isolated, characterized, and expressed [Shworak, J Biol Chem 1999] - highly expressed in placenta and liver, moderately expressed in heart, kidney, and pancreas, lowly expressed in brain, lung, skeletal muscle [Shworak, J Biol Chem 1999] - transfers sulfate to C3 position of IdoA2S-GlcNH2 [Sugahara, IUBMB Life 2002] 9952: - cDNA was isolated and expressed [Shworak, J Biol Chem 1999] - functionality remains to be established [Shworak, J Biol Chem 1999]; assumed to have same functionality as HS3ST3B1 64711:
S3TASE3ly	2	Leder IG	A novel 3-O sulfatase from human urine acting on methyl-2-deoxy-2-sulfamino-alphs-D-glucopyranoside 3-sulfate	Biochem Biophys Res Commun	1980	7396957	- heparan sulfate contains a small amount of glucosamine O-sulfated at the C3 position, but it is not known which sulfatase is responsible for the hydrolysis of this sulfate ester [Winchester 1996] - the 3-O-sulfatase described by [Leder 1980] would not function on this residue since it is not N-sulfated

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S4T1g	3	Evers MR, Xia G, Kang HG, Schachner M, Baenziger JU	Molecular cloning and characterization of a dermatan specific N-acetylgalactosamine 4-O-sulfotransferase	J Biol Chem	2001	1147079	- Golgi localization [Silbert, IUBMB LIfe 2002] - sulfation of Gal residues may be catalyzed byt the same sulfotransferases that add sulfate to GalNAc of chondroitin [Silbert, IUBMB LIfe 2002] 50515: - gene was cloned [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000], [Hiraoka, J Biol Chem 2000] - protein has 29% identity and 48% similarity to rat protein [Yamauchi, J Biol Chem 2000], 96% similarity to mouse protein [Okuda, J Biochem (Tokyo) 2000] - expressed ubiquitously [Okuda, J Biochem (Tokyo) 2000]; mainly expressed in brain and kidney [Yamauchi, J Biol Chem 2000]; predominantly expressed in peripheral leukocytes and hematopoietic tissues [Hiraoka, J Biol Chem 2000] - 4-sulfotransferase activity verified for chondroitin sulfate and desulfated dermatan sulfate [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000] 55501: - gene was cloned and expressed [Hiraoka, J Biol Chem 2000] - widely expressed in various tissues [Hiraoka, J Biol Chem 2000] 166012: - gene was identified based on sequence homology with other HNK-1 family members; was cloned and expressed [Kang, J Bi - expressed in adult liver and at lower levels in adult kidney, ly - similar specificty to C4ST-1 [Kang, J Biol Chem 2002] 113189: - identified based on sequence homology to HNK-1 family [Ev - transfers sulfate to 4 position of GalNAc that is next to IdoA - sulfation occurs after epimerization of GlcA to IdoA [Evers, J
S4T1g	3	Yamauchi S, Mita S, Matsubara T, Fukuta M, Habuchi H, Kimata K, Habuchi O	Molecular cloning and expression of chondroitin 4-sulfotransferase	J Biol Chem	2000	10722746	- Golgi localization [Silbert, IUBMB LIfe 2002] - sulfation of Gal residues may be catalyzed byt the same sulfotransferases that add sulfate to GalNAc of chondroitin [Silbert, IUBMB LIfe 2002] 50515: - gene was cloned [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000], [Hiraoka, J Biol Chem 2000] - protein has 29% identity and 48% similarity to rat protein [Yamauchi, J Biol Chem 2000], 96% similarity to mouse protein [Okuda, J Biochem (Tokyo) 2000] - expressed ubiquitously [Okuda, J Biochem (Tokyo) 2000]; mainly expressed in brain and kidney [Yamauchi, J Biol Chem 2000]; predominantly expressed in peripheral leukocytes and hematopoietic tissues [Hiraoka, J Biol Chem 2000] - 4-sulfotransferase activity verified for chondroitin sulfate and desulfated dermatan sulfate [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000] 55501: - gene was cloned and expressed [Hiraoka, J Biol Chem 2000] - widely expressed in various tissues [Hiraoka, J Biol Chem 2000] 166012: - gene was identified based on sequence homology with other HNK-1 family members; was cloned and expressed [Kang, J Bi - expressed in adult liver and at lower levels in adult kidney, ly - similar specificty to C4ST-1 [Kang, J Biol Chem 2002] 113189: - identified based on sequence homology to HNK-1 family [Ev - transfers sulfate to 4 position of GalNAc that is next to IdoA - sulfation occurs after epimerization of GlcA to IdoA [Evers, J
S4T1g	3	Hiraoka N, Nakagawa H, Ong E, Akama TO, Fukuda MN, Fukuda M	Molecular cloning and expression of two distinct human chondroitin 4-O-sulfotransferases that belong to the HNK-1 sulfotransferase gene family	J Biol Chem	2000	10781601	- Golgi localization [Silbert, IUBMB LIfe 2002] - sulfation of Gal residues may be catalyzed byt the same sulfotransferases that add sulfate to GalNAc of chondroitin [Silbert, IUBMB LIfe 2002] 50515: - gene was cloned [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000], [Hiraoka, J Biol Chem 2000] - protein has 29% identity and 48% similarity to rat protein [Yamauchi, J Biol Chem 2000], 96% similarity to mouse protein [Okuda, J Biochem (Tokyo) 2000] - expressed ubiquitously [Okuda, J Biochem (Tokyo) 2000]; mainly expressed in brain and kidney [Yamauchi, J Biol Chem 2000]; predominantly expressed in peripheral leukocytes and hematopoietic tissues [Hiraoka, J Biol Chem 2000] - 4-sulfotransferase activity verified for chondroitin sulfate and desulfated dermatan sulfate [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000] 55501: - gene was cloned and expressed [Hiraoka, J Biol Chem 2000] - widely expressed in various tissues [Hiraoka, J Biol Chem 2000] 166012: - gene was identified based on sequence homology with other HNK-1 family members; was cloned and expressed [Kang, J Bi - expressed in adult liver and at lower levels in adult kidney, ly - similar specificty to C4ST-1 [Kang, J Biol Chem 2002] 113189: - identified based on sequence homology to HNK-1 family [Ev - transfers sulfate to 4 position of GalNAc that is next to IdoA - sulfation occurs after epimerization of GlcA to IdoA [Evers, J

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S4T1g	3	Okuda T, Mita S, Yamauchi S, Matsubara T, Yagi F, Yamamori D, Fukuta M, Kuroiwa A, Matsuda Y, Habuchi O	Molecular cloning, expression, and chromosomal mapping of human chondroitin 4-sulfotransferase, whose expression pattern in human tissues is different from that of chondroitin 6-sulfotransferase	J Biochem (Tokyo)	2000	11056388	- Golgi localization [Silbert, IUBMB Life 2002] - sulfation of Gal residues may be catalyzed byt the same sulfotransferases that add sulfate to GalNAc of chondroitin [Silbert, IUBMB LIfe 2002] 50515: - gene was cloned [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000], [Hiraoka, J Biol Chem 2000] - protein has 29% identity and 48% similarity to rat protein [Yamauchi, J Biol Chem 2000], 96% similarity to mouse protein [Okuda, J Biochem (Tokyo) 2000] - expressed ubiquitously [Okuda, J Biochem (Tokyo) 2000]; mainly expressed in brain and kidney [Yamauchi, J Biol Chem 2000]; predominantly expressed in peripheral leukocytes and hematopoietic tissues [Hiraoka, J Biol Chem 2000] - 4-sulfotransferase activity verified for chondroitin sulfate and desulfated dermatan sulfate [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000] 55501: - gene was cloned and expressed [Hiraoka, J Biol Chem 2000] - widely expressed in various tissues [Hiraoka, J Biol Chem 2000] 166012: - gene was identified based on sequence homology with other HNK-1 family members; was cloned and expressed [Kang, J Bi - expressed in adult liver and at lower levels in adult kidney, ly - similar specificty to C4ST-1 [Kang, J Biol Chem 2002] 113189: - identified based on sequence homology to HNK-1 family [Ev - transfers sulfate to 4 position of GalNAc that is next to IdoA - sulfation occurs after epimerization of GlcA to IdoA [Evers, J
S4T1g	3	Kang HG, Evers MR, Xia G, Baenziger JU, Schachner M	Molecular cloning and characterization of chondroitin-4-O-sulfotransferase-3. A novel member of the HNK-1 family of sulfotransferases	J Biol Chem	2002	12080076	- Golgi localization [Silbert, IUBMB Life 2002] - sulfation of Gal residues may be catalyzed byt the same sulfotransferases that add sulfate to GalNAc of chondroitin [Silbert, IUBMB LIfe 2002] 50515: - gene was cloned [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000], [Hiraoka, J Biol Chem 2000] - protein has 29% identity and 48% similarity to rat protein [Yamauchi, J Biol Chem 2000], 96% similarity to mouse protein [Okuda, J Biochem (Tokyo) 2000] - expressed ubiquitously [Okuda, J Biochem (Tokyo) 2000]; mainly expressed in brain and kidney [Yamauchi, J Biol Chem 2000]; predominantly expressed in peripheral leukocytes and hematopoietic tissues [Hiraoka, J Biol Chem 2000] - 4-sulfotransferase activity verified for chondroitin sulfate and desulfated dermatan sulfate [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000] 55501: - gene was cloned and expressed [Hiraoka, J Biol Chem 2000] - widely expressed in various tissues [Hiraoka, J Biol Chem 2000] 166012: - gene was identified based on sequence homology with other HNK-1 family members; was cloned and expressed [Kang, J Bi - expressed in adult liver and at lower levels in adult kidney, ly - similar specificty to C4ST-1 [Kang, J Biol Chem 2002] 113189: - identified based on sequence homology to HNK-1 family [Ev - transfers sulfate to 4 position of GalNAc that is next to IdoA - sulfation occurs after epimerization of GlcA to IdoA [Evers, J
S4T1g	3	Mikami T, Mizumoto S, Kago N, Kitagawa H, Sugahara K.	Specificities of three distinct human chondroitin/dermatan N-acetylgalactosamine 4-O-sulfotransferases demonstrated using partially desulfated dermatan sulfate as an acceptor: implication of differential roles in dermatan sulfate biosynthesis.	J Biol Chem	2003	12847091	- Golgi localization [Silbert, IUBMB Life 2002] - sulfation of Gal residues may be catalyzed byt the same sulfotransferases that add sulfate to GalNAc of chondroitin [Silbert, IUBMB LIfe 2002] 50515: - gene was cloned [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000], [Hiraoka, J Biol Chem 2000] - protein has 29% identity and 48% similarity to rat protein [Yamauchi, J Biol Chem 2000], 96% similarity to mouse protein [Okuda, J Biochem (Tokyo) 2000] - expressed ubiquitously [Okuda, J Biochem (Tokyo) 2000]; mainly expressed in brain and kidney [Yamauchi, J Biol Chem 2000]; predominantly expressed in peripheral leukocytes and hematopoietic tissues [Hiraoka, J Biol Chem 2000] - 4-sulfotransferase activity verified for chondroitin sulfate and desulfated dermatan sulfate [Yamauchi, J Biol Chem 2000], [Okuda, J Biochem (Tokyo) 2000] 55501: - gene was cloned and expressed [Hiraoka, J Biol Chem 2000] - widely expressed in various tissues [Hiraoka, J Biol Chem 2000] 166012: - gene was identified based on sequence homology with other HNK-1 family members; was cloned and expressed [Kang, J Bi - expressed in adult liver and at lower levels in adult kidney, ly - similar specificty to C4ST-1 [Kang, J Biol Chem 2002] 113189: - identified based on sequence homology to HNK-1 family [Ev - transfers sulfate to 4 position of GalNAc that is next to IdoA - sulfation occurs after epimerization of GlcA to IdoA [Evers, J

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
S4TASE1ly	3	Schuchman EH, Jackson CE, Desnick RJ	Human arylsulfatase B: MOPAC cloning, nucleotide sequence of a full-length cDNA, and regions of amino acid identity with arylsulfatases A and C	Genomics	1990	1968043	411: - hydrolyzes sulfate groups of GalNAc in chondroitin sulfate and dermatan sulfate [RefSeq] - lysosomal [RefSeq], [Peters 1990] - isolated from hepatoma cell cDNA library; seq used to isolate ORF from human testis library [Schuchman 1990] - isolated and overexpressed in baby hamster kidney cellls [Peters 1990]
S4TASE1ly	3	Peters C, Schmidt B, Rommerskirch W, Rupp K, Zuhlsdorf M, Vingron M, Meyer HE, Pohlmann R, von Figura K	Phylogenetic conservation of arylsulfatases. cDNA cloning and expression of human arylsulfatase B.	J Biol Chem	1990	2303452	411: - hydrolyzes sulfate groups of GalNAc in chondroitin sulfate and dermatan sulfate [RefSeq] - lysosomal [RefSeq], [Peters 1990] - isolated from hepatoma cell cDNA library; seq used to isolate ORF from human testis library [Schuchman 1990] - isolated and overexpressed in baby hamster kidney cellls [Peters 1990]
S6T19g	3	Tsutsumi K, Shimakawa H, Kitagawa H, Sugahara K	Functional expression and genomic structure of human chondroitin 6-sulfotransferase	FEBS Lett	1998	9883891	- Golgi localization [Silbert, IUBMB LIfe 2002] - sulfation of Gal residues may be catalyzed byt the same sulfotransferases that add sulfate to GalNAc of chondroitin [Silbert, IUBMB LIfe 2002] 9469: - Golgi [UniProt] - gene identified and cDNA isolated, has 74% identity to chick ortholog [Fukuda, Biochim Biophys Acta 1998] - highly expressed in sk muscle, lower levels in heart, placenta, pancreas [Fukuda, Biochim Biophys Acta 1998] - keratan sulfate was sulfated by C6ST at position 6 of Gal residues [Habuchi, Glycobiology 1996] - sulfates GalNAc residues in GlcAGalNAc disaccharides, but not IdoAGalNAc or GlcAGalNAc4S [Tsutsumi, FEBS Lett 1998] 56548: - gene identified by BLAST [Kitagawa, J Biol Chem 2000], [Bhakta, J Biol Chem 2000] - gene was cloned and expressed [Kitagawa, J Biol Chem 2000], [Uchimura, Biochem Biophys Res Commun 2000], [Bhakta, J Biol Chem 2000] - predominant activity is 6-sulfation of GalNAc in chondroitin, little to no activity w/ keratan sulfate [Kitagawa, J Biol Chem 2000] - expression varied during development, persisted through adulthood in spleen [Kitagawa, J Biol Chem 2000], primarily in
S6T19g	3	Kitagawa H, Fujita M, Ito N, Sugahara K	Molecular cloning and expression of a novel chondroitin 6-O-sulfotransferase	J Biol Chem	2000	10781596	- Golgi localization [Silbert, IUBMB LIfe 2002] - sulfation of Gal residues may be catalyzed byt the same sulfotransferases that add sulfate to GalNAc of chondroitin [Silbert, IUBMB LIfe 2002] 9469: - Golgi [UniProt] - gene identified and cDNA isolated, has 74% identity to chick ortholog [Fukuda, Biochim Biophys Acta 1998] - highly expressed in sk muscle, lower levels in heart, placenta, pancreas [Fukuda, Biochim Biophys Acta 1998] - keratan sulfate was sulfated by C6ST at position 6 of Gal residues [Habuchi, Glycobiology 1996] - sulfates GalNAc residues in GlcAGalNAc disaccharides, but not IdoAGalNAc or GlcAGalNAc4S [Tsutsumi, FEBS Lett 1998] 56548: - gene identified by BLAST [Kitagawa, J Biol Chem 2000], [Bhakta, J Biol Chem 2000] - gene was cloned and expressed [Kitagawa, J Biol Chem 2000], [Uchimura, Biochem Biophys Res Commun 2000], [Bhakta, J Biol Chem 2000] - predominant activity is 6-sulfation of GalNAc in chondroitin, little to no activity w/ keratan sulfate [Kitagawa, J Biol Chem 2000] - expression varied during development, persisted through adulthood in spleen [Kitagawa, J Biol Chem 2000], primarily in

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S6T19g	3	Bhakta S, Bartes A, Bowman KG, Kao WM, Polsky I, Lee JK, Cook BN, Bruehl RE, Rosen SD, Bertozzi CR, Hemmerich S	Sulfation of N-acetylglucosamine by chondroitin 6-sulfotransferase 2 (GST-5)	J Biol Chem	2000	10956661	- Golgi localization [Silbert, IUBMB LIfe 2002] - sulfation of Gal residues may be catalyzed byt the same sulfotransferases that add sulfate to GalNAc of chondroitin [Silbert, IUBMB LIfe 2002] 9469: - Golgi [UniProt] - gene identified and cDNA isolated, has 74% identity to chick ortholog [Fukuda, Biochim Biophys Acta 1998] - highly expressed in sk muscle, lower levels in heart, placenta, pancreas [Fukuda, Biochim Biophys Acta 1998] - keratan sulfate was sulfated by C6ST at position 6 of Gal residues [Habuchi, Glycobiology 1996] - sulfates GalNAc residues in GlcAGalNAc disaccharides, but not IdoAGalNAc or GlcAGalNAc4S [Tsutsumi, FEBS Lett 1998] 56548: - gene identified by BLAST [Kitagawa, J Biol Chem 2000], [Bhakta, J Biol Chem 2000] - gene was cloned and expressed [Kitagawa, J Biol Chem 2000], [Uchimura, Biochem Biophys Res Commun 2000], [Bhakta, J Biol Chem 2000] - predominant activity is 6-sulfation of GalNAc in chondroitin, little to no activity w/ keratan sulfate [Kitagawa, J Biol Chem 2000] - expression varied during development, persisted through adulthood in spleen [Kitagawa, J Biol Chem 2000], primarily in
S6T1g	3	Uchimura K, Muramatsu H, Kaname T, Ogawa H, Yamakawa T, Fan QW, Mitsuoka C, Kannagi R, Habuchi O, Yokoyama I, Yamamura K, Ozaki T, Nakagawara A, Kadomatsu K, Muramatsu T	Human N-acetylglucosamine-6-O-sulfotransferase involved in the biosynthesis of 6-sulfo sialyl Lewis X: molecular cloning, chromosomal mapping, and expression in various organs and tumor cells	J Biochem (Tokyo)	1998	9722682	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 4166: - gene was cloned [Uchimura, Biochem Biophys Res Commun 2000] and biochemically characterized [Akama, J Biol Chem 2001] - highly expressed in heart, spleen, and ovary [Uchimura, Biochem Biophys Res Commun 2000] 9435: - Golgi [de Graffenried, J Biol Chem 2003] - gene was identified and cloned, 99% identical to mouse ortholog [Uchimura, J Biochem (Tokyo) 1998] - strongly expressed in the bone marrow, peripheral blood leukocytes, spleen, brain, spinal cord, ovary, and placenta, and moderately expressed in many organs including lymph nodes and thymus [Uchimura, J Biochem (Tokyo) 1998] 10164: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned [Bistrup, J Cell Biol 1999] 23563: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned and characterized [Lee Biochem Biophys Res Commun 1999] - expressed in small intestine and colon [Lee Biochem Biophys Res Commun 1999]
S6T1g	3	Bistrup A, Bhakta S, Lee JK, Belov YY, Gunn MD, Zuo FR, Huang CC, Kannagi R, Rosen SD, Hemmerich S	Sulfotransferases of two specificities function in the reconstitution of high endothelial cell ligands for L-selectin	J Cell Biol	1999	10330415	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 4166: - gene was cloned [Uchimura, Biochem Biophys Res Commun 2000] and biochemically characterized [Akama, J Biol Chem 2001] - highly expressed in heart, spleen, and ovary [Uchimura, Biochem Biophys Res Commun 2000] 9435: - Golgi [de Graffenried, J Biol Chem 2003] - gene was identified and cloned, 99% identical to mouse ortholog [Uchimura, J Biochem (Tokyo) 1998] - strongly expressed in the bone marrow, peripheral blood leukocytes, spleen, brain, spinal cord, ovary, and placenta, and moderately expressed in many organs including lymph nodes and thymus [Uchimura, J Biochem (Tokyo) 1998] 10164: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned [Bistrup, J Cell Biol 1999] 23563: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned and characterized [Lee Biochem Biophys Res Commun 1999] - expressed in small intestine and colon [Lee Biochem Biophys Res Commun 1999]

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S6T1g	3	Lee JK, Bhakta S, Rosen SD, Hemmerich S	Cloning and characterization of a mammalian N-acetylglucosamine-6-sulfotransferase that is highly restricted to intestinal tissue	Biochem Biophys Res Commun	1999	10491328	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 4166: - gene was cloned [Uchimura, Biochem Biophys Res Commun 2000] and biochemically characterized [Akama, J Biol Chem 2001] - highly expressed in heart, spleen, and ovary [Uchimura, Biochem Biophys Res Commun 2000] 9435: - Golgi [de Graffenried, J Biol Chem 2003] - gene was identified and cloned, 99% identical to mouse ortholog [Uchimura, J Biochem (Tokyo) 1998] - strongly expressed in the bone marrow, peripheral blood leukocytes, spleen, brain, spinal cord, ovary, and placenta, and moderately expressed in many organs including lymph nodes and thymus [Uchimura, J Biochem (Tokyo) 1998] 10164: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned [Bistrup, J Cell Biol 1999] 23563: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned and characterized [Lee Biochem Biophys Res Commun 1999] - expressed in small intestine and colon [Lee Biochem Biophys Res Commun 1999]
S6T1g	3	Uchimura K, Fasakhany F, Kadomatsu K, Matsukawa T, Yamakawa T, Kurosawa N, Muramatsu T	Diversity of N-acetylglucosamine-6-O-sulfotransferases: molecular cloning of a novel enzyme with different distribution and specificities	Biochem Biophys Res Commun	2000	10913333	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 4166: - gene was cloned [Uchimura, Biochem Biophys Res Commun 2000] and biochemically characterized [Akama, J Biol Chem 2001] - highly expressed in heart, spleen, and ovary [Uchimura, Biochem Biophys Res Commun 2000] 9435: - Golgi [de Graffenried, J Biol Chem 2003] - gene was identified and cloned, 99% identical to mouse ortholog [Uchimura, J Biochem (Tokyo) 1998] - strongly expressed in the bone marrow, peripheral blood leukocytes, spleen, brain, spinal cord, ovary, and placenta, and moderately expressed in many organs including lymph nodes and thymus [Uchimura, J Biochem (Tokyo) 1998] 10164: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned [Bistrup, J Cell Biol 1999] 23563: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned and characterized [Lee Biochem Biophys Res Commun 1999] - expressed in small intestine and colon [Lee Biochem Biophys Res Commun 1999]
S6T1g	3	Akama TO, Nakayama J, Nishida K, Hiraoka N, Suzuki M, McAuliffe J, Hindsgaul O, Fukuda M, Fukuda MN	Human corneal GlcNac 6-O-sulfotransferase and mouse intestinal GlcNac 6-O-sulfotransferase both produce keratan sulfate	J Biol Chem	2001	11278593	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 4166: - gene was cloned [Uchimura, Biochem Biophys Res Commun 2000] and biochemically characterized [Akama, J Biol Chem 2001] - highly expressed in heart, spleen, and ovary [Uchimura, Biochem Biophys Res Commun 2000] 9435: - Golgi [de Graffenried, J Biol Chem 2003] - gene was identified and cloned, 99% identical to mouse ortholog [Uchimura, J Biochem (Tokyo) 1998] - strongly expressed in the bone marrow, peripheral blood leukocytes, spleen, brain, spinal cord, ovary, and placenta, and moderately expressed in many organs including lymph nodes and thymus [Uchimura, J Biochem (Tokyo) 1998] 10164: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned [Bistrup, J Cell Biol 1999] 23563: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned and characterized [Lee Biochem Biophys Res Commun 1999] - expressed in small intestine and colon [Lee Biochem Biophys Res Commun 1999]

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S6T1g	3	Grunwell JR, Bertozzi CR	Carbohydrate sulfotransferases of the GalNAc/Gal/GlcNAc6ST family	Biochemistry	2002	12403612	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 4166: - gene was cloned [Uchimura, Biochem Biophys Res Commun 2000] and biochemically characterized [Akama, J Biol Chem 2001] - highly expressed in heart, spleen, and ovary [Uchimura, Biochem Biophys Res Commun 2000] 9435: - Golgi [de Graffenried, J Biol Chem 2003] - gene was identified and cloned, 99% identical to mouse ortholog [Uchimura, J Biochem (Tokyo) 1998] - strongly expressed in the bone marrow, peripheral blood leukocytes, spleen, brain, spinal cord, ovary, and placenta, and moderately expressed in many organs including lymph nodes and thymus [Uchimura, J Biochem (Tokyo) 1998] 10164: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned [Bistrup, J Cell Biol 1999] 23563: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned and characterized [Lee Biochem Biophys Res Commun 1999] - expressed in small intestine and colon [Lee Biochem Biophys Res Commun 1999]
S6T1g	3	de Graffenried CL, Bertozzi CR	Golgi localization of carbohydrate sulfotransferases is a determinant of L-selectin ligand biosynthesis	J Biol Chem	2003	12855678	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 4166: - gene was cloned [Uchimura, Biochem Biophys Res Commun 2000] and biochemically characterized [Akama, J Biol Chem 2001] - highly expressed in heart, spleen, and ovary [Uchimura, Biochem Biophys Res Commun 2000] 9435: - Golgi [de Graffenried, J Biol Chem 2003] - gene was identified and cloned, 99% identical to mouse ortholog [Uchimura, J Biochem (Tokyo) 1998] - strongly expressed in the bone marrow, peripheral blood leukocytes, spleen, brain, spinal cord, ovary, and placenta, and moderately expressed in many organs including lymph nodes and thymus [Uchimura, J Biochem (Tokyo) 1998] 10164: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned [Bistrup, J Cell Biol 1999] 23563: - Golgi [de Graffenried, J Biol Chem 2003] - gene was cloned and characterized [Lee Biochem Biophys Res Commun 1999] - expressed in small intestine and colon [Lee Biochem Biophys Res Commun 1999]
S6T25g	3	Habuchi H, Kobayashi M, Kimata K	Molecular characterization and expression of heparan sulfate 6-sulfotransferase. Complete cDNA cloning in human and partial cloning in Chinese hamster ovary cells	J Biol Chem	1998	9535912	- transfers sulfate to C6-position of NSGlcNAc residues im HS but not GlcNAc [Sugahara, IUBMB Life 2002] - all 3 isoforms transfer sulfate to heparan sulfate / heparin but not chondoroitin or keratan sulfate [Sugahara, IUBMB Life 2002] 9394: - mouse protein preferentially sulfates IdoA-NSGlcNAc [Sugahara, IUBMB Life 2002] - cDNA was cloned and expressed [Habuchi, J Biol Chem 1998] - expressed ubiquitously, but highest levels in adrenal gland, kidney, liver, intestine, foetal brain and fetal kidney [Habuchi, Biochem J 2003] 90161: - mouse protein preferentially sulfates GlcA residue [Sugahara, IUBMB Life 2002] - cDNA was cloned and expressed [Habuchi, Biochem J 2003] - encodes two transcripts -- a long and short form [Habuchi, Biochem J 2003] - long form xclusively expressed in adult and foetal brain tissues, short form preferentially expressed in ovary, placenta and foetal kidney [Habuchi, Biochem J 2003] 26672: - mouse protein sulfates IdoA-NSGlcNAc and GlcA equally [Sugahara, IUBMB Life 2002] - cDNA was identified through high-throughput study [Ota, Nat Genet 2004]

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S6T25g	3	Habuchi H, Miyake G, Nogami K, Kuroiwa A, Matsuda Y, Kusche-Gullberg M, Habuchi O, Tanaka M, Kimata K	Biosynthesis of heparan sulphate with diverse structures and functions: two alternatively spliced forms of human heparan sulphate 6-O-sulphotransferase-2 having different expression patterns and properties	Biochem J	2003	12492399	- transfers sulfate to C6-position of NSGlcNAc residues im HS but not GlcNAc [Sugahara, IUBMB Life 2002] - all 3 isoforms transfer sulfate to heparan sulfate / heparin but not chondoroitin or keratan sulfate [Sugahara, IUBMB Life 2002] 9394: - mouse protein preferentially sulfates IdoA-NSGlcNAc [Sugahara, IUBMB Life 2002] - cDNA was cloned and expressed [Habuchi, J Biol Chem 1998] - expressed ubiquitously, but highest levels in adrenal gland, kidney, liver, intestine, foetal brain and fetal kidney [Habuchi, Biochem J 2003] 90161: - mouse protein preferentially sulfates GlcA residue [Sugahara, IUBMB Life 2002] - cDNA was cloned and expressed [Habuchi, Biochem J 2003] - encodes two transcripts -- a long and short form [Habuchi, Biochem J 2003] - long form xclusively expressed in adult and foetal brain tissues, short form preferentially expressed in ovary, placenta and foetal kidney [Habuchi, Biochem J 2003] 26672: - mouse protein sulfates IdoA-NSGlcNAc and GlcA equally [Sugahara, IUBMB Life 2002] - cDNA was identified through high-throughput study [Ota, Nat Genet 2004]
S6T25g	3	Ota T, Suzuki Y, Nishikawa T, Otsuki T, Sugiyama T, Irie R, Wakamatsu A, Hayashi K, Sato H, Nagai K, Kimura K, et al	Complete sequencing and characterization of 21,243 full-length human cDNAs.	Nat Genet	2004	14702039	- transfers sulfate to C6-position of NSGlcNAc residues im HS but not GlcNAc [Sugahara, IUBMB Life 2002] - all 3 isoforms transfer sulfate to heparan sulfate / heparin but not chondoroitin or keratan sulfate [Sugahara, IUBMB Life 2002] 9394: - mouse protein preferentially sulfates IdoA-NSGlcNAc [Sugahara, IUBMB Life 2002] - cDNA was cloned and expressed [Habuchi, J Biol Chem 1998] - expressed ubiquitously, but highest levels in adrenal gland, kidney, liver, intestine, foetal brain and fetal kidney [Habuchi, Biochem J 2003] 90161: - mouse protein preferentially sulfates GlcA residue [Sugahara, IUBMB Life 2002] - cDNA was cloned and expressed [Habuchi, Biochem J 2003] - encodes two transcripts -- a long and short form [Habuchi, Biochem J 2003] - long form xclusively expressed in adult and foetal brain tissues, short form preferentially expressed in ovary, placenta and foetal kidney [Habuchi, Biochem J 2003] 26672: - mouse protein sulfates IdoA-NSGlcNAc and GlcA equally [Sugahara, IUBMB Life 2002] - cDNA was identified through high-throughput study [Ota, Nat Genet 2004]
S6T3g	3	Habuchi O, Hirahara Y, Uchimura K, Fukuta M	Enzymatic sulfation of galactose residue of keratan sulfate by chondroitin 6-sulfotransferase	Glycobiology	1996	8991509	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 9469: - Golgi [UniProt] - gene identified and cDNA isolated, has 74% identity to chick ortholog [Fukuda, Biochim Biophys Acta 1998] - highly expressed in sk muscle, lower levels in heart, placenta, pancreas [Fukuda, Biochim Biophys Acta 1998] - keratan sulfate was sulfated by C6ST at position 6 of Gal residues [Habuchi, Glycobiology 1996] 8534: - gene was cloned and characterized [Fukuta, J Biol Chem 1997] - sulfates position 6 of Gal residues [Fukuta, J Biol Chem 1997] - expressed primarily in brain [Fukuta, J Biol Chem 1997]

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S6T3g	3	Fukuta M, Inazawa J, Torii T, Tsuzuki K, Shimada E, Habuchi O	Molecular cloning and characterization of human keratan sulfate Gal-6-sulfotransferase	J Biol Chem	1997	9405439	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 9469: - Golgi [UniProt] - gene identified and cDNA isolated, has 74% identity to chick ortholog [Fukuda, Biochim Biophys Acta 1998] - highly expressed in sk muscle, lower levels in heart, placenta, pancreas [Fukuda, Biochim Biophys Acta 1998] - keratan sulfate was sulfated by C6ST at position 6 of Gal residues [Habuchi, Glycobiology 1996] 8534: - gene was cloned and characterized [Fukuta, J Biol Chem 1997] - sulfates position 6 of Gal residues [Fukuta, J Biol Chem 1997] - expressed primarily in brain [Fukuta, J Biol Chem 1997]
S6T3g	3	Fukuta M, Kobayashi Y, Uchimura K, Kimata K, Habuchi O	Molecular cloning and expression of human chondroitin 6-sulfotransferase	Biochim Biophys Acta	1998	9714738	- refs, substrate specificity, and tissue distribution of GlcNac/Gal-6-O-sulfotransferases [Grunwell, Biochemistry 2002] 9469: - Golgi [UniProt] - gene identified and cDNA isolated, has 74% identity to chick ortholog [Fukuda, Biochim Biophys Acta 1998] - highly expressed in sk muscle, lower levels in heart, placenta, pancreas [Fukuda, Biochim Biophys Acta 1998] - keratan sulfate was sulfated by C6ST at position 6 of Gal residues [Habuchi, Glycobiology 1996] 8534: - gene was cloned and characterized [Fukuta, J Biol Chem 1997] - sulfates position 6 of Gal residues [Fukuta, J Biol Chem 1997] - expressed primarily in brain [Fukuta, J Biol Chem 1997]
S6TASE1ly	3	Robertson DA, Freeman C, Nelson PV, Morris CP, Hopwood JJ	Human glucosamine-6-sulfatase cDNA reveals homology with steroid sulfatase	Biochem Biophys Res Commun	1988	3196333	- lysosomal enzyme; ubiquitous [RefSeq] - cloned [Robertson 1988] - purified from human liver [Freeman 1987] - kinetic characterization [Freeman 1987]
S6TASE1ly	3	Freeman C, Clements PR, Hopwood JJ	Human liver N-acetylglucosamine-6-sulphate sulphatase. Purification and characterization	Biochem J	1987	3689314	- lysosomal enzyme; ubiquitous [RefSeq] - cloned [Robertson 1988] - purified from human liver [Freeman 1987] - kinetic characterization [Freeman 1987]
S6TASE1ly	3	Freeman C, Hopwood JJ	Human liver N-acetylglucosamine-6-sulphate sulphatase. Catalytic properties	Biochem J	1987	3689315	- lysosomal enzyme; ubiquitous [RefSeq] - cloned [Robertson 1988] - purified from human liver [Freeman 1987] - kinetic characterization [Freeman 1987]
SACCD3m	3	Sacksteder,K.A., Biery,B.J., Morrell,J.C., Goodman,B.K., Geisbrecht,B.V., Cox,R.P., Gould,S.J., Geraghty,M.T.,	Identification of the alpha-aminoadipic semialdehyde synthase gene, which is defective in familial hyperlysinemia.		2000	10775527	Mutations in this gene are associated with familial hyperlysinemia.
SADT	3	Fuda H, Shimizu C, Lee YC, Akita H, Strott CA.	Characterization and expression of human bifunctional 3'-phosphoadenosine 5'-phosphosulphate synthase isoforms.	Biochem J	2002	11931637	bifunctional enzyme In brain and skin PAPSS1 is the major expressed isoform, whereas in liver, cartilage and adrenal glands PAPSS2 isoform expression predominates and in various other tissues the proportions of the isoform expressions is purported to vary IT
SADT	3	Venkatachalam KV.	Human 3'-phosphoadenosine 5'-phosphosulfate (PAPS) synthase: biochemistry, molecular biology and genetic deficiency.	IUBMB Life	2003	12716056	bifunctional enzyme In brain and skin PAPSS1 is the major expressed isoform, whereas in liver, cartilage and adrenal glands PAPSS2 isoform expression predominates and in various other tissues the proportions of the isoform expressions is purported to vary IT
SALMCOM2	2	Schluckebier G, O'Gara M, Saenger W, Cheng X.	Universal catalytic domain structure of AdoMet-dependent methyltransferases.	J Mol Biol	1995	7897657	cytosol - uniprot biochem function by seq similarity. Schluckebier - PMID ref 7897657 notes ability of enzyme to methylate wide-rage of catechols NJ
SAMHISTA	3	Yamauchi K, Sekizawa K, Suzuki H, Nakazawa H, Ohkawara Y, Katayose D, Ohtsu H, Tamura G, Shibahara S, Takemura M, et al	Structure and function of human histamine N-methyltransferase: critical enzyme in histamine metabolism in airway	Am J Physiol	1994	7943261	This reaction is important in the airway and probably elsewhere as well.
SARCOXp	2	Dodt G, Kim DG, Reimann SA, Reuber BE, McCabe K, Gould SJ, Mihalik SJ.	L-Pipecolic acid oxidase, a human enzyme essential for the degradation of L-pipecolic acid, is most similar to the monomeric sarcosine oxidases.		2000	10642506	- recombinant enzyme oxidized both L-pipecolic acid and sarcosine. However, PBD patients who lack the enzyme activity accumulate only L-pipecolic acid, suggesting that in humans in vivo, this enzyme is involved mainly in the degradation of L-pipecolic acid (Dodt et. al)

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SARCStex	2	Glorieux FH, Scriver CR, Delvin E, Mohyuddin F.	Transport and metabolism of sarcosine in hypersarcosinemic and normal phenotypes.		1971	5096515	(PMID: 5096515): patients show sarcosine absorption/clearance from plasma MM
SARDHm	3	Eschenbrenner M, Jorns MS.	Cloning and mapping of the cDNA for human sarcosine dehydrogenase, a flavoenzyme defective in patients with sarcosinemia.	Genomics	1999	10444331	0
SBPP1er	3	Johnson KR, Johnson KY, Becker KP, Bielawski J, Mao C, Obeid LM.	Role of human sphingosine-1-phosphate phosphatase 1 in the regulation of intra- and extracellular sphingosine-1-phosphate levels and cell viability.	J Biol Chem	2003	12815058	ER - uniprot and johnson ref Has enzymatic activity against both sphingosine 1 phosphate (S1P) and dihydro-S1P. Regulates intracellular and extracellular S1P levels. NJ
SBTD_D2	0	El-Kabbani O, Darmanin C, Chung RP	Sorbitol dehydrogenase: structure, function and ligand design	Curr Med Chem	2004	14965227	- strictly uses NAD as cofactor [El-Kabbani et al, Curr Med Chem. 2004 Feb;11(4):465-76] - found in the liver, ovaries, sperm, and seminal vesicles [Champe, Biochemistry 2005]
SCP21x	3	Yamamoto R, Kallen CB, Babalola GO, Rennert H, Billheimer JT, Strauss JF 3rd.	Cloning and expression of a cDNA encoding human sterol carrier protein 2.	Proc Natl Acad Sci U S A	1991	1703300	This gene encodes two proteins: sterol carrier protein X (SCPx) and sterol carrier protein 2 (SCP2), as a result of transcription initiation from 2 independently regulated promoters. The transcript initiated from the proximal promoter encodes the longer SCPx protein, and the transcript initiated from the distal promoter encodes the shorter SCP2 protein, with the 2 proteins sharing a common C-terminus. Evidence suggests that the SCPx protein is a peroxisome-associated thiolase that is involved in the oxidation of branched chain fatty acids, while the SCP2 protein is thought to be an intracellular lipid transfer protein. This gene is highly expressed in organs involved in lipid metabolism, and may play a role in Zellweger syndrome, in which cells are deficient in peroxisomes and have impaired bile acid synthesis. Alternative splicing of this gene produces multiple transcript variants, some encoding different isoforms. The full-length nature of all transcript variants has not been determined. NJ
SCPx	3	Mukherji M, Kershaw NJ, Schofield CH, Wierzbicki AS, Lloyd MD	Utilization of sterol carrier protein-2 by phytanoyl-CoA 2-hydroxylase in the peroxisomal alpha oxidation of phytanic acid	Chemistry and Biology	2002		peroxisome: uniprot (SCPx) other isoforms in liver and mit - also catalyzes other reactions specificity: Liver, fibroblasts, and placenta Mediates in vitro the transfer of all common phospholipids, cholesterol and gangliosides between membranes. May play a role in regulating steroidogenesis. NJ
SELADT	1	Xu ZH, Otterness DM, Freimuth RR, Carlini EJ, Wood TC, Mitchell S, Moon E, Kim UJ, Xu JP, Siciliano MJ, Weinshilboum RM	Human 3'-phosphoadenosine 5'-phosphosulfate synthetase 1 (PAPSS1) and PAPSS2: gene cloning, characterization and chromosomal localization	Biochem Biophys Res Commun	2000	10679223	modeling evidence assuming sel is same as sulfur
SELCYSLY	1	Mihara H, Kurihara T, Watanabe T, Yoshimura T, Esaki N.	cDNA cloning, purification, and characterization of mouse liver selenocysteine lyase. Candidate for selenium delivery protein in selenoprotein synthesis	J Biol Chem	2000	10692412	second citation is mostly concerned with mouse other citation says gsh is used modeling evdience only
SELCYSLY2	3	Daher R, Van Lente F.	Characterization of selenocysteine lyase in human tissues and its relationship to tissue selenium concentrations	J Trace Elem Electrolytes Health Dis	1992	1483038	Enzymatic studies on human form state that selcys is the SOLE substrate and pdx5p is the requied cofactor. see PMID: 1483038
SELMETAT	1	Katsuhiko Nakamuro, Tomofumi Okuno, and Tatsuya Hasegawa	Metabolism of Selenoamino Acids and Contribution of Selenium Methylation to Their Toxicity	Journal of Health Science	2000	?	based on assumption that sel isn't different from sulfur, thus modeling evidence
SELNPS	3	Esaki N, Nakamura T, Tanaka H, Soda K	Selenocysteine lyase, a novel enzyme that specifically acts on selenocysteine. Mammalian distribution and purification and properties of pig liver enzyme	J Biol Chem	1982	6461656	0
SELNPS	3	Stadtman TC	Selenocysteine	Annu Rev Biochem	1996	8811175	0
SERGLYexR	2	Fukasawa Y, Segawa H, Kim JY, Chairoungdua A, Kim DK, Matsuo H, Cha SH, Endou H, Kanai Y	Identification and characterization of a Na(+)-independent neutral amino acid transporter that associates with the 4F2 heavy chain and exhibits substrate selectivity for small neutral D- and L-amino acids	J Biol Chem	2000	10734121	homology and is linked to 4F2hc via a disulphide bond [25, 41]. It mediates Na+-independent transport of small neutral amino acids such as Gly, L-Ala, L-Ser, L-Thr, L-Cys, -aminoisobutyric acid and -alanine. Asc-1-4F2hc also transports d-isomers including D-Ser with high apparent affinity. It functions preferentially, but not exclusively, in an exchange mode. These functional properties appear consistent with those of system asc. Heterogeneity in substrate selectivity has been described for this transport system and the existence of at least two subtypes has been proposed. Asc-1 corresponds to the subtype that was characterized originally in trout peripheral blood lymphocytes, which is less stereospecific and transports -aminoisobutyric acid and -alanine. Asc-1 exhibits the highest structural similarity to the L-type transporter light chain LAT2 (66% identity). In contrast to LAT2-4F2hc, which takes neutral amino acids of all sizes, and to LAT1-4F2hc, which transports only large ones, Asc-1 transports only small neutral amino acids and is not inhibited by Asc-1 mRNA is expressed in the brain, lung, small intestine and PMID 14770310

Reaction Abbreviation	Score	Authors	Article or Book Title	Journal	Year	PubMed ID	Curation Notes
SERGLYexR	2	Nakauchi J, Matsuo H, Kim DK, Goto A, Chairoungdua A, Cha SH, Inatomi J, Shiokawa Y, Yamaguchi K, Saito I, Endou H, Kanai Y	Cloning and characterization of a human brain Na(+)-independent transporter for small neutral amino acids that transports D-serine with high affinity	Neurosci Lett	2000	10863037	homology and is linked to 4F2hc via a disulphide bond [25, 41]. It mediates Na+-independent transport of small neutral amino acids such as Gly, L-Ala, L-Ser, L-Thr, L-Cys, -aminoisobutyric acid and -alanine. Asc-1-4F2hc also transports d-isomers including D-Ser with high apparent affinity. It functions preferentially, but not exclusively, in an exchange mode. These functional properties appear consistent with those of system asc. Heterogeneity in substrate selectivity has been described for this transport system and the existence of at least two subtypes has been proposed. Asc-1 corresponds to the subtype that was characterized originally in trout peripheral blood lymphocytes, which is less stereospecific and transports -aminoisobutyric acid and -alanine. Asc-1 exhibits the highest structural similarity to the L-type transporter light chain LAT2 (66% identity). In contrast to LAT2-4F2hc, which takes neutral amino acids of all sizes, and to LAT1-4F2hc, which transports only large ones, Asc-1 transports only small neutral amino acids and is not inhibited by Asc-1 mRNA is expressed in the brain, lung, small intestine and PMID 14770310
SERHL	3	Ogawa H, Gomi T, Konishi K, Date T, Nakashima H, Nose K, Matsuda Y, Peraino C, Pitot HC, Fujioka M.	Human liver serine dehydratase. cDNA cloning and sequence homology with hydroxyamino acid dehydratases from other sources.		1989	2674117	cytosolic according to GeneCards predominantly in liver PMID 14596599: Formation of pyruvate by SDH is a two-step reaction in which the hydroxyl group of serine is cleaved to produce aminoacrylate, and then the aminoacrylate is deaminated by nonenzymatic hydrolysis to produce pyruvate. MM
SERPT	3	Weiss B, Stoffel W	Human and murine serine-palmitoyl-CoA transferase	Eur J Biochem	1997		cytoplasm - actually associated w/ ER membrane, however given the rest of the associated pathway and the lack of specificity of location (inner vs cytoplasmic side) - uniprot NJ
SERtp	2	Xue HH, Sakaguchi T, Fujie M, Ogawa H, Ichiyama A.	Flux of the L-serine metabolism in rabbit, human, and dog livers. Substantial contributions of both mitochondrial and peroxisomal serine:pyruvate/alanine:glyoxylate aminotransferase.		1999	10347152	L-serine catabolism can occur via SPTx pathway in the peroxisome (PMID:10347152) MM
SGPL11r	3	Van Veldhoven PP, Gijsbers S, Mannaerts GP, Vermeesch JR, Brys V.	Human sphingosine-1-phosphate lyase: cDNA cloning, functional expression studies and mapping to chromosome 10q22(1).	Biochim Biophys Acta	2000	11018465	NJ
SGPL11r	3	Reiss U, Oskouian B, Zhou J, Gupta V, Sooriyakumaran P, Kelly S, Wang E, Merrill AH Jr, Saba JD.	Sphingosine-phosphate lyase enhances stress-induced ceramide generation and apoptosis.	J Biol Chem	2004	14570870	NJ
SIAASE	0	Monti E, Preti A, Rossi E, Ballabio A, Borsani G.	Cloning and characterization of NEU2, a human gene homologous to rodent soluble sialidases.	Genomics	1999	10191093	NEU2 belongs to a family of glycohydrolytic enzymes which remove sialic acid residues from glycoproteins and glycolipids. Expression studies in COS7 cells confirmed that this gene encodes a functional sialidase. Its cytosolic localization was demonstrated by cell fractionation experiments. [RefSeq] NEU4 belongs to a family of glycohydrolytic enzymes which remove sialic acid residues from glycoproteins and glycolipids.[RefSeq] NEU4 wasidentified by searching sequence databases for entries showing homologies to the human cytosolic sialidase NEU2. Highest expression in the liver. Associated with inner cell membranes. [Monti et al, Genomics 83(3):445-453 (2004)] Neu2p expressed in sk muscle [Monti et al, Genomics 1999]
SIAT4Bg	3	Giordanengo V, Bannwarth S, Laffont C, Van Miegem V, Harduin-Lepers A, Delannoy P, Lefebvre JC.	Cloning and expression of cDNA for a human Gal(beta1-3)GalNAc alpha2,3-sialyltransferase from the CEM T-cell line.	Eur J Biochem	1997	9266697	Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot The protein encoded by this gene is a type II membrane protein that catalyzes the transfer of sialic acid from CMP-sialic acid to galactose-containing substrates. The encoded protein is normally found in the Golgi but can be proteolytically processed to a soluble form. This protein, which is a member of glycosyltransferase family 29, can use the same acceptor substrates as does sialyltransferase 4A. cloning, biochem, seq - Giordanengo ref NJ

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SIAT9g	3	Ishii A, Ohta M, Watanabe Y, Matsuda K, Ishiyama K, Sakoe K, Nakamura M, Inokuchi J, Sanai Y, Saito M.	Expression cloning and functional characterization of human cDNA for ganglioside GM3 synthase.	J Biol Chem	1998	9822625	Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot See Ishi ref -gene was found in a tissue-specific manner, with predominant expression in brain, skeletal muscle, and testis, and very low expression in liver Ganglioside GM3 is known to participate in the induction of cell differentiation, modulation of cell proliferation, maintenance of fibroblast morphology, signal transduction, and integrin-mediated cell adhesion. The protein encoded by this gene is a type II membrane protein which catalyzes the formation of GM3 using lactosylceramide as the substrate. NJ
SLCBK1	3	Liu H, Sugiura M, Nava VE, Edsall LC, Kono K, Poulton S, Milstien S, Kohama T, Spiegel S.	Molecular cloning and functional characterization of a novel mammalian sphingosine kinase type 2 isoform.	J Biol Chem	2000	10751414	no good localization info available at this point - cyt by default, temporarily - re-review in future. cloning + biochem characterization - Liu ref Sphingosine-1-phosphate (SPP) has diverse biological functions acting inside cells as a second messenger to regulate proliferation and survival, and extracellularly, as a ligand for G protein-coupled receptors of the endothelial differentiation gene 1 subfamily. Based on sequence homology to murine and human sphingosine kinase-1 (SPHK1), which we recently cloned - ref exerpt tissue spec (Liu ref): liver, heart, kideny, testis, brain. NJ
SLDx	2	Weinstein CL, Griffith OW.	Cysteinesulfonate and beta-sulfopyruvate metabolism. Partitioning between decarboxylation, transamination, and reduction pathways.		1988	3346220	- catalysed by mammalian malate dehydrogenase (EC 1.1.1.37), where the resulting sulfolactate is excreted (PMID: 15758220) - direct evidence found in rat (PMID:3346220) MM
SLDx	2	Rein U, Gueta R, Denger K, Ruff J, Hollemeyer K, Cook AM.	Dissimilation of cysteate via 3-sulfolactate sulfo-lyase and a sulfate exporter in Paracoccus pantotrophus NKNCYSA.		2005	15758220	- catalysed by mammalian malate dehydrogenase (EC 1.1.1.37), where the resulting sulfolactate is excreted (PMID: 15758220) - direct evidence found in rat (PMID:3346220) MM
SMPD3l	3	Hofmann K, Tomiuk S, Wolff G, Stoffel W.	Cloning and characterization of the mammalian brain-specific, Mg2+-dependent neutral sphingomyelinase.	Proc Natl Acad Sci	2000	10823942	lysosomal - uniprot
SMPD4	2	Rodriguez-Lafrasse C, Vanier MT.	Sphingosylphosphorylcholine in Niemann-Pick disease brain: accumulation in type A but not in type B.	Neurochem Res	1999	9972865	see PMID: 12069827, 9972865 for evidence - noted accumulation of spc_hs in deficiencies of sphingomyelinase, so although not enough evidence can be inferred for gene assocations, there must be a sphingomyelinase with spc_hs specificity in the cell. IC localization not known, added to cytosol as default and also as part of model gap filling. NJ
SMS	3	Huitema K, van den Dikkenberg J, Brouwers JF, Holthuis JC.	Identification of a family of animal sphingomyelin synthases.	EMBO J	2003	14685263	Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot - also in Huitema ref Note: Huitema notes SMS1 and SMS2 - golgi and cytoplasmic versions respectively TMEM23 = SMS1 SMS2 for humans not found yet in Entrez gene Bidirectional lipid cholinephosphotransferases capable of converting phosphatidylcholine (PC) and ceramide to sphingomyelin (SM) and diacylglycerol (DAG) and vice versa. Direction is dependent on the relative concentrations of DAG and ceramide as phosphocholine acceptors. Directly and specifically recognizes the choline head group on the substrate. Also requires two fatty chains on the choline-P donor molecule in order to be recognized efficiently as a substrate. Does not function strictly as a SM synthase. NJ

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SO4HCOtex	2	Mount DB, Romero MF	The SLC26 gene family of multifunctional anion exchangers	Pflugers Arch	2004	12759755	These anion exchange reactions are all somewhat suspect in some regard. Stoichiometry of exchange is probably least certain. Reversibility is also questionable in some cases, and more substrates almost certainly exist than are indicated in these reactions. From PMID 12759755: The ten-member SLC26 gene family encodes anion exchangers capable of transporting a wide variety of monovalent and divalent anions. The physiological role(s) of individual paralogs is evidently due to variation in both anion specificity and expression pattern. Three members of the gene family are involved in genetic disease; SLC26A2 in chondrodysplasias, SLC26A3 in chloride-losing diarrhea, and SLC26A4 in Pendred syndrome and hereditary deafness (DFNB4). The analysis of Slc26a4-null mice has significantly enhanced the understanding of the roles of this gene in both health and disease. Targeted deletion of Slc26a5 has in turn revealed that this paralog is essential for electromotor activity of cochlear outer hair cells and thus for cochlear amplification. Anions transported by the SLC26 family, with variable specificity, include the chloride, su
SO4t4_2	3	Girard JP, Baekkevold ES, Feliu J, Brandtzaeg P, Amalric F	Molecular cloning and functional analysis of SUT-1, a sulfate transporter from human high endothelial venules	Proc Natl Acad Sci U S A	1999	10535998	26266: - cloned [Girard 1999] -high in placenta and testis; intermediate in brain; low in heart, thymus, liver [Markovich 2005] - 40-50% amino acid identity w/ rat NaS1, human & rat NaC1, NaC3 [Girard 1999] - 2 Na+:1 anion stoichiometry [Markovich 2005]
SO4t4_2	3	Markovich D, Regeer RR, Kunzelmann K, Dawson PA	Functional characterization and genomic organization of the human Na(+)-sulfate cotransporter hNaS2 gene (SLC13A4)	Biochem Biophys Res Commun	2005	15607730	26266: - cloned [Girard 1999] -high in placenta and testis; intermediate in brain; low in heart, thymus, liver [Markovich 2005] - 40-50% amino acid identity w/ rat NaS1, human & rat NaC1, NaC3 [Girard 1999] - 2 Na+:1 anion stoichiometry [Markovich 2005]
SO4t4_3	3	Pajor AM	Molecular properties of sodium/dicarboxylate cotransporters	J Membr Biol	2000	10811962	6561: - cloned [Lee 2000] - ortholog cotransports sulfate, thiosulfate, selenate with Na+; molybdate, tungstate are also competitively transported but not included in model [Lee 2000] - kidney [Lee 2000] - 3 Na+:1 anion stoichiometry [Pajor 2000]
SO4t4_3	3	Lee A, Beck L, Markovich D	The human renal sodium sulfate cotransporter (SLC13A1; hNaSi-1) cDNA and gene: organization, chromosomal localization, and functional characterization	Genomics	2000	11161786	6561: - cloned [Lee 2000] - ortholog cotransports sulfate, thiosulfate, selenate with Na+; molybdate, tungstate are also competitively transported but not included in model [Lee 2000] - kidney [Lee 2000] - 3 Na+:1 anion stoichiometry [Pajor 2000]
SOAT11	3	Chang CC, Huh HY, Cadigan KM, Chang TY.	Molecular cloning and functional expression of human acyl-coenzyme A:cholesterol acyltransferase cDNA in mutant Chinese hamster ovary cells.	J Biol Chem	1993	8407899	Guo et al (PMID: 15850387) have evidence that the transmembrane protein SOAT1 is likely able to catalyze esterification of cholesterol diffusing from the cytosol into the ER membrane or from the ER lumen into the ER membrane. Expression, sequence and function in Change et al (PMID: 8407899). NJ
SOAT11	3	Guo ZY, Chang CC, Lu X, Chen J, Li BL, Chang TY.	The disulfide linkage and the free sulfhydryl accessibility of acyl-coenzyme A:cholesterol acyltransferase 1 as studied by using mPEG5000-maleimide.	Biochemistry	2005	15850387	Guo et al (PMID: 15850387) have evidence that the transmembrane protein SOAT1 is likely able to catalyze esterification of cholesterol diffusing from the cytosol into the ER membrane or from the ER lumen into the ER membrane. Expression, sequence and function in Change et al (PMID: 8407899). NJ
SPH1Ptr	2	Quest AF, Leyton L, Parraga M.	Caveolins, caveolae, and lipid rafts in cellular transport, signaling, and disease.	Biochem Cell Biol	2004	15052333	Unknown mechanism - may or may not be energy dependen - may be vesicular/caveolor/other... However these metabolites must be able to be transported intracellularly and exported outside of the cell. NJ
SPHMDAc	0	Meyer zu Heringdorf D, Himmel HM, Jakobs KH.	Sphingosylphosphorylcholine-biological functions and mechanisms of action.	Biochim Biophys Acta	2002	12069827	Details about localization not known, gene not found. It's biochemical function however has been described. Presently left as cytosolic by default, likely to occur in other compartments. See PMID: 12069827 NJ
SPMDOX	2	Salim EI, Wanibuchi H, Morimura K, Kim S, Yano Y, Yamamoto S, Fukushima S	Inhibitory effects of 1,3-diaminopropane, an ornithine decarboxylase inhibitor, on rat two-stage urinary bladder carcinogenesis initiated by N-butyl-N-(4-hydroxybutyl)nitrosamine	Carcinogenesis	2000	10657958	This reaction added based on the physiological evidence that 13dampp inhibits the ODC reaction, which is upstream.
SPMS	3	Myohanen S, Kauppinen L, Wahlfors J, Alhonen L, Janne J	Human spermidine synthase gene: structure and chromosomal localization	DNA Cell Biol	1991	2069720	Well-accepted gene and enzyme function.
SPMS	3	Kauppinen L, Myohanen S, Halmekyto M, Alhonen L, Janne J	Transgenic mice over-expressing the human spermidine synthase gene	Biochem J	1993	8343131	Well-accepted gene and enzyme function.

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SPODM	3	Crapo JD, Oury T, Rabouille C, Slot JW, Chang LY	Copper,zinc superoxide dismutase is primarily a cytosolic protein in human cells	Proc Natl Acad Sci U S A	1992	1332049	6647: - homodimer, soluble cytosolic protein [UniProt] - widely expressed in cytosol of all mammalian cells - primarily cytosolic, but also found in nucleus and peroxisome [Crapo 1992] - cloned & expressed [Hallewell 1985]
SPODM	3	Hallewell RA, Masiarz FR, Najarian RC, Puma JP, Quiroga MR, Randolph A, Sanchez-Pescador R, Scandella CJ, Smith B, Steimer KS, et al	Human Cu/Zn superoxide dismutase cDNA: isolation of clones synthesising high levels of active or inactive enzyme from an expression library	Nucleic Acids Res	1985	3889846	6647: - homodimer, soluble cytosolic protein [UniProt] - widely expressed in cytosol of all mammalian cells - primarily cytosolic, but also found in nucleus and peroxisome [Crapo 1992] - cloned & expressed [Hallewell 1985]
SPODMe	3	Hjalmarsson K, Marklund SL, Engstrom A, Edlund T	Isolation and sequence of complementary DNA encoding human extracellular superoxide dismutase	Proc Natl Acad Sci U S A	1987	3476950	6649: - homotetrameric protein, secreted [Fridovich 1997] - exhibits affinity for sulfated polysaccharides, such as heparin or heparan sulfate [Fridovich 1997] - cloned [Hjalmarsson 1987] - Highly expressed in alveolar type II cells, proximal renal tubular cells, vascular smooth muscular cells, lung macrophages, cultured fibroblast cell lines (see refs in [Zelko 2002]); also detectable in blood plasma, mostly bound onto the extracellular matrix (see [Fridovich 1997] for refs)
SPODMe	3	Fridovich I	Superoxide anion radical (O2-.), superoxide dismutases, and related matters	J Biol Chem	1997	9228011	6649: - homotetrameric protein, secreted [Fridovich 1997] - exhibits affinity for sulfated polysaccharides, such as heparin or heparan sulfate [Fridovich 1997] - cloned [Hjalmarsson 1987] - Highly expressed in alveolar type II cells, proximal renal tubular cells, vascular smooth muscular cells, lung macrophages, cultured fibroblast cell lines (see refs in [Zelko 2002]); also detectable in blood plasma, mostly bound onto the extracellular matrix (see [Fridovich 1997] for refs)
SPODMe	3	Zelko IN, Mariani TJ, Folz RJ	Superoxide dismutase multigene family: a comparison of the CuZn-SOD (SOD1), Mn-SOD (SOD2), and EC-SOD (SOD3) gene structures, evolution, and expression	Free Radic Biol Med	2002	12126755	6649: - homotetrameric protein, secreted [Fridovich 1997] - exhibits affinity for sulfated polysaccharides, such as heparin or heparan sulfate [Fridovich 1997] - cloned [Hjalmarsson 1987] - Highly expressed in alveolar type II cells, proximal renal tubular cells, vascular smooth muscular cells, lung macrophages, cultured fibroblast cell lines (see refs in [Zelko 2002]); also detectable in blood plasma, mostly bound onto the extracellular matrix (see [Fridovich 1997] for refs)
SPODMm	2	Folz RJ, Guan J, Seldin MF, Oury TD, Enghild JJ, Crapo JD.	Mouse extracellular superoxide dismutase: primary structure, tissue-specific gene expression, chromosomal localization, and lung in situ hybridization.	Am J Respir Cell Mol Biol	1997	9376114	proteome Shlafer M, Myers CL, Adkins S.; Mitochondrial hydrogen peroxide generation and activities of glutathione peroxidase and superoxide dismutase following global ischemia.; J Mol Cell Cardiol. 1987 Dec;19(12):1195-206 Mitochondriral catalase and oxidative injury, Bai J. Catalytic activity: Copper-zinc superoxide dismutase (SOD) is a 32-kDa homodimeric protein that catalyzes the disproportionation of superoxide anion into dioxygen and hydrogen peroxide through redox cycling of its catalytic copper ion. This was according to Elam J. J Biol Chem. 2003 Jun 6;278(23):21032-9. Subcellular localization: 1) Sod1.1 - cytoplasmic & nuclear 2) Sod2.1 - mitochondrial 3) Sod3.1 - extracellular according to Entrez Gene Database Tissue Localization Extracellular SOD (Sod3.1) is found in trace amounts in almost all tissues, but can be found predominantly in the lung and kidney. Tissue localization for the cytoplasmic and mitochondrial was not specified. This was according to Figure 4 in Folz RJ. Am J Respir Cell Mol Biol. 1997 Oct;17(4):393-403. Review.

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SPODMm	2	Elam JS, Malek K, Rodriguez JA, Doucette PA, Taylor AB, Hayward LJ, Cabelli DE, Valentine JS, Hart PJ.	An alternative mechanism of bicarbonate-mediated peroxidation by copper-zinc superoxide dismutase: rates enhanced via proposed enzyme-associated peroxycarbonate intermediate.	J Biol Chem	2003	12649272	proteome Shlafer M, Myers CL, Adkins S.; Mitochondrial hydrogen peroxide generation and activities of glutathione peroxidase and superoxide dismutase following global ischemia.; J Mol Cell Cardiol. 1987 Dec;19(12):1195-206 Mitochondriral catalase and oxidative injury, Bai J. Catalytic activity: Copper-zinc superoxide dismutase (SOD) is a 32-kDa homodimeric protein that catalyzes the disproportionation of superoxide anion into dioxygen and hydrogen peroxide through redox cycling of its catalytic copper ion. This was according to Elam J. J Biol Chem. 2003 Jun 6;278(23):21032-9. Subcellular localization: 1) Sod1.1 - cytoplasmic & nuclear 2) Sod2.1 - mitochondrial 3) Sod3.1 - extracellular according to Entrez Gene Database Tissue Localization Extracellular SOD (Sod3.1) is found in trace amounts in almost all tissues, but can be found predominantly in the lung and kidney. Tissue localization for the cytoplasmic and mitochondrial was not specified. This was according to Figure 4 in Folz RJ. Am J Respir Cell Mol Biol. 1997 Oct;17(4):393-403. Review.
SPRMS	3	Korhonen VP, Halmekyto M, Kauppinen L, Myohanen S, Wahlfors J, Keinanen T, Hyvonen T, Alhonen L, Eloranta T, Janne J	Molecular cloning of a cDNA encoding human spermine synthase	DNA Cell Biol	1995	7546290	Well-accepted gene for this function.
SPRn	3	Elzaouk L, Laufs S, Heerklotz D, Leimbacher W, Blau N, Resibois A, Thony B.	Nuclear localization of tetrahydrobiopterin biosynthetic enzymes.	Biochim Biophys Acta	2004	14729142	IT only ~1% of total SPR could be found in nucleus. however, i included this nuclear reaction. gap: no one knows why GTPCIn, PTHPSn, SPRn are located in nucleus - and what the function of thbpt could be
SPTix	3	Lumb MJ, Danpure CJ.	Functional synergism between the most common polymorphism in human alanine:glyoxylate aminotransferase and four of the most common disease-causing mutations.		2000	10960483	-catalyzed by same gene product as AGTx, although at lower activity (see ref.) MM
SQLSr	3	Cohen LH, Griffioen M, vanRoermund CW, Wanders RJ	Subcellular localization of squalene synthase in human hepatoa cell line Hep G2	Biochim Biophys Acta	1992		ER - according to lit refs (need to check more thoroughly in the literature if on outer side of ER membrane or inside no tissue specificity NJ
SR5ARr	3	Wigley WC, Prihoda JS, Mowaszowicz I, Mendonca BB, New MI, Wilson JD, Russell DW	Natural mutagenesis study of the human steroid 5-alpha-reductase 2 isozyme	Biochemistry	1994		ER/microsomal - uniprot + refs specificity: prostate, liver Converts testosterone into 5-alpha-dihydrotestosterone and progesterone or corticosterone into their corresponding 5- alpha 3-oxosteroids. It plays a central role in sexual differentiation and androgen physiology. NJ
SR5ARr	3	Andersson S, Berman DM, Jenkins EP, Russel DW	Deletion of steroid 5-alpha-reductase 2 gene in male pseudohermaphroditism	Nature	1991		ER/microsomal - uniprot + refs specificity: prostate, liver Converts testosterone into 5-alpha-dihydrotestosterone and progesterone or corticosterone into their corresponding 5- alpha 3-oxosteroids. It plays a central role in sexual differentiation and androgen physiology. NJ
SRTN23OX	3	Okuma M, Tokuyama T, Senoh S, Hirata F, Hayaishi O	Antagonism of 5-hydroxykynurenamine against serotonin action on platelet aggregation	Proc Natl Acad Sci U S A	1976	1061163	Enzyme name connected to reaction in citation.
SRTNACT	3	Coon SL, Mazuruk K, Bernard M, Roseboom PH, Klein DC, Rodriguez IR	The human serotonin N-acetyltransferase (EC 2.3.1.87) gene (AANAT): structure, chromosomal localization, and tissue expression	Genomics	1996	8661026	Enzyme and reaction characterized.
SSALxm	3	Chambliss KL, Caudle DL, Hinson DD, Moomaw CR, Slaughter CA, Jakobs C, Gibson KM.	Molecular cloning of the mature NAD(+)-dependent succinic semialdehyde dehydrogenase from rat and human. cDNA isolation, evolutionary homology, and tissue expression.		1995	7814412	- (SSADH) deficiency causes a rare metabolic disorder of 4-aminobutyric acid degradation (Chambliss et. al. 1998) -mitochondrial (GeneCards) -brain, pancreas, heart, liver, skeletal muscle and kidney. lower in placenta.
SSALxm	3	Chambliss KL, Hinson DD, Trettel F, Malaspina P, Novelletto A, Jakobs C, Gibson KM.	Two exon-skipping mutations as the molecular basis of succinic semialdehyde dehydrogenase deficiency (4-hydroxybutyric aciduria).		1998	9683595	- (SSADH) deficiency causes a rare metabolic disorder of 4-aminobutyric acid degradation (Chambliss et. al. 1998) -mitochondrial (GeneCards) -brain, pancreas, heart, liver, skeletal muscle and kidney. lower in placenta.

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ST3GAL61g	3	Okajima T, Fukumoto S, Miyazaki H, Ishida H, Kiso M, Furukawa K, Urano T, Furukawa K.	Molecular cloning of a novel alpha2,3-sialyltransferase (ST3Gal VI) that sialylates type II lactosamine structures on glycoproteins and glycolipids.	J Biol Chem	1999	10206952	localization: golgi: uniprot and PMID: 10206952 sequence, function, cloning: PMID: 10206952 Sialyltransferases catalyze the transfer of sialic acid from cytidine 5-prime monophospho-N-acetylneuraminic acid (CMP NeuAc) to terminal positions of glycoprotein and glycolipid carbohydrate groups. Terminal NeuAc residues are key determinants of carbohydrate structures, such as the sialyl-Lewis X determinants, and are widely distributed in many cell types.[supplied by OMIM] Involved in the synthesis of sialyl-paragloboside, a precursor of sialyl-Lewis X determinant. Has a alpha-2,3- sialyltransferase activity toward Gal-beta1,4-GlcNAc structure on glycoproteins and glycolipids. Has a restricted substrate specificity, it utilizes Gal-beta1,4-GlcNAc on glycoproteins, and neolactotetraosylceramide and neolactohexaosylceramide, but not lactotetraosylceramide, lactosylceramide or asialo-GM1. NJ
ST8SIA11	3	Kim YJ, Kim KS, Do S, Kim CH, Kim SK, Lee YC.	Molecular cloning and expression of human alpha2,8-sialyltransferase (hST8Sia V).	Biochem Biophys Res Commun	1997	9199191	Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot The protein encoded by this gene is a type II membrane protein that may be present in the Golgi apparatus. The encoded protein, which is a member of glycosyltransferase family 29, may be involved in the synthesis of gangliosides GD1c, GT1a, GQ1b, and GT3 from GD1a, GT1b, GM1b, and GD3, respectively. NJ
ST8SIA11	3	Takashima S, Tsuji S, Tsujimoto	Characterization of second type of human beta-galactoside alpha2,6-sialyltransferase which sialates gal-beta1,4GlcNAc structures on oligosaccharides preferentially	Journal of Biological Chemistry	2002		Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot The protein encoded by this gene is a type II membrane protein that may be present in the Golgi apparatus. The encoded protein, which is a member of glycosyltransferase family 29, may be involved in the synthesis of gangliosides GD1c, GT1a, GQ1b, and GT3 from GD1a, GT1b, GM1b, and GD3, respectively. NJ
ST8SIA12	3	Sasaki K, Kurata K, Kojima N, Kurosawa N, Ohta S, Hanai N, Tsuji S, Nishi T	Expression cloning of a Gm3-specific alpha-2,8-sialyltransferase (GD3 synthase)	Journal of Biological Chemistry	1994		Golgi - lumen side - see ref Kolter and Sanhoff The protein encoded by this gene is a type II membrane protein that may be present in the Golgi apparatus. The encoded protein, which is a member of glycosyltransferase family 29, may be involved in the synthesis of gangliosides GD1c, GT1a, GQ1b, and GT3 from GD1a, GT1b, GM1b, and GD3, respectively. cloning and function: PMID: 9199191 NJ
ST8SIA51g	3	Harduin-Lepers A, Vallejo-Ruiz V, Krzewinski-Recchi MA, Smyn-Petit B, Julien S, Delannoy P	The human sialyltransferase family	Biochimie	2001		Golgi - lumen side - see ref Kolter and Sanhoff; also noted in uniprot The protein encoded by this gene is a type II membrane protein that may be present in the Golgi apparatus. The encoded protein, which is a member of glycosyltransferase family 29, may be involved in the synthesis of gangliosides GD1c, GT1a, GQ1b, and GT3 from GD1a, GT1b, GM1b, and GD3, respectively. cloning and function: PMID: 9199191 NJ
ST8SIA55g	3	Sandhoff K, Kolter T	Biosynthesis and degradation of mammalian glycosphingolipids	Phil Trans R Soc Lond B	2003		golgi inner lumenal side - uniport and refs The protein encoded by this gene is a type II membrane protein that may be present in the Golgi apparatus. The encoded protein, which is a member of glycosyltransferase family 29, may be involved in the synthesis of gangliosides GD1c, GT1a, GQ1b, and GT3 from GD1a, GT1b, GM1b, and GD3, respectively. cloning and function: PMID: 9199191 NJ

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STS1r	3	Migeon BR, Shapiro LJ, Norum RA, Mohandas T, Axelman J, Dabora RL.	Differential expression of steroid sulphatase locus on active and inactive human X chromosome.	Nature	1982		ER membrane - lumen side - ref and uniprot Stein ref also notes: "STS expressed in BHK-21 cells is located predominantly in the endoplasmic reticulum; smaller fractions are found in the Golgi, at the cell surface, multivesicular endosomes, as well as in lysosomes." The protein encoded by this gene catalyzes the conversion of sulfated steroid precursors to estrogens during pregnancy. The encoded protein is found in the endoplasmic reticulum, where it acts as a homodimer. Mutations in this gene are known to cause X-linked ichthyosis (XLI). NJ
STS1r	3	Stein C, Hille A, Seidel J, Rijnbout S, Waheed A, Schmidt B, Geuze H, von Figura K.	Cloning and expression of human steroid-sulfatase. Membrane topology, glycosylation, and subcellular distribution in BHK-21 cells.	J Biol Chem	1989	2668275	ER membrane - lumen side - ref and uniprot Stein ref also notes: "STS expressed in BHK-21 cells is located predominantly in the endoplasmic reticulum; smaller fractions are found in the Golgi, at the cell surface, multivesicular endosomes, as well as in lysosomes." The protein encoded by this gene catalyzes the conversion of sulfated steroid precursors to estrogens during pregnancy. The encoded protein is found in the endoplasmic reticulum, where it acts as a homodimer. Mutations in this gene are known to cause X-linked ichthyosis (XLI). NJ
SUCCt4_3	3	Wang H, Fei YJ, Kekuda R, Yang-Feng TL, Devoe LD, Leibach FH, Prasad PD, Ganapathy V	Structure, function, and genomic organization of human Na(+)-dependent high-affinity dicarboxylate transporter	Am J Physiol Cell Physiol	2000	794676	64849: - cloned [Wang 2000] - high affinty cotransport of Na+ w/ succinate, dimethylsuccinate, aKG [Wang 2000] - basolateral membrane of renal proximal tubular epithelial cells, sinusoidal membrane of hepatocytes, and brain synaptosome [Pajor 1999] - high in kidney, also in placenta, brain, liver, pancreas [Wang 2000] - Na(+)-to-succinate stoichiometry is 3:1 [Wang 2000]
SUCCt4_3	3	Pajor AM	Sodium-coupled transporters for Krebs cycle intermediates	Annu Rev Physiol	1999	10099705	64849: - cloned [Wang 2000] - high affinty cotransport of Na+ w/ succinate, dimethylsuccinate, aKG [Wang 2000] - basolateral membrane of renal proximal tubular epithelial cells, sinusoidal membrane of hepatocytes, and brain synaptosome [Pajor 1999] - high in kidney, also in placenta, brain, liver, pancreas [Wang 2000] - Na(+)-to-succinate stoichiometry is 3:1 [Wang 2000]
SUCOAS1m	0	Johnson JD, Mehus JG, Tews K, Milavetz BI, Lambeth DO	Genetic evidence for the expression of ATP- and GTP-specific succinyl-CoA synthetases in multicellular eucaryotes	J Biol Chem	1998	9765291	- Highly ATP- and GTP-specific isoforms of succinyl-CoA synthetase in pigeon incorporate the same alpha -subunit, but different beta -subunits [Johnson et al, J Biol Chem 1998] - heterodimer of an alpha and a beta chain [UniProt] - mitochondrial [UniProt]
SUCRe	3	Wu GD, Wang W, Traber PG	Isolation and characterization of the human sucrase-isomaltase gene and demonstration of intestine-specific transcriptional elements	J Biol Chem	1992	1560017	8972: - brush border enzyme [RefSeq], [Naim, J Biol Chem 1988] - expressed in small intestine and kidney [UniProt] 6476: - brush border enzyme [GO], [Wu et al, J Biol Chem 1992] -sm intestine [Wu et al, J Biol Chem 1992], [Devlin, Textbook of Biochemistry]
SUCRe	3	Naim HY, Sterchi EE, Lentze MJ	Structure, biosynthesis, and glycosylation of human small intestinal maltase-glucoamylase	J Biol Chem	1988	3143729	8972: - brush border enzyme [RefSeq], [Naim, J Biol Chem 1988] - expressed in small intestine and kidney [UniProt] 6476: - brush border enzyme [GO], [Wu et al, J Biol Chem 1992] -sm intestine [Wu et al, J Biol Chem 1992], [Devlin, Textbook of Biochemistry]
TAURtcx	1	Ramamoorthy S, Leibach FH, Mahesh VB, Han H, Yang-Feng T, Blakely RD, Ganapathy V	Functional characterization and chromosomal localization of a cloned taurine transporter from human placenta	Biochem J	1994	8010975	Mechanism isn't as certain as it could be--sodium symport is commonly thought to be the transport method from outside to inside cell no specific evidence located regarding transport from cytosol to peroxisome, so only modeling confidence for now
TDP	3	Laforenza U, Mazzarello P, Patrini C, Poloni M, Casadei GP, Rindi G.	Different distribution of thiaminpyrophosphatase activity in neuronal and glial cell enriched fractions from human and rat brain: an isoelectric focusing investigation.	Basic Appl Histochem	1990	2171493	IT activity has been measured by Bettendorf et al 1996 vmax=130+-19 nmmol/mg.30 min(at pH=7.4) Laforenza et al, 1990, might have found multiple isoforms of this enzyme extracted from humna neuronal and glial cells - check reference

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TDPDRE	1	Tonetti M, Sturla L, Bisso A, Zanardi D, Benatti U, De Flora A	The metabolism of 6-deoxyhexoses in bacterial and animal cells	Biochimie	1998	9893952	- L-rhamnose has been observed mainly in bacteria; however, there have been a few acttered reports which suggest its presence in animals (see refs in [Tonetti 1998]) - conversion of dTDP-glucose to dTDP-rhamnose is described as a mammalian pathway by Devlin, pgs. 671-672 - these facts together with the putative annotation of a human dTDPglucose 4,6-dehydratase (the first step in dTDP-rhamnose production) prompted the inclusion of this rxn
TDPGDH	2	Strausberg RL, Feingold EA, Grouse LH, Derge JG, Klausner RD, et al.	Generation and initial analysis of more than 15,000 full-length human and mouse cDNA sequences	Proc Natl Acad Sci U S A	2002	12477932	- discovered as part of high-througput study; function inferred from electronic annotation [Strausberg 2002]
TDPm	2	Barile M, Valenti D, Brizio C, Quagliariello E, Passarella S.	Rat liver mitochondria can hydrolyse thiamine pyrophosphate to thiamine monophosphate which can cross the mitochondrial membrane in a carrier-mediated process.	FEBS Lett	1998	9755848	based on rat data (liver cell). It seems that the mitochondrial thmpp is degraded to thmmp in the mitochondria and than exported to be degraded to thiamine in cytosol. IT it seems that thmmp can also be taken up by mitochondria but I do not see the sense in this uptake since there is no possible conversion of this compound reported (rxn has not been included)
THD1m	3	Hatefi Y, Yamaguchi M.	Nicotinamide nucleotide transhydrogenase: a model for utilization of substrate binding energy for proton translocation.	FASEB	1996	8647343	IT - Additional information added by RS/TV: White SA, Peake SJ, McSweeney S, Leonard G, Cotton NP, Jackson JB., The high-resolution structure of the NADP(H)-binding component (dIII) of proton-translocating transhydrogenase from human heart mitochondria., Structure Fold Des. 2000 Jan 15;8(1):1-12 (Jackson, Peake et al. 1999; Peake, Jackson et al. 2000;) There are two isozymes, both of which are mitochondrial, according to Entrez Gene Database. Catalytic Activity: Proton-pumping nicotinamide transhydrogenases are membrane proteins and proton pumps which catalyze the reversible reduction of NADP+ byd NADH linked to proton translocation across the membrane. The NADPH generate may be then used for detoxification of peroxides. According to Arkblad EL. Comp Biochem Physiol B Biochem Mol Biol. 2002 Sep;133(1):13-21.
THD1m	3	Zieger B, Ware J.	Cloning and deduced amino acid sequence of human nicotinamide nucleotide transhydrogenase.	DNA Seq	1997	9524818	IT - Additional information added by RS/TV: White SA, Peake SJ, McSweeney S, Leonard G, Cotton NP, Jackson JB., The high-resolution structure of the NADP(H)-binding component (dIII) of proton-translocating transhydrogenase from human heart mitochondria., Structure Fold Des. 2000 Jan 15;8(1):1-12 (Jackson, Peake et al. 1999; Peake, Jackson et al. 2000;) There are two isozymes, both of which are mitochondrial, according to Entrez Gene Database. Catalytic Activity: Proton-pumping nicotinamide transhydrogenases are membrane proteins and proton pumps which catalyze the reversible reduction of NADP+ byd NADH linked to proton translocation across the membrane. The NADPH generate may be then used for detoxification of peroxides. According to Arkblad EL. Comp Biochem Physiol B Biochem Mol Biol. 2002 Sep;133(1):13-21.
THD1m	3	Arkblad EL, Egorov M, Shakhparonov M, Romanova L, Polzikov M, Rydstrom J.	Expression of proton-pumping nicotinamide nucleotide transhydrogenase in mouse, human brain and C elegans.	Comp Biochem Physiol B Biochem Mol Biol	2002	12223207	IT - Additional information added by RS/TV: White SA, Peake SJ, McSweeney S, Leonard G, Cotton NP, Jackson JB., The high-resolution structure of the NADP(H)-binding component (dIII) of proton-translocating transhydrogenase from human heart mitochondria., Structure Fold Des. 2000 Jan 15;8(1):1-12 (Jackson, Peake et al. 1999; Peake, Jackson et al. 2000;) There are two isozymes, both of which are mitochondrial, according to Entrez Gene Database. Catalytic Activity: Proton-pumping nicotinamide transhydrogenases are membrane proteins and proton pumps which catalyze the reversible reduction of NADP+ byd NADH linked to proton translocation across the membrane. The NADPH generate may be then used for detoxification of peroxides. According to Arkblad EL. Comp Biochem Physiol B Biochem Mol Biol. 2002 Sep;133(1):13-21.
THFtm	2	Suh JR, Herbig AK, Stover PJ.	New perspectives on folate catabolism.	Annu Rev Nutr	2001	11375437	IT

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THMMPt4	3	Dutta B, Huang W, Molero M, Kekuda R, Leibach FH, Devoe LD, Ganapathy V, Prasad PD.	Cloning of the human thiamine transporter, a member of the folate transporter family.	J Biol Chem	1999	10542220	IT reactions is based on the fact that Ganapathy et al mentioned that folate is transported via OH- antiport. Since the same gene product has also been found to transport the thiamine derivates, the same mechanism of transport was concluded
THMMPt4	3	Ganapathy V, Smith SB, Prasad PD.	SLC19: the folate/thiamine transporter family.	Pflugers Arch	2004	14770311	IT reactions is based on the fact that Ganapathy et al mentioned that folate is transported via OH- antiport. Since the same gene product has also been found to transport the thiamine derivates, the same mechanism of transport was concluded
THMP	1	Zhao R, Gao F, Goldman ID.	Reduced folate carrier transports thiamine monophosphate: an alternative route for thiamine delivery into mammalian cells.	Am J Physiol Cell Physiol	2002	11997266	I did not found any biochemical studies on this enzyme, however, Bettendorff et al, 1996, proposed this reaction beyond others. Zhao et al., 2002, reported that the majority of uptaken TMP was hydrolysed to thiamine. IT In kegg they assigned locus 8776 with this function, however, there is no evidence that this is the case
THMt2m	3	Song Q, Singleton CK.	Mitochondria from cultured cells derived from normal and thiamine-responsive megaloblastic anemia individuals efficiently import thiamine diphosphate.	BMC Biochem	2005	12014993	It is thought that this SLC19A2 is also reponsible for the mitochondrial transport since cells of TRMA patients (thiamine responive megaloblastic anemia) in this gene show no mitochondrial transport of thiamine. Role of thiamine in mitochondria is not clear (therefore will be a gap) since there is no mitochondrial thiamine diphosphokinase) IT
THMt3	3	Eudy JD, Spiegelstein O, Barber RC, Wlodarczyk BJ, Talbot J, Finnell RH.	Identification and characterization of the human and mouse SLC19A3 gene: a novel member of the reduced folate family of micronutrient transporter genes.	Mol Genet Metab	2000	11136550	It has been shown that the transport is soudium independent and is stimulated by an outwardly directed H+ gradient. SLC19A2: The gene is highky expressed in skeletal muscle, less in hear and placenta, and very low in intestine and kidney although the uptake there is very high. SLC19A3: most abundant in placenta, follwed by liver, kidney, and heart. IT
THMt3	3	Rajgopal A, Edmondnson A, Goldman ID, Zhao R.	SLC19A3 encodes a second thiamine transporter ThTr2.	Biochim Biophys Acta	2001	11731220	It has been shown that the transport is soudium independent and is stimulated by an outwardly directed H+ gradient. SLC19A2: The gene is highky expressed in skeletal muscle, less in hear and placenta, and very low in intestine and kidney although the uptake there is very high. SLC19A3: most abundant in placenta, follwed by liver, kidney, and heart. IT
THMTP	3	Lakaye B, Makarchikov AF, Wins P, Margineanu I, Roland S, Lins L, Aichour R, Lebeau L, El Moualij B, Zorzi W, Coumans B, Grisar T, Bettendorff L.	Human recombinant thiamine triphosphatase: purification, secondary structure and catalytic properties.	Int J Biochem Cell Biol	2004	15109578	IT In locusLink there is only one isoform marked instead of two as in the gene index based on GeneCards the reaction thakes place in cytoplasm activity has been measured by Bettendorff et al, 1996: vmax= 29+-5 nmol/mg/5min thmtp may modulate ion channel regulation (PMID:11899071). - is dead-end in model
THRD_L	3	Edgar AJ.	The human L-threonine 3-dehydrogenase gene is an expressed pseudogene.		2002	12361482	-catalyzed by same enzyme for serine deaminase -shown in Lehninger Biochemistry (4th ed.) pg. 682 MM
THYMDtm	3	Lai Y, Tse CM, Unadkat JD.	Mitochondrial expression of the human equilibrative nucleoside transporter 1 (hENT1) results in enhanced mitochondrial toxicity of antiviral drugs.	J Biol Chem	2004	14607828	IT liver mitochondria
THYOXt2	3	Lafreniere RG, Carrel L, Willard HF.	A novel transmembrane transporter encoded by the XPCT gene in Xq13.2.	Hum Mol Genet	1994	7981683	- cloned [Lafreniere 1994] - rat ortholog transports T3, T4 by Na- and H-independent facilitated diffusion [Friesema 2003]
THYOXt2	3	Friesema EC, Ganguly S, Abdalla A, Manning Fox JE, Halestrap AP, Visser TJ	Identification of monocarboxylate transporter 8 as a specific thyroid hormone transporter	J Biol Chem	2003	12871948	- cloned [Lafreniere 1994] - rat ortholog transports T3, T4 by Na- and H-independent facilitated diffusion [Friesema 2003]
TMABADH	3	Kikonyogo,A. , Pietruszko,R.	ALdehyde dehydrogenase from adult human brain that dehydrogenates gamma-aminobutyraldehyde: purification, characterization, cloning and distribution.		1996	8645224	-irreversible due to direction in KEGG
TMABADH	3	Vaz,F.M. , Fouchier,S.W. , Ofman,R. , Sommer,M. , Wanders,R.J.	Molecular and biochemical characterization of rat gamma-trimethylaminobutyraldehyde dehydrogenase and evidence for the involvement of human aldehyde dehydrogenase 9 in carnitine biosynthesis.		2000	10702312	-irreversible due to direction in KEGG

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TMDK1	3	Johansson M, Brismar S, Karlsson A.	Human deoxycytidine kinase is located in the cell nucleus.	Proc Natl Acad Sci U S A	1997	9342341	activity of TK2 has been detected in both cytosol and mito. The fluorescence study could not identify a mito TK2, therefore, it is thought that there different transcript. TK2 seems also to act on dCyt but higher concentration is needed --> therefore rxn is not included IT
TMDK1	3	Krawiec K, Kierdaszuk B, Shugar D.	Inorganic tripolyphosphate (PPP(i)) as a phosphate donor for human deoxyribonucleoside kinases.	Biochem Biophys Res Commun	2003	12535661	activity of TK2 has been detected in both cytosol and mito. The fluorescence study could not identify a mito TK2, therefore, it is thought that there different transcript. TK2 seems also to act on dCyt but higher concentration is needed --> therefore rxn is not included IT
TMDPK	3	Zhao R, Gao F, Goldman ID.	Molecular cloning of human thiamin pyrophosphokinase.	Biochim Biophys Acta	2001	11342117	IT activity has been measured by Bettendorff et al, 1996: vmax=11 +- 2pmol/mg/min
TMDPP	3	Kubilus J, Lee LD, Baden HP	Purification of thymidine phosphorylase from human amniochorion	Biochim Biophys Acta	1978	718961	in cytoplasm or in mitochondria - not really clear. In patient with Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE) the thymidine phosphorylase shows only 10% activity:"We hypothesize that, in patients with TP deficiency, increased levels of dThd and dUrd cause mitochondrial nucleotide pool imbalances, which, in turn, lead to mtDNA abnormalities including site-specific point mutations." - not included as mitochondrial reaction yet IT - first purified to homogeneity from human amniochorion [Kubilus 1978], [Gan 1981] - reversible [Brown 1998] - clears thymidine from the cytoplasm [Brown 1998]
TMDPP	3	Gan TE, Hallam L, Pilkington GR, Van der Weyden MB	A rapid and simple radiometric assay for thymidine phosphorylase of human peripheral blood cells	Clin Chim Acta	1981	7028324	in cytoplasm or in mitochondria - not really clear. In patient with Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE) the thymidine phosphorylase shows only 10% activity:"We hypothesize that, in patients with TP deficiency, increased levels of dThd and dUrd cause mitochondrial nucleotide pool imbalances, which, in turn, lead to mtDNA abnormalities including site-specific point mutations." - not included as mitochondrial reaction yet IT - first purified to homogeneity from human amniochorion [Kubilus 1978], [Gan 1981] - reversible [Brown 1998] - clears thymidine from the cytoplasm [Brown 1998]
TMDPP	3	Brown NS, Bicknell R.	Thymidine phosphorylase, 2-deoxy-D-ribose and angiogenesis	Biochem J	1998	9693094	in cytoplasm or in mitochondria - not really clear. In patient with Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE) the thymidine phosphorylase shows only 10% activity:"We hypothesize that, in patients with TP deficiency, increased levels of dThd and dUrd cause mitochondrial nucleotide pool imbalances, which, in turn, lead to mtDNA abnormalities including site-specific point mutations." - not included as mitochondrial reaction yet IT - first purified to homogeneity from human amniochorion [Kubilus 1978], [Gan 1981] - reversible [Brown 1998] - clears thymidine from the cytoplasm [Brown 1998]
TMDPPK	2	Bettendorff L, Mastrogiacomo F, Kish SJ, Grisar T.	Thiamine, thiamine phosphates, and their metabolizing enzymes in human brain.	J Neurochem	1996	8522961	IT proposed by Bettendorf et al., 1996 Such a function has to be pressent based on the fact that intracellular concentration of Thmtp has been measured (Bettendorf et al., 1996) thmtp is used by: - mitochondria: - PDHm - alpha-ketoglutarate deh.(1.2.4.2) - alpha-keto acid deh. (1.2.4.4) - cytosol: - transketolase (2.2.1.1)
TMDS	3	Kaneda S, Nalbantoglu J, Takeishi K, Shimizu K, Gotoh O, Seno T, Ayusawa D.	Structural and functional analysis of the human thymidylate synthase gene.	J Biol Chem	1990	2243092	IT homodimer (Genecards)
TMDS	3	Forsthoefel AM, Pena MM, Xing YY, Rafique Z, Berger FG.	Structural determinants for the intracellular degradation of human thymidylate synthase.	Biochemistry	2004	14967037	IT homodimer (Genecards)

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TMLYSOX	3	Hulse JD, Ellis SR, Henderson LM.	Carnitine biosynthesis. beta-Hydroxylation of trimethyllysine by an alpha-ketoglutarate-dependent mitochondrial dioxygenase.		1978	627563	-paper that states that this enzyme's localisation has not been resolved (may or may not be located in the mitochondrial matrix), so will assume it is in the inner membrane space (PMID:11802770) -sequence similar to purified rat enzyme; mitochondrial (see citation) -from Reactome: Carnitine is synthesized in four steps from trimethyllysine (generated in turn by the S-adenosyl-methionine mediated methylation of lysine residues in proteins, followed by protein hydrolysis). The enzymes that catalyze the first three steps of carnitine synthesis, converting trimethyllysine to gamma-butyrobetaine, are widely distributed in human tissues. The enzyme that catalyzes the last reaction, converting gamma-butyrobetaine to carnitine, is found only in liver and kidney cells, and at very low levels in brain tissues. Other tissues that require carnitine, such as muscle, are dependent on transport systems that mediate its export from the liver and uptake by other tissues. [Kerner & Hoppel 1998] MM
TMLYSOX	3	Vaz,F.M. , Ofman,R. , Westinga,K. , Back,J.W. , Wanders,R.J.	Molecular and Biochemical Characterization of Rat epsilon -N-Trimethyllysine Hydroxylase, the First Enzyme of Carnitine Biosynthesis.		2001	11431483	-paper that states that this enzyme's localisation has not been resolved (may or may not be located in the mitochondrial matrix), so will assume it is in the inner membrane space (PMID:11802770) -sequence similar to purified rat enzyme; mitochondrial (see citation) -from Reactome: Carnitine is synthesized in four steps from trimethyllysine (generated in turn by the S-adenosyl-methionine mediated methylation of lysine residues in proteins, followed by protein hydrolysis). The enzymes that catalyze the first three steps of carnitine synthesis, converting trimethyllysine to gamma-butyrobetaine, are widely distributed in human tissues. The enzyme that catalyzes the last reaction, converting gamma-butyrobetaine to carnitine, is found only in liver and kidney cells, and at very low levels in brain tissues. Other tissues that require carnitine, such as muscle, are dependent on transport systems that mediate its export from the liver and uptake by other tissues. [Kerner & Hoppel 1998] MM
TMLYSOX	3	Vaz FM, Wanders RJ.	Carnitine biosynthesis in mammals.		2002	11802770	-paper that states that this enzyme's localisation has not been resolved (may or may not be located in the mitochondrial matrix), so will assume it is in the inner membrane space (PMID:11802770) -sequence similar to purified rat enzyme; mitochondrial (see citation) -from Reactome: Carnitine is synthesized in four steps from trimethyllysine (generated in turn by the S-adenosyl-methionine mediated methylation of lysine residues in proteins, followed by protein hydrolysis). The enzymes that catalyze the first three steps of carnitine synthesis, converting trimethyllysine to gamma-butyrobetaine, are widely distributed in human tissues. The enzyme that catalyzes the last reaction, converting gamma-butyrobetaine to carnitine, is found only in liver and kidney cells, and at very low levels in brain tissues. Other tissues that require carnitine, such as muscle, are dependent on transport systems that mediate its export from the liver and uptake by other tissues. [Kerner & Hoppel 1998] MM
TRDR	3	Tamura T, Stadtman TC.	A new selenoprotein from human lung adenocarcinoma cells: purification, properties, and thioredoxin reductase activity.	Proc Natl Acad Sci U S A	1996	8577704	Genecards: activity with NADPH as acceptor homordimer acts on a number of substrates - 5,5'-dithiobis(2-nitrobenzoic acid), insulin (both in presence of thioredoxin, NADPH) - Tamura + Stadtman, 1996
TRDR	3	Koishi R, Kawashima I, Yoshimura C, Sugawara M, Serizawa N.	Cloning and characterization of a novel oxidoreductase KDRF from a human bone marrow-derived stromal cell line KM-102.	J Biol Chem	1997	8999974	Genecards: activity with NADPH as acceptor homordimer acts on a number of substrates - 5,5'-dithiobis(2-nitrobenzoic acid), insulin (both in presence of thioredoxin, NADPH) - Tamura + Stadtman, 1996
TRDR2	3	Xia L, Nordman T, Olsson JM, Damdimopoulos A, Bjorkhem-Bergman L, Nalvarte I, Eriksson LC, Arner ES, Spyrou G, Bjornstedt M.	The mammalian cytosolic selenoenzyme thioredoxin reductase reduces ubiquinone. A novel mechanism for defense against oxidative stress.	J Biol Chem	2003	12435734	IT reaction has been experimentally shown. apparently there are 3 important extra-mitochondrial ubiquinone reductase; lipoamide dehydrogenas, glutathionine reductase and thioredoxdin reductase. the question is how ubiquinone come into cytosol (and liposome). There might be a ubiquinol release from mitochondria but i did not found literature on it - will leave as gap for the moment.
TRDRm	3	Miranda-Vizuete A, Damdimopoulos AE, Pedrajas JR, Gustafsson JA, Spyrou G.	Human mitochondrial thioredoxin reductase cDNA cloning, expression and genomic organization.	Eur J Biochem	1999	10215850	IT

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TRDRm	3	Sun QA, Kirnarsky L, Sherman S, Gladyshev VN.	Selenoprotein oxidoreductase with specificity for thioredoxin and glutathione systems.	Proc Natl Acad Sci U S A	2001	11259642	IT
TRDRm	3	Kim MR, Chang HS, Kim BH, Kim S, Baek SH, Kim JH, Lee SR, Kim JR.	Involvements of mitochondrial thioredoxin reductase (TrxR2) in cell proliferation.	Biochem Biophys Res Commun	2003	12705894	IT
TREHe	3	Ishihara R, Taketani S, Sasai-Takedatsu M, Kino M, Tokunaga R, Kobayashi Y	Molecular cloning, sequencing and expression of cDNA encoding human trehalase	Gene	1997	9427547	- extrinsic to plasma membrane, GPI anchored [GO],[UniProt] - trehalose is broken down into glucose at the brush border [Richards et al, Food Chem Toxicol 2002] - mainly found in kidney, liver and small intestine [Ishihara, Gene 1997]
TREHe	3	Richards AB, Krakowka S, Dexter LB, Schmid H, Wolterbeek AP, Waalkens-Berendsen DH, Shigoyuki A, Kurimoto M	Trehalose: a review of properties, history of use and human tolerance, and results of multiple safety studies	Food Chem Toxicol	2002	12065209	- extrinsic to plasma membrane, GPI anchored [GO],[UniProt] - trehalose is broken down into glucose at the brush border [Richards et al, Food Chem Toxicol 2002] - mainly found in kidney, liver and small intestine [Ishihara, Gene 1997]
TRPHYDRO2	3	Wang L, Erlandsen H, Haavik J, Knappskog PM, Stevens RC	Three-dimensional structure of human tryptophan hydroxylase and its implications for the biosynthesis of the neurotransmitters serotonin and melatonin	Biochemistry	2002	12379098	Citations give this reaction.
TRPO2	3	Comings DE, Muhleman D, Dietz G, Sherman M, Forest GL	Sequence of human tryptophan 2,3-dioxygenase (TDO2): presence of a glucocorticoid response-like element composed of a GTT repeat and an intronic CCCCT repeat	Genomics	1995	8666386	Rate-limiting enzyme.
TSTSTERONESULT	3	Chatterjee B, Echchgadda I, Song CS	Vitamin D receptor regulation of the steroid/bile acid sulfotransferase SULT2A1	Methods Enzymol	2005	16399349	localization cytosol by swiss-prot specificity: liver, adrenal glands. Sulfotransferase enzymes catalyze the sulfate conjugation of many hormones, neurotransmitters, drugs, and xenobiotic compounds. These cytosolic enzymes are different in their tissue distributions and substrate specificities. The gene structure (number and length of exons) is similar among family members. This gene is primarily expressed in liver and adrenal tissues where the encoded protein sulfates steroids and bile acids. NJ
TYR3MO2	3	Bodeau-Pean S, Ravassard P, Neuner-Jehle M, Faucheux B, Mallet J, Dumas S	A human tyrosine hydroxylase isoform associated with progressive supranuclear palsy shows altered enzymatic activity	J Biol Chem	1999	9920892	This enzyme is the rate limiting step in catecholamine biosynthesis.
TYRASE	2	Box NF, Wyeth JR, Mayne CJ, O'Gorman LE, Martin NG, Sturm RA	Complete sequence and polymorphism study of the human TYRP1 gene encoding tyrosinase-related protein 1	Mamm Genome	1998	9434945	There may be a parallel pathway (as indicated in KEGG) that is involved in the synthesis of melanin. These reactions should take place exclusively in the melanosomes of melanocytes, but they are localized in the cytoplasm in this reconstruction because that specialized compartment is not available. The reference provides some physiological data. The other tyr gene (7299) may also be involved.
TYRDOPO	3	Pomerantz SH	The tyrosine hydroxylase activity of mammalian tyrosinase	J Biol Chem	1968	5294951	reaction mechanism (2 of each) questionable
TYRDOPO	3	Aaron Bunsen Lerner, Thomas B. Fitzpatrick, Evan Calkins, and William H. Summerson	MAMMALIAN TYROSINASE: PREPARATION AND PROPERTIES	J Biol Chem	1949		reaction mechanism (2 of each) questionable
TYROXDAc	3	Anderson MC, Hasan F, McCrodden JM, Tipton KF	Monoamine oxidase inhibitors and the cheese effect	Neurochem Res	1993	8255365	Biochemical data for this reaction is from the references which discuss the cheese effect. In short, certain Mao inhibitors (with clinical applications in treating depression) also inhibit the degradation of tyramine which is found in cheese, among other foods.
TYROXDAc	3	Humphrey SJ, Curry JT, Turman CN, Stryd RP	Cardiovascular sympathomimetic amine interactions in rats treated with monoamine oxidase inhibitors and the novel oxazolidinone antibiotic linezolid	J Cardiovasc Pharmacol	2001	11336106	Biochemical data for this reaction is from the references which discuss the cheese effect. In short, certain Mao inhibitors (with clinical applications in treating depression) also inhibit the degradation of tyramine which is found in cheese, among other foods.
TYROXDAc	3	Youdim MB, Weinstock M	Therapeutic applications of selective and non-selective inhibitors of monoamine oxidase A and B that do not cause significant tyramine potentiation	Neurotoxicology	2004	14697899	Biochemical data for this reaction is from the references which discuss the cheese effect. In short, certain Mao inhibitors (with clinical applications in treating depression) also inhibit the degradation of tyramine which is found in cheese, among other foods.
TYRTAm	3	Rettenmeier R, Natt E, Zentgraf H, Scherer G.	Isolation and characterization of the human tyrosine aminotransferase gene.	Nucleic Acids Res	1990	1973834	- Additional information added by RS/TV: 1) Tyrosine aminotransferase is a liver-specific enzyme that converts tyrosine to p-hydroxyphenylpyruvate in a pyridoxal phosphate-dependent transamination reaction according to Rettenmeier R, Natt E, Zentgraf H, Scherer G. Isolation and characterization of the human tyrosine aminotransferase gene. Nucleic Acids Res. 1990 Jul 11;18(13):3853-61. PMID: 1973834 mitochondrial according to Entrez gene database catalytic activity specified by GeneCards

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UAGDP	3	Mio T, Yabe T, Arisawa M, Yamada-Okabe H	The eukaryotic UDP-N-acetylglucosamine pyrophosphorylases. Gene cloning, protein expression, and catalytic mechanism	J Biol Chem	1998	9603950	- shown as irreversible in Devlin p. 672, Orten p. 244, Varki p. 74 6675: - cytoplasmic [UniProt] 91373: - only annotated as UDP-GlcNAc pyrophosphorylase in H-Inv Db
UDPDOLPT_L	0	Bossuyt X, Blanckaert N.	Topology of nucleotide-sugar:dolichyl phosphate glycosyltransferases involved in the dolichol pathway for protein glycosylation in native rat liver microsomes	Biochem J	1993	8280060	active centres of the transferases are cytoplasmically oriented Bossuyt X, Blanckaert N. Biochem J. 1993 Dec 15;296 (Pt 3):627-32.
UDPDOLPT_U	0	Imbach T, Burda P, Kuhnert P, Wevers RA, Aebi M, Berger EG, Hennet T.	A mutation in the human ortholog of the Saccharomyces cerevisiae ALG6 gene causes carbohydrate-deficient glycoprotein syndrome type-Ic.	Proc Natl Acad Sci USA	1999	10359825	active centres of the transferases are cytoplasmically oriented Bossuyt X, Blanckaert N. Biochem J. 1993 Dec 15;296 (Pt 3):627-32. Alg5p is ubiquitously expressed [Imbach et al, PNAS 1999]
UDPGALtg	3	Miura N, Ishida N, Hoshino M, Yamauchi M, Hara T, Ayusawa D, Kawakita M	Human UDP-galactose translocator: molecular cloning of a complementary DNA that complements the genetic defect of a mutant cell line deficient in UDP-galactose translocator	J Biochem (Tokyo)	1996	8889805	- cloned [Miura 1996], [Ishida 1996] - complemented UDPGal Golgi transport defect [Miura 1996] - expressed [Yoshioka 1997], [Sun-Wada 1998] - specific for UDPGal [Yoshioka 1997], [Sun-Wada 1998] and UDPGalNAc [Segawa 2002] - localized in Golgi [Yoshioka 1997], [Sun-Wada 1998]
UDPGALtg	3	Yoshioka S, Sun-Wada GH, Ishida N, Kawakita M	Expression of the human UDP-galactose transporter in the Golgi membranes of murine Had-1 cells that lack the endogenous transporter	J Biochem (Tokyo)	1997	9399569	- cloned [Miura 1996], [Ishida 1996] - complemented UDPGal Golgi transport defect [Miura 1996] - expressed [Yoshioka 1997], [Sun-Wada 1998] - specific for UDPGal [Yoshioka 1997], [Sun-Wada 1998] and UDPGalNAc [Segawa 2002] - localized in Golgi [Yoshioka 1997], [Sun-Wada 1998]
UDPGALtg	3	Sun-Wada GH, Yoshioka S, Ishida N, Kawakita M	Functional expression of the human UDP-galactose transporters in the yeast Saccharomyces cerevisiae	J Biochem (Tokyo)	1998	9562625	- cloned [Miura 1996], [Ishida 1996] - complemented UDPGal Golgi transport defect [Miura 1996] - expressed [Yoshioka 1997], [Sun-Wada 1998] - specific for UDPGal [Yoshioka 1997], [Sun-Wada 1998] and UDPGalNAc [Segawa 2002] - localized in Golgi [Yoshioka 1997], [Sun-Wada 1998]
UDPGALtg	3	Segawa H, Kawakita M, Ishida N	Human and Drosophila UDP-galactose transporters transport UDP-N-acetylgalactosamine in addition to UDP-galactose	Eur J Biochem	2002	11784306	- cloned [Miura 1996], [Ishida 1996] - complemented UDPGal Golgi transport defect [Miura 1996] - expressed [Yoshioka 1997], [Sun-Wada 1998] - specific for UDPGal [Yoshioka 1997], [Sun-Wada 1998] and UDPGalNAc [Segawa 2002] - localized in Golgi [Yoshioka 1997], [Sun-Wada 1998]
UDPGD	2	Banhegyi G, Braun L, Csala M, Puskas F, Mandl J	Ascorbate metabolism and its regulation in animals	Free Radic Biol Med	1997	9296457	- reaction described in Devlin pp 611-612 - shown as irreversible in Orten p 241 - thought to be important for hyaluronan biosynthesis, which is occurs on inner side of plasma membrane - occurs in the cytosol [Banhegyi 1997]
UDPGD	2	Spicer AP, Kaback LA, Smith TJ, Seldin MF	Molecular cloning and characterization of the human and mouse UDP-glucose dehydrogenase genes	J Biol Chem	1998	9737970	- reaction described in Devlin pp 611-612 - shown as irreversible in Orten p 241 - thought to be important for hyaluronan biosynthesis, which is occurs on inner side of plasma membrane - occurs in the cytosol [Banhegyi 1997]
UDPGLCAter	3	Muraoka M, Kawakita M, Ishida N	Molecular characterization of human UDP-glucuronic acid/UDP-N-acetylgalactosamine transporter, a novel nucleotide sugar transporter with dual substrate specificity	FEBS Lett	2001	11322953	- cloned [Muraoka 2001] - UDP-GlcA and UDP-GalNAc transport activity determined by expression in yeast [Muraoka 2001] - ER [Muraoka 2001]
UDPGLCter	2	Trombetta ES, Helenius A.	Glycoprotein reglucosylation and nucleotide sugar utilization in the secretory pathway: identification of a nucleoside diphosphatase in the endoplasmic reticulum.	EMBO J	1999	10369669	- there is a UDP-Glc transporter in both the ER and Golgi [Varki, p. 79-80] - UDP-Glc is transported from the cytosol where it is synthesized into the ER lumen where it is used by UDP-Glc:glycoprotein glucosyltransferase (see refs in [Trombetta 1999])

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UDPGLCtg	3	Milla ME, Clairmont CA, Hirschberg CB	Reconstitution into proteoliposomes and partial purification of the Golgi apparatus membrane UDP-galactose, UDP-xylose, and UDP-glucuronic acid transport activities	J Biol Chem	1992	1730575	23443: - cloned [Ishida 1999] - ubiquitous [Ishida 1999] - Golgi [Ishida 1999] - transports UDPGlc [Ishida 1999] 11046: - cloned [Ishida 2005], [Suda 2004] - 50% similarity w/ human SLC35D1, fruitfly fringe connection (frc) transporter, nematode SQV-7 transporter [Ishida 2005] - expression in yeast yielded significant (but only slightly higher) transport of UDPGlcNAc [Ishida 2005], [Suda 2004], UDPGlc & UDPMan [Suda 2004]; in mammalian cells only transported UDPGlcNAc and UDPGlc [Suda 2004] - Golgi [Ishida 2005], [Suda 2004] - high in colon, lung, stomach; moderate in WBC, pancreas; low in thyroid, uterus, placenta, sk muscle, testis, adrenal glands; barely detectable in liver, kidney, mammary, salivary, spinal cord, trachea [Suda 2004] 84912: - Golgi [Ashikov 2005] - demonstrated UDP-Glc, UDP-Xyl transport activity by expressing genes in yeast and measuring transport in vitro from Golgi fractions [Ashikov 2005] - earlier results in rat showed that UDP-Xyl can access Golgi but protein had not been characterized at molecular level [Nuwayhid 1986], [Milla 1992]
UDPGLCtg	3	Nuwayhid N, Glaser JH, Johnson JC, Conrad HE, Hauser SC, Hirschberg CB	Xylosylation and glucuronosylation reactions in rat liver Golgi apparatus and endoplasmic reticulum	J Biol Chem	1986	3093474	23443: - cloned [Ishida 1999] - ubiquitous [Ishida 1999] - Golgi [Ishida 1999] - transports UDPGlc [Ishida 1999] 11046: - cloned [Ishida 2005], [Suda 2004] - 50% similarity w/ human SLC35D1, fruitfly fringe connection (frc) transporter, nematode SQV-7 transporter [Ishida 2005] - expression in yeast yielded significant (but only slightly higher) transport of UDPGlcNAc [Ishida 2005], [Suda 2004], UDPGlc & UDPMan [Suda 2004]; in mammalian cells only transported UDPGlcNAc and UDPGlc [Suda 2004] - Golgi [Ishida 2005], [Suda 2004] - high in colon, lung, stomach; moderate in WBC, pancreas; low in thyroid, uterus, placenta, sk muscle, testis, adrenal glands; barely detectable in liver, kidney, mammary, salivary, spinal cord, trachea [Suda 2004] 84912: - Golgi [Ashikov 2005] - demonstrated UDP-Glc, UDP-Xyl transport activity by expressing genes in yeast and measuring transport in vitro from Golgi fractions [Ashikov 2005] - earlier results in rat showed that UDP-Xyl can access Golgi but protein had not been characterized at molecular level [Nuwayhid 1986], [Milla 1992]
UDPGLCtg	3	Ishida N, Yoshioka S, Chiba Y, Takeuchi M, Kawakita M	Molecular cloning and functional expression of the human Golgi UDP-N-acetylglucosamine transporter	J Biochem (Tokyo)	1999	10393322	23443: - cloned [Ishida 1999] - ubiquitous [Ishida 1999] - Golgi [Ishida 1999] - transports UDPGlc [Ishida 1999] 11046: - cloned [Ishida 2005], [Suda 2004] - 50% similarity w/ human SLC35D1, fruitfly fringe connection (frc) transporter, nematode SQV-7 transporter [Ishida 2005] - expression in yeast yielded significant (but only slightly higher) transport of UDPGlcNAc [Ishida 2005], [Suda 2004], UDPGlc & UDPMan [Suda 2004]; in mammalian cells only transported UDPGlcNAc and UDPGlc [Suda 2004] - Golgi [Ishida 2005], [Suda 2004] - high in colon, lung, stomach; moderate in WBC, pancreas; low in thyroid, uterus, placenta, sk muscle, testis, adrenal glands; barely detectable in liver, kidney, mammary, salivary, spinal cord, trachea [Suda 2004] 84912: - Golgi [Ashikov 2005] - demonstrated UDP-Glc, UDP-Xyl transport activity by expressing genes in yeast and measuring transport in vitro from Golgi fractions [Ashikov 2005] - earlier results in rat showed that UDP-Xyl can access Golgi but protein had not been characterized at molecular level [Nuwayhid 1986], [Milla 1992]

