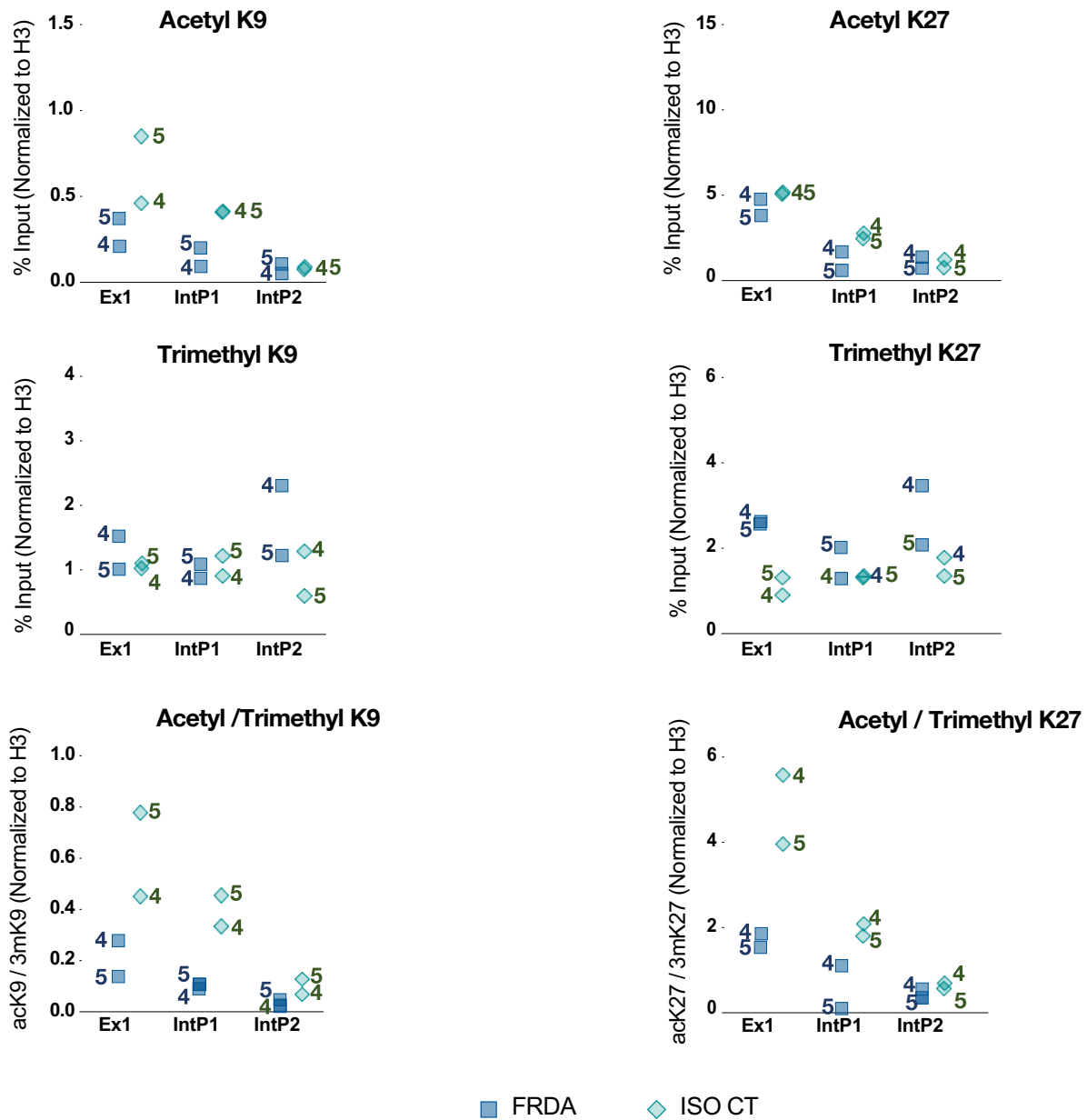
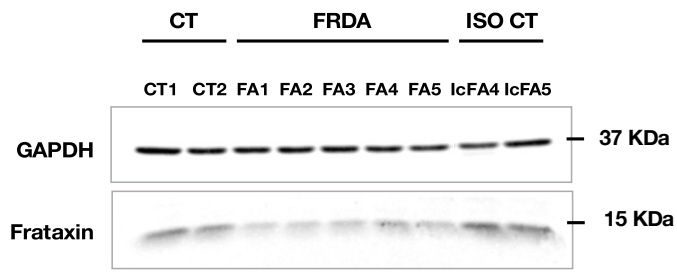


Supplementary Figure 1. Direct comparison of histone post-translational modifications in two FRDA and corresponding ISO CT fully differentiated sensory neuronal cultures

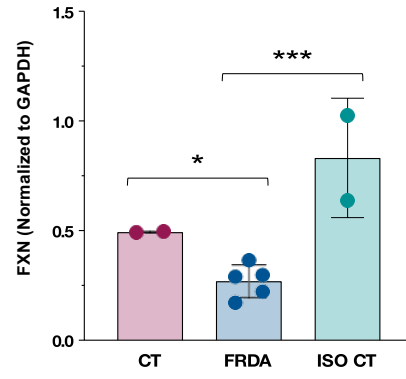


Supplementary Figure 2. Western Blot analysis of frataxin expression

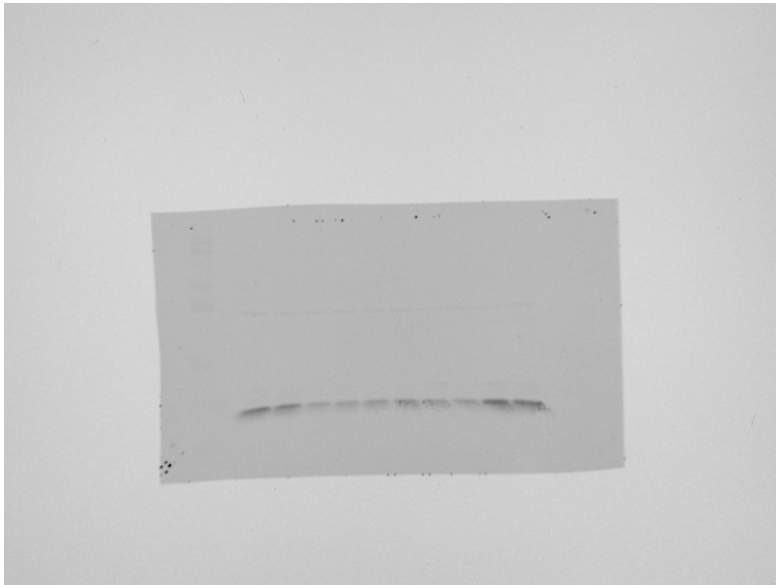
A



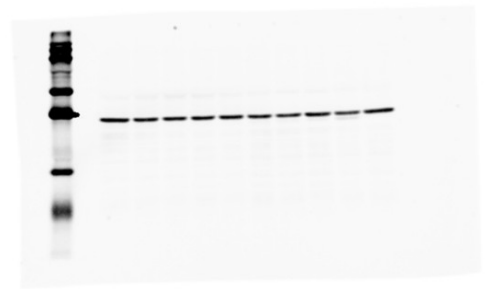
B



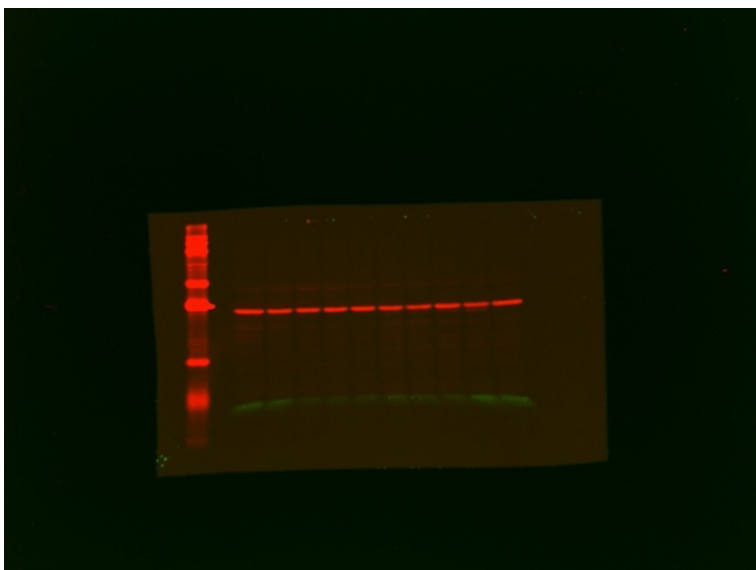
C



D



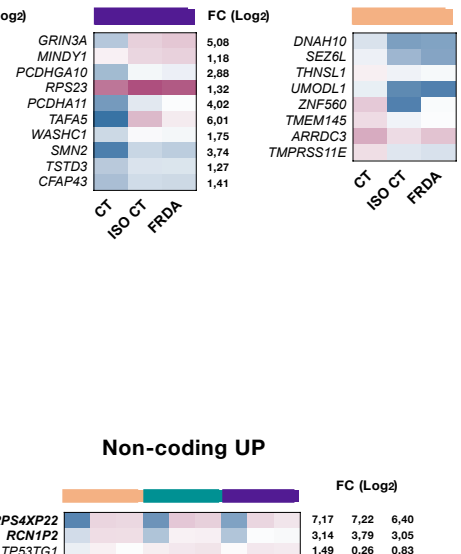
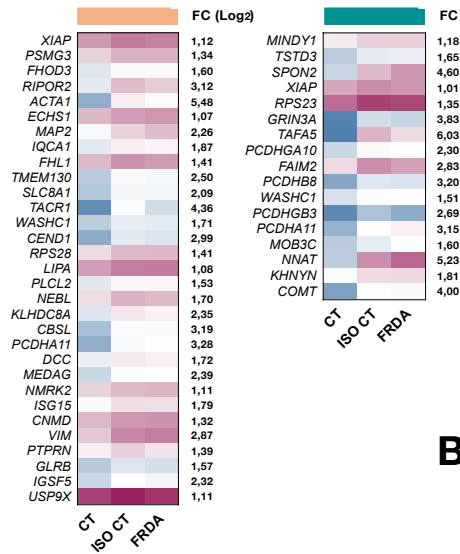
E



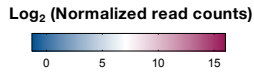
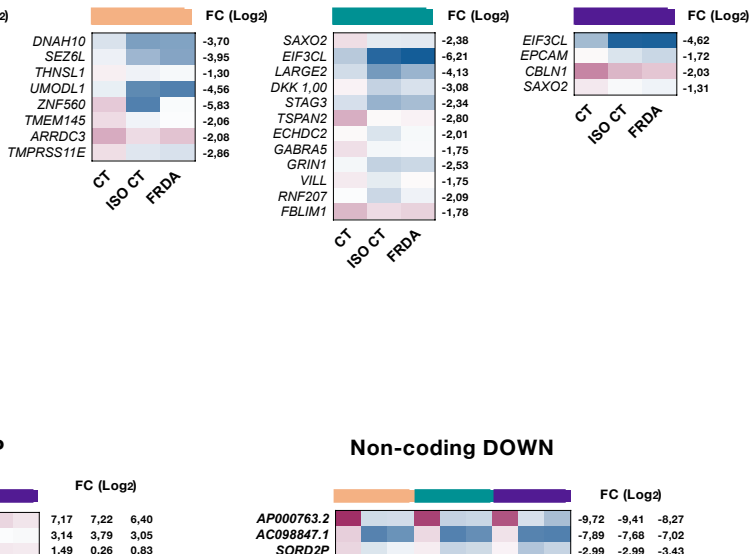
Supplementary Figure 3. Transcriptome profiling of differentiating isogenic control cultures

A

Upregulated ISO CT vs CT



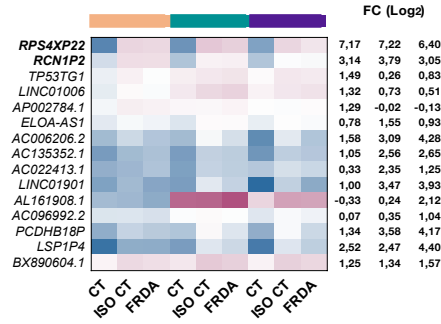
Downregulated ISO CT vs CT



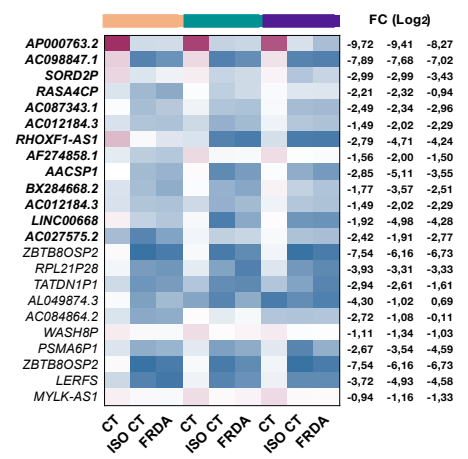
- iPSCs
- Developing neurons
- Mature Neurons

B

Non-coding UP

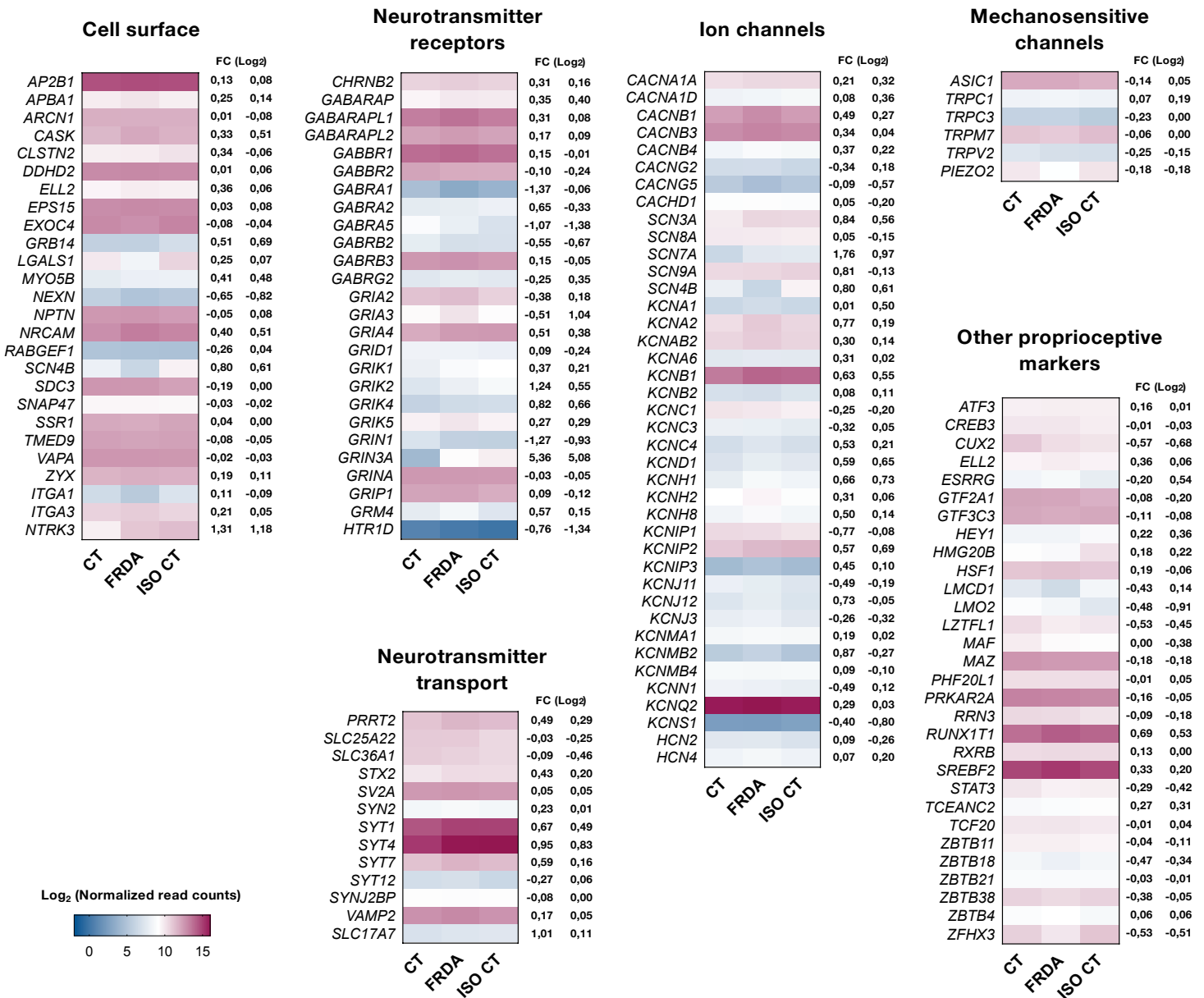


Non-coding DOWN

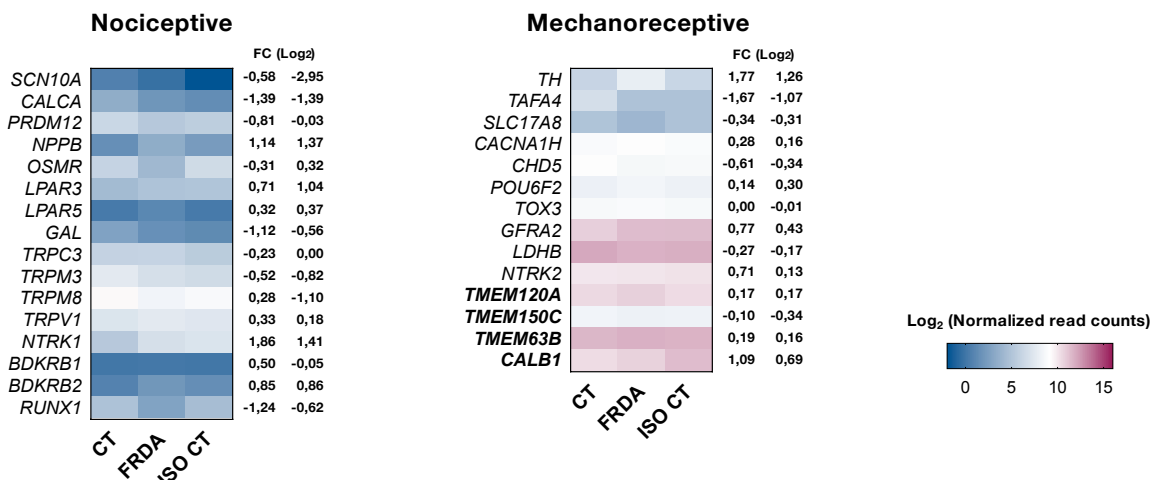


Supplementary Figure 4. Analysis of sensory neuronal markers in differentiated cultures

A Proprioceptive markers



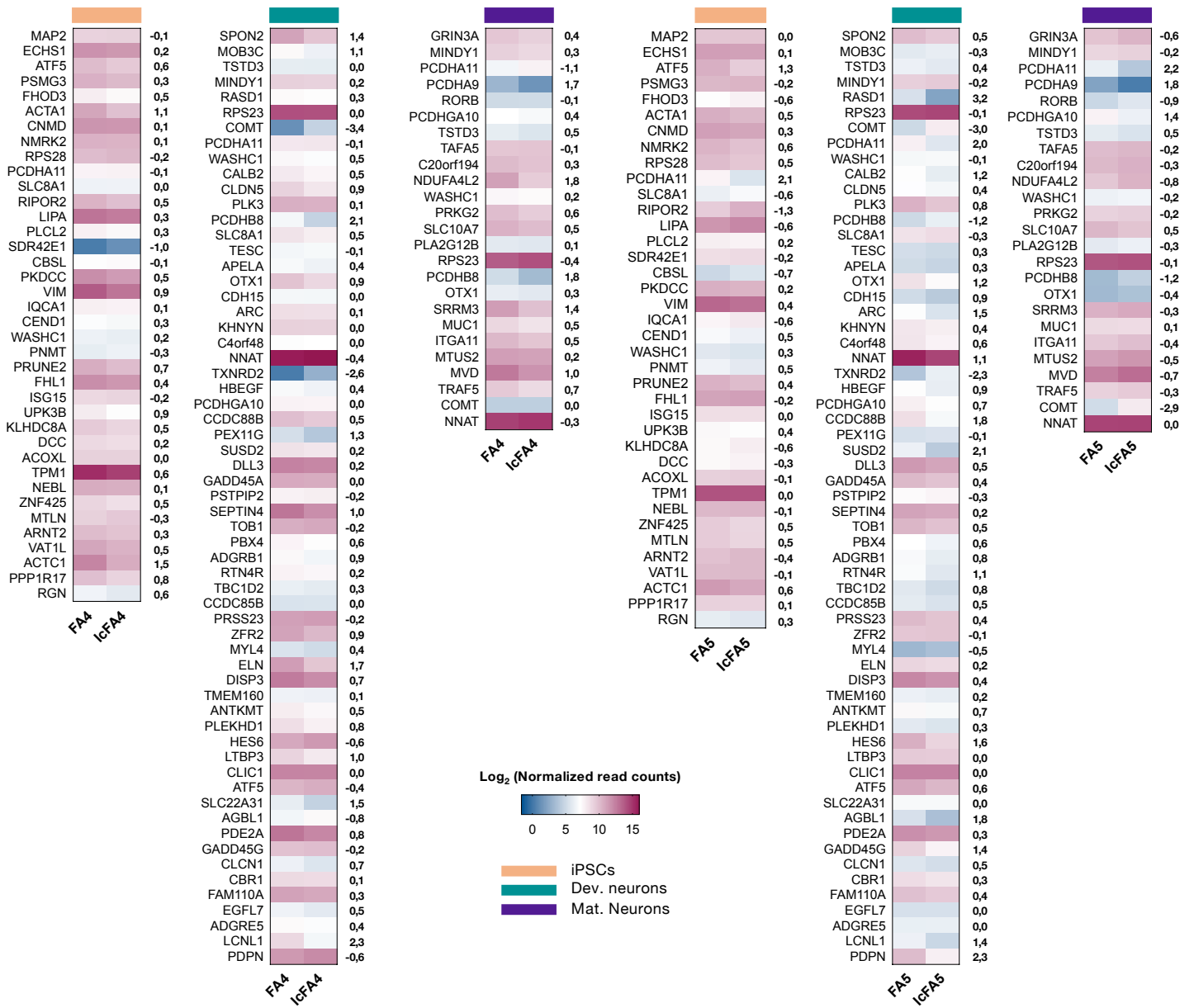
B Nociceptive and mechanoreceptive markers



Supplementary Figure 5. Transcriptome profile comparison of FA4 e FA5 with IcFA4 and IcFA5 for differentially expressed genes (DEGs).

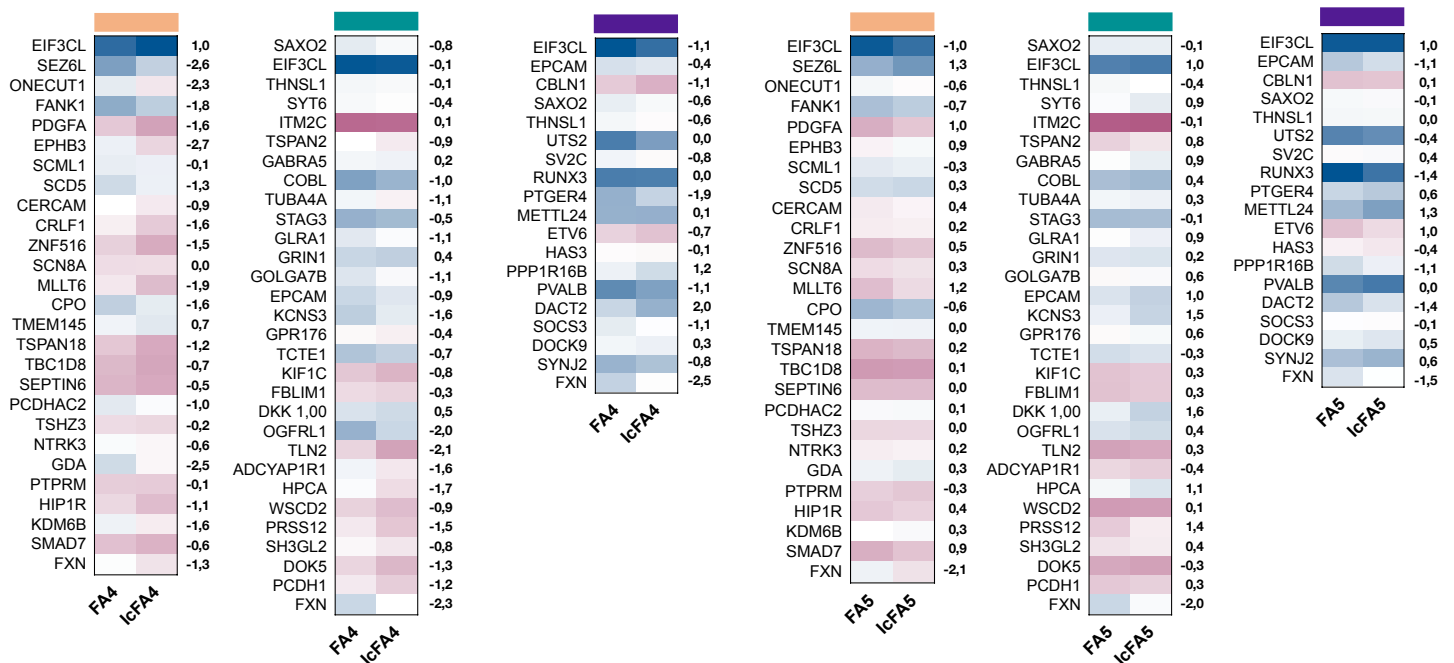
A

Upregulated genes from FRDA vs CT comparison

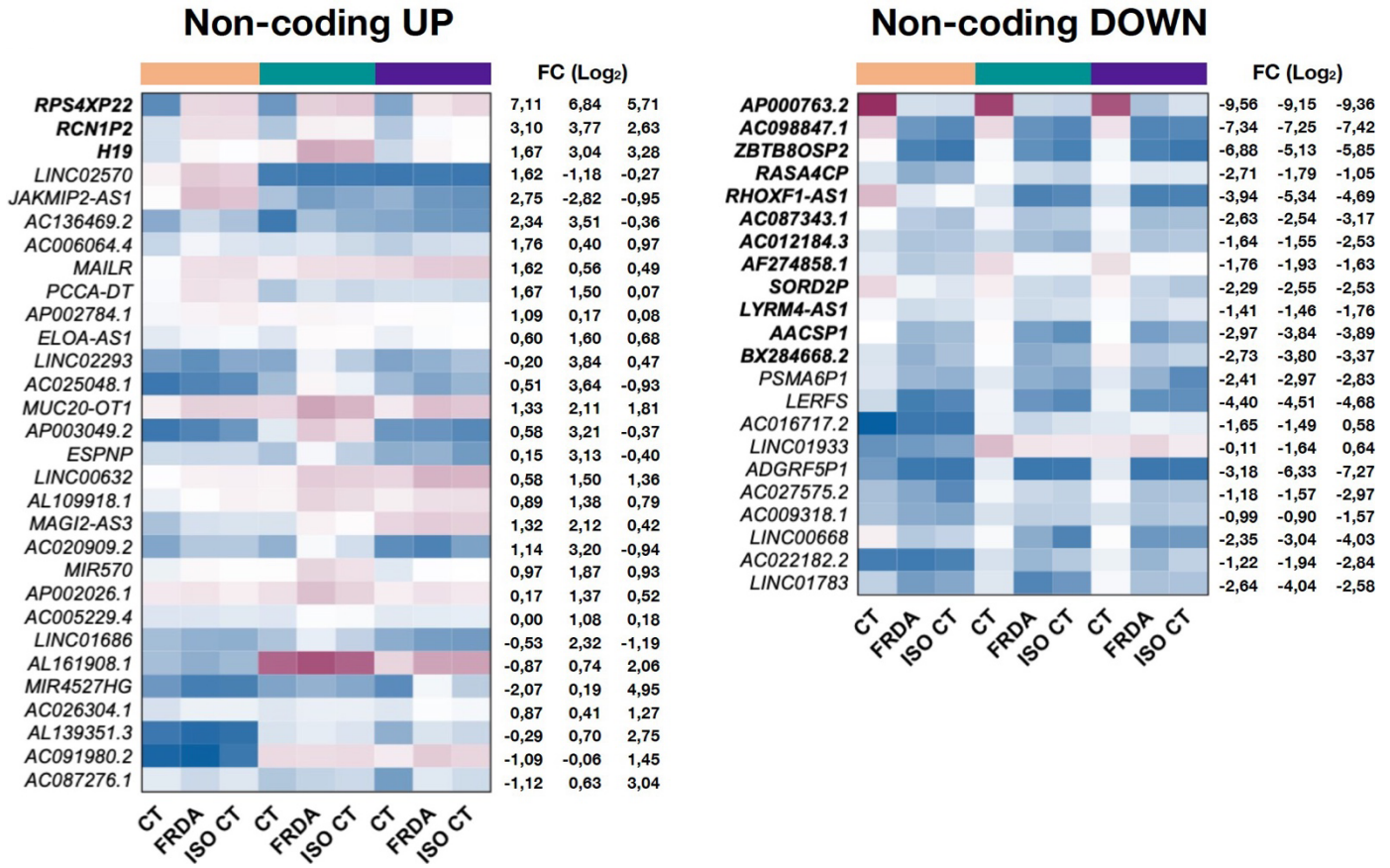


B

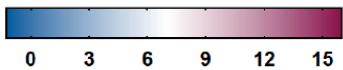
Downregulated genes from FRDA vs CT comparison



Supplementary Figure 6. Transcriptome profile comparison of FRDA, CT and ISO CT lines for non-coding RNAs

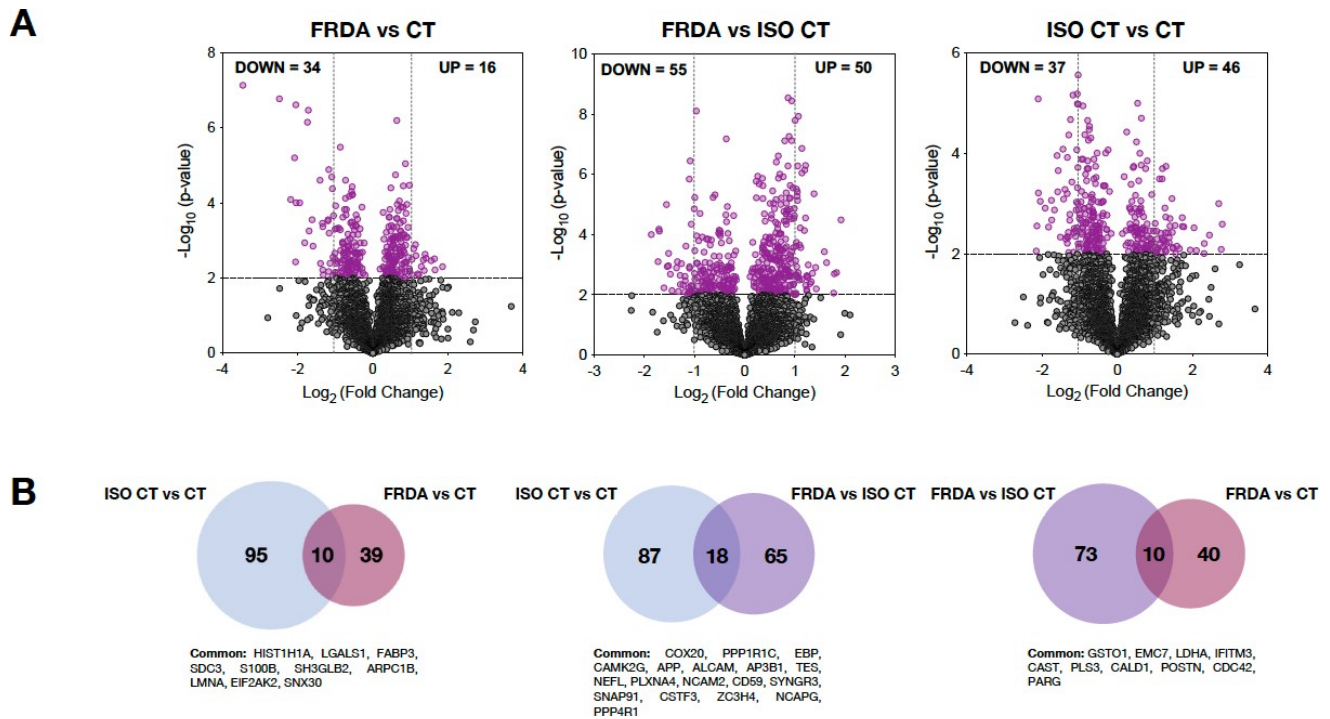


Normalized read counts (Log₂)



- iPSCs
- Developing neurons
- Mature Neurons

Supplementary Figure 7. Volcano plots and Venn diagrams of DEPs in FRDA, CT and ISO CT neurons.



Supplementary Figure 8. Proteome profile comparison of FA4 e FA5 with IcFA4 and IcFA5 for differentially expressed proteins (DEPs).

A



B



Supplementary Figure 1. Direct comparison of histone post-translational modifications in two FRDA and corresponding IOS CT fully differentiated neuronal cultures. Investigation of Histone H3 post-translational modifications of *FXN* 5'-end at exon 1 (Ex1), intronic region (intron 1) upstream (IntP1, IntP2) of GAA expansion as reported in Figure 1. This is an excerpt from Figure 1 in which for clarity results are only shown for ISO CT IcFA4 and IcFA5 (green diamonds) and corresponding FRDA lines FA 4 and FA5 (blue squares), labeled as 4 and 5.

Supplementary Figure 2. Western Blot analysis of frataxin expression. Frataxin expression was evaluated by Western Blot in differentiated fully mature neurons in two independent biological replicates per line (n = 2; A: representative blot). Protein expression was normalized to GAPDH. Mean values obtained for each group (CT, FRDA and ISO CT) were compared (Mean \pm SD). Histograms (B) represent mean values of biological replicates for each group, and error bars represent the standard deviation of mean (SD). Each dot represents the mean of two independent biological replicates per line. A significant decrease in Frataxin expression was observed in FRDA lines compared with CT, along with a recovery of frataxin expression in ISO CT neurons. No significant differences were observed between CT and IC lines (two-way ANOVA with Bonferroni's test for multiple comparisons; * p < 0.05; *** p < 0.001). CT: Control; FRDA: Friedreich Ataxia; ISO CT: Isogenic Control. Original uncropped gel images are in C (Frataxin), D (GAPDH), E (Frataxin+GAPDH two-color image). Uncropped gels contain one extra sample (third lane from the right) corresponding to an FRDA line that was not further utilized in the analyses because of aberrant growth and altered karyotype.

Supplementary Figure 3. Transcriptome profiling of differentiating ISO CT cultures. (A) Heatmaps representing Differentially Expressed Coding Genes between ISO CT and CT ($|\text{Log}_2(\text{Fold Change})| > 1$; adjusted p-value < 0.1), for each stage of differentiation (yellow: iPSCs; green: developing neurons; violet: mature neurons). For each gene, the $\log_2(\text{Fold Change})$ between ISO CT and CT lines is shown on the right side of the corresponding heatmap. For each comparison, the transcriptional level of identified DEGs was represented also for the FRDA group. Colour scale represents $\log_2(\text{Normalized read counts})$ values. **(B)** Heatmaps representing differentially expressed

were analysed and represented as in (B). For each DEG, the $\log_2(\text{Fold Change})$ between ISO CT and CT lines at each stage of differentiation is shown on the right side of the corresponding heatmap (iPSCs: left; developing neurons: middle; mature neurons: right). Non-coding DEGs common to all stages of differentiation are highlighted in bold. (n=2 for CT; n=5 for FRDA; n=3 for ISO CT).

Supplementary Figure 4. Analysis of sensory neuronal markers in differentiated cultures.

Heatmaps representing sensory neuron-specific transcripts detected in differentiated cultures. (A) Proprioceptive-specific genes were schematically separated in categories based on their function or cellular localization. (B) Nociceptive and mechanoreceptive-specific genes expressed in differentiated cultures. Genes common to mechanoreceptive and proprioceptive neurons are highlighted in bold. For each gene, the $\log_2(\text{Fold Change})$ between FRDA and CT neurons (left) or ISO CT and CT neurons (right) is shown on the right side of the corresponding heatmap. Color scale represents $\log_2(\text{normalized read counts})$ of transcripts.

Supplementary Figure 5. Transcriptome profile comparison of FA4 e FA5 with IcFA4 and IcFA5 for differentially expressed genes (DEGs).

A direct comparison between FRDA and their sibling ISO CT lines was performed for all identified DEGs at the transcriptomic analysis. DEG expression was specifically investigated for upregulated (A) and downregulated (B) genes for lines FA4 and IcFA4 and FA5 and IcFA5, to evaluate the effects induced on gene expression by the removal of the GAA expansion mutation in genetically matched cell lines. For each gene, the ratio between FA4 and IcFA4 or FA5 and IcFA5 is represented on the right side of the corresponding heatmap, in \log_2 scale. Color scale represents $\log_2(\text{normalized read counts})$ of transcripts. (n=1 for all lines).

Supplementary Figure 6. Transcriptome profile comparison of FRDA, CT and ISO CT lines for non-coding RNAs.

Heatmaps representing differentially expressed non-coding genes or pseudogenes between FRDA and CT lines, at all stages of differentiation. For each DEG, the $\log_2(\text{Fold Change})$ between FRDA and CT lines at each stage of differentiation is shown on the right side of the corresponding heatmap (iPSCs: left; developing neurons: middle; mature neurons:

Supplementary Figure 7. Volcano plots and Venn diagrams of DEPs in FRDA, CT and ISO CT neurons. **(A)** Volcano plots of significantly differentially expressed proteins in the comparison between FRDA and CT neurons (left), FRDA and ISO CT neurons (middle) and ISO CT and CT neurons (right). Red dots represent proteins exhibiting significant fold changes (p -value < 0.01). Of those, only proteins with absolute $\text{Log}_2(\text{Fold Change}) > 1$ were selected for analysis (left and right sides of vertical broken lines). **(B)** Venn Diagrams of identified differentially expressed proteins in the comparison of FRDA and ISO CT neurons versus CT (left), FRDA and CT neurons versus ISO CT (middle) or CT and ISO CT neurons versus FRDA (right). Common identified proteins are indicated under the corresponding diagram.

Supplementary Figure 8. Proteome profile comparison of FA4 e FA5 with IcFA4 and IcFA5 for differentially expressed proteins (DEPs). A direct comparison between FRDA and their sibling ISO CT lines was performed for all identified DEPs at the proteomic analysis. DEP expression was specifically investigated for lines FA4 and IcFA4 (A) and FA5 and IcFA5 (B), to evaluate the effects induced on protein expression by the removal of the GAA expansion mutation in genetically matched cell lines. Color scale represents $\text{log}_2(\text{Mean Peak Intensity})$ values of identified protein spectra. Exact values of Mean Peak Intensities for each protein are indicated in the heatmaps. ($n=1$ for all lines).

Supplementary Table 1. List of primers used for Chromatin Immunoprecipitation

RT-qPCR Primers	Sequence (5' – 3')
Ex1 Forward	GGAGCAGCATGTGGACTCTC
Ex1 Reverse	CGGCGCGGATACTTACTG
IntP1 Forward	CTCCCGTTGCATTTACT
IntP1 Reverse	GTGACAAGCATGGAGACAGC
IntP2 Forward	CTGACCCGACCTTTATTCCA
IntP2 Reverse	TGGGCGTCACCTTTATCTTC
IntP3 Forward	GAAACCCAAAGAATGGCTGTG
IntP3 Reverse	TTCCCTCCTCGTGAAACACC
IntP4 Forward	CTGGAAAAATAGGCAAGTGTGG
IntP4 Reverse	CAGGGGTGGAAGCCCAATAC
IntP5 Forward	CCCTTGACATCTTGGGTAT
IntP5 Reverse	GAGAAAAGGGTGGGGAAGAG

Supplementary Table 2. List of identified differentially expressed coding and non-coding genes for iPSCs, developing neurons and mature neurons.

DIFFERENTIALLY EXPRESSED GENES – iPSCs																
UPREGULATED GENES																
Normalized read counts									GENE	Description	Chr	GO: Biological process	Log ₂ FC FRDA/CT	FDR	Log ₂ FC ISO CT/CT	FDR
iPSCs			Developing Neurons			Mature Neurons										
CT	FRDA	ISO CT	CT	FRDA	ISO CT	CT	FRDA	ISO CT								
104	1068	554	48000	56547	77763	43223	52417	54091	MAP2	microtubule associated protein 2	2	microtubule bundle formation; central nervous system neuron development; microtubule cytoskeleton organization; neuron projection development; regulation of cellular protein localization; regulation of microtubule polymerization; dendrite development; negative regulation of axon extension; negative regulation of microtubule polymerization; dendrite morphogenesis; positive regulation of anterograde dense core granule transport; regulation of organelle transport along microtubule; positive regulation of anterograde synaptic vesicle transport; negative regulation of microtubule binding; negative regulation of microtubule motor activity; cellular response to organic substance; axonogenesis; establishment of cell polarity	3,16	1,8E-08	2,26	4,8E-03
1077	2726	2265	1583	2731	2455	1204	1847	1961	ECHS1	enoyl-CoA hydratase, short chain 1	10	lipid metabolic process; fatty acid metabolic process; fatty acid beta-oxidation	1,33	8,0E-07	1,07	3,9E-03
328	1210	574	646	1364	1120	568	741	640	ATF5	activating transcription factor 5	19	regulation of transcription, DNA-templated; regulation of transcription by RNA polymerase II; negative regulation of apoptotic process; positive regulation of transcription, DNA-templated; fat cell differentiation; negative regulation of cell population proliferation; positive regulation of transcription by RNA polymerase II; negative regulation of transcription, DNA-templated; regulation of centrosome cycle; negative regulation of cell cycle G2/M phase transition; cerebellar granule cell precursor proliferation; circadian rhythm; post-embryonic development; regulation of gene expression; olfactory bulb interneuron differentiation; olfactory bulb interneuron development; olfactory lobe development; multicellular organism growth	1,86	3,1E-06	0,79	6,6E-01
521	1375	1331	397	795	721	394	754	690	PSMG3	proteasome assembly chaperone 3	7	chaperone-mediated protein complex assembly	1,39	8,8E-06	1,34	4,3E-04
51	163	157	821	743	782	777	765	756	FHOD3	formin homology 2 domain containing 3	18	actin filament organization; cortical actin cytoskeleton organization; cardiac myofibril assembly; sarcomere organization; actin filament network formation; negative regulation of actin filament polymerization	1,64	1,1E-05	1,60	4,3E-04
7	746	721	48	468	76	6	6	5	ACTA1	actin alpha 1, skeletal muscle	1	positive regulation of gene expression; mesenchyme migration; muscle filament sliding; skeletal muscle thin filament assembly; muscle contraction; skeletal muscle fiber development; response to mechanical stimulus; response to extracellular stimulus; response to lithium ion; skeletal muscle fiber adaptation; response to steroid hormone; cellular response to organonitrogen compound	5,68	7,1E-05	5,48	1,6E-03
1063	2973	2627	9	2	7	5	3	4	CNMD	chondromodulin	13	multicellular organism development; cell differentiation; cartilage development; negative regulation of angiogenesis; skeletal system development; endothelial cell morphogenesis; negative regulation of endothelial cell proliferation; proteoglycan metabolic process; negative regulation of vascular endothelial growth factor receptor signaling pathway	1,52	9,3E-04	1,32	4,5E-02
496	1160	1062	5	4	3	0	3	3	NMRK2	nicotinamide riboside kinase 2	19	phosphorylation; NAD biosynthetic process; pyridine nucleotide biosynthetic process; NAD metabolic process; negative regulation of myoblast differentiation	1,25	1,2E-03	1,11	4,1E-02
397	1117	1068	1016	1458	1513	683	1189	1199	RPS28	ribosomal protein S28	19	translation; cytoplasmic translation; rRNA processing; ribosome biogenesis; ribosomal small subunit biogenesis; ribosomal small subunit assembly; viral transcription; translational initiation; maturation of SSU-rRNA; nuclear-transcribed mRNA catabolic process, nonsense-mediated decay; SRP-dependent cotranslational protein targeting to membrane	1,47	1,4E-03	1,41	2,0E-02
10	144	122	15	213	161	4	124	88	PCDHA11	protocadherin alpha 11	5	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion; nervous system development	3,53	1,7E-03	3,28	3,1E-02
20	90	92	105	408	253	515	1063	892	SLC8A1	solute carrier family 8 member A1	2	transmembrane transport; calcium ion transport; cell communication; regulation of cardiac muscle contraction by regulation of the release of sequestered calcium ion; negative regulation of protein serine/threonine kinase activity; cellular response to reactive oxygen species; regulation of heart rate; ion transport; sodium ion transport; calcium ion transmembrane transport; sodium ion transmembrane transport; calcium ion import across plasma membrane; positive regulation of bone mineralization; muscle contraction; regulation of cardiac conduction; regulation of cell communication by electrical coupling; cardiac muscle contraction; cellular response to caffeine; calcium ion import; response to muscle stretch; calcium ion homeostasis; cellular calcium ion homeostasis; membrane depolarization during cardiac muscle cell action potential; cellular sodium ion homeostasis; sodium ion export across plasma membrane; relaxation of cardiac muscle; cell communication by electrical coupling involved in cardiac conduction; regulation of cardiac muscle contraction by calcium ion signaling; regulation of gene expression; regulation of the force of heart contraction; sodium ion import across plasma membrane; positive regulation of the force of heart contraction; vascular associated smooth muscle contraction; metal ion transport; relaxation of smooth muscle; negative regulation of cytosolic calcium ion concentration; cardiac muscle cell development; cytosolic calcium ion transport; calcium ion transport into cytosol; regulation of postsynaptic cytosolic calcium ion concentration; response to hypoxia; regulation of sodium ion transport; positive regulation of cytosolic calcium ion concentration; response to nutrient; response to glucose; positive regulation of fibroblast migration; telencephalon development; response to ATP; response to immobilization stress; response to drug; response to hydrogen peroxide; regulation of calcium ion transport; cellular response to cAMP; cellular response to hypoxia; calcium ion export	2,04	2,0E-03	2,09	9,8E-03
107	696	1051	1689	1424	1415	600	1188	1057	RIPOR2	RHO family interacting cell polarization regulator 2	6	sensory perception of sound; skeletal muscle fiber development; cell adhesion; multicellular organism development; cell differentiation; chemotaxis; negative regulation of cell adhesion; muscle organ development; negative regulation of signal transduction; negative regulation of Rho protein signal transduction; establishment of protein localization; protein homooligomerization; positive regulation of filopodium assembly; auditory receptor cell stereocilium organization; cellular response to	2,54	2,0E-03	3,12	4,3E-04

389	1083	1313	7873	8484	8831	6143	5941	6777	NEBL	nebulette	10	muscle fiber development; cardiac muscle thin filament assembly	1,41	3,9E-02	1,70	2,9E-02
228	466	381	617	719	887	552	674	699	ZNF425	zinc finger protein 425	7	regulation of transcription, DNA-templated; regulation of transcription by RNA polymerase II; negative regulation of transcription by RNA polymerase II; negative regulation of transcription, DNA-templated	1,02	4,6E-02	0,73	6,6E-01
338	726	582	182	233	202	222	228	203	MTLN	mitoregulin	2	positive regulation of protein-containing complex assembly; positive regulation of mitochondrial membrane potential; positive regulation of sequestering of calcium ion	1,10	4,7E-02	0,78	6,8E-01
337	789	831	3001	3313	3472	5298	5382	5450	ARNT2	aryl hydrocarbon receptor nuclear translocator 2	15	regulation of transcription, DNA-templated; positive regulation of transcription, DNA-templated; positive regulation of transcription by RNA polymerase II; response to hypoxia; xenobiotic metabolic process; brain development; in utero embryonic development; central nervous system development; positive regulation of cell population proliferation; response to estradiol; negative regulation of apoptotic process	1,19	6,2E-02	1,27	1,3E-01
246	1400	1158	336	268	332	4458	5744	3670	VAT1L	vesicle amine transport 1 like	16	oxidation-reduction process	2,33	6,2E-02	2,06	3,9E-01
206	1969	1292	66	500	88	15	13	12	ACTC1	actin alpha cardiac muscle 1	15	actin filament organization; positive regulation of gene expression; mesenchyme migration; muscle filament sliding; actin filament-based movement; heart contraction; cardiac myofibril assembly; cardiac muscle tissue morphogenesis; actomyosin structure organization; skeletal muscle thin filament assembly; actin-myosin filament sliding; response to drug; negative regulation of apoptotic process; response to ethanol; cardiac muscle contraction; actin-mediated cell contraction	3,13	6,2E-02	2,45	5,7E-01
72	523	462	937	1930	1552	2181	2080	2477	PPP1R17	protein phosphatase 1 regulatory subunit 17	7	regulation of phosphatase activity; negative regulation of phosphoprotein phosphatase activity; negative regulation of catalytic activity; intracellular signal transduction; central nervous system development	2,50	7,4E-02	2,35	3,2E-01
9	45	47	14	27	26	8	10	15	RGN	regucalcin	X	positive regulation of ATPase activity; cellular calcium ion homeostasis; regulation of calcium-mediated signaling; L-ascorbic acid biosynthetic process; kidney development; liver development; negative regulation of protein phosphorylation; negative regulation of protein kinase activity; spermatogenesis; aging; positive regulation of triglyceride biosynthetic process; positive regulation of glucose metabolic process; positive regulation of phosphatase activity; negative regulation of phosphoprotein phosphatase activity; negative regulation of GTPase activity; negative regulation of apoptotic process; positive regulation of GTPase activity; negative regulation of nitric oxide biosynthetic process; positive regulation of fatty acid biosynthetic process; negative regulation of epithelial cell proliferation; negative regulation of cyclic-nucleotide phosphodiesterase activity; liver regeneration; negative regulation of flagellated sperm motility; positive regulation of superoxide dismutase activity; positive regulation of ATPase-coupled calcium transmembrane transporter activity; negative regulation of RNA biosynthetic process; negative regulation of bone development; positive regulation of proteolysis involved in cellular protein catabolic process; negative regulation of calcium-dependent ATPase activity; negative regulation of DNA catabolic process; positive regulation of dUTP diphosphatase activity; negative regulation of leucine-tRNA ligase activity; negative regulation of DNA biosynthetic process	2,22	7,4E-02	2,24	2,2E-01
39	367	387	14	231	238	15	111	150	RCN1P2	reticulocalbin 1 pseudogene 2	13		3,10	4,9E-04	3,14	3,5E-03
222	653	520	1	0	0	0	0	0	LINC02570	long intergenic non-protein coding RNA 2570	6		1,62	1,2E-02	1,25	3,5E-01
157	1035	884	30	4	5	7	4	2	JAKMIP2-AS1	JAKMIP2 antisense RNA 1	5		2,75	2,0E-02	2,44	2,1E-01
5	28	13	1	12	7	5	4	4	AC136469.2	novel transcript	19		2,34	3,2E-02	1,23	9,3E-01
26	91	49	39	51	31	19	37	40	AC006064.4	novel transcript, antisense to GAPDH	12		1,76	4,3E-02	0,87	1,0E+00
119	382	368	229	343	353	440	616	609	MAILR	macrophage interferon regulatory lncRNA	8		1,62	5,3E-02	1,57	2,4E-01
102	327	276	14	43	30	40	42	45	PCCA-DT	PCCA divergent transcript	13		1,67	7,6E-02	1,41	4,9E-01
91	196	226	165	186	163	133	141	122	AP002784.1	novel transcript	11		1,09	8,8E-02	1,29	7,9E-02

DOWNREGULATED GENES

Normalized read counts									GENE	Description	Chr	GO: Biological process	Log ₂ FC FRDA/CT	FDR	Log ₂ FC ISO CT/CT	FDR
iPSCs			Developing Neurons			Mature Neurons										
CT	FRDA	ISO CT	CT	FRDA	ISO CT	CT	FRDA	ISO CT								
19	0	0	18	0	0	11	0	0	EIF3CL	eukaryotic translation initiation factor 3 subunit C like	16	translational initiation; formation of cytoplasmic translation initiation complex; cytoplasmic translational initiation; translation	-5,08	1,0E-10	-5,10	1,8E-07
224	5	12	339	602	492	1445	2987	2702	SEZ6L	seizure related 6 homolog like	22	adult locomotory behavior; cerebellar Purkinje cell layer development; synapse maturation; regulation of protein kinase C signaling	-5,24	7,0E-10	-3,95	4,3E-04
609	93	234	1481	1516	1841	1778	1618	1845	ONECUT1	one cut homeobox 1	15	positive regulation of transcription by RNA polymerase II; cell differentiation; regulation of transcription, DNA-templated; liver development; regulation of cell-matrix adhesion; epithelial cell development; glucose metabolic process; regulation of transcription by RNA polymerase II; Notch signaling pathway; endoderm development; anatomical structure morphogenesis; B cell differentiation; positive regulation of cell migration; negative regulation of transforming growth factor	-2,68	1,8E-04	-1,34	6,3E-01

											chemotaxis; positive regulation of phospholipase C activity; positive regulation of neurotrophin TRK receptor signaling pathway; neuron migration; circadian rhythm; modulation by virus of host transcription; neuronal action potential propagation; myelination in peripheral nervous system; mechanoreceptor differentiation; positive regulation of apoptotic process; response to ethanol; neuron fate specification; response to axon injury; positive regulation of axon extension involved in regeneration; negative regulation of astrocyte differentiation; response to corticosterone; positive regulation of synapse assembly; negative regulation of cell death; lens fiber cell differentiation; cellular response to retinoic acid; cochlea development; regulation of postsynaptic density assembly; regulation of presynapse assembly					
488	30	223	17	4	9	23	5	15	GDA	guanine deaminase	9	guanine catabolic process; nucleobase-containing compound metabolic process; nervous system development; purine nucleotide catabolic process; guanine metabolic process	-3,45	5,3E-02	-0,87	1,0E+00
1535	635	741	340	546	623	5083	2919	3565	PTPRM	protein tyrosine phosphatase receptor type M	18	protein dephosphorylation; dephosphorylation; cell adhesion; signal transduction; peptidyl-tyrosine dephosphorylation; retina layer formation; negative regulation of angiogenesis; negative regulation of endothelial cell proliferation; negative regulation of endothelial cell migration; homophilic cell adhesion via plasma membrane adhesion molecules; neuron projection development; retinal ganglion cell axon guidance; response to drug; positive regulation of blood vessel diameter	-1,26	5,3E-02	-1,03	4,7E-01
291	115	232	7511	7915	6439	3463	4147	3864	KDM6B	lysine demethylase 6B	17	histone H3-K27 demethylation; positive regulation of transcription by RNA polymerase II; mesodermal cell differentiation; oxidation-reduction process; chromatin organization; inflammatory response; positive regulation of cold-induced thermogenesis; inflammatory response to antigenic stimulus; chromatin remodeling; regulation of gene expression; response to activity; histone demethylation; hippocampus development; cell fate commitment; endothelial cell differentiation; cardiac muscle cell differentiation; response to fungicide; cellular response to hydrogen peroxide	-1,36	7,3E-02	-0,34	1,0E+00
2244	985	876	218	263	243	512	265	358	SMAD7	SMAD family member 7	18	regulation of transcription, DNA-templated; transforming growth factor beta receptor signaling pathway; negative regulation of protein ubiquitination; protein stabilization; negative regulation of cell migration; negative regulation of BMP signaling pathway; negative regulation of DNA-binding transcription factor activity; regulation of ventricular cardiac muscle cell membrane depolarization; ventricular septum morphogenesis; negative regulation of epithelial to mesenchymal transition; negative regulation of transforming growth factor beta receptor signaling pathway; negative regulation of pathway-restricted SMAD protein phosphorylation; positive regulation of transcription by RNA polymerase II; cell differentiation; anatomical structure morphogenesis; protein deubiquitination; BMP signaling pathway; SMAD protein signal transduction; negative regulation of transcription by RNA polymerase II; positive regulation of proteasomal ubiquitin-dependent protein catabolic process; artery morphogenesis; negative regulation of peptidyl-threonine phosphorylation; ureteric bud development; negative regulation of T cell cytokine production; regulation of epithelial to mesenchymal transition; negative regulation of transcription by competitive promoter binding; regulation of transforming growth factor beta receptor signaling pathway; positive regulation of cell-cell adhesion; negative regulation of ossification; positive regulation of protein ubiquitination; regulation of activin receptor signaling pathway; negative regulation of peptidyl-serine phosphorylation; adherens junction assembly; response to laminar fluid shear stress; cellular protein-containing complex localization; negative regulation of ubiquitin-protein transferase activity; ventricular cardiac muscle tissue morphogenesis; regulation of cardiac muscle contraction; pathway-restricted SMAD protein phosphorylation; cellular response to transforming growth factor beta stimulus; negative regulation of chondrocyte proliferation; positive regulation of chondrocyte hypertrophy; cellular response to leukemia inhibitory factor; negative regulation of T-helper 17 type immune response; negative regulation of T-helper 17 cell differentiation	-1,15	8,8E-02	-1,33	1,1E-01
277	92	221	108	31	90	160	46	89	FXN	frataxin	9	iron-sulfur cluster assembly; oxidation-reduction process; positive regulation of cell population proliferation; negative regulation of apoptotic process; ion transport; cellular iron ion homeostasis; oxidative phosphorylation; heme biosynthetic process; mitochondrion organization; adult walking behavior; aerobic respiration; embryo development ending in birth or egg hatching; proprioception; negative regulation of multicellular organism growth; negative regulation of organ growth; iron ion homeostasis; positive regulation of cell growth; cellular response to hydrogen peroxide; negative regulation of release of cytochrome c from mitochondria; protein autophagy; positive regulation of catalytic activity; small molecule metabolic process; response to iron ion; regulation of ferrochelatase activity; iron incorporation into metallo-sulfur cluster; positive regulation of lyase activity; positive regulation of succinate dehydrogenase activity; positive regulation of aconitate hydratase activity	-1,51	2,4E-01	-0,29	1,0E+00
33154	40	35	21340	34	27	14136	20	41	AP000763.2	mitochondrially encoded cytochrome c oxidase I (MT-CO1) pseudogene	11		-9,56	5,9E-130	-9,72	2,8E-94
552	3	2	421	2	1	305	1	2	AC098847.1	ribosomal protein, large, P0 pseudogene	18		-7,34	3,6E-39	-7,89	2,2E-25
153	1	0	102	2	1	86	1	0	ZBTB80SP2	zinc finger and BTB domain containing 8 opposite strand pseudogene 2	2		-6,88	2,4E-10	-7,54	6,3E-09
46	7	9	114	32	22	109	52	56	RASA4CP	RAS p21 protein activator 4C, pseudogene	7		-2,71	2,3E-06	-2,21	3,9E-03
1051	53	127	61	1	1	40	1	1	RHOXF1-AS1	RHOXF1 antisense RNA 1	X		-3,94	1,9E-05	-2,79	6,0E-02
90	4	5	62	1	5	50	2	4	RPL21P28	ribosomal protein L21 pseudogene 28	1		-4,26	2,9E-05	-3,93	2,6E-03
598	284	413	689	652	808	1239	947	1120	NNT-AS1	NNT antisense RNA 1	5		-1,07	1,2E-04	-0,53	6,3E-01
117	18	20	86	14	16	101	10	12	AC087343.1	ribosomal protein L21 (RPL21) pseudogene	8		-2,63	1,7E-04	-2,49	5,5E-03
44	14	15	32	11	8	80	14	16	AC012184.3	novel transcript, antisense to DDX19B and DDX19A	16		-1,64	2,0E-03	-1,49	4,5E-02
60	17	20	404	104	98	364	115	126	AF274858.1	Morf4 family associated protein 1 (MRFAP1) pseudogene	X		-1,76	2,0E-03	-1,56	6,6E-02

442	84	51	223	36	26	185	31	16	SORD2P	sorbitol dehydrogenase 2, pseudogene	15		-2,29	2,4E-03	-2,99	1,7E-04
121	36	50	224	277	213	286	233	184	AL359881.1	novel transcript	1		-1,74	2,6E-03	-1,24	2,9E-01
231	106	106	353	178	138	238	135	116	WASH8P	WAS protein family homolog 8, pseudogene	12		-1,11	7,8E-03	-1,11	4,6E-02
167	34	73	265	146	153	407	220	244	LINC02175	long intergenic non-protein coding RNA 2175	16		-2,24	1,2E-02	-1,16	8,7E-01
72	20	27	3	8	7	5	7	9	AP003170.1	interferon stimulated exonuclease gene 20kDa-like 2 (ISG20L2) pseudogene	11		-1,79	2,0E-02	-1,35	4,5E-01
42	11	18	181	126	105	190	124	116	AL359881.2	novel transcript	1		-1,84	2,2E-02	-1,19	6,6E-01
84	31	35	92	97	105	116	97	124	CDC37L1-DT	CDC37L1 divergent transcript	9		-1,38	2,3E-02	-1,24	2,5E-01
92	34	40	89	32	39	107	31	51	LYRM4-AS1	LYRM4 antisense RNA 1	6		-1,41	2,4E-02	-1,16	3,6E-01
222	93	111	12	7	10	20	12	12	AC022893.1	novel transcript	8		-1,22	3,0E-02	-0,97	4,4E-01
57	10	10	5	2	2	14	1	1	AC034102.1	RAB13 member RAS oncogene family pseudogene	12		-2,41	3,0E-02	-2,42	1,3E-01
64	16	41	1615	1815	1378	411	386	419	SLC26A10	solute carrier family 26 member 10	12		-1,96	6,2E-02	-0,65	1,0E+00
48	9	6	149	108	66	16	22	15	AC084864.2	novel transcript	7		-2,32	6,5E-02	-2,72	8,3E-02
128	13	14	146	8	3	115	6	7	AACSP1	acetoacetyl-CoA synthetase pseudogene 1	5		-2,97	8,0E-02	-2,85	3,1E-01

DIFFERENTIALLY EXPRESSED GENES – DEVELOPING NEURONS

UPREGULATED GENES

Normalized read counts									GENE	Description	Chr	GO: Biological process	Log2FC FRDA/CT	FDR	Log2FC ISO CT/CT	FDR
iPSCs			Developing Neurons			Mature Neurons										
CT	FRDA	ISO CT	CT	FRDA	ISO CT	CT	FRDA	ISO CT								
18	42	36	36	3878	1143	8	73	74	SPON2	spondin 2	4	cell adhesion; immune system process; innate immune response; mast cell mediated immunity; opsonization; response to lipopolysaccharide; positive regulation of interleukin-6 production; positive regulation of tumor necrosis factor production; defense response to bacterium; induction of bacterial agglutination; defense response to fungus; defense response to virus; positive regulation of macrophage cytokine production; cellular response to lipopolysaccharide	6,38	7,9E-12	4,60	1,4E-04
58	92	110	20	127	63	43	99	81	MOB3C	MOB kinase activator 3C	1		2,60	4,2E-08	1,60	5,0E-02
22	40	49	19	69	61	18	48	44	TSTD3	thiosulfate sulfurtransferase like domain containing 3	6		1,83	3,0E-07	1,65	1,3E-04
122	192	190	234	533	531	194	500	441	MINDY1	MINDY lysine 48 deubiquitinase 1	1	proteolysis; protein K48-linked deubiquitination	1,19	1,8E-06	1,18	4,7E-05
22	21	26	13	329	78	28	38	39	RASD1	ras related dexamethasone induced 1	17	signal transduction; G protein-coupled receptor signaling pathway; nitric oxide mediated signal transduction; negative regulation of transcription, DNA-templated	4,29	4,0E-04	2,28	6,8E-01
10886	15191	13263	8254	19031	21163	6392	12889	16069	RPS23	ribosomal protein S23	5	translation; cytoplasmic translation; viral transcription; translational initiation; stress granule assembly; nuclear-transcribed mRNA catabolic process, nonsense-mediated decay; SRP-dependent cotranslational protein targeting to membrane; maintenance of translational fidelity	1,20	7,4E-04	1,35	4,8E-04
371	477	336	4	396	191	17	341	188	COMT	catechol-O-methyltransferase	22	catecholamine metabolic process; neurotransmitter catabolic process; methylation; developmental process; dopamine metabolic process; catecholamine catabolic process; response to organic cyclic compound; response to drug; short-term memory; cellular response to phosphate starvation; dopamine catabolic process; female pregnancy; learning; estrogen metabolic process; response to lipopolysaccharide; negative regulation of renal sodium excretion; response to estrogen; negative regulation of dopamine metabolic process; response to pain; multicellular organismal reproductive process; negative regulation of smooth muscle cell proliferation; positive regulation of homocysteine metabolic process; regulation of sensory perception of pain	4,98	1,1E-03	4,00	1,1E-01
10	144	122	15	213	161	4	124	88	PCDHA11	protocadherin alpha 11	5	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion; nervous system development	3,58	1,4E-03	3,15	4,9E-02
20	62	68	41	130	118	32	99	109	WASHC1	WASH complex subunit 1	9	Arp2/3 complex-mediated actin nucleation; protein transport; exocytosis; extracellular matrix disassembly; positive regulation of cell migration; negative regulation of autophagy; endosomal transport; positive regulation of pseudopodium assembly; regulation of protein ubiquitination;	1,66	1,8E-03	1,51	3,9E-02

																			retrograde transport, endosome to Golgi; negative regulation of phosphatidylinositol 3-kinase activity; retrograde transport, endosome to plasma membrane							
89	115	83	19	337	96	166	121	229	CALB2	calbindin 2	16	regulation of cytosolic calcium ion concentration; regulation of presynaptic cytosolic calcium ion concentration; regulation of long-term synaptic potentiation	3,77	1,8E-03	2,04		8,0E-01									
5	5	4	51	639	174	10	11	6	CLDN5	claudin 5	22	positive regulation of gene expression; negative regulation of gene expression; myelination; response to ethanol; tight junction assembly; negative regulation of cell migration; cell adhesion; bicellular tight junction assembly; outflow tract morphogenesis; cell-cell junction assembly; transforming growth factor beta receptor signaling pathway; learning; positive regulation of cell population proliferation; calcium-independent cell-cell adhesion via plasma membrane cell-adhesion molecules; negative regulation of angiogenesis; positive regulation of protein binding; maintenance of blood-brain barrier; negative regulation of vascular permeability; roof of mouth development; face morphogenesis; positive regulation of establishment of endothelial barrier; positive regulation of bicellular tight junction assembly; establishment of blood-retinal barrier; regulation of bicellular tight junction assembly	3,58	2,1E-03	1,70		9,4E-01									
291	292	319	747	1764	1117	608	670	527	PLK3	polo like kinase 3	1	protein phosphorylation; cell cycle; phosphorylation; negative regulation of apoptotic process; apoptotic process; cellular response to DNA damage stimulus; negative regulation of transcription by RNA polymerase II; DNA damage response, signal transduction by p53 class mediator resulting in cell cycle arrest; mitotic cell cycle; regulation of cell division; regulation of signal transduction by p53 class mediator; regulation of cytokinesis; G1/S transition of mitotic cell cycle; response to osmotic stress; positive regulation of intracellular protein transport; response to radiation; mitotic cell cycle checkpoint; G2/M transition of mitotic cell cycle; protein kinase B signaling; response to reactive oxygen species; Golgi disassembly; cytoplasmic microtubule organization; mitotic G1/S transition checkpoint; positive regulation of proteasomal ubiquitin-dependent protein catabolic process involved in cellular response to hypoxia; endomitotic cell cycle; positive regulation of chaperone-mediated autophagy	1,24	3,3E-03	0,58		9,8E-01									
6	14	17	5	59	58	2	22	20	PCDH8	protocadherin beta 8	5	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion	3,26	4,0E-03	3,20		2,8E-02									
20	90	92	105	408	253	515	1063	892	SLC8A1	solute carrier family 8 member A1	2	transmembrane transport; calcium ion transport; cell communication; regulation of cardiac muscle contraction by regulation of the release of sequestered calcium ion; negative regulation of protein serine/threonine kinase activity; cellular response to reactive oxygen species; regulation of heart rate; ion transport; sodium ion transport; calcium ion transmembrane transport; sodium ion transmembrane transport; calcium ion import across plasma membrane; positive regulation of bone mineralization; muscle contraction; regulation of cardiac conduction; regulation of cell communication by electrical coupling; cardiac muscle contraction; cellular response to caffeine; calcium ion import; response to muscle stretch; calcium ion homeostasis; cellular calcium ion homeostasis; membrane depolarization during cardiac muscle cell action potential; cellular sodium ion homeostasis; sodium ion export across plasma membrane; relaxation of cardiac muscle; cell communication by electrical coupling involved in cardiac conduction; regulation of cardiac muscle contraction by calcium ion signaling; regulation of gene expression; regulation of the force of heart contraction; sodium ion import across plasma membrane; positive regulation of the force of heart contraction; vascular associated smooth muscle contraction; metal ion transport; relaxation of smooth muscle; negative regulation of cytosolic calcium ion concentration; cardiac muscle cell development; cytosolic calcium ion transport; calcium ion transport into cytosol; regulation of postsynaptic cytosolic calcium ion concentration; response to hypoxia; regulation of sodium ion transport; positive regulation of cytosolic calcium ion concentration; response to nutrient; response to glucose; positive regulation of fibroblast migration; telencephalon development; response to ATP; response to immobilization stress; response to drug; response to hydrogen peroxide; regulation of calcium ion transport; cellular response to cAMP; cellular response to hypoxia; calcium ion export	1,92	4,1E-03	1,23		6,1E-01									
106	164	117	17	131	55	20	19	25	TESC	tescalcin	12	protein transport; cell differentiation; negative regulation of catalytic activity; positive regulation of transcription, DNA-templated; negative regulation of protein kinase activity; protein stabilization; positive regulation of gene expression; negative regulation of cell population proliferation; protein maturation; cellular response to retinoic acid; regulation of cell adhesion mediated by integrin; protein localization to plasma membrane; positive regulation of sodium:proton antiporter activity; positive regulation of megakaryocyte differentiation; positive regulation of granulocyte differentiation	2,77	4,4E-03	1,53		8,2E-01									
808	3610	3892	7	156	63	46	24	51	APELA	apelin receptor early endogenous ligand	4	multicellular organism development; cell differentiation; angiogenesis; heart development; positive regulation of ERK1 and ERK2 cascade; positive regulation of angiogenesis; vasculogenesis; gastrulation; endoderm development; mesoderm migration involved in gastrulation; adult heart development; embryonic heart tube development; positive regulation of heart contraction; apelin receptor signaling pathway; placenta blood vessel development; coronary vasculature development; mesendoderm migration; cell migration involved in mesendoderm migration; positive regulation of trophoblast cell migration; positive regulation of blood vessel endothelial cell proliferation involved in sprouting angiogenesis; positive regulation of G protein-coupled receptor internalization	3,72	5,4E-03	2,52		5,4E-01									
14	23	38	54	1074	203	2	45	22	OTX1	orthodenticle homeobox 1	2	regulation of transcription, DNA-templated; multicellular organism development; positive regulation of transcription by RNA polymerase II; regulation of transcription by RNA polymerase II; anterior/posterior pattern specification; metencephalon development; forebrain development; midbrain development; inner ear morphogenesis; diencephalon morphogenesis	4,09	5,4E-03	1,74		1,0E+00									
7	19	20	9	156	42	2	8	27	CDH15	cadherin 15	16	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion; cell-cell adhesion via plasma-membrane adhesion molecules	3,86	6,5E-03	2,01		9,2E-01									
65	136	120	31	312	152	17	44	44	ARC	activity regulated cytoskeleton associated protein	8	mRNA transport; multicellular organism development; endocytosis; cytoskeleton organization; cell migration; protein homooligomerization; regulation of cell morphogenesis; long-term memory; regulation of long-term synaptic potentiation; regulation of neuronal synaptic plasticity; modulation of chemical synaptic transmission; long-term synaptic potentiation; endoderm development; learning; anterior/posterior pattern specification; dendritic spine morphogenesis; regulation of dendritic spine morphogenesis; regulation of postsynaptic neurotransmitter receptor internalization; vesicle-mediated intercellular transport; regulation of long-term synaptic depression; positive regulation of AMPA receptor activity	3,12	8,0E-03	2,10		6,1E-01									
909	861	1004	115	442	422	457	616	601	KHNYN	KH and NYN domain containing	14	RNA phosphodiester bond hydrolysis, endonucleolytic	1,87	9,5E-03	1,81		7,1E-02									
18	41	26	97	862	165	83	232	158	C4orf48	chromosome 4 open reading frame 48	4		3,04	9,5E-03	0,72		1,0E+00									
20	2624	2895	20	37489	38283	25	15558	15743	NNAT	neuronatin	20	brain development; multicellular organism development; positive regulation of insulin secretion; protein lipoylation	5,42	1,1E-02	5,23		5,9E-02									

81	386	341	3	118	49	9	117	71	TXNRD2	thioredoxin reductase 2	22	oxidation-reduction process; cell redox homeostasis; cellular oxidant detoxification; cellular response to oxidative stress; response to oxygen radical	4,22	1,2E-02	3,07	5,1E-01
97	122	131	46	115	71	177	210	181	HBEFG	heparin binding EGF like growth factor	5	positive regulation of protein kinase B signaling; positive regulation of cell migration; MAPK cascade; signal transduction; membrane organization; positive regulation of cell population proliferation; cell chemotaxis; ERBB2 signaling pathway; regulation of cell motility; muscle organ development; epidermal growth factor receptor signaling pathway; negative regulation of epidermal growth factor receptor signaling pathway; positive regulation of epidermal growth factor-activated receptor activity; positive regulation of wound healing; regulation of heart contraction; cell migration; positive regulation of cell growth; wound healing, spreading of epidermal cells; positive regulation of smooth muscle cell proliferation; positive regulation of peptidyl-tyrosine phosphorylation; negative regulation of elastin biosynthetic process; positive regulation of keratinocyte migration	1,30	1,5E-02	0,61	1,0E+00
31	92	85	30	133	157	12	91	94	PCDHGA10	protocadherin gamma subfamily A, 10	5	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion	2,09	1,7E-02	2,30	2,8E-02
210	226	260	230	1292	364	46	57	76	CCDC88B	coiled-coil domain containing 88B	11	cytoskeleton-dependent intracellular transport; positive regulation of cytokine production; cytoplasmic microtubule organization; positive regulation of T cell proliferation; defense response to protozoan; positive regulation of T cell activation	2,44	1,7E-02	0,64	1,0E+00
24	43	37	10	37	27	15	37	41	PEX11G	peroxisomal biogenesis factor 11 gamma	19	peroxisome fission; regulation of peroxisome size	1,81	1,8E-02	1,34	5,5E-01
9	25	26	34	381	133	14	29	21	SUSD2	sushi domain containing 2	22	negative regulation of cell division; negative regulation of cell cycle G1/S phase transition	3,32	1,8E-02	1,82	9,5E-01
94	149	302	1849	8242	4072	432	438	464	DLL3	delta like canonical Notch ligand 3	19	multicellular organism development; cell differentiation; Notch signaling pathway; skeletal system development; somitogenesis; compartment pattern specification; tissue development; paraxial mesoderm development; negative regulation of neurogenesis	2,16	2,0E-02	1,13	9,9E-01
220	228	255	818	1999	1185	319	379	345	GADD45A	growth arrest and DNA damage inducible alpha	1	regulation of cell cycle; cell cycle; cellular response to DNA damage stimulus; cell cycle arrest; DNA damage response, signal transduction by p53 class mediator resulting in cell cycle arrest; positive regulation of apoptotic process; cellular response to mechanical stimulus; apoptotic process; positive regulation of JNK cascade; DNA repair; positive regulation of p38MAPK cascade; regulation of cyclin-dependent protein serine/threonine kinase activity; activation of MAPKK activity; negative regulation of transcription by RNA polymerase II; positive regulation of reactive oxygen species metabolic process; signal transduction in response to DNA damage; cellular response to ionizing radiation; negative regulation of blood vessel endothelial cell migration; negative regulation of protein kinase activity; centrosome cycle; negative regulation of angiogenesis; negative regulation of peptidyl-serine phosphorylation of STAT protein; mitotic cell cycle arrest; negative regulation of protein serine/threonine kinase activity	1,29	2,1E-02	0,53	1,0E+00
124	163	185	24	183	145	155	166	143	PSTPIP2	proline-serine-threonine phosphatase interacting protein 2	18	cytoskeleton organization; cell migration; actin filament polymerization	2,72	2,1E-02	2,39	2,6E-01
142	159	182	1163	4388	2280	623	947	938	SEPTIN4	septin 4	17	cell cycle; apoptotic process; cell division; cell differentiation; spermatogenesis; positive regulation of apoptotic process; positive regulation of protein ubiquitination; cellular protein localization; cytoskeleton-dependent cytokinesis; regulation of apoptotic process; positive regulation of intrinsic apoptotic signaling pathway; regulation of exocytosis	1,90	2,1E-02	0,96	1,0E+00
449	458	571	746	1641	1213	388	299	336	TOB1	transducer of ERBB2, 1	17	negative regulation of cell population proliferation; negative regulation of translation; negative regulation of nucleic acid-templated transcription; regulation of gene expression; positive regulation of nuclear-transcribed mRNA poly(A) tail shortening; positive regulation of nuclear-transcribed mRNA catabolic process, deadenylation-dependent decay; negative regulation of BMP signaling pathway; negative regulation of osteoblast differentiation; negative regulation of nuclear-transcribed mRNA poly(A) tail shortening; regulation of SMAD protein signal transduction	1,13	2,3E-02	0,70	8,6E-01
38	48	47	69	203	100	70	125	94	PBX4	PBX homeobox 4	19	regulation of transcription, DNA-templated; positive regulation of transcription, DNA-templated; regulation of transcription by RNA polymerase II; animal organ morphogenesis; eye development; brain development; embryonic organ development; neuron development	1,54	2,6E-02	0,52	1,0E+00
58	71	44	57	202	71	124	145	119	ADGRB1	adhesion G protein-coupled receptor B1	8	G protein-coupled receptor signaling pathway; cell surface receptor signaling pathway; negative regulation of angiogenesis; muscle organ development; signal transduction; immune system process; innate immune response; nervous system development; phagocytosis; defense response to Gram-negative bacterium; cell adhesion; negative regulation of endothelial cell migration; phagocytosis, recognition; phagocytosis, engulfment; adenylylate cyclase-activating G protein-coupled receptor signaling pathway; axonogenesis; peripheral nervous system development; negative regulation of cell population proliferation; negative regulation of protein ubiquitination; negative regulation of protein catabolic process; apoptotic cell clearance; engulfment of apoptotic cell; regulation of synaptic plasticity; positive regulation of synapse assembly; positive regulation of myoblast fusion; positive regulation of reactive oxygen species biosynthetic process	1,77	2,6E-02	0,29	1,0E+00
118	275	186	68	350	119	63	132	79	RTN4R	reticulon 4 receptor	22	positive regulation of GTPase activity; negative regulation of neuron projection development; negative regulation of axon regeneration; cell surface receptor signaling pathway; positive regulation of Rho protein signal transduction; negative regulation of axonogenesis; negative regulation of axon extension; corpus callosum development; neuronal signal transduction; axonogenesis	2,27	2,6E-02	0,75	1,0E+00
67	74	74	32	80	38	20	32	27	TBC1D2	TBC1 domain family member 2	9	intracellular protein transport; activation of GTPase activity; positive regulation of GTPase activity	1,30	2,8E-02	0,23	1,0E+00
56	126	86	40	320	45	26	39	42	CCDC85B	coiled-coil domain containing 85B	11	cell differentiation; viral process; regulation of growth; negative regulation of cell growth; negative regulation of transcription, DNA-templated; negative regulation of fat cell differentiation	2,85	2,9E-02	0,14	1,0E+00
379	431	442	735	2249	1694	1081	654	571	PRSS23	serine protease 23	11	proteolysis; cellular protein metabolic process; post-translational protein modification	1,59	3,3E-02	1,18	6,5E-01
303	598	485	491	1643	881	513	867	842	ZFR2	zinc finger RNA binding protein 2	19		1,71	4,0E-02	0,82	1,0E+00
1	3	5	6	63	20	6	2	2	MYL4	myosin light chain 4	17	cardiac muscle contraction; muscle filament sliding; positive regulation of ATPase activity; regulation of the force of heart contraction	3,34	4,0E-02	1,69	1,0E+00
347	674	424	239	1561	522	135	249	179	ELN	elastin	7	outflow tract morphogenesis; extracellular matrix organization; animal organ morphogenesis; aortic valve morphogenesis; respiratory gaseous exchange by respiratory system; blood circulation; regulation of smooth muscle cell proliferation; skeletal muscle tissue development; regulation of actin filament polymerization; stress fiber assembly	2,58	4,0E-02	1,04	1,0E+00

215	264	235	2692	6727	4441	1012	1428	1277	DISP3	dispatched RND transporter family member 3	1	lipid metabolic process; cell differentiation; steroid metabolic process; cholesterol metabolic process; smoothened signaling pathway; positive regulation of neural precursor cell proliferation; negative regulation of neuron differentiation; cholesterol homeostasis; regulation of lipid transport; positive regulation of lipid metabolic process	1,33	4,1E-02	0,72	1,0E+00
75	149	113	45	218	77	73	91	98	TMEM160	transmembrane protein 160	19		2,18	4,1E-02	0,72	1,0E+00
34	98	77	136	599	164	71	157	105	ANTKMT	adenine nucleotide translocase lysine methyltransferase	16	methylation; peptidyl-lysine methylation; peptidyl-lysine trimethylation; positive regulation of proton-transporting ATP synthase activity, rotational mechanism; regulation of mitochondrial ATP synthesis coupled proton transport	2,09	4,3E-02	0,26	1,0E+00
12	9	15	33	198	90	59	48	26	PLEKHD1	pleckstrin homology and coiled-coil domain containing D1	14		2,51	4,6E-02	1,38	1,0E+00
1489	740	646	630	3086	1508	1557	933	817	HES6	hes family bHLH transcription factor 6	2	regulation of transcription, DNA-templated; negative regulation of DNA-binding transcription factor activity; multicellular organism development; cell differentiation; regulation of transcription by RNA polymerase II; anterior/posterior pattern specification; regulation of neurogenesis; negative regulation of transcription by RNA polymerase II; nervous system development	2,20	4,8E-02	1,19	1,0E+00
417	305	314	184	764	350	256	228	224	LTBP3	latent transforming growth factor beta binding protein 3	11	positive regulation of mesenchymal stem cell differentiation; elastic fiber assembly; transforming growth factor beta activation; positive regulation of mesenchymal stem cell proliferation; skeletal system development; transforming growth factor beta receptor signaling pathway; negative regulation of bone mineralization; negative regulation of chondrocyte differentiation; positive regulation of bone resorption; bone remodeling; bone morphogenesis; lung sacculle development	1,99	4,8E-02	0,88	1,0E+00
5897	6188	5744	1884	4484	3851	2652	4023	3034	CLIC1	chloride intracellular channel 1	6	chloride transport; regulation of cell cycle; ion transport; ion transmembrane transport; regulation of ion transmembrane transport; signal transduction; chloride transmembrane transport; platelet aggregation; positive regulation of osteoblast differentiation; regulation of mitochondrial membrane potential	1,23	5,3E-02	1,01	5,9E-01
328	1210	574	646	1364	1120	568	741	640	ATF5	activating transcription factor 5	19	regulation of transcription, DNA-templated; regulation of transcription by RNA polymerase II; negative regulation of apoptotic process; positive regulation of transcription, DNA-templated; fat cell differentiation; negative regulation of cell population proliferation; positive regulation of transcription by RNA polymerase II; negative regulation of transcription, DNA-templated; regulation of centrosome cycle; negative regulation of cell cycle G2/M phase transition; cerebellar granule cell precursor proliferation; circadian rhythm; post-embryonic development; regulation of gene expression; olfactory bulb interneuron differentiation; olfactory bulb interneuron development; olfactory lobe development; multicellular organism growth	1,07	5,4E-02	0,79	7,8E-01
541	576	433	23	161	72	23	104	84	SLC22A31	solute carrier family 22 member 31	16	transmembrane transport; ion transport	2,55	5,6E-02	1,45	1,0E+00
0	0	0	16	109	64	1	1	3	AGBL1	ATP/GTP binding protein like 1	15	proteolysis; protein side chain deglutamylation; C-terminal protein deglutamylation	2,82	5,8E-02	1,97	8,2E-01
298	525	509	1431	6026	2888	988	1361	1236	PDE2A	phosphodiesterase 2A	11	signal transduction; adenylate cyclase-inhibiting G protein-coupled receptor signaling pathway; negative regulation of transcription by RNA polymerase II; positive regulation of inflammatory response; G protein-coupled receptor signaling pathway; cellular response to mechanical stimulus; positive regulation of gene expression; heart valve development; ventricular septum development; aorta development; regulation of cGMP-mediated signaling; negative regulation of cGMP-mediated signaling; regulation of mitochondrion organization; cAMP-mediated signaling; cGMP-mediated signaling; cellular response to macrophage colony-stimulating factor stimulus; negative regulation of vascular permeability; positive regulation of vascular permeability; regulation of cAMP-mediated signaling; negative regulation of cAMP-mediated signaling; cGMP catabolic process; establishment of endothelial barrier; cellular response to cAMP; cellular response to cGMP; cellular response to transforming growth factor beta stimulus; cellular response to 2,3,7,8-tetrachlorodibenzodioxine; monocyte differentiation; cellular response to drug; cellular response to granulocyte macrophage colony-stimulating factor stimulus	2,04	6,7E-02	0,98	1,0E+00
77	58	94	363	1704	640	166	142	132	GADD45G	growth arrest and DNA damage inducible gamma	9	regulation of cell cycle; apoptotic process; multicellular organism development; cell differentiation; positive regulation of apoptotic process; positive regulation of cold-induced thermogenesis; positive regulation of JNK cascade; positive regulation of p38MAPK cascade; activation of MAPKKK activity	2,20	6,7E-02	0,80	1,0E+00
43	46	49	24	70	44	10	8	8	CLCN1	chloride voltage-gated channel 1	7	transmembrane transport; chloride transport; chloride transmembrane transport; ion transport; regulation of ion transmembrane transport; ion transmembrane transport; muscle contraction; neuronal action potential propagation	1,53	7,3E-02	0,86	1,0E+00
1418	2773	2640	218	455	344	295	427	442	CBR1	carbonyl reductase 1	21	oxidation-reduction process; epithelial cell differentiation; cyclooxygenase pathway; drug metabolic process; vitamin K metabolic process	1,04	7,7E-02	0,64	9,8E-01
148	104	123	843	2328	1179	234	228	231	FAM110A	family with sequence similarity 110 member A	20		1,47	7,7E-02	0,49	1,0E+00
392	525	491	28	100	51	48	70	51	EGFL7	EGF like domain multiple 7	9	cell adhesion; multicellular organism development; cell differentiation; angiogenesis; anatomical structure development; blood vessel development; vasculogenesis; positive regulation of endothelial cell proliferation; negative regulation of Notch signaling pathway	1,76	7,9E-02	0,80	1,0E+00
268	478	464	55	223	74	40	63	56	ADGRE5	adhesion G protein-coupled receptor E5	19	G protein-coupled receptor signaling pathway; cell surface receptor signaling pathway; cell adhesion; signal transduction; neutrophil degranulation; inflammatory response; immune response; cell-cell signaling; adenylate cyclase-activating G protein-coupled receptor signaling pathway	1,92	8,6E-02	0,38	1,0E+00
4	3	4	54	389	49	2	5	1	LCNL1	lipocalin like 1	9		2,88	9,4E-02	-0,02	1,0E+00
4802	5234	5996	449	3425	3067	1416	492	439	PDPN	podoplanin	1	negative regulation of apoptotic process; multicellular organism development; amino acid transmembrane transport; negative regulation of cell population proliferation; positive regulation of cell migration; Rho protein signal transduction; regulation of cell shape; cell-cell adhesion; platelet activation; positive regulation of epithelial to mesenchymal transition; wound healing, spreading of cells; positive regulation of cellular component movement; folic acid transport; positive regulation of extracellular matrix disassembly; cell morphogenesis; lung development; regulation of lamellipodium morphogenesis; lymphangiogenesis; lymph node development; actin-mediated cell contraction; regulation of substrate adhesion-dependent cell spreading; positive regulation of platelet aggregation; water transport; tube morphogenesis; response to hyperoxia; lymphatic endothelial cell fate commitment; regulation of myofibroblast contraction; invadopodium organization	2,65	9,5E-02	2,48	5,0E-01
2	427	451	4	538	707	5	286	468	RPS4XP22	ribosomal protein S4X pseudogene 22	19		6,84	9,1E-27	7,22	1,8E-28

57	86	98	48	147	142	68	109	130	ELOA-AS1	ELOA antisense RNA 1	1		1,60	3,3E-07	1,55	1,8E-05
39	367	387	14	231	238	15	111	150	RCN1P2	reticulocalbin 1 pseudogene 2	13		3,77	9,3E-06	3,79	1,0E-04
4	3	6	6	99	24	5	7	13	LINC02293	long intergenic non-protein coding RNA 2293	14		3,84	4,3E-04	1,80	8,6E-01
0	1	2	15	196	80	10	5	9	AC025048.1	novel transcript	17		3,64	4,3E-04	2,31	4,0E-01
217	561	532	452	1997	1150	252	914	704	MUC20-OT1	MUC20 overlapping transcript	3		2,11	4,5E-04	1,32	4,7E-01
0	1	3	83	751	389	4	3	2	AP003049.2	novel transcript, antisense to C11orf87	11		3,21	1,2E-02	2,18	6,4E-01
32	35	29	13	127	23	11	9	4	ESPNP	espin pseudogene	1		3,13	1,5E-02	0,77	1,0E+00
131	199	207	210	603	525	483	1245	1220	LINC00632	long intergenic non-protein coding RNA 632	X		1,50	1,8E-02	1,30	2,8E-01
72	135	203	189	496	381	208	361	357	AL109918.1	heterogeneous nuclear ribonucleoprotein A/B pseudogene	6		1,38	2,2E-02	1,00	6,4E-01
17	46	50	45	212	165	465	602	558	MAGI2-AS3	MAGI2 antisense RNA 3	7		2,12	3,9E-02	1,76	5,0E-01
7	17	18	13	134	42	2	1	5	AC020909.2	novel transcript	19		3,20	3,9E-02	1,53	1,0E+00
43	166	124	210	1927	1392	34	441	160	H19	H19 imprinted maternally expressed transcript	11		3,04	4,1E-02	2,51	5,1E-01
81	167	153	134	504	332	60	123	129	MIR570	microRNA 570	3		1,87	4,5E-02	1,27	8,2E-01
260	293	217	340	885	494	224	325	281	AP002026.1	novel transcript, antisense to ADH4, ADH6 & ADH1A	4		1,37	4,6E-02	0,53	1,0E+00
48	49	54	53	114	109	54	62	61	AC005229.4	novel transcript, antisense to CUL1	7		1,08	7,5E-02	1,02	4,3E-01
11	7	6	14	71	43	9	4	3	LINC01686	long intergenic non-protein coding RNA 1686	1		2,32	7,8E-02	1,57	9,1E-01

DOWNREGULATED GENES

Normalized read counts									GENE	Description	Chr	GO: Biological process	Log ₂ FC FRDA/CT	FDR	Log ₂ FC ISO CT/CT	FDR
iPSCs			Developing Neurons			Mature Neurons										
CT	FRDA	ISO CT	CT	FRDA	ISO CT	CT	FRDA	ISO CT								
88	64	65	348	56	65	255	84	102	SAXO2	stabilizer of axonemal microtubules 2	15	microtubule anchoring	-2,61	5,9E-13	-2,38	4,8E-08
19	0	0	18	0	0	11	0	0	EIF3CL	eukaryotic translation initiation factor 3 subunit C like	16	translational initiation; formation of cytoplasmic translation initiation complex; cytoplasmic translational initiation; translation	-5,07	9,7E-11	-6,21	5,8E-07
210	100	85	196	90	120	236	103	128	THNSL1	threonine synthase like 1	10		-1,11	1,2E-03	-0,70	5,1E-01
669	465	626	391	80	101	482	340	465	SYT6	synaptotagmin 6	1	acrosome reaction; vesicle-mediated transport; regulation of dopamine secretion; calcium-ion regulated exocytosis; regulation of calcium ion-dependent exocytosis; calcium ion-regulated exocytosis of neurotransmitter; cellular response to calcium ion; acrosomal vesicle exocytosis; presynaptic dense core vesicle exocytosis	-2,24	2,5E-03	-1,90	1,1E-01
27723	30500	32082	21182	10009	10902	13127	14531	12828	ITM2C	integral membrane protein 2C	2	neuron differentiation; negative regulation of neuron projection development; negative regulation of amyloid precursor protein biosynthetic process; positive regulation of extrinsic apoptotic signaling pathway	-1,08	3,2E-03	-0,95	8,4E-02
1116	1231	1546	1489	219	194	2944	3264	2273	TSPAN2	tetraspanin 2	1	inflammatory response; brain development; astrocyte development; microglia development; myelination; oligodendrocyte differentiation; axon development	-2,68	5,6E-03	-2,80	2,0E-02
1934	1318	1418	348	104	101	431	204	162	GABRA5	gamma-aminobutyric acid type A receptor subunit alpha5	15	ion transport; chloride transport; gamma-aminobutyric acid signaling pathway; ion transmembrane transport; signal transduction; chemical synaptic transmission; regulation of membrane potential; nervous system process; chloride transmembrane transport; synaptic transmission, GABAergic; regulation of postsynaptic membrane potential; behavioral fear response; brain development; sensory perception of sound; associative learning; regulation of neuron apoptotic process; negative regulation of neuron apoptotic process; neuron development; inner ear receptor cell development; innervation; cochlea development	-1,72	6,5E-03	-1,75	3,4E-02

