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Rabbit Anti-CLN5 antibody

SL11714R

Product Name:	CLN5
Chinese Name:	神经细胞蜡样质脂褐质沉积病蛋白CLN5抗体
Alias:	Ceroid lipofuscinosis neuronal 5; Ceroid-lipofuscinosis neuronal protein 5; CLN5; CLN5_HUMAN; NCL; Protein CLN5.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	37kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human CLN5:61-120/358
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:	Neuronal ceroid-lipofuscinose (NCL), also designated Batten disease, comprises a group of recessively inherited, progressive neurodegenerative diseases found in children. NCL is characterized by atrophy of the brain and an accumulation of lysosome derived fluorescent bodies found in many cells, especially neurons. Symptoms of NCL include a failure of psychomotor development, seizures, impaired vision and premature death. The eight genes/proteins associated with NCL are designated CLN1-CLN8. Mutations in six of these genes results in a distinct type of NCL-disease; the six

genes/proteins are CLN1 (encoding PPT1, a protein thiolesterase), CLN2 (encodeing the serine protease TPP1), CLN3, CLN5, CLN6 and CLN8. A single base duplication mutation in dog and cow CLN5 has been shown to cause NCL.

Subcellular Location:
Lysosome.

Tissue Specificity:
Ubiquitous.

Post-translational modifications:
Glycosylated.

DISEASE:
Defects in CLN5 are the cause of neuronal ceroid lipofuscinosis type 5 (CLN5) [MIM:256731]; also known as Finnish variant late-infantile neuronal ceroid lipofuscinosis (vLINCL). 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 5 comprise mixed combinations of granular, curvilinear, and fingerprint profiles.

Similarity:
Belongs to the CLN5 family.

SWISS:
O75503

Gene ID:
1203

Database links:

Entrez Gene: 1203Human

Entrez Gene: 211286Mouse

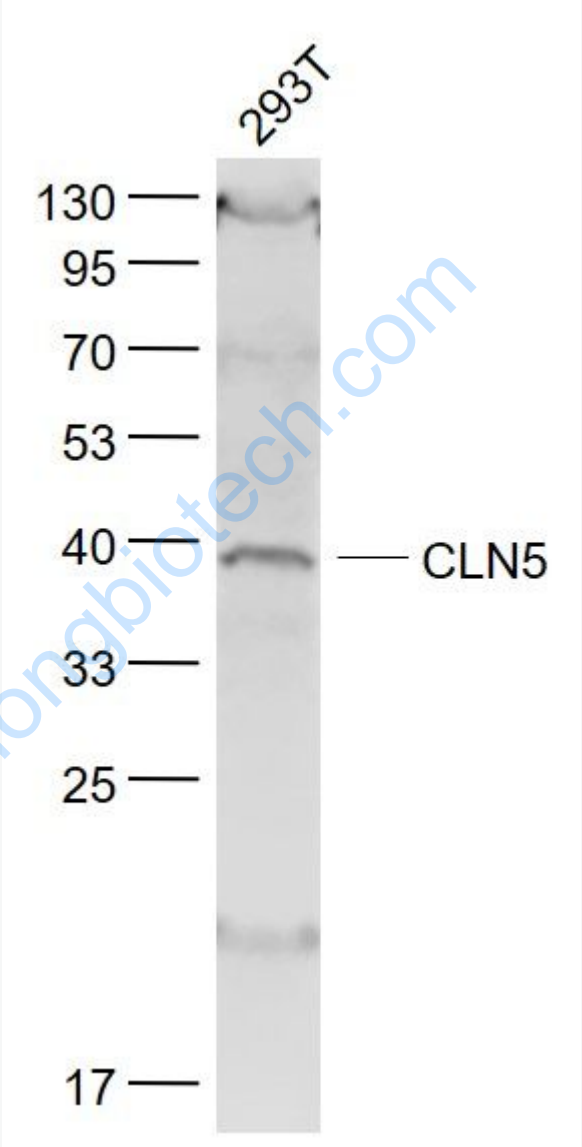
Entrez Gene: 306128Rat

Omim: 608102Human

SwissProt: O75503Human

SwissProt: Q3UMW8Mouse

Unigene: 30213Human

	Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: 293T(Human) Cell Lysate at 30 ug Primary: Anti- CLN5 (SL11714R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

	Predicted band size: 37 kD Observed band size: 37 kD