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CLN8 Rabbit pAb

Catalog # AP97835

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q9UBY8
Other Accession	Q9UBY8
Reactivity	Rat, Human, Mouse
Predicted	Pig, Rabbit, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	32787
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CLN8
Epitope Specificity	201-286/286
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 1 TLC (TRAM/LAG1/CLN8) domain.
Post-translational modifications	Does not seem to be N-glycosylated.
DISEASE	Defects in CLN8 are the cause of neuronal ceroid lipofuscinosis type 8 (CLN8) [MIM:600143]. A form of neuronal ceroid lipofuscinosis with onset in childhood. Neuronal ceroid lipofuscinoses are progressive neurodegenerative, lysosomal storage diseases characterized by intracellular accumulation of autofluorescent liposomal material, and clinically by seizures, dementia, visual loss, and/or cerebral atrophy. The lipopigment patterns observed most often in neuronal ceroid lipofuscinosis type 8 comprise mixed combinations of granular, curvilinear, and fingerprint profiles. Defects in CLN8 are the cause of neuronal ceroid lipofuscinosis type 8 Northern epilepsy variant (CLN8NE) [MIM:610003]. A form of neuronal ceroid lipofuscinosis clinically characterized by epilepsy that presents between 5 and 10 years of age with frequent tonic-clonic seizures followed by progressive mental retardation. Visual loss is not a prominent feature. Intracellular accumulation of autofluorescent material results in curvilinear and granular profiles on ultrastructural analysis.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	CLN8, a 286 amino acid transmembrane protein, localizes mainly to the endoplasmic reticulum, but also partially to the ER-Golgi intermediate compartment (ERGIC). Mutations in the CLN8 gene cause neuronal ceroid lipofuscinosis 8 and progressive epilepsy with mental retardation (EPMR). Both disorders are forms of neuronal ceroid-lipofuscinose (NCL), a group of progressive neurodegenerative diseases found in children, characterized by failure of psychomotor development, impaired vision, seizures and premature death. The CLN8 protein is one of eight proteins in the CLN family, including CLN1-CLN7, which are associated with NCL.

Additional Information

Gene ID	2055
Other Names	C8orf61; EPMR; TLCD6; mnd; CLN8_CANLF; CLN8; CLN8_HUMAN; CLN8_MOUSE; CLN8 transmembrane ER and ERGIC protein; chromosome 8 open reading frame 61; epilepsy, progressive with mental retardation; ceroid-lipofuscinosis, neuronal 8; CLN8, transmembrane ER and ERGIC protein
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	CLN8
Synonyms	C8orf61
Function	Could play a role in cell proliferation during neuronal differentiation and in protection against cell death.
Cellular Location	Endoplasmic reticulum membrane; Multi-pass membrane protein. Endoplasmic reticulum-Golgi intermediate compartment membrane; Multi-pass membrane protein. Endoplasmic reticulum

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.