588	572	837	45	8	15	4	1	3	COBL	cordon-bleu WH2 repeat protein	7	digestive tract development; embryonic axis specification; neural tube closure; actin filament polymerization; somite specification; liver development; notochord development; floor plate development; collateral sprouting in absence of injury; actin filament network formation; positive regulation of dendrite development; actin cytoskeleton organization; positive regulation of ruffle assembly	-2,41	7,9E-03	-1,53	6,8E-01
2189	2598	2261	429	72	113	110	32	94	TUBA4A	tubulin alpha 4a	2	microtubule-based process; microtubule cytoskeleton organization; mitotic cell cycle; G2/M transition of mitotic cell cycle; regulation of G2/M transition of mitotic cell cycle; ciliary basal body-plasma membrane docking; platelet degranulation; cytoskeleton organization	-2,49	8,0E-03	-1,84	4,6E-01
284	164	191	44	12	8	67	29	24	STAG3	stromal antigen 3	7	cell cycle; chromosome segregation; meiotic cell cycle; sister chromatid cohesion; synaptonemal complex assembly; synapsis; protein localization to chromosome	-1,84	1,7E-02	-2,34	1,1E-02
1	1	0	353	64	81	207	283	195	GLRA1	glycine receptor alpha 1	5	ion transport; chloride transport; ion transmembrane transport; neuropeptide signaling pathway; signal transduction; excitatory postsynaptic potential; cellular response to amino acid stimulus; chemical synaptic transmission; regulation of membrane potential; nervous system process; chloride transmembrane transport; positive regulation of acrosome reaction; response to amino acid; synaptic transmission, glycinergic; cellular response to zinc ion; muscle contraction; startle response; negative regulation of transmission of nerve impulse; inhibitory postsynaptic potential; cellular response to ethanol; response to alcohol; action potential; regulation of respiratory gaseous exchange by nervous system process; anion transport; acrosome reaction; visual perception; adult walking behavior; regulation of respiratory gaseous exchange; neuromuscular process controlling posture; neuromuscular process; righting reflex	-2,30	1,8E-02	-2,00	2,7E-01
31	14	28	166	31	26	129	51	65	GRIN1	glutamate ionotropic receptor NMDA type subunit 1	9	ion transport; positive regulation of reactive oxygen species biosynthetic process; chemical synaptic transmission; regulation of membrane potential; positive regulation of transcription by RNA polymerase II; MAPK cascade; ephrin receptor signaling pathway; brain development; calcium-mediated signaling; regulation of synaptic plasticity; calcium ion transmembrane import into cytosol; excitatory chemical synaptic transmission; visual learning; protein heterotetramerization; ionotropic glutamate receptor signaling pathway; calcium ion homeostasis; response to ethanol; positive regulation of cysteine-type endopeptidase activity; regulation of NMDA receptor activity; cation transport; excitatory postsynaptic potential; positive regulation of excitatory postsynaptic potential; positive regulation of calcium ion transport into cytosol; propylene metabolic process; response to glycine; conditioned taste aversion; startle response; suckling behavior; response to amphetamine; calcium ion transport; cellular calcium ion homeostasis; learning or memory; learning; memory; long-term memory; associative learning; adult locomotory behavior; olfactory learning; regulation of cell communication; sensory perception of pain; pons maturation; cerebral cortex development; social behavior; synaptic transmission, glutamatergic; positive regulation of apoptotic process; response to morphine; regulation of neuron apoptotic process; negative regulation of neuron apoptotic process; regulation of respiratory gaseous exchange; regulation of neuronal synaptic plasticity; regulation of long-term neuronal synaptic plasticity; regulation of dendrite morphogenesis; regulation of axonogenesis; neuromuscular process; regulation of synapse assembly; prepulse inhibition; male mating behavior; protein localization to postsynaptic membrane; ion transmembrane transport	-2,29	2,2E-02	-2,53	3,9E-02
2	1	1	283	76	97	249	207	205	GOLGA7B	golgin A7 family member B	10	protein targeting to membrane; peptidyl-L-cysteine S-palmitoylation	-1,81	2,8E-02	-1,47	5,0E-01
8433	8351	7939	75	28	39	156	30	47	EPCAM	epithelial cell adhesion molecule	2	ureteric bud development; negative regulation of apoptotic process; cell-cell adhesion via plasma-membrane adhesion molecules; positive regulation of cell motility; leukocyte migration; positive regulation of transcription by RNA polymerase II; stem cell differentiation; positive regulation of cell population proliferation; positive regulation of stem cell proliferation; signal transduction involved in regulation of gene expression; negative regulation of cell-cell adhesion mediated by cadherin	-1,43	3,1E-02	-0,93	8,2E-01
1897	1552	1820	163	32	37	114	63	54	KCNS3	potassium voltage-gated channel modifier subfamily S member 3	2	ion transport; transmembrane transport; potassium ion transport; protein homooligomerization; regulation of ion transmembrane transport; potassium ion transmembrane transport; regulation of insulin secretion; ion transmembrane transport	-2,30	3,3E-02	-2,04	3,5E-01
4304	6165	5752	322	120	171	268	143	231	GPR176	G protein-coupled receptor 176	15	G protein-coupled receptor signaling pathway; signal transduction; rhythmic process; circadian behavior; chemical synaptic transmission; adenylate cyclase-inhibiting G protein-coupled receptor signaling pathway	-1,42	4,0E-02	-0,90	8,9E-01
2	2	4	97	17	36	50	33	47	TCTE1	t-complex-associated-testis-expressed 1	6	flagellated sperm motility	-2,37	4,5E-02	-1,31	1,0E+00
10188	10035	9391	1603	613	799	3064	1699	1654	KIF1C	kinesin family member 1C	17	microtubule-based movement; vesicle-mediated transport; cytoskeleton-dependent intracellular transport; retrograde vesicle-mediated transport, Golgi to endoplasmic reticulum; anterograde neuronal dense core vesicle transport; retrograde neuronal dense core vesicle transport	-1,38	4,5E-02	-0,99	7,8E-01
5648	5579	5380	1526	513	432	1292	932	911	FBLIM1	filamin binding LIM protein 1	1	cell adhesion; regulation of cell shape; cell-cell adhesion; regulation of integrin activation; cell junction assembly	-1,55	5,4E-02	-1,78	7,8E-02
9	9	13	244	47	25	41	33	13	DKK 1,00	dickkopf WNT signaling pathway inhibitor 1	10	multicellular organism development; negative regulation of Wnt signaling pathway; positive regulation of gene expression; negative regulation of cardiac muscle cell differentiation; negative regulation of canonical Wnt signaling pathway; positive regulation of tau-protein kinase activity; negative regulation of pathway-restricted SMAD protein phosphorylation; negative regulation of apoptotic process; Wnt signaling pathway; negative regulation of signaling receptor activity; negative regulation of transcription by RNA polymerase II; cell morphogenesis involved in differentiation; endoderm formation; mesoderm formation; hair follicle development; endoderm development; learning or memory; negative regulation of neuron projection development; regulation of Wnt signaling pathway; negative regulation of ossification; embryonic limb morphogenesis; negative regulation of BMP signaling pathway; forebrain development; response to retinoic acid; regulation of synapse organization; regulation of synaptic transmission, glutamatergic; limb development; head morphogenesis; face morphogenesis; motor learning; positive regulation of heart induction by negative regulation of canonical Wnt signaling pathway; Wnt signaling pathway involved in somitogenesis; modulation of age-related behavioral decline; positive regulation of midbrain dopaminergic neuron differentiation; negative regulation of presynapse assembly; negative regulation of peptidyl-serine phosphorylation; positive regulation of neuron death; positive regulation of cell death; regulation of receptor internalization; negative regulation of protein binding; negative regulation of mesodermal cell fate specification; regulation of endodermal cell fate specification; positive regulation of JUN kinase activity; positive regulation of Wnt signaling pathway, calcium modulating pathway; synapse pruning; negative regulation of canonical Wnt signaling pathway involved in cardiac muscle cell fate commitment; regulation of dopaminergic neuron differentiation; negative regulation of Wnt-Frizzled-LRP5/6 complex assembly; positive regulation of Wnt signaling pathway, planar cell polarity pathway	-2,21	5,4E-02	-3,08	9,7E-03
99	122	147	63	17	57	244	161	226	OGFRL1	opioid growth factor receptor like 1	6	opioid receptor signaling pathway	-1,87	6,0E-02	-0,14	1,0E+00

2109	2353	2101	2755	900	1988	3012	2409	2757	TLN2	talin 2	15	cell adhesion; obsolete cytoskeletal anchoring at plasma membrane; cytoskeleton organization; cell-cell adhesion; cell-cell junction assembly; endocytosis	-1,58	6,1E-02	-0,46	1,0E+00
706	957	933	1012	219	357	2332	2570	2178	ADCYAP1R1	ADCYAP receptor type 1	7	G protein-coupled receptor signaling pathway; cell surface receptor signaling pathway; multicellular organism development; cell differentiation; signal transduction; spermatogenesis; adenylate cyclase-modulating G protein-coupled receptor signaling pathway; activation of phospholipase C activity; positive regulation of calcium ion transport into cytosol; cAMP-mediated signaling; response to estradiol; multicellular organismal response to stress; response to drug; positive regulation of cAMP-mediated signaling; response to ethanol; development of primary female sexual characteristics; positive regulation of small GTPase mediated signal transduction; negative regulation of cell death; positive regulation of inositol phosphate biosynthetic process	-2,15	6,7E-02	-1,43	9,1E-01
67	79	93	441	87	218	269	172	297	HPCA	hippocalcin	1	regulation of voltage-gated calcium channel activity; cellular response to calcium ion; brain development; positive regulation of phospholipase activity; response to organic cyclic compound; calcium-mediated signaling; negative regulation of guanylate cyclase activity; activation of phospholipase D activity; positive regulation of adenylate cyclase activity; inner ear development; retina development in camera-type eye; cellular response to electrical stimulus; positive regulation of protein targeting to membrane; regulation of postsynaptic neurotransmitter receptor internalization; response to ketamine; response to L-glutamate; cellular response to monosodium glutamate; response to Aroclor 1254	-2,21	7,9E-02	-0,94	1,0E+00
84	121	102	5291	1142	1677	3101	1858	2804	WSCD2	WSC domain containing 2	12		-2,07	7,9E-02	-1,54	8,0E-01
1427	505	675	1093	273	491	2460	2989	2585	PRSS12	serine protease 12	4	proteolysis; exocytosis; endocytosis; zymogen activation	-1,95	8,4E-02	-1,11	1,0E+00
209	162	169	438	180	221	760	695	625	SH3GL2	SH3 domain containing GRB2 like 2, endophilin A1	9	negative regulation of gene expression; negative regulation of protein phosphorylation; endocytosis; antigen processing and presentation of exogenous peptide antigen via MHC class II; signal transduction; central nervous system development; membrane organization; neuron projection development; negative regulation of epidermal growth factor receptor signaling pathway; negative regulation of blood-brain barrier permeability; dendrite extension; cellular response to brain-derived neurotrophic factor stimulus; regulation of receptor internalization; synaptic vesicle uncoating; synaptic vesicle endocytosis; lipid tube assembly; membrane tubulation; membrane bending; vesicle scission; positive regulation of membrane tubulation; regulation of clathrin-dependent endocytosis	-1,26	8,9E-02	-0,97	8,0E-01
313	258	282	2409	827	1240	3613	4047	3832	DOK5	docking protein 5	20	axon guidance; transmembrane receptor protein tyrosine kinase signaling pathway; neuron differentiation; positive regulation of MAPK cascade; regulation of neurotrophin TRK receptor signaling pathway	-1,50	9,9E-02	-0,92	1,0E+00
4643	5228	4547	812	331	504	1390	1150	1184	PCDH1	protocadherin 1	5	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion; nervous system development; cell-cell signaling	-1,29	1,1E-01	-0,68	1,0E+00
277	92	221	108	31	90	160	46	89	FXN	frataxin	9	iron-sulfur cluster assembly; oxidation-reduction process; positive regulation of cell population proliferation; negative regulation of apoptotic process; ion transport; cellular iron ion homeostasis; oxidative phosphorylation; heme biosynthetic process; mitochondrion organization; adult walking behavior; aerobic respiration; embryo development ending in birth or egg hatching; proprioception; negative regulation of multicellular organism growth; negative regulation of organ growth; iron ion homeostasis; positive regulation of cell growth; cellular response to hydrogen peroxide; negative regulation of release of cytochrome c from mitochondria; protein autoprocessing; positive regulation of catalytic activity; small molecule metabolic process; response to iron ion; regulation of ferredoxin activity; iron incorporation into metallo-sulfur cluster; positive regulation of lyase activity; positive regulation of succinate dehydrogenase activity; positive regulation of aconitate hydratase activity	-1,77	1,2E-01	-0,26	1,0E+00
33154	40	35	21340	34	27	14136	20	41	AP000763.2	mitochondrially encoded cytochrome c oxidase I (MT-COI) pseudogene	11		-9,15	6,0E-113	-9,41	9,7E-75
552	3	2	421	2	1	305	1	2	AC098847.1	ribosomal protein, large, P0 pseudogene	18		-7,25	2,4E-34	-7,68	8,2E-21
153	1	0	102	2	1	86	1	0	ZBTB8OSP2	zinc finger and BTB domain containing 8 opposite strand pseudogene 2	2		-5,13	4,2E-08	-6,16	6,0E-06
48	9	7	56	7	4	55	7	2	PSMA6P1	proteasome subunit alpha 6 pseudogene 1	Y		-2,97	3,1E-07	-3,54	6,0E-06
1051	53	127	61	1	1	40	1	1	RHOXF1-AS1	RHOXF1 antisense RNA 1	X		-5,34	7,2E-07	-4,71	2,8E-04
30	1	2	81	3	2	77	2	3	LERFS	lncRNA	9		-4,51	8,9E-07	-4,93	1,4E-05
50	7	14	111	7	8	186	16	29	BX284668.2	novel transcript	1		-3,80	3,9E-06	-3,57	4,1E-04
60	17	20	404	104	98	364	115	126	AF274858.1	Morf4 family associated protein 1 (MRFAP1) pseudogene	X		-1,93	4,8E-06	-2,00	4,7E-05
442	84	51	223	36	26	185	31	16	SORD2P	sorbitol dehydrogenase 2, pseudogene	15		-2,55	5,3E-04	-2,99	1,9E-04
117	18	20	86	14	16	101	10	12	AC087343.1	ribosomal protein L21 (RPI21) pseudogene	8		-2,54	7,4E-04	-2,34	2,8E-02
46	7	9	114	32	22	109	52	56	RASA4CP	RAS p21 protein activator 4C, pseudogene	7		-1,79	3,0E-03	-2,32	2,3E-04
128	13	14	146	8	3	115	6	7	AACSP1	acetoacetyl-CoA synthetase pseudogene 1	5		-3,84	6,2E-03	-5,11	2,8E-04
0	0	0	82	28	41	52	76	64	AC016717.2	novel transcript	2		-1,49	2,0E-02	-0,96	8,0E-01

92	34	40	89	32	39	107	31	51	LYRM4-AS1	LYRM4 antisense RNA 1	6		-1,46	2,1E-02	-1,17	4,7E-01
3	4	4	1036	310	267	315	482	255	LINC01933	long intergenic non-protein coding RNA 1933	5		-1,64	7,2E-02	-1,87	1,1E-01
MATURE NEURONS																
UPREGULATED GENES																
Normalized read counts									GENE	Description	Chr	GO: Biological process	Log₂FC FRDA/CT	FDR	Log₂FC ISO CT/CT	FDR
iPSCs			Developing Neurons			Mature Neurons										
CT	FRDA	ISO CT	CT	FRDA	ISO CT	CT	FRDA	ISO CT								
0	0	0	2	31	38	17	735	646	GRIN3A	glutamate ionotropic receptor NMDA type subunit 3A	9	ion transport; ion transmembrane transport; calcium ion transport; calcium ion transmembrane transport; regulation of postsynaptic membrane potential; ionotropic glutamate receptor signaling pathway; response to ethanol; dendrite development; prepulse inhibition; regulation of synaptic vesicle exocytosis	5,36	3,1E-12	5,08	5,7E-09
122	192	190	234	533	531	194	500	441	MINDY1	MINDY lysine 48 deubiquitinase 1	1	proteolysis; protein K48-linked deubiquitination	1,36	1,1E-08	1,18	5,5E-05
10	144	122	15	213	161	4	124	88	PCDHA11	protocadherin alpha 11	5	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion; nervous system development	4,52	6,9E-06	4,02	1,7E-03
2	6	1	6	35	4	1	27	2	PCDHA9	protocadherin alpha 9	5	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion	4,46	7,7E-06	0,82	1,0E+00
51	79	89	11	30	22	4	57	27	RORB	RAR related orphan receptor B	9	regulation of transcription, DNA-templated; multicellular organism development; intracellular receptor signaling pathway; rhythmic process; visual perception; positive regulation of transcription, DNA-templated; response to stimulus; G protein-coupled receptor signaling pathway; positive regulation of transcription by RNA polymerase II; cellular response to retinoic acid; negative regulation of transcription, DNA-templated; transcription initiation from RNA polymerase II promoter; eye photoreceptor cell development; retina development in camera-type eye; regulation of circadian rhythm; negative regulation of osteoblast differentiation; retinal cone cell development; retinal rod cell development; amacrine cell differentiation	3,50	9,6E-05	2,47	1,5E-01
31	92	85	30	133	157	12	91	94	PCDHGA10	protocadherin gamma subfamily A, 10	5	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion	2,83	9,6E-05	2,88	7,8E-04
22	40	49	19	69	61	18	48	44	TSTD3	thiosulfate sulfurtransferase like domain containing 3	6		1,39	5,9E-04	1,27	2,2E-02
1	399	429	1	1224	1163	1	739	1068	TAFA5	TAFA chemokine like family member 5	22	signal transduction	5,68	9,3E-04	6,01	1,8E-03
127	118	141	768	1061	1160	516	1117	966	C20orf194	chromosome 20 open reading frame 194	20		1,11	2,3E-03	0,90	1,7E-01
14	5	6	960	1634	1042	161	899	679	NDUFA4L2	NDUFA4 mitochondrial complex associated like 2	12	proton transmembrane transport; electron transport chain	2,44	3,4E-03	2,01	1,7E-01
20	62	68	41	130	118	32	99	109	WASHC1	WASH complex subunit 1	9	Arp2/3 complex-mediated actin nucleation; protein transport; exocytosis; extracellular matrix disassembly; positive regulation of cell migration; negative regulation of autophagy; endosomal transport; positive regulation of pseudopodium assembly; regulation of protein ubiquitination; retrograde transport, endosome to Golgi; negative regulation of phosphatidylinositol 3-kinase activity; retrograde transport, endosome to plasma membrane	1,62	3,5E-03	1,75	6,3E-03
22	42	65	575	426	468	192	615	452	PRKG2	protein kinase cGMP-dependent 2	4	protein phosphorylation; phosphorylation; signal transduction; peptidyl-serine autophosphorylation; protein localization to plasma membrane; negative regulation of chloride transport	1,67	7,3E-03	1,22	5,3E-01
178	179	178	1167	1158	935	383	1016	716	SLC10A7	solute carrier family 10 member 7	4	ion transport; sodium ion transport; transmembrane transport; bone development; heparin biosynthetic process; Golgi vesicle transport; cellular calcium ion homeostasis; bile acid and bile salt transport; glycoprotein transport	1,40	7,3E-03	0,89	8,5E-01
1	1	0	11	16	17	4	54	43	PLA2G12B	phospholipase A2 group XIIB	10	phospholipid metabolic process; lipid catabolic process; arachidonic acid secretion; cholesterol homeostasis; triglyceride homeostasis	3,68	8,0E-03	3,29	1,3E-01
10886	15191	13263	8254	19031	21163	6392	12889	16069	RPS23	ribosomal protein S23	5	translation; cytoplasmic translation; viral transcription; translational initiation; stress granule assembly; nuclear-transcribed mRNA catabolic process, nonsense-mediated decay; SRP-dependent cotranslational protein targeting to membrane; maintenance of translational fidelity	1,00	1,7E-02	1,32	1,1E-03
6	14	17	5	59	58	2	22	20	PCDHB8	protocadherin beta 8	5	homophilic cell adhesion via plasma membrane adhesion molecules; cell adhesion	3,00	2,6E-02	2,90	1,3E-01
14	23	38	54	1074	203	2	45	22	OTX1	orthodenticle homeobox 1	2	regulation of transcription, DNA-templated; multicellular organism development; positive regulation of transcription by RNA polymerase II; regulation of transcription by RNA polymerase II; anterior/posterior pattern specification; metencephalon development; forebrain development; midbrain development; inner ear morphogenesis; diencephalon morphogenesis	3,71	4,3E-02	2,81	7,6E-01
288	267	294	2101	2305	1412	737	1701	1095	SRRM3	serine/arginine repetitive matrix 3	7		1,20	4,4E-02	0,56	1,0E+00
176	157	185	1218	2469	1422	189	440	308	MUC1	mucin 1, cell surface associated	1	stimulatory C-type lectin receptor signaling pathway; cytokine-mediated signaling pathway; negative regulation of transcription by competitive promoter binding; O-glycan processing; negative regulation of intrinsic apoptotic signaling pathway in response to DNA damage by p53 class mediator; DNA damage response, signal transduction by p53 class mediator resulting in transcription of p21 class	1,20	6,2E-02	0,69	1,0E+00

																	mediator; DNA damage response; signal transduction by p53 class mediator resulting in cell cycle arrest; negative regulation of cell adhesion mediated by integrin; positive regulation of transcription from RNA polymerase II promoter in response to stress; regulation of transcription from RNA polymerase II promoter in response to stress; positive regulation of histone H4 acetylation					
24	20	23	111	232	621	60	607	627	ITGA11	integrin subunit alpha 11	15						cell adhesion; integrin-mediated signaling pathway; extracellular matrix organization; cell-matrix adhesion; muscle organ development; osteoblast differentiation; cell adhesion mediated by integrin; substrate-dependent cell migration; collagen-activated signaling pathway	3,10	6,4E-02	3,08	2,3E-01	
43	54	47	2235	1598	1850	938	2485	1836	MTUS2	microtubule associated scaffold protein 2	13							1,40	6,4E-02	0,96	1,0E+00	
2767	3382	2412	3207	3835	3013	2531	6633	4239	MVD	mevalonate diphosphate decarboxylase	16						isoprenoid biosynthetic process; isopentenyl diphosphate biosynthetic process; mevalonate pathway; lipid metabolic process; steroid biosynthetic process; cholesterol biosynthetic process; steroid metabolic process; cholesterol metabolic process; sterol biosynthetic process; positive regulation of cell population proliferation; regulation of cholesterol biosynthetic process; dolichyl diphosphate biosynthetic process	1,37	6,4E-02	0,73	1,0E+00	
543	399	472	151	442	264	119	384	375	TRAF5	TNF receptor associated factor 5	1						signal transduction; regulation of apoptotic process; positive regulation of I-kappaB kinase/NF-kappaB signaling; positive regulation of DNA-binding transcription factor activity; apoptotic process; regulation of I-kappaB kinase/NF-kappaB signaling; positive regulation of NF-kappaB transcription factor activity; protein K63-linked ubiquitination; tumor necrosis factor-mediated signaling pathway; positive regulation of JNK cascade	1,64	8,3E-02	1,60	3,3E-01	
371	477	336	4	396	191	17	341	188	COMT	catechol-O-methyltransferase	22						catecholamine metabolic process; neurotransmitter catabolic process; methylation; developmental process; dopamine metabolic process; catecholamine catabolic process; response to organic cyclic compound; response to drug; short-term memory; cellular response to phosphate starvation; dopamine catabolic process; female pregnancy; learning; estrogen metabolic process; response to lipopolysaccharide; negative regulation of renal sodium excretion; response to estrogen; negative regulation of dopamine metabolic process; response to pain; multicellular organismal reproductive process; negative regulation of smooth muscle cell proliferation; positive regulation of homocysteine metabolic process; regulation of sensory perception of pain	3,48	1,1E-01	2,68	9,4E-01	
20	2624	2895	20	37489	38283	25	15558	15743	NNAT	neuronatin	20						brain development; multicellular organism development; positive regulation of insulin secretion; protein lipoylation	4,30	1,5E-01	4,17	4,5E-01	
2	427	451	4	538	707	5	286	468	RPS4XP22	ribosomal protein S4X pseudogene 22	19							5,71	2,3E-23	6,40	5,4E-28	
11	7	9	9502	15271	10959	465	1969	2083	AL161908.1	novel transcript, antisense to LIM1B	9							2,06	1,1E-03	2,12	5,7E-03	
4	1	1	9	10	23	2	131	47	MIR4527HG	MIR4527 host gene	18							4,95	1,5E-02	3,47	6,9E-01	
39	73	73	52	69	69	49	118	98	AC026304.1	novel transcript	3							1,27	2,0E-02	0,99	5,8E-01	
43	166	124	210	1927	1392	34	441	160	H19	H19 imprinted maternally expressed transcript	11							3,28	3,2E-02	1,91	1,0E+00	
0	0	1	43	65	51	7	50	49	AL139351.3	novel transcript	20							2,75	4,6E-02	2,67	2,1E-01	
1	0	0	410	381	413	236	639	514	AC091980.2	novel transcript, antisense to TLX3	5							1,45	7,5E-02	1,12	8,7E-01	
72	31	54	15	25	30	5	58	31	AC087276.1	novel transcript, sense overlapping TTC17	11							3,04	9,4E-02	2,17	1,0E+00	

DOWNREGULATED GENES

Normalized read counts									GENE	Description	Chr	GO: Biological process	Log ₂ FC FRDA/CT	FDR	Log ₂ FC ISO CT/CT	FDR
iPSCs			Developing Neurons			Mature Neurons										
CT	FRDA	ISO CT	CT	FRDA	ISO CT	CT	FRDA	ISO CT								
19	0	0	18	0	0	11	0	0	EIF3CL	eukaryotic translation initiation factor 3 subunit C like	16	translational initiation; formation of cytoplasmic translation initiation complex; cytoplasmic translational initiation; translation	-5,82	2,4E-08	-4,62	4,2E-05
8433	8351	7939	75	28	39	156	30	47	EPCAM	epithelial cell adhesion molecule	2	ureteric bud development; negative regulation of apoptotic process; cell-cell adhesion via plasma-membrane adhesion molecules; positive regulation of cell motility; leukocyte migration; positive regulation of transcription by RNA polymerase II; stem cell differentiation; positive regulation of cell population proliferation; positive regulation of stem cell proliferation; signal transduction involved in regulation of gene expression; negative regulation of cell-cell adhesion mediated by cadherin	-2,35	4,3E-07	-1,72	1,5E-02
243	285	225	598	568	810	5040	773	1177	CBLN1	cerebellin 1 precursor	16	nervous system development; chemical synaptic transmission; synapse organization; heterophilic cell-cell adhesion via plasma membrane cell adhesion molecules; cerebellar granule cell differentiation; positive regulation of long-term synaptic depression; protein secretion; positive regulation of synapse assembly; negative regulation of excitatory postsynaptic potential; regulation of postsynaptic density assembly; maintenance of synapse structure; regulation of presynapse assembly; negative regulation of inhibitory synapse assembly	-2,62	3,2E-06	-2,03	1,6E-02
88	64	65	348	56	65	255	84	102	SAXO2	stabilizer of axonemal microtubules 2	15	microtubule anchoring	-1,58	2,1E-04	-1,31	5,0E-02
210	100	85	196	90	120	236	103	128	THNSL1	threonine synthase like 1	10		-1,18	3,0E-04	-0,87	1,7E-01

3	5	8	0	0	0	46	1	4	UTS2	urotensin 2	1	regulation of blood vessel diameter; regulation of blood pressure; G protein-coupled receptor signaling pathway; muscle contraction; chemical synaptic transmission	-4,38	7,3E-03	-3,17	4,2E-01
3	4	6	314	90	97	1055	130	453	SV2C	synaptic vesicle glycoprotein 2C	5	transmembrane transport; chemical synaptic transmission; neurotransmitter transport	-2,79	8,7E-03	-1,09	1,0E+00
19	6	13	0	0	0	4	0	1	RUNX3	RUNX family transcription factor 3	1	regulation of transcription, DNA-templated; positive regulation of transcription, DNA-templated; regulation of transcription by RNA polymerase II; neuron differentiation; regulation of cell differentiation; negative regulation of transcription by RNA polymerase II; negative regulation of cell cycle; protein phosphorylation; hemopoiesis; negative regulation of epithelial cell proliferation; ossification; negative regulation of CD4-positive, alpha-beta T cell differentiation; chondrocyte differentiation; peripheral nervous system neuron development; positive regulation of CD8-positive, alpha-beta T cell differentiation; response to transforming growth factor beta	-3,49	1,5E-02	-1,81	1,0E+00
45	65	84	4	2	3	297	22	99	PTGER4	prostaglandin E receptor 4	5	G protein-coupled receptor signaling pathway; signal transduction; immune response; bone development; positive regulation of inflammatory response; negative regulation of inflammatory response; cellular response to mechanical stimulus; inflammatory response; ERK1 and ERK2 cascade; positive regulation of cytosolic calcium ion concentration; adenylate cyclase-activating G protein-coupled receptor signaling pathway; JNK cascade; negative regulation of cytokine secretion; regulation of stress fiber assembly; positive regulation of cytokine secretion; adenylate cyclase-modulating G protein-coupled receptor signaling pathway; T-helper cell differentiation; response to mechanical stimulus; regulation of ossification; response to lipopolysaccharide; negative regulation of integrin activation; cellular response to prostaglandin E stimulus; negative regulation of eosinophil extravasation	-3,34	3,3E-02	-1,37	1,0E+00
3	3	8	6	4	6	47	8	6	METTL24	methyltransferase like 24	6	methylation	-2,42	4,3E-02	-2,90	4,6E-02
1971	1862	1855	893	1052	955	1355	529	603	ETV6	ETS variant transcription factor 6	12	regulation of transcription, DNA-templated; regulation of transcription by RNA polymerase II; vitellogenesis; neurogenesis; cell differentiation; positive regulation of transcription by RNA polymerase II; mesenchymal cell apoptotic process; negative regulation of transcription by RNA polymerase II; hematopoietic stem cell proliferation	-1,34	4,3E-02	-1,15	5,0E-01
6180	5493	5431	166	146	162	498	219	221	HAS3	hyaluronan synthase 3	16	positive regulation of transcription, DNA-templated; carbohydrate metabolic process; hyaluronan biosynthetic process; extracellular polysaccharide biosynthetic process; extracellular matrix assembly; positive regulation of hyaluronan cable assembly	-1,18	6,4E-02	-1,16	2,7E-01
294	332	304	13	23	24	187	51	102	PPP1R16B	protein phosphatase 1 regulatory subunit 16B	20	regulation of phosphoprotein phosphatase activity; regulation of phosphatidylinositol 3-kinase signaling; negative regulation of protein dephosphorylation; positive regulation of endothelial cell proliferation; regulation of filopodium assembly; positive regulation of protein dephosphorylation; regulation of protein dephosphorylation; establishment of endothelial barrier; negative regulation of peptidyl-serine dephosphorylation; positive regulation of blood vessel endothelial cell proliferation involved in sprouting angiogenesis; regulation of sprouting angiogenesis	-1,82	6,4E-02	-0,84	1,0E+00
38	61	79	2	1	1	47	4	11	PVALB	parvalbumin	22	cochlea development	-3,05	7,8E-02	-1,88	1,0E+00
71	80	58	78	49	40	71	17	37	DACT2	dishevelled binding antagonist of beta catenin 2	6	negative regulation of cell adhesion; epithelial cell morphogenesis; skin development; inner medullary collecting duct development; negative regulation of nodal signaling pathway; hematopoietic progenitor cell differentiation	-1,99	8,4E-02	-0,92	1,0E+00
553	901	925	70	50	53	284	97	116	SOCS3	suppressor of cytokine signaling 3	17	intracellular signal transduction; negative regulation of apoptotic process; receptor signaling pathway via JAK-STAT; protein ubiquitination; negative regulation of signal transduction; regulation of growth; regulation of phosphatidylinositol 3-kinase activity; phosphatidylinositol phosphorylation; negative regulation of protein kinase activity; post-translational protein modification; cytokine-mediated signaling pathway; regulation of protein phosphorylation; positive regulation of tyrosine phosphorylation of STAT protein; negative regulation of tyrosine phosphorylation of STAT protein; regulation of cell differentiation; positive regulation of cell differentiation; negative regulation of receptor signaling pathway via JAK-STAT; negative regulation of insulin receptor signaling pathway; negative regulation of inflammatory response; regulation of interferon-gamma-mediated signaling pathway; branching involved in labyrinthine layer morphogenesis; placenta blood vessel development; trophoblast giant cell differentiation; spongiotrophoblast differentiation; interleukin-6-mediated signaling pathway; cellular response to leukemia inhibitory factor	-1,52	8,7E-02	-1,27	7,4E-01
447	526	642	47	25	40	157	58	83	DOCK9	dedicator of cytokinesis 9	13	small GTPase mediated signal transduction; positive regulation of GTPase activity; blood coagulation	-1,40	1,4E-01	-0,89	1,0E+00
376	434	335	5	3	6	51	14	14	SYNJ2	synaptojanin 2	6	phosphatidylinositol dephosphorylation; membrane organization; phosphatidylinositol biosynthetic process; synaptic vesicle endocytosis; brain development; inositol phosphate dephosphorylation	-1,82	1,5E-01	-1,80	4,9E-01
277	92	221	108	31	90	160	46	89	FXN	frataxin	9	iron-sulfur cluster assembly; oxidation-reduction process; positive regulation of cell population proliferation; negative regulation of apoptotic process; ion transport; cellular iron ion homeostasis; oxidative phosphorylation; heme biosynthetic process; mitochondrion organization; adult walking behavior; aerobic respiration; embryo development ending in birth or egg hatching; proprioception; negative regulation of multicellular organism growth; negative regulation of organ growth; iron ion homeostasis; positive regulation of cell growth; cellular response to hydrogen peroxide; negative regulation of release of cytochrome c from mitochondria; protein autoprocessing; positive regulation of catalytic activity; small molecule metabolic process; response to iron ion; regulation of ferrochelatase activity; iron incorporation into metallo-sulfur cluster; positive regulation of lyase activity; positive regulation of succinate dehydrogenase activity; positive regulation of aconitate hydratase activity	-1,75	2,1E-01	-0,81	1,0E+00
33154	40	35	21340	34	27	14136	20	41	AP000763.2	mitochondrially encoded cytochrome c oxidase I (MT-COI) pseudogene	11		-9,36	3,0E-95	-8,27	3,5E-72
552	3	2	421	2	1	305	1	2	AC098847.1	ribosomal protein, large, P0 pseudogene	18		-7,42	3,9E-29	-7,02	1,2E-19
44	14	15	32	11	8	80	14	16	AC012184.3	novel transcript, antisense to DDX19B and DDX19A	16		-2,53	6,5E-10	-2,29	4,7E-06
153	1	0	102	2	1	86	1	0	ZBTB80SP2	zinc finger and BTB domain containing 8 opposite strand pseudogene 2	2		-5,85	2,4E-08	-6,73	7,6E-07
4	0	0	59	0	0	55	0	0	ADGRF5P1	adhesion G protein-coupled receptor F5 pseudogene 1	9		-7,27	1,8E-07	-6,87	4,2E-05

12	5	2	87	28	22	109	13	15	AC027575.2	novel transcript	18		-2,97	2,6E-07	-2,77	1,4E-04
117	18	20	86	14	16	101	10	12	AC087343.1	ribosomal protein L21 (RPI21) pseudogene	8		-3,17	2,3E-06	-2,96	7,5E-04
1051	53	127	61	1	1	40	1	1	RHOXF1-AS1	RHOXF1 antisense RNA 1	X		-4,69	2,6E-05	-4,24	3,9E-03
50	7	14	111	7	8	186	16	29	BX284668.2	novel transcript	1		-3,37	4,5E-05	-2,51	6,8E-02
60	17	20	404	104	98	364	115	126	AF274858.1	Morf4 family associated protein 1 (MRFAP1) pseudogene	X		-1,63	4,0E-04	-1,50	1,7E-02
442	84	51	223	36	26	185	31	16	SORD2P	sorbitol dehydrogenase 2, pseudogene	15		-2,53	6,7E-04	-3,43	8,3E-06
92	34	40	89	32	39	107	31	51	LYRM4-AS1	LYRM4 antisense RNA 1	6		-1,76	1,2E-03	-1,05	7,8E-01
12	6	5	41	21	22	43	14	18	AC009318.1	novel transcript, antisense to FAR2	12		-1,57	4,4E-03	-1,24	2,8E-01
128	13	14	146	8	3	115	6	7	AACSP1	acetoacetyl-CoA synthetase pseudogene 1	5		-3,89	7,0E-03	-3,55	9,5E-02
219	30	44	102	9	1	140	6	4	LINC00668	long intergenic non-protein coding RNA 668	18		-4,03	4,0E-02	-4,28	8,6E-02
1	0	1	50	11	22	74	8	27	AC022182.2	novel transcript	8		-2,84	7,5E-02	-1,27	1,0E+00
27	4	5	50	2	3	96	14	18	LINC01783	long intergenic non-protein coding RNA 1783	1		-2,58	9,7E-02	-2,17	7,3E-01

Supplementary Table 3. Gene and protein expression of commonly identified markers in FRDA.

Gene name	Description	Normalized read counts			Log ₂ FC (FRDA vs CT)	Adj p-value	Mean peak intensity			Log ₂ FC (FRDA vs CT)	p-value
		Mean CT	Mean FRDA	Mean ISO CT			Mean CT	Mean FRDA	Mean ISO CT		
IRON HOMEOSTASIS											
ACO1 (IRP1)	aconitase 1	376,17	396,17	377,87	0,071	0,968	14441,8	12354,5	21312,1	-0,225	0,424
IREB2 (IRP2)	iron responsive element binding protein 2	9898,82	10504,92	9772,08	0,085	0,946					
FTH1	ferritin heavy chain 1	6297,04	8423,56	8313,49	0,417	0,718	13712,5	12471,7	7112,4	-0,137	0,654
FTL	ferritin light chain	13635,7	16389,20	16557,82	0,261	0,849	10441,2	14490,8	17710,8	0,473	0,231
IREG1	ferroportin-1	130,72	116,48	172,49	-0,019	0,999					
TFRC	transferrin receptor	4778,88	4439,17	5277,34	-0,116	0,962	14091,0	16383,5	22994,0	0,217	0,429
FBXL5	F-box and leucine rich repeat protein 5	4546,78	4483,85	4778,91	-0,020	0,989					
TF	transferrin	56,52	106,76	133,34	0,905	0,720	66054,1	53408,9	41030,9	-0,307	0,171
TFRC	transferrin receptor	4778,88	4439,17	5277,34	-0,116	0,962					
TFR2	transferrin receptor 2	85,24	136,71	133,94	0,674	0,716					
SLC25A28	mitoferrin-2	695,91	771,53	693,24	0,149	0,922					
DMT1	solute carrier family 11 member 2	2843,03	2777,61	2615,55	-0,035	0,980					
HIF1A	hypoxia inducible factor 1 subunit alpha	8431,72	8814,44	8051,17	0,062	0,968					
HIF1AN	hypoxia inducible factor 1 subunit alpha inhibitor	4709,07	4254,29	4498,54	-0,146	0,790					
EPAS1 (HIF2A)	endothelial PAS domain protein 1	42,40	25,51	17,35	-0,696	0,869					
Fe-S CLUSTER METABOLISM											
NFS1	NFS1 cysteine desulfurase	519,05	445,19	488,85	-0,221	0,782	4592,0	3321,8	3360,0	-0,467	0,148
ISCU	iron-sulfur cluster assembly enzyme	1542,55	1898,36	1801,57	0,298	0,570					
LYRM4 (ISD11)	LYR motif containing 4	1063,58	1088,79	1135,61	0,033	0,984					
ISCA1	iron-sulfur cluster assembly 1	1556,89	1490,26	1549,56	-0,063	0,980	5923,7	7379,9	5711,2	0,317	0,607
ISCA2	iron-sulfur cluster assembly 2	516,06	440,48	502,23	-0,228	0,835					
FDX1	ferredoxin 1	114,96	102,59	95,46	-0,166	0,913					
FDXR	ferredoxin reductase	169,32	181,38	148,10	0,093	0,974	18131,2	15805,7	17247,4	-0,198	0,635
HSPA9	heat shock protein family A (Hsp70) member 9	2157,18	1818,66	1837,56	-0,247	0,809	667919,4	770581,7	788489	0,206	0,226
HSCB	HscB mitochondrial iron-sulfur cluster cochaperone	190,57	180,39	183,92	-0,083	0,962					
GLRX	glutaredoxin	806,28	1035,14	847,88	0,356	0,819	18840,9	16460,3	17493,4	-0,195	0,578
GLRX2	glutaredoxin 2	332,32	336,17	327,15	0,016	0,994					
GLRX3	glutaredoxin 3	2524,20	2666,78	2548,81	0,078	0,962	105363,3	121356,2	131291	0,204	0,135
NUBP1	nucleotide binding protein 1	855,26	712,76	649,25	-0,262	0,825					
NUBP2	nucleotide binding protein 2	656,82	932,51	747,65	0,495	0,750	3785,5	4910,2	8705,8	0,375	0,384
NARF	nuclear prelamin A recognition factor	5655,53	5591,14	5798,12	-0,014	0,994					
CIAO1	cytosolic iron-sulfur assembly component 1	2094,92	2110,34	2053,77	0,011	0,991	1429,7	4026	3406,1	1,494	0,011
MITOCHONDRIAL Fe-S PROTEINS											
NDUFS1	NADH:ubiquinone oxidoreductase core subunit S1	2626,51	3040,60	2826,87	0,211	0,795	112035,3	133029,9	116340	0,248	0,088
NDUFS2	NADH:ubiquinone oxidoreductase core subunit S2	1993,35	1919,07	2013,20	-0,054	0,963	121309,6	120737,1	112029	-0,007	0,958
NDUFS7	NADH:ubiquinone oxidoreductase core subunit S7	479,92	849,93	586,45	0,802	0,669	34396,8	46721,1	44989,1	0,442	0,021
NDUFS8	NADH:ubiquinone oxidoreductase core subunit S8	1213,85	1702,99	1390,34	0,479	0,745	50235,0	78825,7	63470,6	0,650	0,136
NDUFV1	NADH:ubiquinone oxidoreductase core subunit V1	4511,76	5241,70	4802,20	0,215	0,790	71843,9	64869,2	50453,0	-0,147	0,141
NDUFV2	NADH:ubiquinone oxidoreductase core subunit V2	34,28	60,87	39,30	0,794	0,791	107372,6	139448,4	136270	0,377	0,153
SDHB	succinate dehydrogenase complex iron sulfur subunit B	1270,98	1061,55	1073,89	-0,260	0,779	48769,6	45546,4	46100,7	-0,099	0,645
UQCRCFS1	Rieske iron-sulfur protein	492,89	382,46	450,67	-0,363	0,851	135660,2	121098,9	123410	-0,164	0,306
ACO2	aconitase 2	1333,28	1344,84	1372,49	0,012	0,993	212450,1	195053,9	174644	-0,123	0,213
OXIDATIVE STRESS											
SOD2	superoxide dismutase 2	8828,19	8381,57	8668,55	-0,070	0,974	132451,8	143580,6	104841	0,116	0,153
CAT	catalase	770,77	1152,69	945,46	0,572	0,772	54048,4	63429,7	30772,2	0,231	0,497
NFE2L2	nuclear factor, erythroid 2 like 2	1237,90	1194,35	1209,02	-0,051	0,982					
DNA DAMAGE											
MSH2	mutS homolog 2	1341,66	1213,53	1149,30	-0,146	0,919	36452,7	46858,0	65600,6	0,362	0,244
MSH3	mutS homolog 3	258,46	235,38	221,80	-0,136	0,918	1906,6	3248,7	2683,7	0,769	0,298
MSH6	mutS homolog 6	1900,62	1402,77	1608,32	-0,435	0,765	23783,5	40868,3	81032,6	0,781	0,091

Supplementary Table 4. List of identified differentially expressed proteins in FRDA and ISO CT mature neurons.

DIFFERENTIALLY EXPRESSED PROTEINS – FRDA vs CT									
UPREGULATED									
PROTEIN	GROUP AND RELATED GENE (GN)	GO: BIOLOGICAL FUNCTION	P-value	Protein Mean Peak Intensity					Log ₂ FC FRDA/CT
				Mean CT	Mean FRDA	Mean ISO CT	Median CT	Median FRDA	
sp Q9H4L7-3 SMRCD_HUMAN	Isoform 3 of SWI/SNF-related matrix-associated actin-dependent regulator of chromatin subfamily A containing DEAD/H box 1 OS=Homo sapiens OX=9606 GN=SMARCAD1	ATPase activity, acting on DNA; ATP binding; chromatin binding; DNA binding; DNA helicase activity; nucleosome-dependent ATPase activity; ubiquitin bindin; ATP-dependent chromatin remodeling; chromatin remodeling; chromosome separation; DNA double-strand break processing; histone H3 deacetylation; histone H4 deacetylation; regulation of DNA recombination	0,003	2383,1	6056,7	4878,7	2501,5	6254,7	1,35
sp Q9BRJ2 RM45_HUMAN	39S ribosomal protein L45, mitochondrial OS=Homo sapiens OX=9606 GN=MRPL45 PE=1 SV=2	RNA binding; mitochondrial translational elongation; mitochondrial translational termination	0,006	3162,0	8172,4	6993,8	2309,5	8422,2	1,37
sp P19525 E2AK2_HUMAN	Interferon-induced, double-stranded RNA-activated protein kinase OS=Homo sapiens OX=9606 GN=EIF2AK2 PE=1 SV=2	ATP binding; double-stranded RNA binding; eukaryotic translation initiation factor 2alpha kinase activity; identical protein binding; non-membrane spanning protein tyrosine kinase activity; protein kinase activity; protein serine kinase activity; protein threonine kinase activity; transmembrane receptor protein tyrosine kinase activity; activation of MAPKK activity; cellular response to amino acid starvation; defense response to virus; endoplasmic reticulum unfolded protein response, innate immune response; negative regulation of apoptotic process; negative regulation of osteoblast proliferation; negative regulation of translation; negative regulation of viral genome replication; positive regulation of apoptotic process; positive regulation of chemokine production; positive regulation of cytokine production; positive regulation of NF-kappaB transcription factor activity; positive regulation of NIK/NF-kappaB signaling; positive regulation of stress-activated MAPK cascade; protein autophosphorylation; protein phosphorylation; regulation of hematopoietic progenitor cell differentiation; regulation of hematopoietic stem cell differentiation; regulation of hematopoietic stem cell proliferation; regulation of NLRP3 inflammasome complex assembly; response to interferon-alpha; response to lipopolysaccharide; response to toxic substance; response to virus; response to vitamin E; translation	0,005	3072,5	11122,9	11814,6	2972,5	8138,0	1,86
sp A1L0T0 ILVBL_HUMAN	Acetolactate synthase-like protein OS=Homo sapiens OX=9606 GN=ILVBL PE=1 SV=2	acetylactate synthase activity; flavin adenine dinucleotide binding; lyase activity; magnesium ion binding; thiamine pyrophosphate binding; acetolactate synthase activity; flavin adenine dinucleotide binding; lyase activity; magnesium ion binding; thiamine pyrophosphate binding; fatty acid alpha-oxidation; isoleucine biosynthetic process; valine biosynthetic process	0,003	14797,6	30474,7	29355,4	14787,4	27224,7	1,04
sp Q16822 PCKGM_HUMAN	Phosphoenolpyruvate carboxykinase [GTP], mitochondrial OS=Homo sapiens OX=9606 GN=PCK2 PE=1 SV=4	GTP binding; manganese ion binding; cellular response to dexamethasone stimulus; cellular response to glucose stimulus; cellular response to insulin stimulus; cellular response to tumor necrosis; gluconeogenesis; glycerol biosynthetic process from pyruvate; hepatocyte differentiation; NADH oxidation; oxaloacetate metabolic process; positive regulation of insulin secretion; propionate catabolic process; response to lipid; response to lipopolysaccharide; response to starvation	0,008	15690,9	32336,3	29279,3	17062,8	36017,7	1,04
sp Q9UQ16-2 DYN3_HUMAN	Isoform 2 of Dynamin-3 OS=Homo sapiens OX=9606 GN=DNM3	GTPase activity; GTP binding; identical protein binding; microtubule binding; nitric-oxide synthase binding; structural constituent of postsynapse; type 1 metabotropic glutamate receptor binding; type 5 metabotropic glutamate receptor binding; dynamin family protein polymerization involved in mitochondrial fission; endocytosis; filopodium assembly; membrane fusion; mitochondrial fission; negative regulation of dendritic spine; morphogenesis; positive regulation of filopodium assembly; positive regulation of synaptic vesicle recycling; postsynaptic neurotransmitter receptor internalization; receptor internalization; regulation of synapse structure or activity; synapse assembly; synaptic vesicle budding from presynaptic endocytic zone membrane	0,002	34483,6	73319,7	39624,3	39350,4	76370,3	1,09
sp Q86VN1 VPS36_HUMAN	Vacuolar protein-sorting-associated protein 36 OS=Homo sapiens OX=9606 GN=VPS36 PE=1 SV=1	phosphatidylinositol-3-phosphate binding; protein C-terminus binding; ubiquitin binding; endosomal transport; macroautophagy; multivesicular body assembly; protein transport to vacuole involved in ubiquitin-dependent protein catabolic process via the multivesicular body sorting pathway	0,001	6324,8	13900,4	9997,8	6734,2	13474,7	1,14
sp Q9UQ13 SHOC2_HUMAN	Leucine-rich repeat protein SHOC-2 OS=Homo sapiens OX=9606 GN=SHOC2 PE=1 SV=2	protein phosphatase 1 binding; protein phosphatase regulator activity; fibroblast growth factor receptor signaling pathway; MAPK cascade; positive regulation of Ras protein signal transduction; Ras protein signal transduction; signal transduction	0,002	8455,5	18748,0	11569,1	9299,4	17982,7	1,15
sp P60953-1 CDC42_HUMAN	Isoform 1 of Cell division control protein 42 homolog OS=Homo sapiens OX=9606 GN=CDC42	G protein activity; GTPase activity; GTP binding; GTP-dependent protein binding; identical protein binding; mitogen-activated protein kinase kinase binding; protein kinase binding; thioesterase binding; ubiquitin protein ligase activity; actin cytoskeleton organization; actin filament branching; actin filament organization; adherens junction organization; cardiac conduction system development; Cdc42 protein signal transduction; cell migration; cell projection assembly; cellular protein localization; cellular response to interferon-gamma; dendritic cell migration; dendritic spine morphogenesis; endocytosis; establishment of Golgi localization; establishment or maintenance of cell polarity; Fc-gamma receptor signaling pathway involved in phagocytosis; filopodium assembly; Golgi organization; heart contraction; integrin-mediated signaling pathway; modification of synaptic structure; negative regulation of epidermal growth factor receptor signaling pathway; negative regulation of protein-containing complex assembly; neuron fate determination; neuropilin signaling pathway; nuclear migration; organelle transport along microtubule; positive regulation of filopodium assembly; positive regulation of intracellular protein transport; positive regulation of lamellipodium assembly; positive regulation of muscle cell differentiation; positive regulation of neuron apoptotic process; positive regulation of stress fiber assembly; positive regulation of substrate adhesion-dependent cell spreading; positive regulation of synapse structural plasticity; regulation of attachment of spindle microtubules to kinetochore; viral RNA genome replication; Wnt signaling pathway, planar cell polarity pathway	0,007	8005,6	18426,8	8752,9	4504,4	21819,9	1,20
sp Q5VWJ9 SNX30_HUMAN	Sorting nexin-30 OS=Homo sapiens OX=9606 GN=SNX30 PE=1 SV=1	Phosphatidylinositol binding; protein transport; intracellular trafficking	0,002	6938,5	18207,1	17019,6	7531,6	18044,3	1,39

sp Q96M27 PRRC1_HUMAN	Protein PRRC1 OS=Homo sapiens OX=9606 GN=PRRC1 PE=1 SV=1	Identical protein binding; activation of PKA activity	0,003	3420,4	9135,2	10360,5	3245,2	8698,4	1,42	
sp Q9Y4P1-2 ATG4B_HUMAN	Isoform 2 of Cysteine protease ATG4B OS=Homo sapiens OX=9606 GN=ATG4B	cysteine-type endopeptidase activity; autophagy; macroautophagy; positive regulation of protein catabolic process; protein delipidation; protein transport; proteolysis	0,003	971,0	2853,9	1851,7	745,8	3439,5	1,56	
sp Q86W56-4 PARG_HUMAN	Isoform 4 of Poly(ADP-ribose) glycohydrolase OS=Homo sapiens OX=9606 GN=PARG	poly(ADP-ribose) glycohydrolase activity; ATP generation from poly-ADP-D-ribose; base-excision repair; gap-filling; carbohydrate metabolic process; nucleotide-sugar metabolic process; regulation of DNA repair; regulation of histone modification	0,006	6888,5	21685,1	8766,8	2726,1	24338,9	1,65	
sp Q7Z3J2 VP35L_HUMAN	VPS35 endosomal protein sorting factor-like OS=Homo sapiens OX=9606 GN=VPS35L PE=1 SV=2	endocytic recycling; Golgi to plasma membrane transport; neutrophil degranulation; protein transport	0,005	2641,4	6463,8	6618,5	2207,2	6745,8	1,29	
sp P10909-5 CLUS_HUMAN	Isoform 5 of Clusterin OS=Homo sapiens OX=9606 GN=CLU	amyloid-beta binding; chaperone binding; low-density lipoprotein particle receptor binding; misfolded protein binding; protein-containing complex binding; protein heterodimerization activity; signaling receptor binding; tau protein binding; ubiquitin protein ligase binding; unfolded protein binding; antimicrobial humoral response; cell morphogenesis; central nervous system myelin maintenance; chaperone-mediated protein complex assembly; chaperone-mediated protein folding; complement activation; immune response; regulation of amyloid-beta formation; regulation of neuronal signal transduction; regulation of neuron death; release of cytochrome c from mitochondria	0,008	51063,8	119029,0	79311,2	50662,5	117686,8	1,22	
sp P07942 LAMB1_HUMAN	Laminin subunit beta-1 OS=Homo sapiens OX=9606 GN=LAMB1 PE=1 SV=2	Cell Adhesion; cell migration; extracellular matrix organization; neuron projection development; neuronal interaction involved in cerebral cortex radial migration	0,003	24084,1	75103,3	50825,5	25640,4	93515,5	1,64	
DOWNREGULATED										
PROTEIN	GROUP AND RELATED GENE (GN)	GO: BIOLOGICAL FUNCTION	P-value	Protein Mean Peak Intensity					Log ₂ FC FRDA/CT	
				Mean CT	Mean FRDA	Mean ISO CT	Median CT	Median FRDA		
sp Q02539 H11_HUMAN	Histone H1.1 OS=Homo sapiens OX=9606 GN=HIST1H1A PE=1 SV=3	chromosome condensation; negative regulation of chromatin silencing; negative regulation of DNA recombination; chromatin binding; double-stranded DNA binding	0,000	162206,4	14742,2	7956,5	152890,7	12918,8	-3,46	
sp Q96S19-5 MTL26_HUMAN	Isoform 5 of Methyltransferase-like 26 OS=Homo sapiens OX=9606 GN=METTL26	methyltransferase	0,004	26797,8	6447,1	6426,1	26698,2	1487,3	-2,06	
sp P54198-2 HIRA_HUMAN	Isoform Short of Protein HIRA OS=Homo sapiens OX=9606 GN=HIRA	histone binding; RNA polymerase II transcription factor binding; transcription corepressor activity anatomical structure morphogenesis; DNA replication-independent nucleosome assembly; regulation of transcription by RNA polymerase II; transcription, DNA-templated	0,004	8658,0	3299,6	2955,7	7770,3	2667,1	-1,39	
sp P52926 HMGA2_HUMAN	High mobility group protein HMGI-C OS=Homo sapiens OX=9606 GN=HMGA2 PE=1 SV=1	DNA binding; nucleosomal binding; SMAD binding; transcription coregulator activity; transcription corepressor activity; transcription factor binding; transcription regulatory region sequence-specific DNA binding; base-excision repair; cell division; chondrocyte differentiation; chromatin organization; chromosome breakage; chromosome condensation; DNA damage response, detection of DNA damage; endodermal cell differentiation; epithelial to mesenchymal transition; fat cell differentiation; heterochromatin assembly; histone H2A-S139 phosphorylation; mesenchymal cell differentiation; mesodermal cell differentiation; mesodermal-endodermal cell signaling; mitotic G2 DNA damage checkpoint; multicellular organism development; negative regulation by host of viral transcription; negative regulation of apoptotic process; negative regulation of cellular senescence; negative regulation of DNA binding; negative regulation of double-strand break repair via nonhomologous end joining; negative regulation of single stranded viral RNA replication via double stranded DNA intermediate; negative regulation of transcription, DNA-templated; negative regulation of transcription by RNA polymerase II; oncogene-induced cell senescence; positive regulation of angiogenesis; positive regulation of apoptotic process; positive regulation of cell proliferation in bone marrow; positive regulation of cellular response to X-ray; positive regulation of cellular senescence; positive regulation of gene expression; positive regulation of protein serine/threonine kinase activity; positive regulation of response to DNA damage stimulus; positive regulation of stem cell proliferation; positive regulation of transcription, DNA-templated; positive regulation of transcription by RNA polymerase II; regulation of cell cycle process; regulation of cellular response to drug; regulation of growth regulation of stem cell population maintenance; regulation of transcription, DNA-templated; response to virus; senescence-associated heterochromatin focus assembly; stem cell differentiation	0,000	94730,2	37407,7	54202,3	79679,0	34183,4	-1,34	
sp Q6BDS2 URFB1_HUMAN	UHRF1-binding protein 1 OS=Homo sapiens OX=9606 GN=UHRF1BP1 PE=1 SV=1	histone deacetylase binding; identical protein binding; may act as a negative regulator of cell growth	0,008	3695,0	1768,2	2984,7	3780,3	1514,2	-1,06	
sp Q9H0H0 INT2_HUMAN	Integrator complex subunit 2 OS=Homo sapiens OX=9606 GN=INTS2 PE=1 SV=2	snRNA 3'-end processing; snRNA processing; snRNA transcription by RNA polymerase II	0,002	5561,2	2767,0	3406,5	5858,2	2663,4	-1,01	
sp P09382 LEG1_HUMAN	Galectin-1 OS=Homo sapiens OX=9606 GN=LGALS1 PE=1 SV=2	carbohydrate binding; identical protein binding; lactose binding; laminin binding; RNA binding apoptotic process; cellular protein metabolic process; cellular response to glucose stimulus; cellular response to organic cyclic compound; myoblast differentiation; negative regulation of cell-substrate adhesion; negative regulation of neuron projection development; plasma cell differentiation; positive regulation of erythrocyte aggregation; positive regulation of I-kappaB kinase/NF-kappaB signaling; positive regulation of viral entry into host cell; post-translational protein modification; regulation of apoptotic process; response to axon injury; response to drug; response to isolation stress; T cell costimulation	0,000	42609,4	7614,2	17767,9	43715,6	7357,6	-2,48	
sp P78417 GSTO1_HUMAN	Glutathione S-transferase omega-1 OS=Homo sapiens OX=9606 GN=GSTO1 PE=1 SV=2	glutathione dehydrogenase (ascorbate) activity; glutathione transferase activity; methylarsenate reductase activity; oxidoreductase activity cellular response to arsenic-containing substance; glutathione derivative biosynthetic process; glutathione metabolic process; interleukin-12-mediated signaling pathway; L-ascorbic acid metabolic process; methylation; negative regulation of ryanodine-sensitive calcium-release channel activity; positive regulation of ryanodine-sensitive calcium-release channel activity; positive regulation of skeletal muscle contraction by regulation of release of sequestered calcium ion; regulation of cardiac muscle contraction by regulation of the release of	0,000	54244,4	12811,0	41800,8	51628,2	11358,4	-2,08	

		sequestered calcium ion; regulation of release of sequestered calcium ion into cytosol by sarcoplasmic reticulum; xenobiotic catabolic process									
sp Q9NPA0 EMC7_HUMAN	ER membrane protein complex subunit 7 OS=Homo sapiens OX=9606 GN=EMC7 PE=1 SV=1	carbohydrate binding; protein insertion into ER membrane by stop-transfer membrane-anchor sequence; tail-anchored membrane protein insertion into ER membrane; part of the endoplasmic reticulum membrane protein complex (EMC) that enables the energy-independent insertion into endoplasmic reticulum membranes of newly synthesized membrane proteins	0,000	98207,5	23769,4	59332,4	106419,1	27504,3			-2,05
sp P00338 LDHA_HUMAN	L-lactate dehydrogenase A chain OS=Homo sapiens OX=9606 GN=LDHA PE=1 SV=2	L-lactate dehydrogenase activity; carbohydrate metabolic process; carboxylic acid metabolic process	0,000	728496,5	221896,9	568446,7	728226,0	225622,6			-1,72
sp Q01628 IFM3_HUMAN	Interferon-induced transmembrane protein 3 OS=Homo sapiens OX=9606 GN=IFITM3 PE=1 SV=2	defense response to virus; immune response; negative regulation of viral entry into host cell; negative regulation of viral genome replication; negative regulation of viral transcription; response to interferon-alpha; response to interferon-beta; response to interferon-gamma; response to virus; type I interferon signaling pathway	0,001	6776,0	2249,2	6132,4	6805,8	1148,2			-1,59
sp O75056 SDC3_HUMAN	Syndecan-3 OS=Homo sapiens OX=9606 GN=SDC3 PE=1 SV=2	identical protein binding; cell migration; glycosaminoglycan biosynthetic process; glycosaminoglycan catabolic process; leukocyte migration; retinoid metabolic process; may have a role in the organization of cell shape by affecting the actin cytoskeleton, possibly by transferring signals from the cell surface in a sugar-dependent mechanism	0,006	29250,0	11609,7	6666,0	29752,7	11238,0			-1,33
sp Q9H1A4 APC1_HUMAN	Anaphase-promoting complex subunit 1 OS=Homo sapiens OX=9606 GN=ANAPC1 PE=1 SV=1	molecular adaptor activity; anaphase-promoting complex-dependent catabolic process; cell division; metaphase/anaphase transition of mitotic cell cycle; protein K11-linked ubiquitination; regulation of exit from mitosis; regulation of mitotic cell cycle phase transition; ubiquitin-dependent protein catabolic process	0,001	13038,2	5830,0	6525,8	13409,8	5737,5			-1,16
sp Q9H497-2 TOR3A_HUMAN	Isoform 2 of Torsin-3A OS=Homo sapiens OX=9606 GN=TOR3A	ATP binding; ATPase activity	0,004	9201,2	4179,3	5398,0	8838,6	3972,4			-1,14
sp P00491 PNPH_HUMAN	Purine nucleoside phosphorylase OS=Homo sapiens OX=9606 GN=PNP PE=1 SV=2	guanosine phosphorylase activity; dAMP catabolic process	0,007	68926,3	34322,0	40639,6	52149,7	34474,3			-1,01
sp P48539 PCP4_HUMAN	Calmodulin regulator protein PCP4 OS=Homo sapiens OX=9606 GN=PCP4 PE=1 SV=3	calcium ion binding; calmodulin binding; calmodulin dependent kinase signaling pathway; positive regulation of neuron differentiation	0,007	76299,6	33823,1	38857,0	78335,5	41455,1			-1,17
sp Q9584 S10AD_HUMAN	Protein S100-A13 OS=Homo sapiens OX=9606 GN=S100A13 PE=1 SV=1	calcium-dependent protein binding; calcium ion binding; copper ion binding; fibroblast growth factor binding; lipid binding; protein homodimerization activity; RAGE receptor binding; zinc ion binding; mast cell degranulation; positive regulation of cell population proliferation; positive regulation of cytokine production; positive regulation of I-kappaB kinase/NF-kappaB signaling; positive regulation of interleukin-1 alpha production; protein transport; regulation of cell shape; response to copper ion; response to electrical stimulus	0,000	44553,1	16816,7	42855,2	45934,5	17453,9			-1,41
sp P04271 S100B_HUMAN	Protein S100-B OS=Homo sapiens OX=9606 GN=S100B PE=1 SV=2	Calcium-dependent protein binding; calcium ion binding; identical protein binding; protein homodimerization activity; RAGE receptor binding; S100 protein binding; tau protein binding; zinc ion binding axonogenesis; central nervous system development; innate immune response; learning or memory; positive regulation of cell population proliferation; positive regulation of I-kappaB kinase/NF-kappaB signaling	0,000	4858,3	1181,2	1452,5	5091,4	913,5			-2,04
sp P07602-3 SAP_HUMAN	Isoform Sap-mu-9 of Prosaposin OS=Homo sapiens OX=9606 GN=PSAP	identical protein binding; phospholipid binding; protease binding; protein homodimerization activity; enzyme activator activity; adenylate cyclase-inhibiting G protein-coupled receptor signaling pathway; epithelial cell differentiation involved in prostate gland development; ganglioside GM1 transport to membrane; glycosphingolipid metabolic process; G protein-coupled receptor signaling pathway; lysosomal transport; neutrophil degranulation; platelet degranulation; positive regulation of beta-galactosidase activity; prostate gland growth; regulation of autophagy; regulation of lipid metabolic process	0,008	121325,2	48654,8	60125,4	122082,2	47824,0			-1,32
sp Q9NR46 SHLB2_HUMAN	Endophilin-B2 OS=Homo sapiens OX=9606 GN=SH3GLB2 PE=1 SV=1	Cadherin binding; identical protein binding	0,000	29053,1	6385,6	7041,0	26340,0	4314,2			-2,19
sp P07951-2 TPM2_HUMAN	Isoform 2 of Tropomyosin beta chain OS=Homo sapiens OX=9606 GN=TPM2	actin binding; actin filament binding; identical protein binding; protein heterodimerization activity; protein homodimerization activity; structural constituent of muscle; actin filament organization; muscle contraction ; muscle filament sliding; regulation of ATPase activity	0,000	20198,9	5246,2	12062,3	20606,7	3876,3			-1,94
sp P20810-6 ICAL_HUMAN	Isoform 6 of Calpastatin OS=Homo sapiens OX=9606 GN=CAST	cadherin binding; calcium-dependent cysteine-type endopeptidase inhibitor activity; endopeptidase inhibitor activity; RNA binding inhibition of cysteine-type endopeptidase activity; negative regulation of type B; pancreatic cell apoptotic process; presynaptic active zone organization	0,001	6050,0	1722,6	4798,6	6818,9	860,0			-1,81
sp O15143 ARC1B_HUMAN	Actin-related protein 2/3 complex subunit 1B OS=Homo sapiens OX=9606 GN=ARPC1B PE=1 SV=3	Arp2/3 complex-mediated actin nucleation; ephrin receptor signaling pathway; Fc-gamma receptor signaling pathway involved in phagocytosis; response to estradiol; response to estrogen; actin binding; structural constituent of cytoskeleton	0,000	19370,0	5806,3	9225,2	17823,9	5933,2			-1,74
sp P05413 FABPH_HUMAN	Fatty acid-binding protein, heart OS=Homo sapiens OX=9606 GN=FABP3 PE=1 SV=4	cytoskeletal protein binding; icosatetraenoic acid binding; long-chain fatty acid binding; long-chain fatty acid transporter activity; oleic acid binding brown fat cell differentiation; cholesterol homeostasis; fatty acid metabolic process; intracellular lipid transport; long-chain fatty acid transport; negative regulation of cell population proliferation; phospholipid homeostasis; positive regulation of long-chain fatty acid import into cell; positive regulation of phospholipid biosynthetic process; regulation of fatty acid oxidation; regulation of phosphatidylcholine biosynthetic process; response to drug; response to fatty acid; response to insulin; triglyceride catabolic process	0,000	35322,0	11533,1	9372,6	33570,8	9986,9			-1,61
sp P13797 PLST_HUMAN	Plastin-3 OS=Homo sapiens OX=9606 GN=PLS3 PE=1 SV=4	actin filament binding; calcium ion binding actin filament bundle assembly; actin filament network formation; bone development	0,000	128506,5	51227,6	150688,1	146194,4	48804,9			-1,33
sp Q05682-5 CALD1_HUMAN	Isoform 5 of Caldesmon OS=Homo sapiens OX=9606 GN=CALD1	actin binding; cadherin binding; calmodulin binding; myosin binding; tropomyosin binding; actin filament bundle assembly; angiogenesis; muscle contraction	0,000	31728,9	13931,4	29765,1	28902,0	12843,1			-1,19
sp P09493-4 TPM1_HUMAN	Isoform 4 of Tropomyosin alpha-1 chain OS=Homo sapiens OX=9606 GN=TPM1	actin binding; actin filament binding ; cytoskeletal protein binding; identical protein binding; protein heterodimerization activity; protein homodimerization activity; structural constituent of cytoskeleton; structural constituent of muscle	0,000	59520,2	26441,3	32966,9	58235,6	27690,7			-1,17
sp P06396-2 GELS_HUMAN	Isoform 2 of Gelsolin OS=Homo sapiens OX=9606 GN=GSN	actin binding; calcium ion binding; myosin II binding; phosphatidylinositol 3-kinase catalytic subunit binding; phosphatidylinositol-4,5-bisphosphate binding; actin filament capping; actin filament depolymerization; actin filament polymerization; actin filament reorganization; actin filament severing; actin nucleation; aging; amyloid fibril formation; cardiac muscle cell contraction; cell projection	0,000	108015,2	50728,5	100901,0	100845,1	52184,5			-1,09

