

UniProt ID	Protein Name	Protein itself aggregates	Homologous protein aggregate	Involved in aggregation pathology	Reference
ALBU_HUMAN	Serum albumin		Bovine Serum Albumin		[44]
HBB_HUMAN	Hemoglobin subunit beta	in vitro evidence			[45]
HBA_HUMAN	Hemoglobin subunit alpha	in vitro evidence			[45]
ANT3_HUMAN	Antithrombin-III			Shows reduced activity in AL amyloidosis	[46]
ACTG_HUMAN	Actin, cytoplasmic 2	Component of amyloid deposit of localized cutaneous amyloidosis			[47]
IC1_HUMAN	Serpин G1		The entire serpin family is known to be aggregation prone via domain swapping		[48]
LDHB_HUMAN	Lactate dehydrogenase		Homolog from rabbit aggregates		[49]
HBG2_HUMAN	Hemoglobin subunit gamma-2	in vitro evidence			[45]
ACTC_HUMAN	Actin, alpha cardiac muscle 1	in vitro evidence			[50]
ACTS_HUMAN	Actin, alpha skeletal muscle	in vitro evidence			[50]
G6PI_HUMAN	Glucose-6-phosphate isomerase		The homolog from yeast aggregates during refolding in vitro		[51]

THBG_HUMAN	Serpин A7		The entire serpin family is known to be aggregation prone via domain swapping		[48]
NB5R3_HUMAN	NADH-cytochrome b5 reductase 3				
RAC2_HUMAN	Ras-related C3 botulinum toxin substrate 2				
CAH2_HUMAN	Carbonic anhydrase 2	in vitro evidence			[52]
GDIA_HUMAN	Rab GDP dissociation inhibitor alpha				
PUR9_HUMAN	Bifunctional purine biosynthesis protein PURH				
THIL_HUMAN	Acetyl-CoA acetyltransferase, mitochondrial	in vitro evidence			[53]
PROC_HUMAN	Vitamin K-dependent protein C				
CP19A_HUMAN	Aromatase				
CHLE_HUMAN	Cholinesterase				
OAT_HUMAN	Ornithine aminotransferase, mitochondrial	in vitro evidence			[54]
ACADM_HUMAN	Medium-chain specific acyl-CoA dehydrogenase, mitochondrial	in vitro evidence			[55]

KPYR_HUMAN	Pyruvate kinase PKLR				
GSHB_HUMAN	Glutathione synthetase				
IGLL1_HUMAN	Immunoglobulin lambda-like polypeptide 1			Light Chain Amyloidosis	
STAT3_HUMAN	Signal transducer and activator of transcription 3			Involved in Abeta toxicity	
PUR8_HUMAN	Adenylosuccinate lyase	in vitro evidence			[56]
RASH_HUMAN	GTPase HRas				
ASSY_HUMAN	Argininosuccinate synthase				
HEMH_HUMAN	Ferrochelatase, mitochondrial	in vitro evidence			[57]
RASN_HUMAN	GTPase NRas				
SH21A_HUMAN	SH2 domain-containing protein 1A				
ATLA1_HUMAN	Atlastin-1	in vitro evidence			[58]
P53_HUMAN	Cellular tumor antigen p53	in vitro evidence			[4]
ARGI1_HUMAN	Arginase-1				

STS_HUMAN	Steryl-sulfatase				
MSH2_HUMAN	DNA mismatch repair protein Msh2				
HEXA_HUMAN	Beta-hexosaminidase subunit alpha	in vitro evidence		Tay–Sachs disease	[59]
MSH6_HUMAN	DNA mismatch repair protein Msh6				
SCO1_HUMAN	Protein SCO1 homolog, mitochondrial				
CASQ2_HUMAN	Calsequestrin-2	in vitro evidence			[60]
ODBA_HUMAN	2-oxoisovalerate dehydrogenase subunit alpha, mitochondrial				
XRP2_HUMAN	Protein XRP2			Retinitis pigmentosa	
GCDH_HUMAN	Glutaryl-CoA dehydrogenase, mitochondrial	in vitro evidence			[61]
RNH2B_HUMAN	Ribonuclease H2 subunit B				
PMM2_HUMAN	Phosphomannomutase 2	in vitro evidence			[62]
MUTA_HUMAN	Methylmalonyl-CoA mutase, mitochondrial				

