Product Name: CLN1 Rabbit Polyclonal Antibody
Catalog #: APRab09055

Summary

Production Name	CLN1 Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	IHC,ELISA
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	PPT1
Alternative Names	PPT1; PPT; Palmitoyl-protein thioesterase 1; PPT-1; Palmitoyl-protein hydrolase 1
Gene ID	5538.0
SwissProt ID	P50897.The antiserum was produced against synthesized peptide derived from human CLN1. AA range:16-65

Application

Dilution Ratio	IHC 1:100-1:300 ELISA: 1:10000
Molecular Weight	37kD

Background

The protein encoded by this gene is a small glycoprotein involved in the catabolism of lipid-modified proteins during

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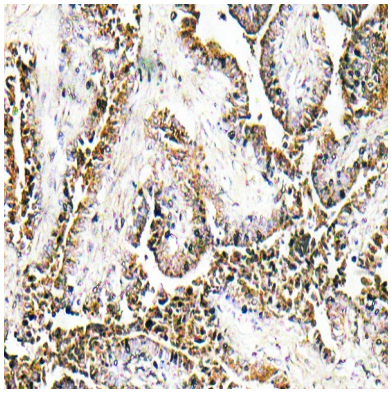
lysosomal degradation. The encoded enzyme removes thioester-linked fatty acyl groups such as palmitate from cysteine residues. Defects in this gene are a cause of infantile neuronal ceroid lipofuscinosis 1 (CLN1, or INCL) and neuronal ceroid lipofuscinosis 4 (CLN4). Two transcript variants encoding different isoforms have been found for this gene.[provided by RefSeq, Dec 2008],catalytic activity:Palmitoyl-protein + H(2)O = palmitate + protein.,disease:Defects in PPT1 are a cause of neuronal ceroid lipofuscinosis 4 (CLN4) [MIM:204300]; also known as adult type neuronal ceroid lipofuscinosis (NCL) or Kufs disease.,disease:Defects in PPT1 are the cause of infantile neuronal ceroid lipofuscinosis 1 (CLN1) [MIM:256730]; also called infantile neuronal ceroid lipofuscinosis (INCL). The neuronal ceroid lipofuscinosis are a group of progressive neurodegenerative diseases characterized by the intracellular accumulation of autofluorescent lipopigment storage material in different patterns ultrastructurally. The lipopigment pattern seen most often in CLN1 is referred to as granular osmiophilic deposits (GROD). There is a core group of four major clinical forms, the infantile, the late-infantile, the juvenile, and the adult forms. The infantile forms are characterized by progressive visual impairment, seizure, motor disturbances, dementia and premature death (8-11 years of age).,function:Removes thioester-linked fatty acyl groups such as palmitate from modified cysteine residues in proteins or peptides during lysosomal degradation. Prefers acyl chain lengths of 14 to 18 carbons.,online information:Neural Ceroid Lipofuscinoses mutation db,online information:Retina International's Scientific Newsletter,similarity:Belongs to the palmitoyl-protein thioesterase family.,

Research Area

Fatty acid elongation in mitochondria;Lysosome;

Image Data



Immunohistochemistry analysis of CLN1 antibody in paraffin-embedded human prostate carcinoma tissue.

Note

For research use only.