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Rabbit Anti-CLN8 antibody

SL11715R

Product Name:	CLN8
Chinese Name:	神经细胞蜡样质脂褐质沉积病蛋白CLN8抗体
Alias:	Ceroid-lipofuscinosis, neuronal 8 (epilepsy, progressive with mental retardation); Cln8; CLN8_HUMAN; EPMR; Protein CLN8.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Horse,Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=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:	33kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human CLN8:201-286/286
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:	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.

Function:
Could play a role in cell proliferation during neuronal differentiation and in protection against cell death.

Subcellular Location:
Endoplasmic reticulum membrane. Endoplasmic reticulum-Golgi intermediate compartment membrane.

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.

Similarity:
Contains 1 TLC (TRAM/LAG1/CLN8) domain.

SWISS:
Q9UBY8

Gene ID:
2055

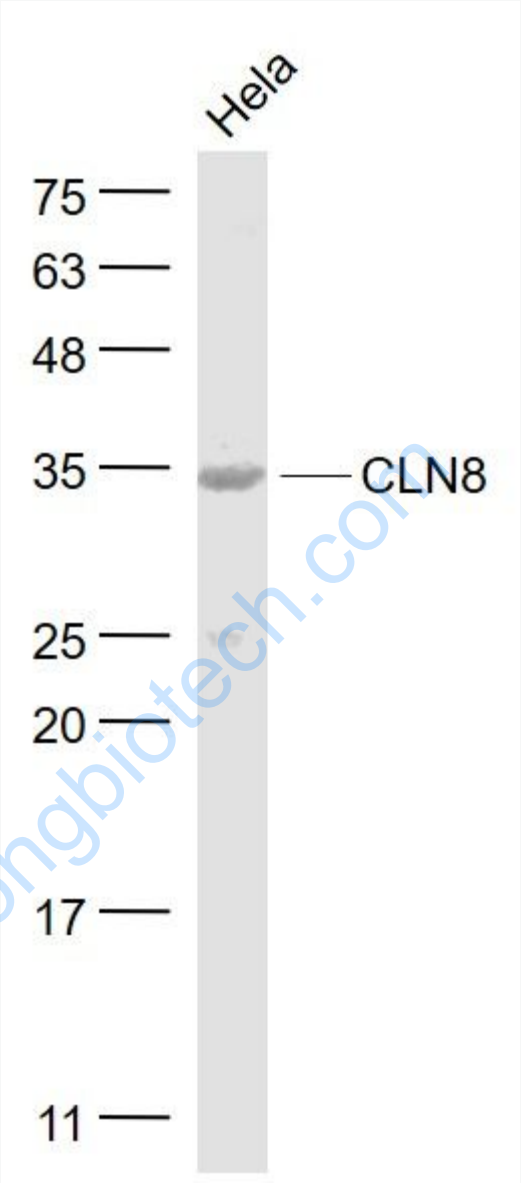
Database links:

Entrez Gene: 488558Dog

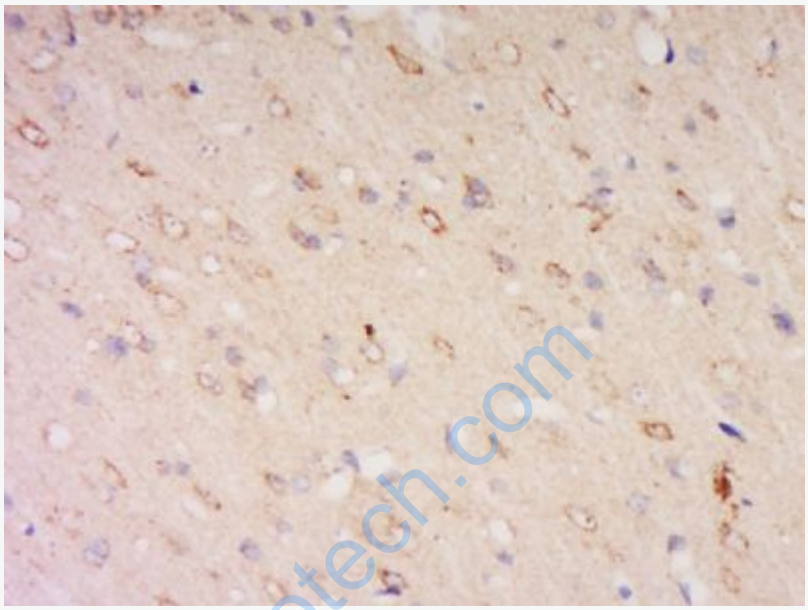
Entrez Gene: 2055Human

Entrez Gene: 26889Mouse

	Omim: 607837Human SwissProt: Q5JZQ7Dog SwissProt: Q9UBY8Human SwissProt: Q9QUK3Mouse Unigene: 127675Human Unigene: 254027Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Picture:	 Sample: Hela(Human) Cell Lysate at 30 ug Primary: Anti- CLN8 (SL11715R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 33 kD

Observed band size: 33 kD



Tissue/cell: Rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;

Incubation: Anti-CLN8 Polyclonal Antibody, Unconjugated(SL11715R) 1:500, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining