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Rabbit Anti-CLN6 antibody

SL8017R

Product Name:	CLN6
Chinese Name:	神经细胞蜡样质脂褐质沉积病蛋白CLN6抗体
Alias:	Ceroid lipofuscinosis, neuronal 6, late infantile, variant; CLN6 protein; FLJ20561; Nclf; Protein CLN6; CLN6_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	36kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human CLN6:101-200/311
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	· CLN6, a 311 amino acid protein, has seven predicted transmembrane domains and is conserved across vertebrates. The CLN6 protein localizes to the endoplasmic reticulum but contributes to lysosomal function. Mutations in the CLN6 gene cause variant late-onset infantile neuronal ceroid lipofuscinosis (vLINCL), a lysosomal storage disorder marked by progressive mental deterioration and blindness; part of a group of severe inherited neurodegenerative disorders affecting children wherein lysosomes accumulate

storage material, causing the death of neurons. CLN6 is one of eight proteins, including CLN1-8, that are associated with NCL.

Function:
Defects in CLN6 are the cause of variant late onset infantile neuronal ceroid lipofuscinosis (vLINCL).

Subunit:
Endoplasmic reticulum membrane; Multi pass membrane protein.

Subcellular Location:
Endoplasmic reticulum membrane; Multi-pass membrane protein.

DISEASE:
Defects in CLN6 are the cause of neuronal ceroid lipofuscinosis type 6 (CLN6) [MIM:601780]. A form of neuronal ceroid lipofuscinosis. 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 6 comprise mixed combinations of granular, curvilinear, and fingerprint profiles.
Defects in CLN6 are the cause of neuronal ceroid lipofuscinosis type 4A (CLN4A) [MIM:204300]. An adult-onset neuronal ceroid lipofuscinosis. 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. CLN4A has no visual involvement and is characterized by progressive myoclonic epilepsy.

SWISS:
Q9NWW5

Gene ID:
54982

Database links:

Entrez Gene: 497068Dog

Entrez Gene: 54982Human

Entrez Gene: 76524Mouse

Entrez Gene: 315746Rat

Omim: 606725Human

SwissProt: Q5JZQ8Dog

	SwissProt: Q9NWW5Human SwissProt: A0PJN2Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.