HGD_HUMAN	Homogentisate 1,2-dioxygenase			Alkaptonuria, novel type II AA amyloidosis	
SPYA_HUMAN	Serine-pyruvate aminotransferase	in vitro evidence			[63]
GLCM_HUMAN	Glucosylceramidase				
UNG_HUMAN	Uracil-DNA glycosylase		Poxvirus-encoded uracil DNA glycosylase shown to aggregate		[64]
JAK3_HUMAN	Tyrosine-protein kinase JAK3				
TY3H_HUMAN	Tyrosine 3-monooxygenase				
METK1_HUMAN	S-adenosylmethionine synthase isoform type-1				
ARSB_HUMAN	Arylsulfatase B				
BGLR_HUMAN	Beta-glucuronidase				
OTC_HUMAN	Ornithine carbamoyltransferase, mitochondrial				
SOS1_HUMAN	Son of sevenless homolog 1				
ARL6_HUMAN	ADP-ribosylation factor-like protein 6				

KHK_HUMAN	Ketohexokinase				
SUIS_HUMAN	Sucrase-isomaltase, intestinal	in vitro evidence			[65]
TGFR1_HUMAN	TGF-beta receptor type-1				
CXB2_HUMAN	Gap junction beta-2 protein				
INSR_HUMAN	Insulin receptor	in vitro evidence			[66]
FA8_HUMAN	Coagulation factor VIII	in vitro evidence			[67]
CAH4_HUMAN	Carbonic anhydrase 4				
SUMF1_HUMAN	Sulfatase-modifying factor 1				
ATTY_HUMAN	Tyrosine aminotransferase				
NEUS_HUMAN	Neuroserpin	Neuroserpin inclusion bodies in Familial encephalopathy			[68]
CD40L_HUMAN	CD40 ligand			CD40-CD40L interaction associated with AD pathology	[69]
PTPS_HUMAN	6-pyruvoyl tetrahydrobiopterin synthase				

CHK2_HUMAN	Serine/threonine-protein kinase Chk2				
ACY2_HUMAN	Aspartoacylase	in vitro evidence			[70]
TTPA_HUMAN	Alpha-tocopherol transfer protein	in vitro evidence			[71]
CLAT_HUMAN	Choline O-acetyltransferase	in vitro evidence			[72]
RLBP1_HUMAN	Retinaldehyde-binding protein 1				
RPE65_HUMAN	Retinoid isomerohydrolase				
CP2R1_HUMAN	Vitamin D 25-hydroxylase				

References:

4. Xu J, Reumers J, Couceiro JR, De Smet F, Gallardo R, et al. (2011) Gain of function of mutant p53 by coaggregation with multiple tumor suppressors. *Nat Chem Biol* 7: 285-295.
44. Holm NK, Jespersen SK, Thomassen LV, Wolff TY, Sehgal P, et al. (2007) Aggregation and fibrillation of bovine serum albumin. *Biochim Biophys Acta* 1774: 1128-1138.
45. McGovern P, Reisberg P, Olson JS (1976) Aggregation of deoxyhemoglobin subunits. *J Biol Chem* 251: 7871-7879.
46. Gamba G, Montani N, Anesi E, Palladini G, Lorenzutti F, et al. (1999) Abnormalities in thrombin-antithrombin pathway in AL amyloidosis. *Amyloid* 6: 273-277.
47. Miura Y, Harumiya S, Ono K, Fujimoto E, Akiyama M, et al. (2013) Galectin-7 and actin are components of amyloid deposit of localized cutaneous amyloidosis. *Exp Dermatol* 22: 36-40.
48. Huntington JA, Sendall TJ, Yamasaki M (2009) New insight into serpin polymerization and aggregation. *Prion* 3: 12-14.