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UDPGLCtg	3	Suda T, Kamiyama S, Suzuki M, Kikuchi N, Nakayama K, Narimatsu H, Jigami Y, Aoki T, Nishihara S	Molecular cloning and characterization of a human multisubstrate specific nucleotide-sugar transporter homologous to Drosophila fringe connection	J Biol Chem	2004	15082721	23443: - cloned [Ishida 1999] - ubiquitous [Ishida 1999] - Golgi [Ishida 1999] - transports UDPGlc [Ishida 1999] 11046: - cloned [Ishida 2005], [Suda 2004] - 50% similarity w/ human SLC35D1, fruitfly fringe connection (frc) transporter, nematode SQV-7 transporter [Ishida 2005] - expression in yeast yielded significant (but only slightly higher) transport of UDPGlcNAc [Ishida 2005], [Suda 2004], UDPGlc & UDPMan [Suda 2004]; in mammalian cells only transported UDPGlcNAc and UDPGlc [Suda 2004] - Golgi [Ishida 2005], [Suda 2004] - high in colon, lung, stomach; moderate in WBC, pancreas; low in thyroid, uterus, placenta, sk muscle, testis, adrenal glands; barely detectable in liver, kidney, mammary, salivary, spinal cord, trachea [Suda 2004] 84912: - Golgi [Ashikov 2005] - demonstrated UDP-Glc, UDP-Xyl transport activity by expressing genes in yeast and measuring transport in vitro from Golgi fractions [Ashikov 2005] - earlier results in rat showed that UDP-Xyl can access Golgi but protein had not been characterized at molecular level [Nuwayhid 1986], [Milla 1992]
UDPGLCtg	3	Ishida N, Kuba T, Aoki K, Miyatake S, Kawakita M, Sanai Y	Identification and characterization of human Golgi nucleotide sugar transporter SLC35D2, a novel member of the SLC35 nucleotide sugar transporter family	Genomics	2005	15607426	23443: - cloned [Ishida 1999] - ubiquitous [Ishida 1999] - Golgi [Ishida 1999] - transports UDPGlc [Ishida 1999] 11046: - cloned [Ishida 2005], [Suda 2004] - 50% similarity w/ human SLC35D1, fruitfly fringe connection (frc) transporter, nematode SQV-7 transporter [Ishida 2005] - expression in yeast yielded significant (but only slightly higher) transport of UDPGlcNAc [Ishida 2005], [Suda 2004], UDPGlc & UDPMan [Suda 2004]; in mammalian cells only transported UDPGlcNAc and UDPGlc [Suda 2004] - Golgi [Ishida 2005], [Suda 2004] - high in colon, lung, stomach; moderate in WBC, pancreas; low in thyroid, uterus, placenta, sk muscle, testis, adrenal glands; barely detectable in liver, kidney, mammary, salivary, spinal cord, trachea [Suda 2004] 84912: - Golgi [Ashikov 2005] - demonstrated UDP-Glc, UDP-Xyl transport activity by expressing genes in yeast and measuring transport in vitro from Golgi fractions [Ashikov 2005] - earlier results in rat showed that UDP-Xyl can access Golgi but protein had not been characterized at molecular level [Nuwayhid 1986], [Milla 1992]
UDPGLCtg	3	Ashikov A, Routier F, Fuhlrott J, Helmus Y, Wild M, Gerardy-Schahn R, Bakker H	The Human Solute Carrier Gene SLC35B4 Encodes a Bifunctional Nucleotide Sugar Transporter with Specificity for UDP-Xylose and UDP-N-Acetylglucosamine	J Biol Chem	2005	15911612	23443: - cloned [Ishida 1999] - ubiquitous [Ishida 1999] - Golgi [Ishida 1999] - transports UDPGlc [Ishida 1999] 11046: - cloned [Ishida 2005], [Suda 2004] - 50% similarity w/ human SLC35D1, fruitfly fringe connection (frc) transporter, nematode SQV-7 transporter [Ishida 2005] - expression in yeast yielded significant (but only slightly higher) transport of UDPGlcNAc [Ishida 2005], [Suda 2004], UDPGlc & UDPMan [Suda 2004]; in mammalian cells only transported UDPGlcNAc and UDPGlc [Suda 2004] - Golgi [Ishida 2005], [Suda 2004] - high in colon, lung, stomach; moderate in WBC, pancreas; low in thyroid, uterus, placenta, sk muscle, testis, adrenal glands; barely detectable in liver, kidney, mammary, salivary, spinal cord, trachea [Suda 2004] 84912: - Golgi [Ashikov 2005] - demonstrated UDP-Glc, UDP-Xyl transport activity by expressing genes in yeast and measuring transport in vitro from Golgi fractions [Ashikov 2005] - earlier results in rat showed that UDP-Xyl can access Golgi but protein had not been characterized at molecular level [Nuwayhid 1986], [Milla 1992]

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UDPGLDCg	3	Silbert JE, DeLuca S	The synthesis of uridine diphosphate xylose by particulate preparations from mouse mast-cell tumors	Biochim Biophys Acta	1967	4293108	- reaction described in Devlin p. 672, Varki p.74 - occurs in the lumen of ER or Golgi [Silbert, Biochim Biophy Acta, 1967], [Kearns, J Biol Chem 1993] - cloned and expressed, had UDP-glucuronate decarboxylase activity [Moriarity 2002] - perinuclear Golgi [Moriarity 2002] - highest expression levels in heart, brain, and testes; moderate levels in kidney, liver, lung, lower levels in spleen, sk muscle; immunochemical studies showed significant staining in kidney, liver, and brain [Moriarity 2002] - 75-80% amino acid sequence identity and 90% similarity between plants and mammals [Moriarity 2002]
UDPGLDCg	3	Moriarity JL, Hurt KJ, Resnick AC, Storm PB, Laroy W, Schnaar RL, Snyder SH	UDP-glucuronate decarboxylase, a key enzyme in proteoglycan synthesis: cloning, characterization, and localization	J Biol Chem	2002	11877387	- reaction described in Devlin p. 672, Varki p.74 - occurs in the lumen of ER or Golgi [Silbert, Biochim Biophy Acta, 1967], [Kearns, J Biol Chem 1993] - cloned and expressed, had UDP-glucuronate decarboxylase activity [Moriarity 2002] - perinuclear Golgi [Moriarity 2002] - highest expression levels in heart, brain, and testes; moderate levels in kidney, liver, lung, lower levels in spleen, sk muscle; immunochemical studies showed significant staining in kidney, liver, and brain [Moriarity 2002] - 75-80% amino acid sequence identity and 90% similarity between plants and mammals [Moriarity 2002]
UGCG	3	Ichikawa S, Sakiyama H, Suzuki G, Hidari KI, Hirabayashi Y.	Expression cloning of a cDNA for human ceramide glucosyltransferase that catalyzes the first glycosylation step of glycosphingolipid synthesis.	Proc Natl Acad Sci	1996	8643456	ER - uniprot May serve as a "flippase" as well as a glucosyltransferase that transfers glucose to ceramide. --> dual fxn as transporter also Glycosphingolipids (GSLs) are a group of membrane components that contain lipid and sugar moieties. They are present in essentially all animal cells and are believed to have important roles in various cellular processes. UDP-glucose ceramide glucosyltransferase catalyzes the first glycosylation step in glycosphingolipid biosynthesis. The product, glucosylceramide, is the core structure of more than 300 GSLs. UGCG is widely expressed and transciption is upregulated during keratinocyte differentiation. NJ
UGLT	0	Holden HM, Rayment I, Thoden JB	Structure and function of enzymes of the Leloir pathway for galactose metabolism	J Biol Chem	2003	12923184	- UGLT reaction is reversible [Holden et al, J Biol Chem, 2003]
UGT1A1r	3	Basu NK, Kubota S, Meselhy MR, Ciotti M, Chowdhury B, Hartori M, Owens IS.	Gastrointestinally distributed UDP-glucuronosyltransferase 1A10, which metabolizes estrogens and nonsteroidal anti-inflammatory drugs, depends upon phosphorylation.	J Biol Chem	2004		specificity: microsomal - uniprot specificity: Liver and colon. This gene encodes a UDP-glucuronosyltransferase, an enzyme of the glucuronidation pathway that transforms small lipophilic molecules, such as steroids, bilirubin, hormones, and drugs, into water-soluble, excretable metabolites. This gene is part of a complex locus that encodes several UDP-glucuronosyltransferases. The locus includes thirteen unique alternate first exons followed by four common exons. Four of the alternate first exons are considered pseudogenes. Each of the remaining nine 5' exons may be spliced to the four common exons, resulting in nine proteins with different N-termini and identical C-termini. Each first exon encodes the substrate binding site, and is regulated by its own promoter. The enzyme encoded by this gene has glucuronidase activity on mycophenolic acid, coumarins, and quinolines. NJ
UGT1A1r	3	Mackenzie PI, Owens IS, Burchell B, Bock KW, Bairoch A, Belanger A, Fournel-Gigleux S, Green M, Hum DW, Iyanagi T, Lancet D, Louisot P, Magdalou J, Chowdhury JR, Ritter JK, Schachter H, Tephly TR, Tipton KF, Nebert DW.	The UDP glycosyltransferase gene superfamily: recommended nomenclature update based on evolutionary divergence.	Pharmacogenetics	1997	9295054	specificity: microsomal - uniprot specificity: Liver and colon. This gene encodes a UDP-glucuronosyltransferase, an enzyme of the glucuronidation pathway that transforms small lipophilic molecules, such as steroids, bilirubin, hormones, and drugs, into water-soluble, excretable metabolites. This gene is part of a complex locus that encodes several UDP-glucuronosyltransferases. The locus includes thirteen unique alternate first exons followed by four common exons. Four of the alternate first exons are considered pseudogenes. Each of the remaining nine 5' exons may be spliced to the four common exons, resulting in nine proteins with different N-termini and identical C-termini. Each first exon encodes the substrate binding site, and is regulated by its own promoter. The enzyme encoded by this gene has glucuronidase activity on mycophenolic acid, coumarins, and quinolines. NJ

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UGT1A3r	3	Levesque E, Turgeon D, Carrier JS, Montminy V, Beaulieu M, Belanger A.	Isolation and characterization of the UGT2B28 cDNA encoding a novel human steroid conjugating UDP-glucuronosyltransferase.	Biochemistry	2001	11300766	specificity: microsomal - uniprot specificity: Liver and colon. This gene encodes a UDP-glucuronosyltransferase, an enzyme of the glucuronidation pathway that transforms small lipophilic molecules, such as steroids, bilirubin, hormones, and drugs, into water-soluble, excretable metabolites. This gene is part of a complex locus that encodes several UDP-glucuronosyltransferases. The locus includes thirteen unique alternate first exons followed by four common exons. Four of the alternate first exons are considered pseudogenes. Each of the remaining nine 5' exons may be spliced to the four common exons, resulting in nine proteins with different N-termini and identical C-termini. Each first exon encodes the substrate binding site, and is regulated by its own promoter. The enzyme encoded by this gene has glucuronidase activity on mycophenolic acid, coumarins, and quinolines. UGT2B28: PMID: 11300766. TISSUE SPECIFICITY: High expression in the liver and pancreas, lower in the skeletal muscle and kidney. See also PMID: 14643063 and 15666817 - for more about UGT2B7/17 - high expression in prostate gland (epithelium). NJ
UGT1A3r	3	Belanger A, Pelletier G, Labrie F, Barbier O, Chouinard S.	Inactivation of androgens by UDP-glucuronosyltransferase enzymes in humans.	Trends Endocrinol Metab	2003	14643063	specificity: microsomal - uniprot specificity: Liver and colon. This gene encodes a UDP-glucuronosyltransferase, an enzyme of the glucuronidation pathway that transforms small lipophilic molecules, such as steroids, bilirubin, hormones, and drugs, into water-soluble, excretable metabolites. This gene is part of a complex locus that encodes several UDP-glucuronosyltransferases. The locus includes thirteen unique alternate first exons followed by four common exons. Four of the alternate first exons are considered pseudogenes. Each of the remaining nine 5' exons may be spliced to the four common exons, resulting in nine proteins with different N-termini and identical C-termini. Each first exon encodes the substrate binding site, and is regulated by its own promoter. The enzyme encoded by this gene has glucuronidase activity on mycophenolic acid, coumarins, and quinolines. UGT2B28: PMID: 11300766. TISSUE SPECIFICITY: High expression in the liver and pancreas, lower in the skeletal muscle and kidney. See also PMID: 14643063 and 15666817 - for more about UGT2B7/17 - high expression in prostate gland (epithelium). NJ
UGT1A5r2	2	Barua AB, Sidell N.	Retinoyl beta-glucuronide: a biologically active interesting retinoid.	J Nutr	2004	14704335	IT
UGT1A8r	3	Jin CJ, Miners JO, Lillywhite KJ, Mackenzie PI.	cDNA cloning and expression of two new members of the human liver UDP-glucuronosyltransferase 2B subfamily.	Biochem Biophys Res Commun	1993	8333863	specificity: microsomal - uniprot specificity: Liver and colon. This gene encodes a UDP-glucuronosyltransferase, an enzyme of the glucuronidation pathway that transforms small lipophilic molecules, such as steroids, bilirubin, hormones, and drugs, into water-soluble, excretable metabolites. This gene is part of a complex locus that encodes several UDP-glucuronosyltransferases. The locus includes thirteen unique alternate first exons followed by four common exons. Four of the alternate first exons are considered pseudogenes. Each of the remaining nine 5' exons may be spliced to the four common exons, resulting in nine proteins with different N-termini and identical C-termini. Each first exon encodes the substrate binding site, and is regulated by its own promoter. The enzyme encoded by this gene has glucuronidase activity on mycophenolic acid, coumarins, and quinolines. UGT2B28: PMID: 11300766. TISSUE SPECIFICITY: High expression in the liver and pancreas, lower in the skeletal muscle and kidney. UGT2B4: PMID: 11300766. PMID: 8333863 NJ
UPP3S	0	Tsai SF, Bishop DF, Desnick RJ.	Human uroporphyrinogen III synthase: molecular cloning, nucleotide sequence, and expression of a full-length cDNA.	Proc Natl Acad Sci U S A	1988	3174619	- Added by RS/TV Proteome - Uroporphyrinogen-III synthase (UROS), hmb is rapidly converted to uroporphyrinogen III by an intramolecular rearrangment of the D-pyrrole group and ring closure. (Tsai SF, Bishop DF, Desnick RJ. Proc Natl Acad Sci U S A. 1988 Oct;85(19):7049-53) - Although expressed in multiple tissues, prominently expressed in liver, heart, and skeletal muscle. (Aizencang G, Solis C, Bishop DF, Warner C, Desnick RJ. Genomics. 2000 Dec 1;70(2):223-31.) - Catalytic activity also specified by GeneCards.

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UPPDC1	3	Romeo PH, Raich N, Dubart A, Beaupain D, Pryor M, Kushner J, Cohen-Solal M, Goossens M.	Molecular cloning and nucleotide sequence of a complete human uroporphyrinogen decarboxylase cDNA.	J Biol Chem	1986	3015909	- Added by RS/TV Biochem textbook - Cytoplasmic according to GeneCards - Uroporphyrinogen decarboxylase is a cytosolic enzyme involved in the biosynthesis of heme. It catalyzes the sequential removal of the four carboxyl groups of the carboxymethyl side chains of uroporphyrinogen to yield coproporphyrinogen. (Romeo PH, Raich N, Dubart A, Beaupain D, Pryor M, Kushner J, Cohen-Solal M, Goossens M. J Biol Chem. 1986 Jul 25;261(21):9825-31)
UPPN	3	Vreken P, van Kuilenburg AB, Hamajima N, Meinsma R, van Lenthe H, Gohlich-Ratmann G, Assmann BE, Wevers RA, van Gennip AH	cDNA cloning, genomic structure and chromosomal localization of the human BUP-1 gene encoding beta-ureidopropionase	Biochim Biophys Acta	1999	10542323	Standard degradation pathway of uracil, which also works for a related drug according to the first citation. Pyrimidine Catabolism
UPPN	3	Kuhara T	Diagnosis and monitoring of inborn errors of metabolism using urease-pretreatment of urine, isotope dilution, and gas chromatography-mass spectrometry	J Chromatogr B Analyt Technol Biomed Life Sci	2002	12450676	Standard degradation pathway of uracil, which also works for a related drug according to the first citation. Pyrimidine Catabolism
URCN	3	Kessler D, Retey J, Schulz GE	Structure and action of urocanase	J Mol Biol	2004	15313616	Common enzyme.
UREAt	3	Dai G, Levy O, Carrasco N	Cloning and characterization of the thyroid iodide transporter	Nature	1996	8559252	-cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6563,8170 - In mammalian cells, urea is the chief end-product of nitrogen catabolism and plays an important role in the urinary concentration mechanism. Thus, the plasma membrane of erythrocytes and some renal epithelial cells exhibit an elevated urea permeability that is mediated by highly selective urea transporters. In mammals, 2 urea transporters have been identified: the renal tubular urea transporter, UT2, and the erythrocyte urea transporter, UT11 (SLC14A1; MIM 111000).[supplied by OMIM] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandar - primarily in thyroid gland [De La Vieja 2000], also expressed - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs
UREAt	3	Smanik PA, Liu Q, Furminger TL, Ryu K, Xing S, Mazzaferri EL, Jhiang SM	Cloning of the human sodium lodide symporter	Biochem Biophys Res Commun	1996	8806637	-cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6563,8170 - In mammalian cells, urea is the chief end-product of nitrogen catabolism and plays an important role in the urinary concentration mechanism. Thus, the plasma membrane of erythrocytes and some renal epithelial cells exhibit an elevated urea permeability that is mediated by highly selective urea transporters. In mammals, 2 urea transporters have been identified: the renal tubular urea transporter, UT2, and the erythrocyte urea transporter, UT11 (SLC14A1; MIM 111000).[supplied by OMIM] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandar - primarily in thyroid gland [De La Vieja 2000], also expressed - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs

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UREAt	3	Eskandari S, Loo DD, Dai G, Levy O, Wright EM, Carrasco N.	Thyroid Na+/I- symporter. Mechanism, stoichiometry, and specificity	J Biol Chem	1997	9341168	-cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6563,8170 - In mammalian cells, urea is the chief end-product of nitrogen catabolism and plays an important role in the urinary concentration mechanism. Thus, the plasma membrane of erythrocytes and some renal epithelial cells exhibit an elevated urea permeability that is mediated by highly selective urea transporters. In mammals, 2 urea transporters have been identified: the renal tubular urea transporter, UT2, and the erythrocyte urea transporter, UT11 (SLC14A1; MIM 111000).[supplied by OMIM] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandar - primarily in thyroid gland [De La Vieja 2000], also expressed - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs
UREAt	3	De La Vieja A, Dohan O, Levy O, Carrasco N	Molecular analysis of the sodium/iodide symporter: impact on thyroid and extrathyroid pathophysiology	Physiol Rev	2000	10893432	-cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6563,8170 - In mammalian cells, urea is the chief end-product of nitrogen catabolism and plays an important role in the urinary concentration mechanism. Thus, the plasma membrane of erythrocytes and some renal epithelial cells exhibit an elevated urea permeability that is mediated by highly selective urea transporters. In mammals, 2 urea transporters have been identified: the renal tubular urea transporter, UT2, and the erythrocyte urea transporter, UT11 (SLC14A1; MIM 111000).[supplied by OMIM] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandar - primarily in thyroid gland [De La Vieja 2000], also expressed - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs
UREAt	3	Wapnir IL, van de Rijn M, Nowels K, Amenta PS, Walton K, Montgomery K, Greco RS, Dohan O, Carrasco N	Immunohistochemical profile of the sodium/iodide symporter in thyroid, breast, and other carcinomas using high density tissue microarrays and conventional sections	J Clin Endocrinol Metab	2003	12679487	-cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6563,8170 - In mammalian cells, urea is the chief end-product of nitrogen catabolism and plays an important role in the urinary concentration mechanism. Thus, the plasma membrane of erythrocytes and some renal epithelial cells exhibit an elevated urea permeability that is mediated by highly selective urea transporters. In mammals, 2 urea transporters have been identified: the renal tubular urea transporter, UT2, and the erythrocyte urea transporter, UT11 (SLC14A1; MIM 111000).[supplied by OMIM] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandar - primarily in thyroid gland [De La Vieja 2000], also expressed - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs

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UREAt	3	Shayakul C, Hediger MA.	The SLC14 gene family of urea transporters.		2004	12856182	-cloned [Hediger 1989] - cotransports Glc/2 Na+, Gal/2 Na+ [Quick 2001] - H+ can replace Na+ [Hirayama 1994] - behaves as urea channel in the absence of substrates; cotransports urea under substrate-transporting conditions [Leung 2000] - Na+ transport occurs by a saturable uniport mechanism; water permeation is through a low conductance water channel [Loo 1999] - brush border membrane [Wright 1994] - plasma membrane; see [Wright 2004] for refs 6563,8170 - In mammalian cells, urea is the chief end-product of nitrogen catabolism and plays an important role in the urinary concentration mechanism. Thus, the plasma membrane of erythrocytes and some renal epithelial cells exhibit an elevated urea permeability that is mediated by highly selective urea transporters. In mammals, 2 urea transporters have been identified: the renal tubular urea transporter, UT2, and the erythrocyte urea transporter, UT11 (SLC14A1; MIM 111000).[supplied by OMIM] 6528: - cloned [Dai 1996] - gene has 84% identity to the rat homolog [Smanik 1996] - sodium iodide cotransport [Dai 1996]; 2 Na+ per I- [Eskandar - primarily in thyroid gland [De La Vieja 2000], also expressed - also transports ClO3-, SCN-, SeCN-, NO3-, Br-, BF4-, IO4-, I - basolateral plasma membrane; see [Wright 2004] for refs
UREAtm	2	Tsukaguchi H, Shayakul C, Berger UV, Mackenzie B, Devidas S, Guggino WB, van Hoek AN, Hediger MA	Molecular characterization of a broad selectivity neutral solute channel	J Biol Chem	1998	9733774	Citations indicate that this aquaporin can transport urea into the mitochondria. There is no explicit information that it is reversible, but the reversibility fills a gap in the model and is thus assumed. This transporter can supposedly also transport lactate into the mitochondria, but that functionality has been omitted for now due to loop issues. Also, this transporter takes many other neutral solutes, probably more prominently into and out of the cell rather than the mitochondria.
UREAtm	2	Agre P, King LS, Yasui M, Guggino WB, Ottersen OP, Fujiyoshi Y, Engel A, Nielsen S	Aquaporin water channels--from atomic structure to clinical medicine	J Physiol	2002	12096044	Citations indicate that this aquaporin can transport urea into the mitochondria. There is no explicit information that it is reversible, but the reversibility fills a gap in the model and is thus assumed. This transporter can supposedly also transport lactate into the mitochondria, but that functionality has been omitted for now due to loop issues. Also, this transporter takes many other neutral solutes, probably more prominently into and out of the cell rather than the mitochondria.
UREAtm	2	Amiry-Moghaddam M, Lindland H, Zelenin S, Roberg BA, Gundersen BB, Petersen P, Rinvik E, Torgner IA, Ottersen OP	Brain mitochondria contain aquaporin water channels: evidence for the expression of a short AQP9 isoform in the inner mitochondrial membrane	FASEB J	2005	16126913	Citations indicate that this aquaporin can transport urea into the mitochondria. There is no explicit information that it is reversible, but the reversibility fills a gap in the model and is thus assumed. This transporter can supposedly also transport lactate into the mitochondria, but that functionality has been omitted for now due to loop issues. Also, this transporter takes many other neutral solutes, probably more prominently into and out of the cell rather than the mitochondria.
URIK1	3	Ozaki K, Kuroki T, Hayashi S, Nakamura Y.	Isolation of three testis-specific genes (TSA303, TSA806, TSA903) by a differential mRNA display method.	Genomics	1996	8812458	IT Kashuba 2002 found 7371 product predominately in cytoplasme but when cell was infected with Epstein-Barr virus, the protein was located in nucleus --> I did not account for this observation yet. Found protein expressed in most tissues. Ozaki et al, 1996 found mRNA expressed in testis UCK2: interestingly the papers differ in their results of expression of the corresponding mRNA. While Ozaki et al, 1996, found the mRNA only in testis, and Van Rompay et al 2001 only in placenta, Kashuba et al 2002 detected the mRNA in most tissues they investigated ...??? UCK1,UCK2: Van Rompay et al., 2001:The enzymes phosphorylated several of the analogs, such as 6-azauridine, 5-fluorouridine, 4-thiouridine, 5-bromouridine, N(4)-acetylcytidine, N(4)-benzoylcytidine, 5-fluorocytidine, 2-thiocytidine, 5-methylcytidine, and N(4)-anisoylcytidine

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URIK1	3	Van Rompay AR, Norda A, Linden K, Johansson M, Karlsson A.	Phosphorylation of uridine and cytidine nucleoside analogs by two human uridine-cytidine kinases.	Mol Pharmacol	2001	11306702	IT Kashuba 2002 found 7371 product predominately in cytoplasme but when cell was infected with Epstein-Barr virus, the protein was located in nucleus --> I did not account for this observation yet. Found protein expressed in most tissues. Ozaki et al, 1996 found mRNA expressed in testis UCK2: interestingly the papers differ in their results of expression of the corresponding mRNA. While Ozaki et al, 1996, found the mRNA only in testis, and Van Rompay et al 2001 only in placenta, Kashuba et al 2002 detected the mRNA in most tissues they investigated ...??? UCK1,UCK2: Van Rompay et al., 2001:The enzymes phosphorylated several of the analogs, such as 6-azauridine, 5-fluorouridine, 4-thiouridine, 5-bromouridine, N(4)-acetylcytidine, N(4)-benzoylcytidine, 5-fluorocytidine, 2-thiocytidine, 5-methylcytidine, and N(4)-anisoylcytidine
URIK1	3	Kashuba E, Kashuba V, Sandalova T, Klein G, Szekely L	Epstein-Barr virus encoded nuclear protein EBNA-3 binds a novel human uridine kinase/uracil phosphoribosyltransferase.	BMC Cell Biol	2002	12199906	IT Kashuba 2002 found 7371 product predominately in cytoplasme but when cell was infected with Epstein-Barr virus, the protein was located in nucleus --> I did not account for this observation yet. Found protein expressed in most tissues. Ozaki et al, 1996 found mRNA expressed in testis UCK2: interestingly the papers differ in their results of expression of the corresponding mRNA. While Ozaki et al, 1996, found the mRNA only in testis, and Van Rompay et al 2001 only in placenta, Kashuba et al 2002 detected the mRNA in most tissues they investigated ...??? UCK1,UCK2: Van Rompay et al., 2001:The enzymes phosphorylated several of the analogs, such as 6-azauridine, 5-fluorouridine, 4-thiouridine, 5-bromouridine, N(4)-acetylcytidine, N(4)-benzoylcytidine, 5-fluorocytidine, 2-thiocytidine, 5-methylcytidine, and N(4)-anisoylcytidine
Uritl	3	Pisoni RL, Thoene JG.	Detection and characterization of a nucleoside transport system in human fibroblast lysosomes.	J Biol Chem	1989	2925670	IT it is not clear whether H+ is co-transported or not. since influx has low affinity it is more likely that transporter serve to remove nucleosides from lysosome rather then importing them (baldwin, 2005)
Uritl	3	Baldwin SA, Yao SY, Hyde RJ, Ng AM, Foppolo S, Barnes K, Ritzel MW, Cass CE, Young JD.	Functional characterization of novel human and mouse equilibrative nucleoside transporters (hENT3 and mENT3) located in intracellular membranes.	J Biol Chem	2005	15701636	IT it is not clear whether H+ is co-transported or not. since influx has low affinity it is more likely that transporter serve to remove nucleosides from lysosome rather then importing them (baldwin, 2005)
UROLACer	1	Winkelman J, Lehninger AL	Aldono- and uronolactonases of animal tissues.	J Biol Chem	1958	13587494	-found in the ER of rat livers; also found in several other animals (including monkey) [Winkelman, J Biol Chem 1958]
VALt5m	3	Porter RK.	Mammalian mitochondrial inner membrane cationic and neutral amino acid carriers.		2000	11004451	PMID 11004451: Non-respiring mitochondria swell when suspended in isotonic solutions of each of several different amino acids, indicating that these compounds can penetrate the mitochondrial inner membrane [5 and 6]. Gamble and Lehninger [12] showed that rat liver mitochondria swell in glycine, alanine, proline, valine and citrulline, but rat heart mitochondria do not swell in citrulline. Halling et al. [13] also showed that a variety of neutral amino acids, including non-metabolisable amino acids, could penetrate the mitochondrial inner membrane; Further advances were made when Cybulski and Fisher [15] found that mitochondrial swelling in neutral amino acids showed L-stereoisomer specificity
VITD3Hm	3	Guo YD, Strugnell S, Back DW, Jones G.	Transfected human liver cytochrome P-450 hydroxylates vitamin D analogs at different side-chain positions.	Proc Natl Acad Sci U S A	1993	7690968	This reaction takes place only in liver moeglich dass gleiche reaktion auch in microsome stattfindet based on Vitamins, G.F.M. Ball,2004, Blackwell publishing, 1st ed (book) pg.194 IT plasma conc: 10-40 ng/ml blood most of 25hvitd is in blood since tissue uptake is small
VLCSp	3	Mihalik SJ, Steinberg SJ, Pei Z, Park J, Kim DG, Heinzer AK, Dacremont G, Wanders JAR, Cuebas DA, Smith KD, Watkins PA	Participation of two members of the veryl long-chain acyl-CoA synthetase family in bile acid synthesis and recycling	Journal of Biological Chemistry	2002		peroxisome + ER: uniprot + literature references integral membrane protein specificity: Expressed in liver, kidney, placenta and pancreas NJ
VLCSp	3	Kelley M, Vessey DA	Dual role of diavlent cations in the bile acid: CoA ligase catalyzed reaction	Biochimica et Biophysica Acta	1994		peroxisome + ER: uniprot + literature references integral membrane protein specificity: Expressed in liver, kidney, placenta and pancreas NJ
VLCSp	3	Steinberg SJ, Mihalik SJ, Kim DG, Cuebas DA, Watkins PA	The human liver-specific homolog of very long-chain acyl-CoA synthetase is cholate: CoA ligase	Journal of Biological Chemistry	2000		peroxisome + ER: uniprot + literature references integral membrane protein specificity: Expressed in liver, kidney, placenta and pancreas NJ

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VLCSp	3	Mihalik SJ, Steinberg SJ, Pei Z, Park J, Kim DG, Heinzer AK, Dacremont G, Wanders JAR, Cuebas DA, Smith KD, Watkins PA	Participation of two members of the veryl long-chain acyl-CoA synthetase family in bile acid synthesis and recycling	Journal of Biological Chemistry	2002		peroxisome + ER: uniprot + literature references integral membrane protein specificity: Expressed in liver, kidney, placenta and pancreas NJ
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XANDp	3	Saksela M, Raivio KO.	Cloning and expression in vitro of human xanthine dehydrogenase/oxidase.	Biochem J	1996	8670112	GeneCards: peroxisome homodimer. dehydrogenase form can be reversibly converted oxidase through oxidation of sulhydryl group needs molybdenum as cofactor IT
XANDp	3	Linder N, Martelin E, Lapatto R, Raivio KO.	Posttranslational inactivation of human xanthine oxidoreductase by oxygen under standard cell culture conditions.	Am J Physiol Cell Physiol	2003	12637268	GeneCards: peroxisome homodimer. dehydrogenase form can be reversibly converted oxidase through oxidation of sulhydryl group needs molybdenum as cofactor IT
XYLt	3	Kayano T, Fukumoto H, Eddy RL, Fan YS, Byers MG, Shows TB, Bell GI	Evidence for a family of human glucose transporter-like proteins. Sequence and gene localization of a protein expressed in fetal skeletal muscle and other tissues	J Biol Chem	1988	3170580	6515: - major substrates are Glc (high affinity), Gal, Man, Xyl, maltose, dehydroascorbic acid [Uldry, Pflugers Arch 2004] - protein localized to brain (neurons), testis (spermatozoa) [Haber, Endocrinology 1993], [Uldry, Pflugers Arch 2004], sk muscle (slow twitch fibers) [Stuart, Metabolism 1999], platelets (alpha-granules) [Heijen, J Cell Biol 1997] - cDNA was cloned [Kayano, J Biol Chem 1988]
XYLt	3	Haber RS, Weinstein SP, O'Boyle E, Morgello S	Tissue distribution of the human GLUT3 glucose transporte	Endocrinology	1993	8504756	6515: - major substrates are Glc (high affinity), Gal, Man, Xyl, maltose, dehydroascorbic acid [Uldry, Pflugers Arch 2004] - protein localized to brain (neurons), testis (spermatozoa) [Haber, Endocrinology 1993], [Uldry, Pflugers Arch 2004], sk muscle (slow twitch fibers) [Stuart, Metabolism 1999], platelets (alpha-granules) [Heijen, J Cell Biol 1997] - cDNA was cloned [Kayano, J Biol Chem 1988]
XYLt	3	Heijnen HF, Oorschot V, Sixma JJ, Slot JW, James DE	Thrombin stimulates glucose transport in human platelets via the translocation of the glucose transporter GLUT-3 from alpha-granules to the cell surface	J Cell Biol	1997	9230074	6515: - major substrates are Glc (high affinity), Gal, Man, Xyl, maltose, dehydroascorbic acid [Uldry, Pflugers Arch 2004] - protein localized to brain (neurons), testis (spermatozoa) [Haber, Endocrinology 1993], [Uldry, Pflugers Arch 2004], sk muscle (slow twitch fibers) [Stuart, Metabolism 1999], platelets (alpha-granules) [Heijen, J Cell Biol 1997] - cDNA was cloned [Kayano, J Biol Chem 1988]
XYLt	3	Stuart CA, Wen G, Jiang J	GLUT3 protein and mRNA in autopsy muscle specimens	Metabolism	1999	10421229	6515: - major substrates are Glc (high affinity), Gal, Man, Xyl, maltose, dehydroascorbic acid [Uldry, Pflugers Arch 2004] - protein localized to brain (neurons), testis (spermatozoa) [Haber, Endocrinology 1993], [Uldry, Pflugers Arch 2004], sk muscle (slow twitch fibers) [Stuart, Metabolism 1999], platelets (alpha-granules) [Heijen, J Cell Biol 1997] - cDNA was cloned [Kayano, J Biol Chem 1988]

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XYLt	3	Uldry M, Thorens B	The SLC2 family of facilitated hexose and polyol transporters	Pflugers Arch	2004	12750891	6515: - major substrates are Glc (high affinity), Gal, Man, Xyl, maltose, dehydroascorbic acid [Uldry, Pflugers Arch 2004] - protein localized to brain (neurons), testis (spermatozoa) [Haber, Endocrinology 1993], [Uldry, Pflugers Arch 2004], sk muscle (slow twitch fibers) [Stuart, Metabolism 1999], platelets (alpha-granules) [Heijen, J Cell Biol 1997] - cDNA was cloned [Kayano, J Biol Chem 1988]
XYLTer	3	Kearns AE, Vertel BM, Schwartz NB	Topography of glycosylation and UDP-xylose production	J Biol Chem	1993	8496172	- immunocytochemistry, subcellular fractionation, and electron microscopy have demonstrated that xylosylation begins in the ER and continues in the early Golgi [Kearns, J Biol Chem 1993] - ER localization [Silbert, IUBMB LIfe 2002] 64131: - catalyzes the transfer of UDP-xylose to serine residues within XT recognition sequences of target proteins [OMIM] - protein was isolated, partial aa seq identified [Kuhn, J Biol Chem 2001] - cDNA was cloned and expressed, 94% identity to rat homolog [Gotting, J Mol Biol 2000] - ubiquitously expressed [Gotting, J Mol Biol 2000] 64132: - transfers xylose from UDP-xylose to specific serine residues of the core protein [RefSeq] - cDNA was isolated [Gotting, J Mol Biol 2000] - ubiquitously expressed [Gotting, J Mol Biol 2000] ** May need to also add a Golgi reaction since UDPXyl is now produced in Golgi instead of ER ***
XYLTer	3	Kuhn J, Gotting C, Schnolzer M, Kempf T, Brinkmann T, Kleesiek K	First isolation of human UDP-D-xylose: proteoglycan core protein beta-D-xylosyltransferase secreted from cultured JAR choriocarcinoma cells	J Biol Chem	2001	11087729	- immunocytochemistry, subcellular fractionation, and electron microscopy have demonstrated that xylosylation begins in the ER and continues in the early Golgi [Kearns, J Biol Chem 1993] - ER localization [Silbert, IUBMB LIfe 2002] 64131: - catalyzes the transfer of UDP-xylose to serine residues within XT recognition sequences of target proteins [OMIM] - protein was isolated, partial aa seq identified [Kuhn, J Biol Chem 2001] - cDNA was cloned and expressed, 94% identity to rat homolog [Gotting, J Mol Biol 2000] - ubiquitously expressed [Gotting, J Mol Biol 2000] 64132: - transfers xylose from UDP-xylose to specific serine residues of the core protein [RefSeq] - cDNA was isolated [Gotting, J Mol Biol 2000] - ubiquitously expressed [Gotting, J Mol Biol 2000] ** May need to also add a Golgi reaction since UDPXyl is now produced in Golgi instead of ER ***
XYLTer	3	Gotting C, Kuhn J, Zahn R, Brinkmann T, Kleesiek K	Molecular cloning and expression of human UDP-d-Xylose:proteoglycan core protein beta-d-xylosyltransferase and its first isoform XT-II	J Mol Biol	2000	11099377	- immunocytochemistry, subcellular fractionation, and electron microscopy have demonstrated that xylosylation begins in the ER and continues in the early Golgi [Kearns, J Biol Chem 1993] - ER localization [Silbert, IUBMB LIfe 2002] 64131: - catalyzes the transfer of UDP-xylose to serine residues within XT recognition sequences of target proteins [OMIM] - protein was isolated, partial aa seq identified [Kuhn, J Biol Chem 2001] - cDNA was cloned and expressed, 94% identity to rat homolog [Gotting, J Mol Biol 2000] - ubiquitously expressed [Gotting, J Mol Biol 2000] 64132: - transfers xylose from UDP-xylose to specific serine residues of the core protein [RefSeq] - cDNA was isolated [Gotting, J Mol Biol 2000] - ubiquitously expressed [Gotting, J Mol Biol 2000] ** May need to also add a Golgi reaction since UDPXyl is now produced in Golgi instead of ER ***

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XYLTer	3	Silbert JE, Sugumaran G	Biosynthesis of chondroitin/dermatan sulfate	IUBMB Life	2002	12512856	- immunocytochemistry, subcellular fractionation, and electron microscopy have demonstrated that xylosylation begins in the ER and continues in the early Golgi [Kearns, J Biol Chem 1993] - ER localization [Silbert, IUBMB LIfe 2002] 64131: - catalyzes the transfer of UDP-xylose to serine residues within XT recognition sequences of target proteins [OMIM] - protein was isolated, partial aa seq identified [Kuhn, J Biol Chem 2001] - cDNA was cloned and expressed, 94% identity to rat homolog [Gotting, J Mol Biol 2000] - ubiquitously expressed [Gotting, J Mol Biol 2000] 64132: - transfers xylose from UDP-xylose to specific serine residues of the core protein [RefSeq] - cDNA was isolated [Gotting, J Mol Biol 2000] - ubiquitously expressed [Gotting, J Mol Biol 2000] ** May need to also add a Golgi reaction since UDPXyl is now produced in Golgi instead of ER ***
XYLTt	1	Wang YM, van Eys J	Nutritional significance of fructose and sugar alcohols	Annu Rev Nutr	1981	6821187	- most likely absorbed by intestinal mucosa through passive or facilitated diffusion (see refs in [Wang 1981])
XYLUR	3	Nakagawa J, Ishikura S, Asami J, Isaji T, Usami N, Hara A, Sakurai T, Tsuritani K, Oda K, Takahashi M, Yoshimoto M, Otsuka N, Kitamura K	Molecular characterization of mammalian dicarbonyl/L-xylulose reductase and its localization in kidney	J Biol Chem	2002	11882650	- rxn described in Devlin p. 676, Orten p. 243, [Wang 1981] - pathway operates in adipose tissue [Devlin, Textbook of Biochem 2001] - highly expressed in kidney and liver [Nakagawa, J Biol Chem 2002]