



CLN1 Polyclonal Antibody

Catalog No	BYab-03775
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	IHC;IF;ELISA
Gene Name	PPT1
Protein Name	Palmitoyl-protein thioesterase 1
Immunogen	The antiserum was produced against synthesized peptide derived from human CLN1. AA range:16-65
Specificity	CLN1 Polyclonal Antibody detects endogenous levels of CLN1 protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	IHC: 1/100 - 1/300. ELISA: 1/10000.. IF 1:50-200
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	PPT1; PPT; Palmitoyl-protein thioesterase 1; PPT-1; Palmitoyl-protein hydrolase 1
Observed Band	37kD
Cell Pathway	Lysosome . Secreted .
Tissue Specificity	Brain,Cerebellum,Liver,Prostate,Testis,
Function	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

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forms, the infantile, the late-infantile, the juvenile, and the adult forms. The infantile forms are characterized by p

Background

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

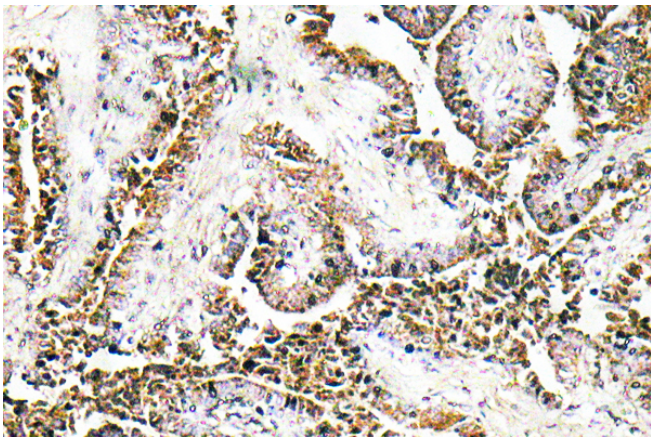
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



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