49. Goyal K, Walton LJ, Tunnacliffe A (2005) LEA proteins prevent protein aggregation due to water stress. *Biochem J* 388: 151-157.
50. Mazhul VM, Zaitseva EM, Shavlovskii MM, Povarova OI, Kuznetsova IM, et al. (2005) [Room temperature phosphorescence of amorphous aggregates and amyloid fibrils resulting from protein misfolding]. *Tsitologiya* 47: 978-987.
51. Li J, Zhang S, Wang C (2001) Effects of macromolecular crowding on the refolding of glucose- 6-phosphate dehydrogenase and protein disulfide isomerase. *J Biol Chem* 276: 34396-34401.
52. Wetlaufer DB, Xie Y (1995) Control of aggregation in protein refolding: a variety of surfactants promote renaturation of carbonic anhydrase II. *Protein Sci* 4: 1535-1543.
53. Miyazaki A, Sakashita N, Lee O, Takahashi K, Horiuchi S, et al. (1998) Expression of ACAT-1 protein in human atherosclerotic lesions and cultured human monocytes-macrophages. *Arterioscler Thromb Vasc Biol* 18: 1568-1574.
54. Boernke WE, Stevens FJ, Peraino C (1981) Effects of self-association of ornithine aminotransferase on its physicochemical characteristics. *Biochemistry* 20: 115-121.
55. Jank JM, Maier EM, Reibeta DD, Haslbeck M, Kemter KF, et al. (2014) The domain-specific and temperature-dependent protein misfolding phenotype of variant medium-chain acyl-CoA dehydrogenase. *PLoS One* 9: e93852.
56. Sivendran S, Patterson D, Spiegel E, McGown I, Cowley D, et al. (2004) Two novel mutant human adenylosuccinate lyases (ASLs) associated with autism and characterization of the equivalent mutant *Bacillus subtilis* ASL. *J Biol Chem* 279: 53789-53797.
57. Najahi-Missaoui W, Dailey HA (2005) Production and characterization of erythropoietic protoporphyrinic heterodimeric ferrochelatases. *Blood* 106: 1098-1104.
58. Zhao J, Hedera P (2013) Hereditary spastic paraplegia-causing mutations in atlastin-1 interfere with BMPRII trafficking. *Mol Cell Neurosci* 52: 87-96.
59. Weitz G, Proia RL (1992) Analysis of the glycosylation and phosphorylation of the alpha-subunit of the lysosomal enzyme, beta-hexosaminidase A, by site-directed mutagenesis. *J Biol Chem* 267: 10039-10044.
60. He Z, Dunker AK, Wesson CR, Trumble WR (1993) Ca(2+)-induced folding and aggregation of skeletal muscle sarcoplasmic reticulum calsequestrin. The involvement of the trifluoperazine-binding site. *J Biol Chem* 268: 24635-24641.
61. Westover JB, Goodman SI, Frerman FE (2003) Pathogenic mutations in the carboxyl-terminal domain of glutaryl-CoA dehydrogenase: effects on catalytic activity and the stability of the tetramer. *Mol Genet Metab* 79: 245-256.
62. Vega AI, Perez-Cerda C, Abia D, Gamez A, Briones P, et al. (2011) Expression analysis revealing destabilizing mutations in phosphomannomutase 2 deficiency (PMM2-CDG): expression analysis of PMM2-CDG mutations. *J Inherit Metab Dis* 34: 929-939.
63. Santana A, Salido E, Torres A, Shapiro LJ (2003) Primary hyperoxaluria type 1 in the Canary Islands: a conformational disease due to I244T mutation in the P11L-containing alanine:glyoxylate aminotransferase. *Proc Natl Acad Sci U S A* 100: 7277-7282.

64. Stuart DT, Upton C, Higman MA, Niles EG, McFadden G (1993) A poxvirus-encoded uracil DNA glycosylase is essential for virus viability. *J Virol* 67: 2503-2512.
65. Sigrist H, Ronner P, Semenza G (1975) A hydrophobic form of the small-intestinal sucrase-isomaltase complex. *Biochim Biophys Acta* 406: 433-446.
66. Kohanski RA (1989) Insulin receptor aggregation and autophosphorylation in the presence of cationic polyamino acids. *J Biol Chem* 264: 20984-20991.
67. Tagliavacca L, Wang Q, Kaufman RJ (2000) ATP-dependent dissociation of non-disulfide-linked aggregates of coagulation factor VIII is a rate-limiting step for secretion. *Biochemistry* 39: 1973-1981.
68. Ying Z, Wang H, Fan H, Wang G (2011) The endoplasmic reticulum (ER)-associated degradation system regulates aggregation and degradation of mutant neuroserpin. *J Biol Chem* 286: 20835-20844.
69. Tan J, Town T, Crawford F, Mori T, DelleDonne A, et al. (2002) Role of CD40 ligand in amyloidosis in transgenic Alzheimer's mice. *Nat Neurosci* 5: 1288-1293.
70. Le Coq J, An HJ, Lebrilla C, Viola RE (2006) Characterization of human aspartoacylase: the brain enzyme responsible for Canavan disease. *Biochemistry* 45: 5878-5884.
71. Min KC, Kovall RA, Hendrickson WA (2003) Crystal structure of human alpha-tocopherol transfer protein bound to its ligand: implications for ataxia with vitamin E deficiency. *Proc Natl Acad Sci U S A* 100: 14713-14718.
72. Froissart C, Basset P, Mandel P, Massarelli R (1979) Choline acetyltransferase aggregates from human placenta and rat brain. *FEBS Lett* 100: 276-280.