		assembly; cellular response to cadmium ion; cellular response to interferon-gamma; central nervous system development; cilium assembly; negative regulation of viral entry into host cell; neutrophil degranulation; oligodendrocyte development; phagocytosis, engulfment; phosphatidylinositol-mediated signaling; positive regulation of actin nucleation; positive regulation of cysteine-type endopeptidase activity involved in apoptotic signaling pathway; positive regulation of gene expression; positive regulation of keratinocyte apoptotic process; positive regulation of protein processing in phagocytic vesicle; protein destabilization; regulation of cell adhesion; regulation of establishment of T cell polarity; regulation of plasma membrane raft polarization; regulation of podosome assembly; regulation of receptor clustering; relaxation of cardiac muscle; renal protein absorption; response to ethanol; response to folic acid; response to muscle stretch; sequestering of actin monomers; striated muscle atrophy; tissue regeneration; wound healing							
sp P02545 LMNA_HUMAN	Prelamin-A/C OS=Homo sapiens OX=9606 GN=LMNA PE=1 SV=1	identical protein binding; structural molecule activity; cellular protein localization; cellular response to hypoxia; DNA double-strand break attachment to nuclear envelope; establishment or maintenance of microtubule; cytoskeleton polarity; IRE1-mediated unfolded protein response; mitotic nuclear envelope reassembly; muscle organ development; negative regulation of cardiac muscle hypertrophy in response to stress; negative regulation of cell population proliferation; nuclear envelope organization; positive regulation of cell aging; protein localization to nucleus; regulation of cell migration; regulation of telomere maintenance; nuclear assembly; chromatin organization, nuclear membrane and telomere dynamics; required for normal development of peripheral nervous system and skeletal muscle and for muscle satellite cell proliferation	0,000	50695,3	24063,7	24589,4	46463,5	23290,2	-1,07
sp Q9NUM4 T106B_HUMAN	Transmembrane protein 106B OS=Homo sapiens OX=9606 GN=TMEM106B PE=1 SV=2	dendrite morphogenesis and branching; lysosomal transport; lysosome localization; lysosome organization	0,003	11092,9	5332,1	7955,7	10581,2	4901,5	-1,06
sp Q15063-5 POSTN_HUMAN	Isoform 5 of Periostin OS=Homo sapiens OX=9606 GN=POSTN	cell adhesion molecule binding; heparin binding; metal ion binding; bone regeneration; cell adhesion; cellular response to fibroblast growth factor stimulus; cellular response to transforming growth factor beta; cellular response to tumor necrosis factor; cellular response to vitamin K; extracellular matrix organization; negative regulation of cell-matrix adhesion; negative regulation of substrate adhesion-dependent cell spreading; neuron projection extension; positive regulation of chemokine (C-X-C motif) ligand 2 production; positive regulation of smooth muscle cell migration; regulation of Notch signaling pathway; regulation of systemic arterial blood pressure; response to hypoxia; response to mechanical stimulus; response to muscle activity; tissue development; wound healing	0,000	52190,0	25267,4	83386,8	52547,3	18971,6	-1,05
sp Q43491-2 E41L2_HUMAN	Isoform 2 of Band 4.1-like protein 2 OS=Homo sapiens OX=9606 GN=EPB41L2	actin binding; PH domain binding; spectrin binding; structural molecule activity; actomyosin structure organization; cell cycle; cell division; cortical actin cytoskeleton organization; positive regulation of protein localization to cell cortex	0,000	34255,4	17115,9	34051,0	34673,3	16833,9	-1,00
sp Q15427 MOT4_HUMAN	Monocarboxylate transporter 4 OS=Homo sapiens OX=9606 GN=SLC16A3 PE=1 SV=1	lactate transmembrane transporter activity; monocarboxylic acid transmembrane transporter activity; RNA binding; symporter activity leukocyte migration; monocarboxylic acid transport; plasma membrane lactate transport; pyruvate metabolic process	0,001	16322,5	5038,0	9766,9	16549,7	3905,9	-1,70
sp Q96ER9 CCDC51_HUMAN	Coiled-coil domain-containing protein 51 OS=Homo sapiens OX=9606 GN=CCDC51 PE=1 SV=2	mitochondrial ATP-gated potassium channel activity; potassium ion transmembrane transport; may contribute to the homeostatic control of cellular metabolism under stress conditions by regulating the mitochondrial matrix volume	0,003	10002,8	4990,9	6763,4	10016,5	4178,0	-1,00

DIFFERENTIALLY EXPRESSED PROTEINS – FRDA vs ISO CT

UPREGULATED

PROTEIN	GROUP AND RELATED GENE (GN)	GO: BIOLOGICAL FUNCTION	P-value	Protein Mean Peak Intensity					Log ₂ FC FRDA/ ISO CT
				Mean ISO CT	Mean FRDA	Mean CT	Median ISO CT	Median FRDA	
sp Q96MU7-2 YTDC1_HUMAN	Isoform 2 of YTH domain-containing protein 1 OS=Homo sapiens OX=9606 GN=YTDC1	dosage compensation by inactivation of X chromosome; mRNA export from the nucleus; mRNA splicing, via spliceosome; posttranscriptional regulation of gene expression	0,000	3945,5	8453,7	16266,7	4221,9	8767,0	1,10
sp Q9HCJ6 VAT1L_HUMAN	Synaptic vesicle membrane protein VAT-1 homolog-like OS=Homo sapiens OX=9606 GN=VAT1L PE=1 SV=2	oxidoreductase activity; zinc ion binding	0,000	75389,8	153541,5	89811,6	85987,6	168354,1	1,03
sp Q43301 HS12A_HUMAN	Heat shock 70 kDa protein 12A OS=Homo sapiens OX=9606 GN=HSPA12A PE=1 SV=2	ATP binding	0,000	57363,0	117513,2	75095,8	54891,2	106579,2	1,03
sp Q6PCE3 PGM2L_HUMAN	Glucose 1,6-bisphosphate synthase OS=Homo sapiens OX=9606 GN=PGM2L1 PE=1 SV=3	glucose-1,6-bisphosphate synthase activity; intramolecular transferase activity, phosphotransferases; galactose catabolic process; glucose 6-phosphate metabolic process; glucose metabolic process; glycogen biosynthetic process; glycogen catabolic process	0,002	77243,3	158411,6	129424,1	70848,5	145826,2	1,04
sp P04040 CATA_HUMAN	Catalase OS=Homo sapiens OX=9606 GN=CAT PE=1 SV=3	cellular response to growth factor stimulus; cellular response to oxidative stress; cholesterol metabolic process; negative regulation of apoptotic process; response to hypoxia; antioxidant activity	0,003	30772,1	63429,7	54048,4	31104,6	61985,0	1,04
sp Q05639 EF1A2_HUMAN	Elongation factor 1-alpha 2 OS=Homo sapiens OX=9606 GN=EEF1A2 PE=1 SV=1	positive regulation of apoptotic process; positive regulation of lipid kinase activity; regulation of chaperone-mediated autophagy; response to electrical stimulus; translation; translation elongation	0,000	21916,3	45294,8	32298,4	21960,5	48219,7	1,05
sp P05067-3 A4_HUMAN	Isoform L-APP677 of Amyloid-beta precursor protein OS=Homo sapiens OX=9606 GN=APP	acetylcholine receptor activator activity; chaperone; chemoattractant activity; chromatin binding; DNA binding; enzyme binding; enzyme binding; G protein-coupled receptor binding; growth factor receptor binding; heparan sulfate binding; integrin binding; activation of MAPK activity; adult locomotory behavior; axo-dendritic transport; axon midline choice point recognition; axonogenesis; calcium-mediated signaling; cell adhesion; cellular copper ion homeostasis; cellular process; cellular protein metabolic process; cellular response to amyloid-beta; cellular response to cAMP; dendrite development; endocytosis; extracellular matrix organization; locomotory behavior; modulation of excitatory postsynaptic potential; negative regulation of gene expression; negative regulation of long-term synaptic potentiation; negative regulation of mitochondrion organization; negative regulation of neuron death; neuromuscular process controlling balance; neuron projection	0,000	33268,8	69069,6	79564,4	30510,9	67344,0	1,05

		development; neuron projection maintenance; neuron remodeling; synapse organization; synaptic growth at neuromuscular junction									
sp P60953-1 CDC42_HUMAN	Isoform 1 of Cell division control protein 42 homolog OS=Homo sapiens OX=9606 GN=CDC42	G protein activity; GTPase activity; GTP binding; GTP-dependent protein binding; identical protein binding; mitogen-activated protein kinase kinase binding; protein kinase binding; thioesterase binding; ubiquitin protein ligase activity; actin cytoskeleton organization; actin filament branching; actin filament organization; adherens junction organization; cardiac conduction system development; Cdc42 protein signal transduction; cell migration; cell projection assembly; cellular protein localization; cellular response to interferon-gamma; dendritic cell migration; dendritic spine morphogenesis; endocytosis; establishment of Golgi localization; establishment or maintenance of cell polarity; Fc-gamma receptor signaling pathway involved in phagocytosis; filopodium assembly; Golgi organization; heart contraction; integrin-mediated signaling pathway; modification of synaptic structure; negative regulation of epidermal growth factor receptor signaling pathway; negative regulation of protein-containing complex assembly; neuron fate determination; neuropilin signaling pathway; nuclear migration; organelle transport along microtubule; positive regulation of filopodium assembly; positive regulation of intracellular protein transport; positive regulation of lamellipodium assembly; positive regulation of muscle cell differentiation; positive regulation of neuron apoptotic process; positive regulation of stress fiber assembly; positive regulation of substrate adhesion-dependent cell spreading; positive regulation of synapse structural plasticity; regulation of attachment of spindle microtubules to kinetochore; viral RNA genome replication; Wnt signaling pathway, planar cell polarity pathway	0,003	8752,9	18426,8	8005,6	7597,3	21819,9	1,07		
sp Q9UHG2 PCSK1_HUMAN	ProSAS OS=Homo sapiens OX=9606 GN=PCSK1N PE=1 SV=1	neuropeptide signaling pathway; endopeptidase inhibitor activity	0,000	99290,2	219291,2	169430,5	83346,9	236706,5	1,14		
sp Q15125 EBP_HUMAN	3-beta-hydroxysteroid-Delta(8),Delta(7)-isomerase OS=Homo sapiens OX=9606 GN=EBP PE=1 SV=3	cholesterol biosynthetic process; cholesterol metabolic process; skeletal system development	0,002	4857,6	11475,3	13202,2	4735,5	12372,6	1,24		
sp Q9UJX2 CDC23_HUMAN	Cell division cycle protein 23 homolog OS=Homo sapiens OX=9606 GN=CDC23 PE=1 SV=3	ubiquitin-protein transferase activity; anaphase-promoting complex-dependent catabolic process; cell division	0,002	6416,0	15171,1	12493,9	6898,5	15424,3	1,24		
sp Q86W56-4 PARG_HUMAN	Isoform 4 of Poly(ADP-ribose) glycohydrolase OS=Homo sapiens OX=9606 GN=PARG	poly(ADP-ribose) glycohydrolase activity; ATP generation from poly-ADP-D-ribose; base-excision repair; gap-filling; carbohydrate metabolic process; nucleotide-sugar metabolic process; regulation of DNA repair; regulation of histone modification	0,004	8766,8	21685,1	6888,5	3421,7	24338,9	1,31		
sp Q7L7X3 TAOK1_HUMAN	Serine/threonine-protein kinase TAO1 OS=Homo sapiens OX=9606 GN=TAOK1 PE=1 SV=1	tubulin binding; protein kinase activity; DNA repair; microtubule cytoskeleton organization; mitotic spindle organization; neuron projection morphogenesis; regulation of actin cytoskeleton organization; regulation of microtubule cytoskeleton organization	0,004	25920,7	65189,6	36856,9	18497,0	72282,0	1,33		
sp P17252 KPCA_HUMAN	Protein kinase C alpha type OS=Homo sapiens OX=9606 GN=PRKCA PE=1 SV=4	ATP binding; calcium-dependent protein kinase C activity; enzyme binding; integrin binding; protein kinase activity; apoptotic signaling pathway; axon guidance; cell adhesion; intracellular signal transduction; positive regulation of cardiac muscle hypertrophy; positive regulation of cell adhesion; positive regulation of ERK1 and ERK2 cascade	0,002	5216,4	13320,6	7554,7	4863,3	15984,1	1,35		
sp Q8WV17 PPR1C_HUMAN	Protein phosphatase 1 regulatory subunit 1C OS=Homo sapiens OX=9606 GN=PPP1R1C PE=3 SV=1	protein phosphatase inhibitor activity; intracellular signal transduction; cell cycle; cell division	0,001	14664,0	39211,4	43879,7	10588,7	38670,9	1,42		
sp Q5R115 COX20_HUMAN	Cytochrome c oxidase assembly protein COX20, mitochondrial OS=Homo sapiens OX=9606 GN=COX20 PE=1 SV=2	mitochondrial cytochrome c oxidase assembly	0,009	2947,2	10087,9	10517,7	1673,7	8871,8	1,78		
sp Q9UGI8 TES_HUMAN	Testin OS=Homo sapiens OX=9606 GN=TES PE=1 SV=1	cadherin binding; RNA binding; negative regulation of cell population proliferation; may play a role in cell adhesion, cell spreading and in the reorganization of the actin cytoskeleton	0,000	10767,2	40885,9	45960,6	9723,2	40036,8	1,92		
sp O14639-2 ABL1_HUMAN	Isoform 2 of Actin-binding LIM protein 1 OS=Homo sapiens OX=9606 GN=ABL1M1	actin binding; cytoskeleton organization; animal organ morphogenesis	0,001	7838,7	15892,2	13327,7	7255,2	13520,5	1,02		
sp Q02410-2 APBA1_HUMAN	Isoform 2 of Amyloid-beta A4 precursor protein-binding family A member 1 OS=Homo sapiens OX=9606 GN=APBA1	axo-dendritic transport; cell adhesion; chemical synaptic transmission; gamma-aminobutyric acid secretion; glutamate secretion; intracellular protein transport; in utero embryonic development; locomotory behavior; multicellular organism growth; nervous system development; neurotransmitter transport; protein-containing complex assembly; regulation of gene expression; regulation of synaptic vesicle exocytosis	0,009	1878,5	3828,4	3679,8	1196,0	4287,1	1,03		
sp P07196 NFL_HUMAN	Neurofilament light polypeptide OS=Homo sapiens OX=9606 GN=NEFL PE=1 SV=3	protein binding; phospholipase binding; structural constituent of cytoskeleton; structural constituent of postsynaptic intermediate filament cytoskeleton; anterograde axonal transport; axonal transport of mitochondrion; intermediate filament organization; intermediate filament polymerization or depolymerization; locomotion; MAPK cascade; microtubule cytoskeleton organization; negative regulation of neuron apoptotic process; neurofilament bundle assembly; neurofilament cytoskeleton organization; neuromuscular process controlling balance; neuron projection morphogenesis; peripheral nervous system axon regeneration; positive regulation of axonogenesis; protein polymerization; regulation of axon diameter; regulation of NMDA receptor activity; response to acrylamide; response to corticosterone; response to peptide hormone; response to sodium arsenite; response to toxic substance; retrograde axonal transport; spinal cord development; synapse maturation	0,001	2300996,1	4732646,2	7321839,6	1986276,0	3747057,2	1,04		
sp Q12860 CNTN1_HUMAN	Contactin-1 OS=Homo sapiens OX=9606 GN=CNTN1 PE=1 SV=1	carbohydrate binding; cell adhesion; cerebellum development; neuron projection development; positive regulation of sodium ion transport	0,000	97306,6	202911,5	165656,2	106492,8	209852,8	1,06		
sp Q9UDT6 CLIP2_HUMAN	CAP-Gly domain-containing linker protein 2 OS=Homo sapiens OX=9606 GN=CLIP2 PE=1 SV=1	cytoplasmic microtubule organization	0,002	27022,7	56784,0	49352,5	16534,7	56345,1	1,07		
sp P78310 CXAR_HUMAN	Coxsackievirus and adenovirus receptor OS=Homo sapiens OX=9606 GN=CXADR PE=1 SV=1	cell adhesion molecule binding; beta-catenin binding; actin cytoskeleton reorganization; AV node cell-bundle of His cell adhesion involved in cell communication; AV node cell to bundle of His cell communication; cardiac muscle fiber development; cell-cell junction organization; heart development; heterophilic cell-cell adhesion via plasma membrane cell adhesion molecules; homotypic cell-cell adhesion; regulation of AV node cell action potential; mitochondrion organization	0,000	121187,0	255101,9	188907,3	123193,0	249588,0	1,07		
sp Q9Y6R0 NUMBL_HUMAN	Numb-like protein OS=Homo sapiens OX=9606 GN=NUMBL PE=1 SV=1	adherens junction organization; axonogenesis; cytokine-mediated signaling pathway; lateral ventricle development; nervous system development; positive regulation of neurogenesis	0,003	9418,8	20118,9	17084,1	7112,2	20772,1	1,09		

sp P31146 COR1A_HUMAN	Coronin-1A OS=Homo sapiens OX=9606 GN=CORO1A PE=1 SV=4	actin binding; actin filament binding; actin monomer binding; cytoskeletal protein binding; myosin heavy chain binding; actin cytoskeleton organization; actin filament organization; calcium ion transport; cell-substrate adhesion; negative regulation of neuron apoptotic process; negative regulation of vesicle fusion	0,000	49341,0	107414,9	92671,3	39579,1	119923,2	1,12
sp Q93045-2 STMN2_HUMAN	Isoform 2 of Stathmin-2 OS=Homo sapiens OX=9606 GN=STMN2	calcium-dependent protein binding; tubulin binding; cellular response to nerve growth factor stimulus; microtubule depolymerization; negative regulation of microtubule depolymerization; negative regulation of microtubule polymerization; negative regulation of neuron projection development; neuron projection development; positive regulation of microtubule depolymerization; positive regulation of neuron projection development; regulation of microtubule polymerization or depolymerization	0,000	19646,8	43423,5	23141,8	19731,8	41561,3	1,14
sp Q9H4G0 E41L1_HUMAN	Band 4.1-like protein 1 OS=Homo sapiens OX=9606 GN=EPB41L1 PE=1 SV=2	actin binding; structural molecule activity; actomyosin structure organization; actin cytoskeleton organization	0,000	37100,4	84858,3	57427,8	35669,8	76778,8	1,19
sp Q99719 SEPT5_HUMAN	Septin-5 OS=Homo sapiens OX=9606 GN=SEPTIN5 PE=1 SV=1	GTPase activity; structure molecule activity; adult behavior; cellular protein localization; cytoskeleton-dependent cytokinesis; regulation of exocytosis; regulation of synaptic vesicle exocytosis; social behavior; synaptic vesicle targeting	0,000	57280,1	131721,2	110599,9	58600,3	129382,9	1,20
sp Q6UXK2 ISLR2_HUMAN	Immunoglobulin superfamily containing leucine-rich repeat protein 2 OS=Homo sapiens OX=9606 GN=ISLR2 PE=2 SV=1	positive regulation of axon extension	0,000	43179,1	100245,9	61334,5	49339,3	94547,6	1,22
sp P10636-8 TAU_HUMAN	Isoform Tau-F of Microtubule-associated protein tau OS=Homo sapiens OX=9606 GN=MAPT	actin binding; DNA binding; enzyme binding; RNA binding; microtubule binding; axonal transport; axonal transport of mitochondrion; axon development; cell-cell signaling; central nervous system neuron development; cytoplasmic microtubule organization; generation of neurons; intracellular distribution of mitochondria; microtubule cytoskeleton organization; neuron projection development; positive regulation of axon extension; positive regulation of microtubule polymerization; positive regulation of neuron death; positive regulation of protein localization to synapse; positive regulation of superoxide anion generation; regulation of long-term synaptic depression; regulation of microtubule cytoskeleton organization; regulation of microtubule polymerization; regulation of mitochondrial fission; regulation of synaptic plasticity; synapse organization	0,000	397463,6	923887,1	660284,3	297999,9	800202,7	1,22
sp Q06481-4 APLP2_HUMAN	Isoform 4 of Amyloid-like protein 2 OS=Homo sapiens OX=9606 GN=APLP2	DNA binding; protein binding; cellular protein metabolic process; G protein-coupled receptor signaling pathway; post-translational protein modification	0,005	5299,8	12366,1	11731,2	3580,6	12767,6	1,22
sp Q9HCM2 PLXA4_HUMAN	Plexin-A4 OS=Homo sapiens OX=9606 GN=PLXNA4 PE=1 SV=4	semaphorin receptor activity; negative regulation of cell adhesion; positive regulation of axonogenesis; regulation of axon extension involved in axon guidance; regulation of cell migration; regulation of cell shape; semaphorin-plexin signaling pathway involved in axon guidance	0,001	25731,0	62063,4	64963,2	29293,1	48946,9	1,27
sp Q13740 CD166_HUMAN	CD166 antigen OS=Homo sapiens OX=9606 GN=ALCAM PE=1 SV=2	protein binding; axon extension involved in axon guidance; cell adhesion; heterophilic cell-cell adhesion via plasma membrane cell adhesion molecules; neuron projection extension; signal transduction	0,006	53443,3	144044,7	237315,0	53646,2	96957,5	1,43
sp O15394 NCAM2_HUMAN	Neural cell adhesion molecule 2 OS=Homo sapiens OX=9606 GN=NCAM2 PE=1 SV=2	identical protein binding; axonal fasciculation; neuron cell-cell adhesion	0,001	27533,9	85653,3	67221,9	25818,9	69555,9	1,64
sp O00203-3 AP3B1_HUMAN	Isoform 2 of AP-3 complex subunit beta-1 OS=Homo sapiens OX=9606 GN=AP3B1	GTP-dependent protein binding; anterograde axonal transport; anterograde synaptic vesicle transport; establishment of protein localization to mitochondrial membrane involved in mitochondrial fission; mRNA transcription by RNA polymerase II	0,002	54492,8	188646,5	233981,0	36883,8	173241,8	1,79
sp Q86YM7 HOME1_HUMAN	Homer protein homolog 1 OS=Homo sapiens OX=9606 GN=HOMER1 PE=1 SV=2	G protein-coupled glutamate receptor binding; ion channel binding; positive regulation of calcium ion transport; positive regulation of signal transduction; regulation of dendritic spine maintenance; regulation of postsynaptic neurotransmitter receptor activity; regulation of store-operated calcium entry; regulation of synaptic transmission, glutamatergic; response to calcium ion; skeletal muscle contraction; skeletal muscle fiber development	0,002	9597,6	19589,1	13986,9	10032,5	19667,5	1,03
sp P13987 CD59_HUMAN	CD59 glycoprotein OS=Homo sapiens OX=9606 GN=CD59 PE=1 SV=1	complement binding; cell surface receptor signaling pathway; endoplasmic reticulum to Golgi vesicle-mediated transport	0,000	20512,1	42075,7	43373,9	14890,7	40261,9	1,04
sp Q43854 EDIL3_HUMAN	EGF-like repeat and discoilin I-like domain-containing protein 3 OS=Homo sapiens OX=9606 GN=EDIL3 PE=1 SV=1	calcium ion binding; integrin binding; cell adhesion; multicellular organism development; positive regulation of cell-substrate adhesion	0,000	288180,5	599921,7	502832,4	291210,4	572882,1	1,06
sp Q99567 NUP88_HUMAN	Nuclear pore complex protein Nup88 OS=Homo sapiens OX=9606 GN=NUP88 PE=1 SV=2	structural constituent of nuclear pore; transporter activity; regulation of gene silencing by miRNA; mRNA export from nucleus; protein import into nucleus; ribosomal large subunit export from nucleus; ribosomal small subunit export from nucleus	0,008	1249,6	2956,2	3302,8	727,1	3158,5	1,24
sp P60880 SNP25_HUMAN	Synaptosomal-associated protein 25 OS=Homo sapiens OX=9606 GN=SNAP25 PE=1 SV=1	voltage-gated potassium channel activity; calcium-dependent protein binding; exocytic insertion of neurotransmitter receptor to postsynaptic membrane; locomotory behavior; long-term synaptic potentiation; neurotransmitter receptor internalization; neurotransmitter secretion; neurotransmitter transport; neurotransmitter uptake; regulation of neuron projection development; synaptic vesicle exocytosis; synaptic vesicle fusion to presynaptic active zone membrane	0,000	71406,0	154173,7	105881,6	68096,2	139767,8	1,11
sp Q13555-6 KCC2G_HUMAN	Isoform 6 of Calcium/calmodulin-dependent protein kinase type II subunit gamma OS=Homo sapiens OX=9606 GN=CAMK2G	nervous system development; regulation of calcium ion transport; regulation of neuron projection development; regulation of skeletal muscle adaptation	0,003	9449,8	20862,8	24381,5	9497,4	21467,5	1,14
sp P05937 CALB1_HUMAN	Calbindin OS=Homo sapiens OX=9606 GN=CALB1 PE=1 SV=2	calcium ion binding involved in regulation of pre- and post-synaptic cytosolic calcium ion concentration; locomotory behavior; long-term memory; regulation of cytosolic calcium ion concentration; regulation of long-term synaptic potentiation	0,007	28268,5	66681,0	55406,1	17414,9	71872,2	1,24
sp Q06064-1 AP180_HUMAN	Clathrin coat assembly protein AP180 OS=Homo sapiens OX=9606 GN=SNAP91 PE=1 SV=2	1-phosphatidylinositol binding; clathrin heavy chain binding; clathrin coat assembly; clathrin-dependent endocytosis; protein transport; regulation of clathrin-dependent endocytosis; synaptic vesicle budding from presynaptic endocytic zone membrane; vesicle budding from membrane	0,003	21985,7	52741,6	55052,5	19703,7	48265,5	1,26
sp P21579 SYT1_HUMAN	Synaptotagmin-1 OS=Homo sapiens OX=9606 GN=SYT1 PE=1 SV=1	calcium-dependent protein binding; calcium ion binding; calcium ion sensor activity; calmodulin binding; clathrin binding; brain development; calcium-dependent activation of synaptic vesicle fusion; calcium-ion regulated exocytosis; calcium ion-regulated exocytosis of neurotransmitter; chemical synaptic transmission; positive regulation of dendrite extension; positive regulation of synaptic transmission; regulation of synaptic transmission, glutamatergic; spontaneous neurotransmitter secretion; synaptic vesicle endocytosis; vesicle-mediated transport	0,000	85024,8	222114,8	146817,8	80410,7	210481,1	1,39
sp Q43761 SNG3_HUMAN	Synaptogyrin-3 OS=Homo sapiens OX=9606 GN=SYNGR3 PE=1 SV=2	positive regulation of transporter activity; regulated exocytosis	0,000	752,7	2269,3	2530,7	800,3	2115,4	1,59

sp Q43581-4 SYT7_HUMAN	Isoform 4 of Synaptotagmin-7 OS=Homo sapiens OX=9606 GN=SYT7	calcium-dependent phospholipid binding; calcium ion binding; calmodulin binding; clathrin binding; calcium ion-regulated exocytosis of neurotransmitter; calcium ion regulated lysosome exocytosis; plasma membrane repair; synaptic vesicle recycling; vesicle-mediated transport	0,002	1668,9	5949,6	4496,1	1472,3	6035,5	1,83
DOWNREGULATED									
PROTEIN	GROUP AND RELATED GENE (GN)	GO: BIOLOGICAL FUNCTION	P-value	Protein Mean Peak Intensity					Log ₂ FC FRDA/ISO CT
				Mean ISO CT	Mean FRDA	Mean CT	Median ISO CT	Median FRDA	
sp Q9UPT8 ZC3H4_HUMAN	Zinc finger CCCH domain-containing protein 4 OS=Homo sapiens OX=9606 GN=ZC3H4 PE=1 SV=3	DNA-binding transcription factor activity, RNA polymerase II-specific; metal ion binding; RNA binding	0,000	7219,2	1992,1	1752,3	9209,7	1779,8	-1,86
sp Q96EK4 THA11_HUMAN	THAP domain-containing protein 11 OS=Homo sapiens OX=9606 GN=THA11 PE=1 SV=2	regulation of transcription by RNA polymerase II; DNA binding; DNA-binding transcription factor activity, RNA polymerase II-specific; DNA-binding transcription repressor activity, RNA polymerase II-specific; RNA polymerase II cis-regulatory region sequence-specific DNA binding; zinc ion binding	0,001	6104,8	2096,7	3061,8	5267,6	1601,4	-1,54
sp Q12849 GRSF1_HUMAN	G-rich sequence factor 1 OS=Homo sapiens OX=9606 GN=GRSF1 PE=1 SV=3	mRNA binding; anterior/posterior pattern specification; morphogenesis of embryonic epithelium; mRNA polyadenylation; positive regulation of mitochondrial RNA catabolic process; regulation of RNA splicing; tRNA processing	0,002	3080,5	1266,3	1091,3	2875,0	948,6	-1,28
sp Q95347 SMC2_HUMAN	Structural maintenance of chromosomes protein 2 OS=Homo sapiens OX=9606 GN=SMC2 PE=1 SV=2	chromatin binding; single-stranded DNA binding; ATP binding; ATPase activity; cell division; kinetochore organization; meiotic chromosome condensation; meiotic chromosome segregation; mitotic chromosome condensation	0,005	33131,1	13658,1	16917,0	20526,9	12703,2	-1,28
sp Q9UPN6 SCAF8_HUMAN	SR-related and CTD-associated factor 8 OS=Homo sapiens OX=9606 GN=SCAF8 PE=1 SV=1	mRNA polyadenylation; negative regulation of termination of RNA polymerase II transcription, poly(A)-coupled; positive regulation of DNA-templated transcription, elongation; termination of RNA polymerase II transcription	0,005	6800,5	2912,6	4955,3	5924,8	2149,4	-1,22
sp Q12996 CSTF3_HUMAN	Cleavage stimulation factor subunit 3 OS=Homo sapiens OX=9606 GN=CSTF3 PE=1 SV=1	mRNA binding; mRNA splicing; mRNA cleavage	0,000	33377,1	14846,0	14948,7	38310,5	13344,9	-1,17
sp Q9BZ95-4 NSD3_HUMAN	Isoform 4 of Histone-lysine N-methyltransferase NSD3 OS=Homo sapiens OX=9606 GN=NSD3	histone methylation; regulation of transcription	0,007	5253,3	2477,1	1777,6	6421,0	2959,2	-1,08
sp Q04206 TF65_HUMAN	Transcription factor p65 OS=Homo sapiens OX=9606 GN=RELA PE=1 SV=2	regulation of transcription; regulation of apoptotic process; regulation of protein catabolic process; response to muscle stretch	0,005	5156,7	2460,3	2291,6	5285,2	2055,6	-1,07
sp P84243 H33_HUMAN	Histone H3.3 OS=Homo sapiens OX=9606 GN=H3F3A PE=1 SV=2	regulation of gene expression; nucleosome assembly; regulation of gene silencing by miRNA; muscle cell differentiation; negative regulation of chromosome condensation	0,009	594098,9	287200,1	214567,1	697003,4	312905,9	-1,05
sp Q9C005 DPY30_HUMAN	Protein dpy-30 homolog OS=Homo sapiens OX=9606 GN=DPY30 PE=1 SV=1	chromatin silencing at telomere; endosomal transport; histone H3-K4 methylation	0,000	34138,8	17047,7	20985,8	36409,2	18642,1	-1,00
sp P32521 RFC1_HUMAN	Replication factor C subunit 1 OS=Homo sapiens OX=9606 GN=RFC1 PE=1 SV=4	base-excision repair, gap-filling; DNA clamp unloading; DNA damage response, detection of DNA damage; DNA-dependent DNA replication	0,009	5027,6	2286,0	1961,6	5306,0	1794,9	-1,14
sp Q9BPX3 CND3_HUMAN	Condensin complex subunit 3 OS=Homo sapiens OX=9606 GN=NCAPG PE=1 SV=1	cell division; mitotic chromosome condensation	0,008	25872,6	12065,7	5220,9	26157,9	10559,9	-1,10
sp Q9H773 DCTP1_HUMAN	dCTP pyrophosphatase 1 OS=Homo sapiens OX=9606 GN=DCTPP1 PE=1 SV=1	dCTP catabolic process; DNA protection; nucleoside triphosphate catabolic process	0,000	26644,2	12760,9	13429,2	23715,3	13443,4	-1,06
sp P24941 CDK2_HUMAN	Cyclin-dependent kinase 2 OS=Homo sapiens OX=9606 GN=CDK2 PE=1 SV=2	cell division; DNA repair; DNA replication; regulation of gene expression; histone phosphorylation; protein phosphorylation; signal transduction	0,005	7986,1	3903,5	5040,0	7434,9	2737,7	-1,03
sp Q9H9T3 ELP3_HUMAN	Elongator complex protein 3 OS=Homo sapiens OX=9606 GN=ELP3 PE=1 SV=2	4 iron, 4 sulfur cluster binding; acetyltransferase activity; metal ion binding; N-acetyltransferase activity; phosphorylase kinase regulator activity; tRNA binding; central nervous system development; neuron migration; positive regulation of cell migration; regulation of transcription by RNA polymerase II; tRNA uridine modification	0,001	12866,3	5387,1	10526,5	13666,2	5514,9	-1,26
sp Q96PU8 QKI_HUMAN	Protein quaking OS=Homo sapiens OX=9606 GN=QKI PE=1 SV=1	long-chain fatty acid biosynthetic process; mRNA processing; mRNA transport; muscle cell differentiation; myelination; positive regulation of gene expression; regulation of mRNA splicing, via spliceosome; regulation of translation; RNA splicing	0,008	13689,2	5991,9	6472,7	16831,0	4707,9	-1,19
sp P56545 CTBP2_HUMAN	C-terminal-binding protein 2 OS=Homo sapiens OX=9606 GN=CTBP2 PE=1 SV=1	chromatin binding; identical protein binding; NAD binding; oxidoreductase activity; negative regulation of transcription by RNA polymerase II; positive regulation of chromatin binding; synaptic vesicle docking	0,006	25731,1	11960,5	12653,4	28305,8	10288,4	-1,11
sp P78417 GSTO1_HUMAN	Glutathione S-transferase omega-1 OS=Homo sapiens OX=9606 GN=GSTO1 PE=1 SV=2	glutathione dehydrogenase (ascorbate) activity; glutathione transferase activity; methylarsenate reductase activity; oxidoreductase activity cellular response to arsenic-containing substance; glutathione derivative biosynthetic process; glutathione metabolic process; interleukin-12-mediated signaling pathway; L-ascorbic acid metabolic process; methylation; negative regulation of ryanodine-sensitive calcium-release channel activity; positive regulation of ryanodine-sensitive calcium-release channel activity; positive regulation of skeletal muscle contraction by regulation of release of sequestered calcium ion; regulation of cardiac muscle contraction by regulation of the release of sequestered calcium ion; regulation of release of sequestered calcium ion into cytosol by sarcoplasmic reticulum; xenobiotic catabolic process	0,000	41800,8	12811,0	54244,4	52632,9	11358,4	-1,71
sp Q5EB52 MEST_HUMAN	Mesoderm-specific transcript homolog protein OS=Homo sapiens OX=9606 GN=MEST PE=2 SV=2	hydrolase activity; regulation of lipid storage	0,000	17592,1	5417,0	9789,7	14071,4	5591,8	-1,70

sp Q7Z4H3 HDDC2_HUMAN	HD domain-containing protein 2 OS=Homo sapiens OX=9606 GN=HDDC2 PE=1 SV=1	5'-deoxynucleotidase activity; metal ion binding	0,006	16916,4	5552,1	2785,5	13045,3	3963,6	-1,61
sp O75976 CBPD_HUMAN	Carboxypeptidase D OS=Homo sapiens OX=9606 GN=CPD PE=1 SV=2	metallocarboxypeptidase activity; serine-type carboxypeptidase activity; zinc ion binding; peptide metabolic process; protein processing	0,000	59589,1	20881,7	37782,7	71819,7	17344,6	-1,51
sp Q01628 IFM3_HUMAN	Interferon-induced transmembrane protein 3 OS=Homo sapiens OX=9606 GN=IFITM3 PE=1 SV=2	defense response to virus; immune response; negative regulation of viral entry into host cell; negative regulation of viral genome replication; negative regulation of viral transcription; response to interferon-alpha; response to interferon-beta; response to interferon-gamma; response to virus; type I interferon signaling pathway	0,007	6132,4	2249,2	6776,0	4602,2	1148,2	-1,45
sp P00338 LDHA_HUMAN	L-lactate dehydrogenase A chain OS=Homo sapiens OX=9606 GN=LDHA PE=1 SV=2	L-lactate dehydrogenase activity; carbohydrate metabolic process; carboxylic acid metabolic process	0,001	568446,7	221896,9	728496,5	356305,2	225622,6	-1,36
sp Q9NPH2 INO1_HUMAN	Inositol-3-phosphate synthase 1 OS=Homo sapiens OX=9606 GN=ISYNA1 PE=1 SV=1	inositol biosynthetic process; phospholipid biosynthetic process	0,001	128507,3	50811,9	65935,4	157117,1	48164,8	-1,34
sp Q9NPA0 EMC7_HUMAN	ER membrane protein complex subunit 7 OS=Homo sapiens OX=9606 GN=EMC7 PE=1 SV=1	carbohydrate binding; protein insertion into ER membrane by stop-transfer membrane-anchor sequence; tail-anchored membrane protein insertion into ER membrane; part of the endoplasmic reticulum membrane protein complex (EMC) that enables the energy-independent insertion into endoplasmic reticulum membranes of newly synthesized membrane proteins	0,008	59332,4	23769,4	98207,5	43973,7	27504,3	-1,32
sp Q8TF05-2 PP4R1_HUMAN	Isoform 2 of Serine/threonine-protein phosphatase 4 regulatory subunit 1 OS=Homo sapiens OX=9606 GN=PPP4R1	protein dephosphorylation; protein phosphorylation; signal transduction	0,002	5013,9	2202,2	772,1	5676,0	1692,9	-1,19
sp P10415 BCL2_HUMAN	Apoptosis regulator Bcl-2 OS=Homo sapiens OX=9606 GN=BCL2 PE=1 SV=2	BH3 domain binding; channel activity; channel inhibitor activity; identical protein binding; protease binding; actin filament organization; apoptotic process; axonogenesis; axon regeneration; cell aging; cell-cell adhesion; cellular response to DNA damage stimulus; cellular response to glucose starvation; cellular response to hypoxia; extrinsic apoptotic signaling pathway in absence of ligand; extrinsic apoptotic signaling pathway via death domain receptors; focal adhesion assembly; negative regulation of apoptotic process ; negative regulation of apoptotic signaling pathway; negative regulation of autophagy; negative regulation of calcium ion transport into cytosol; negative regulation of cell growth; negative regulation of cell migration; negative regulation of mitochondrial depolarization; negative regulation of neuron apoptotic process; positive regulation of neuron maturation; reactive oxygen species metabolic process; regulation of calcium ion transport; regulation of cell-matrix adhesion; regulation of gene expression; regulation of glycoprotein biosynthetic process; regulation of mitochondrial membrane permeability; regulation of mitochondrial membrane potential	0,003	4949,0	2180,9	2347,1	6176,9	1731,0	-1,18
sp Q15118 PDK1_HUMAN	[Pyruvate dehydrogenase (acetyl-transferring)] kinase isozyme 1, mitochondrial OS=Homo sapiens OX=9606 GN=PDK1 PE=1 SV=1	3-phosphoinositide-dependent protein kinase activity; apoptotic process; cell differentiation; cell surface receptor signaling pathway; positive regulation of cell growth; synaptic growth at neuromuscular junction	0,009	17280,9	8028,8	9184,7	17490,6	7620,5	-1,11
sp Q06323 PSME1_HUMAN	Proteasome activator complex subunit 1 OS=Homo sapiens OX=9606 GN=PSME1 PE=1 SV=1	anaphase-promoting complex-dependent catabolic process; antigen processing; positive regulation of endopeptidase activity; positive regulation of canonical Wnt signaling pathway; regulation of mitotic cell cycle phase transition; regulation of mRNA stability; regulation of proteasomal protein catabolic process; transmembrane transport	0,000	52119,8	24295,1	38792,3	51167,0	24383,3	-1,10
sp O00483 NDUFA4_HUMAN	Cytochrome c oxidase subunit NDUFA4 OS=Homo sapiens OX=9606 GN=NDUFA4 PE=1 SV=1	mitochondrial electron transport, cytochrome c to oxygen; mitochondrial electron transport, NADH to ubiquinone; positive regulation of cytochrome-c oxidase activity; proton transmembrane transport	0,000	71796,7	33872,1	59137,1	68944,8	31637,4	-1,08
sp Q8NFI3 ENASE_HUMAN	Cytosolic endo-beta-N-acetylglucosaminidase OS=Homo sapiens OX=9606 GN=ENGASE PE=1 SV=1	hydrolase activity, hydrolyzing O-glycosyl compounds; protein deglycosylation	0,007	3286,7	1640,6	3219,5	3014,1	1280,7	-1,00
sp Q15063-5 POSTN_HUMAN	Isoform 5 of Periostin OS=Homo sapiens OX=9606 GN=POSTN	cell adhesion molecule binding; heparin binding; metal ion binding; bone regeneration; cell adhesion; cellular response to fibroblast growth factor stimulus; cellular response to transforming growth factor beta; cellular response to tumor necrosis factor; cellular response to vitamin K; extracellular matrix organization; negative regulation of cell-matrix adhesion; negative regulation of substrate adhesion-dependent cell spreading; neuron projection extension; positive regulation of chemokine (C-X-C motif) ligand 2 production; positive regulation of smooth muscle cell migration; regulation of Notch signaling pathway; regulation of systemic arterial blood pressure; response to hypoxia; response to mechanical stimulus; response to muscle activity; tissue development; wound healing	0,001	83386,8	25267,4	52190,0	99875,1	18971,6	-1,72
sp P13797 PLST_HUMAN	Plastin-3 OS=Homo sapiens OX=9606 GN=PLS3 PE=1 SV=4	actin filament binding; calcium ion binding actin filament bundle assembly; actin filament network formation; bone development	0,000	150688,1	51227,6	128506,5	181379,2	48804,9	-1,56
sp Q9Y490 TLN1_HUMAN	Talin-1 OS=Homo sapiens OX=9606 GN=TLN1 PE=1 SV=3	actin filament binding; cadherin binding; integrin binding; LIM domain binding; phosphatidylinositol binding; phosphatidylserine binding; structural constituent of cytoskeleton; vinculin binding; cell-cell adhesion; cell-cell junction assembly; cell-substrate junction assembly; cortical actin cytoskeleton organization; integrin activation; integrin-mediated signaling pathway; IRE1-mediated unfolded protein response; muscle contraction; platelet aggregation; platelet degranulation; viral process	0,001	37101,1	12961,9	18862,4	49559,7	13137,3	-1,52
sp P20810-6 ICAL_HUMAN	Isoform 6 of Calpastatin OS=Homo sapiens OX=9606 GN=CAST	cadherin binding; calcium-dependent cysteine-type endopeptidase inhibitor activity; endopeptidase inhibitor activity; RNA binding inhibition of cysteine-type endopeptidase activity; negative regulation of type B; pancreatic cell apoptotic process; presynaptic active zone organization	0,002	4798,6	1722,6	6050,0	4572,8	860,0	-1,48
sp Q05682-5 CALD1_HUMAN	Isoform 5 of Caldesmon OS=Homo sapiens OX=9606 GN=CALD1	actin binding; cadherin binding; calmodulin binding; myosin binding; tropomyosin binding; actin filament bundle assembly; angiogenesis; muscle contraction	0,001	29765,1	13931,4	31728,9	28676,5	12843,1	-1,10
sp Q9BVC6 TM109_HUMAN	Transmembrane protein 109 OS=Homo sapiens OX=9606 GN=TMEM109 PE=1 SV=1	voltage-gated ion channel activity; intrinsic apoptotic signaling pathway in response to DNA damage by p53 class mediator; negative regulation of cell death; regulation of ion transmembrane transport	0,001	57317,4	26796,1	21339,2	59240,8	25369,8	-1,10

DIFFERENTIALLY EXPRESSED PROTEINS – ISO CT vs CT

UPREGULATED

PROTEIN	GROUP AND RELATED GENE (GN)	GO: BIOLOGICAL FUNCTION	P-value	Protein Mean Peak Intensity					Log ₂ FC ISO CT/CT
				Mean CT	Mean ISO CT	Mean FRDA	Median CT	Median ISO CT	
sp Q9Y3B4 SF3B6_HUMAN	Splicing factor 3B subunit 6 OS=Homo sapiens OX=9606 GN=SF3B6 PE=1 SV=1	mRNA binding; RNA binding; mRNA splicing, via spliceosome	0,009	16933,8	34646,0	31681,3	16717,6	39571,4	1,03
sp Q14119 VEZF1_HUMAN	Vascular endothelial zinc finger 1 OS=Homo sapiens OX=9606 GN=VEZF1 PE=1 SV=2	DNA-binding transcription activator activity, RNA polymerase II-specific; cellular defense response; endothelial cell development	0,007	1866,0	3818,9	3181,7	1519,7	3928,4	1,03
sp Q12996 CSTF3_HUMAN	Cleavage stimulation factor subunit 3 OS=Homo sapiens OX=9606 GN=CSTF3 PE=1 SV=1	mRNA binding; mRNA splicing; mRNA cleavage	0,004	14948,7	33377,1	14846,0	15526,2	38310,5	1,16
sp P46782 RS5_HUMAN	40S ribosomal protein S5 OS=Homo sapiens OX=9606 GN=RPS5 PE=1 SV=4	mRNA binding; structural constituent of ribosome; cytoplasmic translation; nuclear-transcribed mRNA catabolic process, nonsense-mediated decay	0,009	115082,3	260217,7	195136,9	125754,9	285223,7	1,18
sp Q9Y333 LSM2_HUMAN	U6 snRNA-associated Sm-like protein LSm2 OS=Homo sapiens OX=9606 GN=LSM2 PE=1 SV=1	RNA binding; small GTPase binding; U6 snRNA binding; exonucleolytic catabolism of deadenylated mRNA; mRNA catabolic process; mRNA splicing, via spliceosome; spliceosomal tri-snRNP complex assembly	0,000	16206,8	37123,1	21222,2	14042,3	34627,0	1,20
sp Q13895 BYST_HUMAN	Bystin OS=Homo sapiens OX=9606 GN=BYSL PE=1 SV=3	RNA binding; snoRNA binding; maturation of SSU-rRNA from tricistronic rRNA transcript (SSU-rRNA, 5.8S rRNA, LSU-rRNA); regulation of protein localization to nucleolus; ribosome biogenesis; rRNA processing	0,010	3741,0	10966,5	6589,5	3379,0	11715,0	1,55
sp P55209 NP1L1_HUMAN	Nucleosome assembly protein 1-like 1 OS=Homo sapiens OX=9606 GN=NAP1L1 PE=1 SV=1	chromatin binding; histone binding; RNA binding; DNA replication; nucleosome assembly; positive regulation of cell population proliferation; positive regulation of neural precursor cell proliferation; positive regulation of neurogenesis	0,002	87163,6	288853,7	168816,9	91911,8	285655,0	1,73
sp Q9H0U6 RM18_HUMAN	39S ribosomal protein L18, mitochondrial OS=Homo sapiens OX=9606 GN=MRPL18 PE=1 SV=1	structural constituent of ribosome; mitochondrial translational elongation; mitochondrial translational termination; rRNA import into mitochondrion	0,003	5059,9	17288,0	12073,3	4338,3	16813,5	1,77
sp Q9UPT8 ZC3H4_HUMAN	Zinc finger CCCH domain-containing protein 4 OS=Homo sapiens OX=9606 GN=ZC3H4 PE=1 SV=3	DNA-binding transcription factor activity, RNA polymerase II-specific; metal ion binding; RNA binding	0,003	1752,3	7219,2	1992,1	1184,0	9209,7	2,04
sp Q9BPX3 CND3_HUMAN	Condensin complex subunit 3 OS=Homo sapiens OX=9606 GN=NCAPG PE=1 SV=1	cell division; mitotic chromosome condensation	0,010	5220,9	25872,6	12065,7	6048,9	26157,9	2,31
sp Q6PD62 CTR9_HUMAN	RNA polymerase-associated protein CTR9 homolog OS=Homo sapiens OX=9606 GN=CTR9 PE=1 SV=1	RNA polymerase II complex binding; SH2 domain binding; histone H2B ubiquitination; histone H3-K4 trimethylation; histone monoubiquitination; negative regulation of mRNA polyadenylation; negative regulation of transcription by RNA polymerase II; positive regulation of histone H2B ubiquitination; positive regulation of histone H3-K4 methylation; positive regulation of histone H3-K79 methylation; regulation of transcription, DNA-templated; stem cell population maintenance; transcription by RNA polymerase II; Wnt signaling pathway	0,006	2959,0	14734,8	7027,5	2451,6	16092,6	2,32
sp Q16540 RM23_HUMAN	39S ribosomal protein L23, mitochondrial OS=Homo sapiens OX=9606 GN=MRPL23 PE=1 SV=1	RNA binding; structural constituent of ribosome; mitochondrial translation; mitochondrial translational elongation; mitochondrial translational termination	0,008	1338,5	9098,9	6535,8	1035,0	6856,3	2,77
sp Q9NQ48-3 LZTL1_HUMAN	Isoform 3 of Leucine zipper transcription factor-like protein 1 OS=Homo sapiens OX=9606 GN=LZTL1	identical protein binding; protein-containing complex binding; cilium assembly; negative regulation of protein localization to ciliary membrane; may play a role in neurite outgrowth	0,003	1836,8	12707,1	7955,7	1863,4	15702,5	2,79
sp P55039 DRG2_HUMAN	Developmentally-regulated GTP-binding protein 2 OS=Homo sapiens OX=9606 GN=DRG2 PE=1 SV=1	GTPase activity; GTP binding; metal ion binding; RNA binding; signal transduction; cytoplasmic translation	0,002	20701,8	41598,2	25918,6	21229,7	38004,0	1,01
sp P42167 LAP2B_HUMAN	Lamina-associated polypeptide 2, isoforms beta/gamma OS=Homo sapiens OX=9606 GN=TMPO PE=1 SV=2	cadherin binding; DNA binding; lamin binding; regulation of transcription, DNA-templated	0,005	63865,6	132618,8	82154,4	69386,3	151657,4	1,05
sp Q86TG7-2 PEG10_HUMAN	Isoform 2 of Retrotransposon-derived protein PEG10 OS=Homo sapiens OX=9606 GN=PEG10	DNA binding; RNA binding; zinc binding; apoptotic process; cell differentiation; negative regulation of transforming growth factor beta receptor signaling pathway	0,009	6148,6	15058,4	8272,1	4703,3	13574,0	1,29
sp P35244 RFA3_HUMAN	Replication protein A 14 kDa subunit OS=Homo sapiens OX=9606 GN=RPA3 PE=1 SV=1	damaged DNA binding; single-stranded DNA binding; base-excision repair; base-excision repair, gap-filling; DNA damage response; detection of DNA damage; DNA replication; DNA replication initiation; double-strand break repair via homologous recombination; regulation of signal transduction by p53 class mediator; telomere maintenance	0,007	19194,8	42012,3	25628,1	19383,0	44564,9	1,13
sp Q9Y5B9 SP16H_HUMAN	FACT complex subunit SPT16 OS=Homo sapiens OX=9606 GN=SUPT16H PE=1 SV=1	nucleosome binding; RNA binding; DNA repair; DNA replication; DNA replication-independent nucleosome organization; nucleosome disassembly; positive regulation of DNA-templated transcription, elongation; positive regulation of transcription elongation from RNA polymerase II promoter; regulation of signal transduction by p53 class mediator	0,008	58300,9	142036,4	84854,9	59834,9	119219,3	1,28
sp A6NHR9 SMHD1_HUMAN	Structural maintenance of chromosomes flexible hinge domain-containing protein 1 OS=Homo sapiens OX=9606 GN=SMCHD1 PE=1 SV=2	ATPase activity; ATP binding; DNA binding; dosage compensation by inactivation of X chromosome; double-strand break repair; heterochromatin organization involved in chromatin silencing; inactivation of X chromosome by DNA methylation; inactivation of X chromosome by heterochromatin assembly; negative regulation of double-strand break repair via homologous recombination; positive regulation of DNA repair; positive regulation of double-strand break repair via nonhomologous end joining	0,005	10049,4	23057,4	13469,3	10017,8	24380,7	1,20
sp P30084 ECHM_HUMAN	Enoyl-CoA hydratase, mitochondrial OS=Homo sapiens OX=9606 GN=ECHS1 PE=1 SV=4	branched-chain amino acid catabolic process; fatty acid beta-oxidation	0,000	142273,3	290415,1	279863,0	142049,3	294716,6	1,03
sp Q709C8-4 VP13C_HUMAN	Isoform 4 of Vacuolar protein sorting-associated protein 13C OS=Homo sapiens OX=9606 GN=VPS13C	Golgi to endosome transport; mitochondrion organization; negative regulation of parkin-mediated stimulation of mitophagy in response to mitochondrial depolarization; necessary for proper mitochondrial function and maintenance of mitochondrial transmembrane potential	0,008	3194,1	6658,3	5478,2	2497,3	6467,4	1,06

sp Q95486 SC24A_HUMAN	Protein transport protein Sec24A OS=Homo sapiens OX=9606 GN=SEC24A PE=1 SV=2	COPII vesicle coating; endoplasmic reticulum to Golgi vesicle-mediated transport; intracellular protein transport	0,008	12196,8	25741,8	21727,3	12893,1	27058,1	1,08
sp P61289-2 PSME3_HUMAN	Isoform 2 of Proteasome activator complex subunit 3 OS=Homo sapiens OX=9606 GN=PSME3	endopeptidase activator activity; anaphase-promoting complex-dependent catabolic process; apoptotic process; regulation of Wnt signaling; positive regulation of endopeptidase activity; post-translational protein modification; pre-replicative complex assembly; proteasome-mediated ubiquitin-dependent protein catabolic process; protein deubiquitination; protein polyubiquitination; regulation of cellular amino acid metabolic process; regulation of G1/S transition of mitotic cell cycle; transmembrane transport	0,000	36081,7	78630,4	60058,4	37450,0	77805,1	1,12
sp Q9Y5Y2 NUBP2_HUMAN	Cytosolic Fe-S cluster assembly factor NUBP2 OS=Homo sapiens OX=9606 GN=NUBP2 PE=1 SV=1	4 iron, 4 sulfur cluster binding; ATPase activity; ATP binding; iron-sulfur cluster binding; metal ion binding; nucleotide binding; cell projection organization; iron-sulfur cluster assembly	0,000	3785,6	8705,8	4910,2	3698,2	9136,3	1,20
sp Q8NFF5 FAD1_HUMAN	FAD synthase OS=Homo sapiens OX=9606 GN=FLAD1 PE=1 SV=1	ATP binding; FMN adenylyltransferase activity; FAD biosynthetic process; riboflavin metabolic process	0,005	6221,1	14397,0	11685,2	7286,2	15276,9	1,21
sp Q5MNZ6 WIPI3_HUMAN	WD repeat domain phosphoinositide-interacting protein 3 OS=Homo sapiens OX=9606 GN=WDR45B PE=1 SV=2	phosphatidylinositol-3,5-bisphosphate binding; phosphatidylinositol-3-phosphate binding; TSC1-TSC2 complex binding; autophagosome assembly; autophagy of mitochondrion; autophagy of nucleus; cellular response to starvation; protein lipidation; protein localization to phagophore assembly site	0,007	4137,0	9678,1	7092,9	3501,1	10986,8	1,23
sp Q9BXW7 HDHD5_HUMAN	Haloacid dehalogenase-like hydrolase domain-containing 5 OS=Homo sapiens OX=9606 GN=HDHD5 PE=1 SV=1	glycerophospholipid biosynthetic process	0,001	11048,8	26605,3	20260,1	10986,2	21798,5	1,27
sp P49915 GUAA_HUMAN	GMP synthase [glutamine-hydrolyzing] OS=Homo sapiens OX=9606 GN=GMPS PE=1 SV=1	GMP synthase activity; ATP binding; glutamine metabolic process	0,002	51049,9	124581,2	66104,5	57323,3	102939,4	1,29
sp P13807-2 GYS1_HUMAN	Isoform 2 of Glycogen [starch] synthase, muscle OS=Homo sapiens OX=9606 GN=GYS1	glycogen biosynthetic process; heart development	0,004	12251,2	29912,4	17632,9	11898,6	29817,2	1,29
sp O15260 SURF4_HUMAN	Surfeit locus protein 4 OS=Homo sapiens OX=9606 GN=SURF4 PE=1 SV=3	Golgi organization; retrograde vesicle-mediated transport, Golgi to endoplasmic reticulum	0,006	46752,8	115458,8	88125,4	49770,1	133919,2	1,30
sp Q9ULS5 TMCC3_HUMAN	Transmembrane and coiled-coil domain protein 3 OS=Homo sapiens OX=9606 GN=TMCC3 PE=1 SV=3	14-3-3 protein binding; identical protein binding (Endoplasmic reticulum)	0,004	2366,2	6022,3	4093,0	1593,9	6012,2	1,35
sp Q9NXE4-3 NSMA3_HUMAN	Isoform 3 of Sphingomyelin phosphodiesterase 4 OS=Homo sapiens OX=9606 GN=SMPD4	metal ion binding; sphingomyelin phosphodiesterase activity; endoplasmic reticulum organization; glycerophospholipid catabolic process	0,006	1828,5	4753,5	3687,2	1615,8	3958,1	1,38
sp O60725 CMT_HUMAN	Protein-S-isoprenylcysteine O-methyltransferase OS=Homo sapiens OX=9606 GN=ICMT PE=1 SV=1	cellular protein modification process; C-terminal protein methylation; MAPK cascade; post-translational protein modification; protein targeting to membrane	0,005	2297,6	5974,0	4904,6	2073,7	6358,6	1,38
sp Q9Y6A9 SPCS1_HUMAN	Signal peptidase complex subunit 1 OS=Homo sapiens OX=9606 GN=SPCS1 PE=1 SV=4	peptidase activity; ribosome binding; protein targeting to ER; proteolysis; signal peptide processing	0,002	3672,7	9710,4	9448,5	1874,7	9472,9	1,40
sp Q6YP21 KAT3_HUMAN	Kynurenine-oxoglutarate transaminase 3 OS=Homo sapiens OX=9606 GN=KYAT3 PE=1 SV=1	2-oxoglutarate metabolic process; biosynthetic process; cellular amino acid metabolic process; kynurenine metabolic process; L-kynurenine catabolic process	0,008	5112,8	14240,7	10108,1	5867,1	15612,3	1,48
sp Q12770-4 SCAP_HUMAN	Isoform 4 of Sterol regulatory element-binding protein cleavage-activating protein OS=Homo sapiens OX=9606 GN=SCAP	sterol binding; unfolded protein binding; aging; cellular lipid metabolic process; cholesterol metabolic process; COPII-coated vesicle cargo loading; regulation of fatty acid biosynthetic process; response to hypoxia; response to insulin; SREBP signaling pathway	0,003	1609,9	4676,5	3651,0	1680,9	5473,3	1,54
sp Q6NUM9 RETSAT_HUMAN	All-trans-retinol 13,14-reductase OS=Homo sapiens OX=9606 GN=RETSAT PE=1 SV=2	saturation of all-trans-retinol to all-trans-13,14-dihydroretinol	0,007	3047,6	9164,2	6747,3	2858,6	10129,3	1,59
sp Q70UQ0 IKIP_HUMAN	Inhibitor of nuclear factor kappa-B kinase-interacting protein OS=Homo sapiens OX=9606 GN=IKBIP PE=1 SV=1		0,008	2866,4	9035,8	4910,2	2727,2	10587,8	1,66
sp O14618 CCS_HUMAN	Copper chaperone for superoxide dismutase OS=Homo sapiens OX=9606 GN=CCS PE=1 SV=1	cadherin binding; copper ion binding; cellular response to oxidative stress; positive regulation of oxidoreductase activity; protein maturation by copper ion transfer; removal of superoxide radicals; superoxide metabolic process	0,003	2894,2	9613,7	5014,0	1340,9	9058,7	1,73
sp P19525 E2AK2_HUMAN	Interferon-induced, double-stranded RNA-activated protein kinase OS=Homo sapiens OX=9606 GN=EIF2AK2 PE=1 SV=2	ATP binding; double-stranded RNA binding; eukaryotic translation initiation factor 2alpha kinase activity; identical protein binding; non-membrane spanning protein tyrosine kinase activity; protein kinase activity; protein serine kinase activity; protein threonine kinase activity; transmembrane receptor protein tyrosine kinase activity; activation of MAPKK activity; cellular response to amino acid starvation; defense response to virus; endoplasmic reticulum unfolded protein response; innate immune response; negative regulation of apoptotic process; negative regulation of osteoblast proliferation; negative regulation of translation; negative regulation of viral genome replication; positive regulation of apoptotic process; positive regulation of chemokine production; positive regulation of cytokine production; positive regulation of NF-kappaB transcription factor activity; positive regulation of NIK/NF-kappaB signaling; positive regulation of stress-activated MAPK cascade; protein autophosphorylation; protein phosphorylation; regulation of hematopoietic progenitor cell differentiation; regulation of hematopoietic stem cell differentiation; regulation of hematopoietic stem cell proliferation; regulation of NLRP3 inflammasome complex assembly; response to interferon-alpha; response to lipopolysaccharide; response to toxic substance; response to virus; response to vitamin E; translation	0,008	3072,5	11814,6	11122,9	2972,5	10882,5	1,94
sp Q6P1X6 CH082_HUMAN	UPF0598 protein C8orf82 OS=Homo sapiens OX=9606 GN=C8orf82 PE=1 SV=2		0,004	1452,9	7950,9	4706,9	977,1	10441,7	2,45
sp Q8TF05-2 PP4R1_HUMAN	Isoform 2 of Serine/threonine-protein phosphatase 4 regulatory subunit 1 OS=Homo sapiens OX=9606 GN=PPP4R1	protein dephosphorylation; protein phosphorylation; signal transduction	0,001	772,1	5013,9	2202,2	554,6	5676,0	2,70

