

产品名称: CLN1 Rabbit Polyclonal Antibody
产品货号: APRab09055



产品概述 (Summary)

产品名称 (Production Name)	CLN1 Rabbit Polyclonal Antibody
描述 (Description)	Rabbit polyclonal Antibody
宿主 (Host)	Rabbit
应用 (Application)	IHC, ICC, IF, ELISA
种属反应性 (Reactivity)	Human, Mouse, Rat

产品性能 (Performance)

偶联物 (Conjugation)	Unconjugated
修饰 (Modification)	Unmodified
同种型 (Isotype)	IgG
克隆 (Clonality)	Polyclonal
形式 (Form)	Liquid
存放说明 (Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液 (Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化方式 (Purification)	Affinity purification

免疫原信息 (Immunogen)

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

产品应用 (Application)

稀释比 (Dilution Ratio)	IHC 1:100-1:300, ICC/IF 1:50-1:200, ELISA 1:5000-1:20000
蛋白分子量 (Molecular Weight)	37kDa

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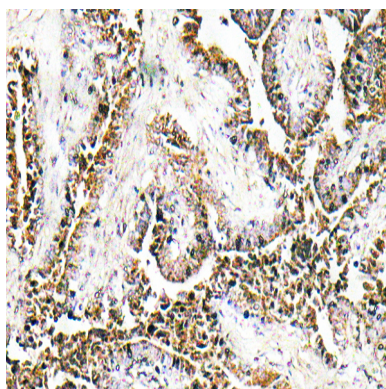
研究背景 (Background)

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],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,.

研究领域 (Research Area)

Fatty acid elongation in mitochondria;Lysosome;

图片 (Image Data)



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

注意事项 (Note)

For research use only .