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CLN3 Antibody (Center)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP13304c

Product Information

Application	WB, E
Primary Accession	Q13286
Other Accession	Q60HH0, NP_001035897.1, NP_000077.1
Reactivity	Human
Predicted	Monkey
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB33286
Calculated MW	47623
Antigen Region	235-263

Additional Information

Gene ID	1201
Other Names	Battenin, Batten disease protein, Protein CLN3, CLN3, BTS
Target/Specificity	This CLN3 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 235-263 amino acids from the Central region of human CLN3.
Dilution	WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	CLN3 Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	CLN3 (HGNC:2074)
Synonyms	BTS
Function	Mediates microtubule-dependent, anterograde transport connecting the

	Golgi network, endosomes, autophagosomes, lysosomes and plasma membrane, and participates in several cellular processes such as regulation of lysosomal pH, lysosome protein degradation, receptor- mediated endocytosis, autophagy, transport of proteins and lipids from the TGN, apoptosis and synaptic transmission (PubMed:10924275, PubMed:15471887, PubMed:18317235, PubMed:18817525, PubMed:20850431, PubMed:22261744). Facilitates the proteins transport from trans-Golgi network (TGN)-to other membrane compartments such as transport of microdomain-associated proteins to the plasma membrane, IGF2R transport to the lysosome where it regulates the CTSD release leading to regulation of CTSD maturation and thereby APP intracellular processing (PubMed:10924275, PubMed:18817525). Moreover regulates CTSD activity in response to osmotic stress (PubMed:23840424, PubMed:28390177). Also binds galactosylceramide and transports it from the trans Golgi to the rafts, which may have immediate and downstream effects on cell survival by modulating ceramide synthesis (PubMed:18317235). At the plasma membrane, regulates actin-dependent events including filopodia formation, cell migration, and pinocytosis through ARF1-CDC42 pathway and also the cytoskeleton organization through interaction with MYH10 and fodrin leading to the regulation of the plasma membrane association of Na+, K+ ATPase complex (PubMed:20850431). Regulates synaptic transmission in the amygdala, hippocampus, and cerebellum through regulation of synaptic vesicles density and their proximity to active zones leading to modulation of short-term plasticity and age-dependent anxious behavior, learning and memory (By similarity). Regulates autophagic vacuoles (AVs) maturation by modulating the trafficking between endocytic and autophagolysosomal/lysosomal compartments, which involves vesicle fusion leading to regulation of degradation process (By similarity). Also participates in cellular homeostasis of compounds such as, water, ions, amino acids, proteins and lipids in several tissue namely in brain and kidney through regulation of their transport and synthesis (PubMed:17482562).
Cellular Location	Lysosome membrane; Multi-pass membrane protein. Late endosome. Lysosome. Golgi apparatus. Golgi apparatus membrane. Golgi apparatus, Golgi stack. Golgi apparatus, trans-Golgi network. Cell membrane Recycling endosome. Membrane raft. Membrane, caveola. Early endosome membrane. Synapse, synaptosome {ECO:0000250|UniProtKB:Q61124}. Late endosome membrane {ECO:0000250|UniProtKB:Q61124}. Cytoplasmic vesicle, autophagosome {ECO:0000250|UniProtKB:Q61124}. Note=CLN3 is not present in late endosomes/lysosomes in fibroblasts and neurons (PubMed:15240864) Trafficks from cell membrane to Golgi via endosomes (PubMed:15240864) Osmotic stress changes the subcellular localization of CLN3 (PubMed:23840424). Trafficks to intracellular compartments via the plasma membranet through AP3M1-dependent mechanisms (PubMed:14644441) Excluded from the synaptic vesicles (By similarity) {ECO:0000250|UniProtKB:Q61124, ECO:0000269|PubMed:14644441, ECO:0000269|PubMed:15240864, ECO:0000269|PubMed:23840424}
Tissue Location	Expressed in the cortical brain, pancreas, spleen, and testis with weaker expression in the peripheral nerve (at protein level). Highly expressed in gray matter (at protein level)

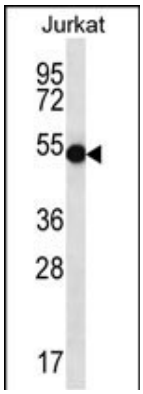
Background

This gene encodes a protein that is involved in lysosomal function. Mutations in this, as well as other neuronal ceroid-lipofuscinosis (CLN) genes, cause neurodegenerative diseases commonly known as Batten disease or collectively known as neuronal ceroid lipofuscinoses (NCLs). Many alternatively spliced transcript variants have been found for this gene.

References

Adams, H.R., et al. Dev Med Child Neurol 52(7):637-643(2010)
Imielinski, M., et al. Nat. Genet. 41(12):1335-1340(2009)
Sarpong, A., et al. Clin. Genet. 76(1):38-45(2009)
Codlin, S., et al. J. Cell. Sci. 122 (PT 8), 1163-1173 (2009) :
Tuxworth, R.I., et al. Hum. Mol. Genet. 18(4):667-678(2009)

Images



CLN3 Antibody (Center) (Cat. #AP13304c) western blot analysis in Jurkat cell line lysates (35ug/lane).This demonstrates the CLN3 antibody detected the CLN3 protein (arrow).

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