sp Q8NCE2 MTMRE_HUMAN	Myotubularin-related protein 14 OS=Homo sapiens OX=9606 GN=MTMR14 PE=1 SV=2	phosphatidylinositol-3-phosphatase activity; protein tyrosine phosphatase activity; macroautophagy; phosphatidylinositol biosynthetic process	0,001	1190,4	3264,9	2363,0	1063,7	3113,3	1,46
sp O15540-2 FABP7_HUMAN	Isoform 2 of Fatty acid-binding protein, brain OS=Homo sapiens OX=9606 GN=FABP7	lipid binding; epithelial cell proliferation; negative regulation of cell population proliferation; nervous system development; triglyceride catabolic process	0,004	1412,6	3898,4	2191,3	1063,7	4279,7	1,46
sp Q92643 GPI8_HUMAN	GPI-anchor transamidase OS=Homo sapiens OX=9606 GN=PIGK PE=1 SV=2	attachment of GPI anchor to protein; protein localization to cell surface	0,004	7850,3	16234,2	12577,3	7826,5	18328,4	1,05
sp Q8WUW1-2 BRK1_HUMAN	Isoform 2 of Protein BRICK1 OS=Homo sapiens OX=9606 GN=BRK1	protein binding; actin filament organization; cell motility; positive regulation of Arp2/3 complex-mediated actin nucleation; Rac protein signal transduction; regulation of actin polymerization or depolymerization	0,005	16534,1	39471,7	36667,9	17642,2	40625,2	1,26
sp Q5VWJ9 SNX30_HUMAN	Sorting nexin-30 OS=Homo sapiens OX=9606 GN=SNX30 PE=1 SV=1	Phosphatidylinositol binding; protein transport; intracellular trafficking	0,000	6938,5	17019,6	18207,1	7531,6	16649,3	1,29
sp Q96GZ6-9 S41A3_HUMAN	Isoform 9 of Solute carrier family 41 member 3 OS=Homo sapiens OX=9606 GN=SLC41A3	cation transmembrane transporter activity	0,003	2616,0	6919,2	4617,7	2581,4	7174,6	1,40
sp Q14728 MFS10_HUMAN	Major facilitator superfamily domain-containing protein 10 OS=Homo sapiens OX=9606 GN=MFS10 PE=1 SV=1	organic anion transmembrane transporter activity; tetracycline transmembrane transporter activity; apoptotic process; sodium-independent organic anion transport	0,007	5507,8	15339,6	14750,7	6755,2	16007,8	1,48
sp Q9P0S9 TM14C_HUMAN	Transmembrane protein 14C OS=Homo sapiens OX=9606 GN=TMEM14C PE=1 SV=1	erythrocyte differentiation; heme biosynthetic process; mitochondrial transport; regulation of heme biosynthetic process	0,009	2950,1	12698,8	7440,0	3196,0	14938,4	2,11

DOWNREGULATED

PROTEIN	GROUP AND RELATED GENE (GN)	GO: BIOLOGICAL FUNCTION	P-value	Protein Mean Peak Intensity					Log ₂ FC ISO CT/CT
				Mean CT	Mean ISO CT	Mean FRDA	Median CT	Median ISO CT	
sp Q02539 H11_HUMAN	Histone H1.1 OS=Homo sapiens OX=9606 GN=HIST1H1A PE=1 SV=3	chromosome condensation; negative regulation of chromatin silencing; negative regulation of DNA recombination; chromatin binding; double-stranded DNA binding	0,000	162206,4	7956,5	14742,2	152890,7	7701,6	-4,35
sp P40425 PBX2_HUMAN	Pre-B-cell leukemia transcription factor 2 OS=Homo sapiens OX=9606 GN=PBX2 PE=1 SV=2	chromatin binding; DNA-binding transcription activator activity, RNA polymerase II-specific; transcription factor binding; brain development; embryonic limb morphogenesis; embryonic organ development; neuron development; proximal/distal pattern formation; regulation of transcription by RNA polymerase II	0,006	13857,9	6746,6	9320,9	13178,8	5136,5	-1,04
sp Q7Z5J4 RAI1_HUMAN	Retinoic acid-induced protein 1 OS=Homo sapiens OX=9606 GN=RAI1 PE=1 SV=2	metal ion binding; circadian regulation of gene expression; negative regulation of multicellular organism growth; positive regulation of transcription, DNA-templated; regulation of transcription by RNA polymerase II; skeletal system development	0,000	5210,9	1722,1	3389,7	5262,5	1525,6	-1,60
sp Q59GN2 R39L5_HUMAN	Putative 60S ribosomal protein L39-like 5 OS=Homo sapiens OX=9606 GN=RPL39P5 PE=5 SV=2	structural constituent of ribosome; translation	0,009	52098,5	21273,6	29642,0	44741,9	22690,5	-1,29
sp P80723 BASP1_HUMAN	Brain acid soluble protein 1 OS=Homo sapiens OX=9606 GN=BASP1 PE=1 SV=2	protein domain specific binding; transcription corepressor activity;transcription regulatory region sequence-specific DNA binding; mesenchymal to epithelial transition; negative regulation of transcription, DNA-templated; positive regulation of heart growth; substantia nigra development; thorax and anterior abdomen determination; regulate actin dynamics and presynaptic vesicle cycling at axon terminals, thereby facilitating axonal growth, regeneration, and plasticity	0,000	1687939,3	809983,4	1252682,1	1646542,2	743255,5	-1,06
sp Q8N111 CEND_HUMAN	Cell cycle exit and neuronal differentiation protein 1 OS=Homo sapiens OX=9606 GN=CEND1 PE=1 SV=1	adult walking behavior; cerebellar granular layer maturation; cerebellar Purkinje cell differentiation; negative regulation of cerebellar granule cell precursor proliferation; radial glia guided migration of cerebellar granule cell	0,003	43229,6	19533,9	34504,5	42246,7	12176,8	-1,15
sp Q5R115 COX20_HUMAN	Cytochrome c oxidase assembly protein COX20, mitochondrial OS=Homo sapiens OX=9606 GN=COX20 PE=1 SV=2	mitochondrial cytochrome c oxidase assembly	0,010	10517,7	2947,2	10087,9	8609,5	1673,7	-1,84
sp Q8WV17 PPR1C_HUMAN	Protein phosphatase 1 regulatory subunit 1C OS=Homo sapiens OX=9606 GN=PPP1R1C PE=3 SV=1	protein phosphatase inhibitor activity; intracellular signal transduction; cell cycle; cell division	0,001	43879,7	14664,0	39211,4	43812,9	10588,7	-1,58
sp Q15125 EBP_HUMAN	3-beta-hydroxysteroid-Delta(8),Delta(7)-isomerase OS=Homo sapiens OX=9606 GN=EBP PE=1 SV=3	cholesterol biosynthetic process; cholesterol metabolic process; skeletal system development	0,000	13202,2	4857,6	11475,3	12039,3	4735,5	-1,44
sp P56181 NDUV3_HUMAN	NADH dehydrogenase [ubiquinone] flavoprotein 3, mitochondrial OS=Homo sapiens OX=9606 GN=NDUV3 PE=1 SV=2	mitochondrial ATP synthesis coupled electron transport; mitochondrial electron transport, NADH to ubiquinone; mitochondrial respiratory chain complex I assembly	0,001	21572,7	8248,4	16136,6	19767,9	9730,8	-1,39
sp Q13555-6 KCC2G_HUMAN	Isoform 6 of Calcium/calmodulin-dependent protein kinase type II subunit gamma OS=Homo sapiens OX=9606 GN=CAMK2G	nervous system development; regulation of calcium ion transport; regulation of neuron projection development; regulation of skeletal muscle adaptation	0,005	24381,5	9449,8	20862,8	29545,1	9497,4	-1,37
sp P54803 GALC_HUMAN	Galactocerebrosidase OS=Homo sapiens OX=9606 GN=GALC PE=1 SV=3	galactosylceramide catabolic process; glycosphingolipid metabolic process; myelination	0,000	20556,2	8195,8	15326,0	19685,1	6534,5	-1,33
sp P09382 LEG1_HUMAN	Galectin-1 OS=Homo sapiens OX=9606 GN=LGALS1 PE=1 SV=2	carbohydrate binding; identical protein binding; lactose binding; laminin binding; RNA binding apoptotic process; cellular protein metabolic process; cellular response to glucose stimulus; cellular response to organic cyclic compound; myoblast differentiation; negative regulation of cell-substrate adhesion; negative regulation of neuron projection development; plasma cell differentiation; positive regulation of erythrocyte aggregation; positive regulation of I-kappaB kinase/NF-kappaB signaling; positive	0,002	42609,4	17767,9	7614,2	43715,6	11256,9	-1,26

		regulation of viral entry into host cell; post-translational protein modification; regulation of apoptotic process; response to axon injury; response to drug ; response to isolation stress; T cell costimulation								
sp O75781-2 PALM_HUMAN	Isoform 2 of Paralemmin-1 OS=Homo sapiens OX=9606 GN=PALM	adenylate cyclase-inhibiting G protein-coupled receptor signaling pathway; cellular response to electrical stimulus; cytoskeleton organization; synapse maturation	0,000	148105,6	62681,7	124981,8	142596,9	66614,7	-1,24	
sp P45954 ACADSB_HUMAN	Short/branched chain specific acyl-CoA dehydrogenase, mitochondrial OS=Homo sapiens OX=9606 GN=ACADSB PE=1 SV=1	branched-chain amino acid catabolic process; fatty acid metabolic process; isoleucine catabolic process	0,004	1252,2	563,4	873,1	1314,8	405,0	-1,15	
sp O75323 NIPSN2_HUMAN	Protein NipSnap homolog 2 OS=Homo sapiens OX=9606 GN=NIPSNAP2 PE=1 SV=1	mitochondrion organization; oxidative phosphorylation; positive regulation of high voltage-gated calcium channel activity	0,003	8121,5	3755,6	4617,7	7638,6	3625,7	-1,11	
sp O75663 TIPRL_HUMAN	TIP41-like protein OS=Homo sapiens OX=9606 GN=TIPRL PE=1 SV=2	DNA damage checkpoint; negative regulation of phosphoprotein phosphatase activity; regulation of phosphoprotein phosphatase activity; TOR signaling	0,004	63340,2	29390,8	38318,2	57436,2	18339,7	-1,11	
sp Q96GY0 ZC21A_HUMAN	Zinc finger C2HC domain-containing protein 1A OS=Homo sapiens OX=9606 GN=ZC2HC1A PE=1 SV=2	metal ion binding	0,010	105556,2	50530,2	91155,5	127768,2	48731,4	-1,06	
sp O15069 NACAD_HUMAN	NAC-alpha domain-containing protein 1 OS=Homo sapiens OX=9606 GN=NACAD PE=1 SV=3	unfolded protein binding; protein targeting to membrane	0,000	20376,3	9873,1	15387,7	19560,1	11527,6	-1,05	
sp P05413 FABPH_HUMAN	Fatty acid-binding protein, heart OS=Homo sapiens OX=9606 GN=FABP3 PE=1 SV=4	cytoskeletal protein binding; icosatetraenoic acid binding; long-chain fatty acid binding; long-chain fatty acid transporter activity; oleic acid binding brown fat cell differentiation; cholesterol homeostasis; fatty acid metabolic process; intracellular lipid transport; long-chain fatty acid transport; negative regulation of cell population proliferation; phospholipid homeostasis; positive regulation of long-chain fatty acid import into cell; positive regulation of phospholipid biosynthetic process; regulation of fatty acid oxidation; regulation of phosphatidylcholine biosynthetic process; response to drug; response to fatty acid; response to insulin; triglyceride catabolic process	0,001	35322,0	9372,6	11533,1	33570,8	8549,3	-1,91	
sp O75056 SDC3_HUMAN	Syndecan-3 OS=Homo sapiens OX=9606 GN=SDC3 PE=1 SV=2	identical protein binding; cell migration; glycosaminoglycan biosynthetic process; glycosaminoglycan catabolic process; leukocyte migration; retinoid metabolic process; may have a role in the organization of cell shape by affecting the actin cytoskeleton, possibly by transferring signals from the cell surface in a sugar-dependent mechanism	0,003	29250,0	6666,0	11609,7	29752,7	2764,7	-2,13	
sp O75061-4 AUX1_HUMAN	Isoform 4 of Putative tyrosine-protein phosphatase auxilin OS=Homo sapiens OX=9606 GN=DNAJC6	clathrin binding; protein tyrosine phosphatase activity; clathrin coat disassembly; clathrin-dependent endocytosis; membrane organization; post-Golgi vesicle-mediated transport; regulation of clathrin-dependent endocytosis; synaptic vesicle uncoating	0,003	46130,1	13027,2	30196,2	42984,9	9906,2	-1,82	
sp P04271 S100B_HUMAN	Protein S100-B OS=Homo sapiens OX=9606 GN=S100B PE=1 SV=2	Calcium-dependent protein binding; calcium ion binding; identical protein binding; protein homodimerization activity; RAGE receptor binding; S100 protein binding; tau protein binding; zinc ion binding axonogenesis; central nervous system development; innate immune response; learning or memory; positive regulation of cell population proliferation; positive regulation of I-kappaB kinase/NF-kappaB signaling	0,002	4858,3	1452,5	1181,2	5091,4	1199,2	-1,74	
sp P05067-3 A4_HUMAN	Isoform L-APP677 of Amyloid-beta precursor protein OS=Homo sapiens OX=9606 GN=APP	acetylcholine receptor activator activity; chaperone; chemoattractant activity; chromatin binding; DNA binding; enzyme binding; G protein-coupled receptor binding; growth factor receptor binding; heparan sulfate binding; integrin binding; activation of MAPK activity; adult locomotory behavior; axo-dendritic transport; axon midline choice point recognition; axonogenesis; calcium-mediated signaling; cell adhesion; cellular copper ion homeostasis; cellular process; cellular protein metabolic process; cellular response to amyloid-beta; cellular response to cAMP; dendrite development; endocytosis; extracellular matrix organization; locomotory behavior; modulation of excitatory postsynaptic potential; negative regulation of gene expression; negative regulation of long-term synaptic potentiation; negative regulation of mitochondrion organization; negative regulation of neuron death; neuromuscular process controlling balance; neuron projection development; neuron projection maintenance; neuron remodeling; synapse organization; synaptic growth at neuromuscular junction	0,002	79564,4	33268,8	69069,6	71795,5	30510,9	-1,26	
sp P17677 NEUM_HUMAN	Neuromodulin OS=Homo sapiens OX=9606 GN=GAP43 PE=1 SV=1	calmodulin binding; phosphatidylinositol phosphate binding; phosphatidylserine binding; axon choice point recognition; axon regeneration; cell fate commitment; glial cell differentiation; protein kinase C-activating G protein-coupled receptor signaling pathway; regulation of filopodium assembly; regulation of growth; regulation of postsynaptic specialization assembly; response to wounding; tissue regeneration	0,000	1235395,2	546870,5	1034495,6	1253315,8	566231,3	-1,18	
sp Q02952-3 AKA12_HUMAN	Isoform 3 of A-kinase anchor protein 12 OS=Homo sapiens OX=9606 GN=AKAP12	adenylate cyclase binding; calmodulin binding; protein kinase A binding; modulation of chemical synaptic transmission; positive regulation of ERK1 and ERK2 cascade; response to electrical stimulus; response to lipopolysaccharide; positive regulation of oligodendrocyte apoptotic process	0,000	98199,7	47148,5	79823,6	99442,5	45654,0	-1,06	
sp O75351 VPS4B_HUMAN	Vacuolar protein sorting-associated protein 4B OS=Homo sapiens OX=9606 GN=VPS4B PE=1 SV=2	ATPase activity; ATP binding; identical protein binding; microtubule-severing ATPase activity; protein homodimerization activity	0,001	16493,4	8083,9	10462,6	17530,9	7044,3	-1,03	
sp O60636 TSN2_HUMAN	Tetraspanin-2 OS=Homo sapiens OX=9606 GN=TSPAN2 PE=1 SV=2	astrocyte development; axon development; brain development; inflammatory response; microglia development; myelination; oligodendrocyte differentiation	0,009	1806,3	808,5	1340,4	1759,7	759,1	-1,16	
sp Q13740 CD166_HUMAN	CD166 antigen OS=Homo sapiens OX=9606 GN=ALCAM PE=1 SV=2	protein binding; axon extension involved in axon guidance; cell adhesion; heterophilic cell-cell adhesion via plasma membrane cell adhesion molecules; neuron projection extension; signal transduction	0,009	237315,0	53443,3	144044,7	228995,8	53646,2	-2,15	
sp O00203-3 AP3B1_HUMAN	Isoform 2 of AP-3 complex subunit beta-1 OS=Homo sapiens OX=9606 GN=AP3B1	GTP-dependent protein binding; anterograde axonal transport; anterograde synaptic vesicle transport; establishment of protein localization to mitochondrial membrane involved in mitochondrial fission; mRNA transcription by RNA polymerase II	0,000	233981,0	54492,8	188646,5	228019,4	36883,8	-2,10	
sp Q9UGI8 TES_HUMAN	Testin OS=Homo sapiens OX=9606 GN=TES PE=1 SV=1	cadherin binding; RNA binding; negative regulation of cell population proliferation; may play a role in cell adhesion, cell spreading and in the reorganization of the actin cytoskeleton	0,001	45960,6	10767,2	40885,9	47701,8	9723,2	-2,09	
sp Q9NR46 SHLB2_HUMAN	Endophilin-B2 OS=Homo sapiens OX=9606 GN=SH3GLB2 PE=1 SV=1	Cadherin binding; identical protein binding	0,001	29053,1	7041,0	6385,6	26340,0	2752,1	-2,04	

sp P07196 NFL_HUMAN	Neurofilament light polypeptide OS=Homo sapiens OX=9606 GN=NEFL PE=1 SV=3	protein binding; phospholipase binding; structural constituent of cytoskeleton; structural constituent of postsynaptic intermediate filament cytoskeleton; anterograde axonal transport; axonal transport of mitochondrion; intermediate filament organization; intermediate filament polymerization or depolymerization; locomotion; MAPK cascade; microtubule cytoskeleton organization; negative regulation of neuron apoptotic process; neurofilament bundle assembly; neurofilament cytoskeleton organization; neuromuscular process controlling balance; neuron projection morphogenesis; peripheral nervous system axon regeneration; positive regulation of axonogenesis; protein polymerization; regulation of axon diameter; regulation of NMDA receptor activity; response to acrylamide; response to corticosterone; response to peptide hormone; response to sodium arsenite; response to toxic substance; retrograde axonal transport; spinal cord development; synapse maturation	0,001	7321839,6	2300996,1	4732646,2	6625586,3	1986276,0	-1,67
sp O00151 PDL1_HUMAN	PDZ and LIM domain protein 1 OS=Homo sapiens OX=9606 GN=PDLIM1 PE=1 SV=4	actin binding; cadherin binding involved in cell-cell adhesion; metal ion binding; muscle alpha-actinin binding; transcription coactivator activity; actin cytoskeleton organization; establishment or maintenance of actin cytoskeleton polarity; heart development; maintenance of cell polarity; muscle structure development; regulation of transcription by RNA polymerase II; response to hypoxia; response to oxidative stress; stress fiber assembly	0,000	90040,9	30387,5	50060,9	85171,0	21241,2	-1,57
sp P09493-5 TPM1_HUMAN	Isoform 5 of Tropomyosin alpha-1 chain OS=Homo sapiens OX=9606 GN=TPM1	actin binding; actin filament binding; cytoskeletal protein binding; identical protein binding; protein heterodimerization activity; protein homodimerization activity; structural constituent of cytoskeleton; structural constituent of muscle	0,005	37645,8	13192,8	22935,8	35785,7	14387,8	-1,51
sp Q9UM54-6 MYO6_HUMAN	Isoform 6 of Unconventional myosin-VI OS=Homo sapiens OX=9606 GN=MYO6	actin binding; actin-dependent ATPase activity; actin filament binding; ADP binding; ATP binding; calmodulin binding; microfilament motor activity; actin filament-based movement; actin filament organization; DNA damage response, signal transduction by p53 class mediator; endocytosis; vesicle transport along actin filament	0,001	19060,2	7091,1	12330,3	17728,2	6665,2	-1,43
sp Q9HCM2 PLXA4_HUMAN	Plexin-A4 OS=Homo sapiens OX=9606 GN=PLXNA4 PE=1 SV=4	semaphorin receptor activity; negative regulation of cell adhesion; positive regulation of axonogenesis; regulation of axon extension involved in axon guidance; regulation of cell migration; regulation of cell shape; semaphorin-plexin signaling pathway involved in axon guidance	0,005	64963,2	25731,0	62063,4	56792,2	29293,1	-1,34
sp Q9UHC6 CNTP2_HUMAN	Contactin-associated protein-like 2 OS=Homo sapiens OX=9606 GN=CNTNAP2 PE=1 SV=1	enzyme binding; brain development; cell adhesion; clustering of voltage-gated potassium channels; learning; limbic system development; neuron projection development; neuron projection morphogenesis; neuron recognition; positive regulation of gap junction assembly; protein localization to juxtaparanode region of axon; transmission of nerve impulse	0,004	35970,4	14632,0	27178,2	35855,8	13537,0	-1,30
sp O15394 NCAM2_HUMAN	Neural cell adhesion molecule 2 OS=Homo sapiens OX=9606 GN=NCAM2 PE=1 SV=2	identical protein binding; axonal fasciculation; neuron cell-cell adhesion	0,000	67221,9	27533,9	85653,3	72958,3	25818,9	-1,29
sp Q7Z2K8 GRIN1_HUMAN	G protein-regulated inducer of neurite outgrowth 1 OS=Homo sapiens OX=9606 GN=GPRIN1 PE=1 SV=2	phosphoprotein binding; neuron projection development	0,000	104499,0	45276,8	74697,8	104184,8	35785,5	-1,21
sp O15143 ARC1B_HUMAN	Actin-related protein 2/3 complex subunit 1B OS=Homo sapiens OX=9606 GN=ARPC1B PE=1 SV=3	Arp2/3 complex-mediated actin nucleation; ephrin receptor signaling pathway; Fc-gamma receptor signaling pathway involved in phagocytosis; response to estradiol; response to estrogen; actin binding; structural constituent of cytoskeleton	0,003	19370,0	9225,2	5806,3	17823,9	7009,7	-1,07
sp P04216 THY1_HUMAN	Thy-1 membrane glycoprotein OS=Homo sapiens OX=9606 GN=THY1 PE=1 SV=2	GPI anchor binding; GTPase activator activity; integrin binding; protein kinase binding; cell-cell adhesion; cell-cell signaling; cytoskeleton organization; focal adhesion assembly; integrin-mediated signaling pathway; regulation of axonogenesis; negative regulation of neuron projection regeneration; positive regulation of focal adhesion assembly; positive regulation of heterotypic cell-cell adhesion; positive regulation of release of sequestered calcium ion into cytosol; regulation of cell-matrix adhesion calcium ion binding; extracellular matrix binding; glycosaminoglycan binding; metalloendopeptidase inhibitor activity; extracellular matrix organization; positive regulation of cell motility; positive regulation of cell-substrate adhesion; regulation of cell differentiation; synapse assembly	0,000	136683,3	65449,2	131625,4	129246,4	62074,0	-1,06
sp Q92563 TICN2_HUMAN	Testican-2 OS=Homo sapiens OX=9606 GN=SPOCK2 PE=1 SV=1	identical protein binding; structural molecule activity; cellular protein localization; cellular response to hypoxia; DNA double-strand break attachment to nuclear envelope; establishment or maintenance of microtubule; cytoskeleton polarity; IRE1-mediated unfolded protein response; mitotic nuclear envelope reassembly; muscle organ development; negative regulation of cardiac muscle hypertrophy in response to stress; negative regulation of cell population proliferation; nuclear envelope organization; positive regulation of cell aging; protein localization to nucleus; regulation of cell migration; regulation of telomere maintenance; nuclear assembly, chromatin organization, nuclear membrane and telomere dynamics; required for normal development of peripheral nervous system and skeletal muscle and for muscle satellite cell proliferation	0,010	15479,4	7427,4	9968,4	14369,5	7028,8	-1,06
sp P02545 LMNA_HUMAN	Prelamin-A/C OS=Homo sapiens OX=9606 GN=LMNA PE=1 SV=1	identical protein binding; structural molecule activity; cellular protein localization; cellular response to hypoxia; DNA double-strand break attachment to nuclear envelope; establishment or maintenance of microtubule; cytoskeleton polarity; IRE1-mediated unfolded protein response; mitotic nuclear envelope reassembly; muscle organ development; negative regulation of cardiac muscle hypertrophy in response to stress; negative regulation of cell population proliferation; nuclear envelope organization; positive regulation of cell aging; protein localization to nucleus; regulation of cell migration; regulation of telomere maintenance; nuclear assembly, chromatin organization, nuclear membrane and telomere dynamics; required for normal development of peripheral nervous system and skeletal muscle and for muscle satellite cell proliferation	0,001	50695,3	24589,4	24063,7	46463,5	24973,7	-1,04
sp Q9Y639 NPTN_HUMAN	Neuroplastin OS=Homo sapiens OX=9606 GN=NPTN PE=1 SV=2	cell adhesion molecule binding; cell-cell adhesion mediator activity; ion channel binding; axon guidance; cellular calcium ion homeostasis; dendrite self-avoidance; excitatory synapse assembly; homophilic cell adhesion via plasma membrane adhesion molecules; long-term synaptic potentiation; negative regulation of cytokine production; positive regulation of cellular protein localization; positive regulation of cytosolic calcium ion concentration; positive regulation of ERK1 and ERK2 cascade; positive regulation of fibroblast growth factor receptor signaling pathway; positive regulation of long-term neuronal synaptic plasticity; positive regulation of long-term synaptic potentiation; positive regulation of neuron projection development; positive regulation of protein phosphorylation; regulation of receptor localization to synapse; trans-synaptic signaling by trans-synaptic complex, modulating synaptic transmission	0,000	134619,3	65472,8	91972,8	134484,9	59869,0	-1,04
sp P32004 L1CAM_HUMAN	Neural cell adhesion molecule L1 OS=Homo sapiens OX=9606 GN=L1CAM PE=1 SV=2	axon guidance receptor activity; protein domain specific binding; axon development; axon guidance; cell adhesion; cell-matrix adhesion; chemotaxis; homophilic cell adhesion via plasma membrane adhesion molecules; nervous system development; neuron projection development; positive regulation of axon extension; synapse organization	0,000	394117,0	192357,9	342555,6	371148,3	167235,0	-1,03
sp Q92572 AP3S1_HUMAN	AP-3 complex subunit sigma-1 OS=Homo sapiens OX=9606 GN=AP3S1 PE=1 SV=1	anterograde axonal transport; anterograde synaptic vesicle transport; insulin receptor signaling pathway; intracellular protein transport; vesicle-mediated transport	0,007	14932,6	7338,2	10737,1	14273,4	8550,7	-1,02
sp P13987 CD59_HUMAN	CD59 glycoprotein OS=Homo sapiens OX=9606 GN=CD59 PE=1 SV=1	complement binding; cell surface receptor signaling pathway; endoplasmic reticulum to Golgi vesicle-mediated transport	0,001	43373,9	20512,1	42075,7	41121,0	14890,7	-1,08
sp O95297 MPZL1_HUMAN	Myelin protein zero-like protein 1 OS=Homo sapiens OX=9606 GN=MPZL1 PE=1 SV=1	structural molecule activity; cell-cell signaling; transmembrane receptor protein tyrosine kinase signaling pathway	0,002	31014,3	15436,6	26992,5	26668,5	14530,2	-1,01
sp O43761 SNG3_HUMAN	Synaptogyrin-3 OS=Homo sapiens OX=9606 GN=SYNGR3 PE=1 SV=2	positive regulation of transporter activity; regulated exocytosis	0,001	2530,7	752,7	2269,3	2330,9	800,3	-1,75

sp Q4VC31 CCD58_HUMAN	Coiled-coil domain-containing protein 58 OS=Homo sapiens OX=9606 GN=CCDC58 PE=1 SV=1	Mitochondrial Matrix Import Factor 23	0,001	24128,2	9611,4	13886,8	21924,3	8024,0	-1,33
sp O60641 AP180_HUMAN	Clathrin coat assembly protein AP180 OS=Homo sapiens OX=9606 GN=SNAP91 PE=1 SV=2	1-phosphatidylinositol binding; clathrin heavy chain binding; clathrin coat assembly; clathrin-dependent endocytosis; protein transport; regulation of clathrin-dependent endocytosis; synaptic vesicle budding from presynaptic endocytic zone membrane; vesicle budding from membrane	0,006	55052,5	21985,7	52741,6	60405,0	19703,7	-1,32
sp Q13433 S39A6_HUMAN	Zinc transporter ZIP6 OS=Homo sapiens OX=9606 GN=SLC39A6 PE=1 SV=3	amino acid transmembrane transport; glutamine transport; sodium ion transport	0,005	7822,4	3180,8	6332,6	6754,3	3005,9	-1,30
sp Q8TAC9 SCAM5_HUMAN	Secretory carrier-associated membrane protein 5 OS=Homo sapiens OX=9606 GN=SCAMP5 PE=1 SV=1	protein-containing complex binding; exocytosis; negative regulation of endocytosis; positive regulation of calcium ion-dependent exocytosis; positive regulation of cytokine production; protein transport; response to endoplasmic reticulum stress	0,001	29310,3	13752,6	22589,0	28484,3	10957,2	-1,09
sp Q8TC26 TM163_HUMAN	Transmembrane protein 163 OS=Homo sapiens OX=9606 GN=TMEM163 PE=2 SV=1	zinc ion import into synaptic vesicle	0,000	38717,6	18369,4	27010,1	36727,5	15172,3	-1,08