

产品名称: CLN1 Rabbit Polyclonal Antibody

产品货号: APRab09055

产品概述 (Summary)

描述(Description)	Rabbit polyclonal Antibody
宿主(Host)	Rabbit
应用(Application)	IHC, ICC/IF, ELISA
种属反应(Reactivity)	Human, Mouse, Rat
偶联物(Conjugation)	Unconjugated
修饰(Modification)	Unmodified
同种型(Isotype)	IgG
克隆(Clonality)	Polyclonal
剂型(Form)	Liquid
保存条件(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 (Purification)	Affinity purification

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
分子量(Molecular Weight)	37kDa

抗原信息(Antigen Information)

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
SwissProt ID	P50897
免疫原(Immunogen)	The antiserum was produced against synthesized peptide derived from human CLN1. AA range:16-65

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.,

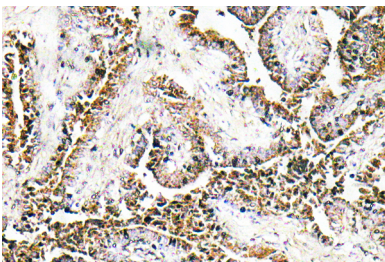
研究领域 (Research Area)

Fatty acid elongation in mitochondria;Lysosome;

注意事项 (Note)

For research use only.

图片 (Image Data)



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

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产品类别	产品货号	产品名称
WB 解决方案	RA10020	2h 极速 WB 即用型全流程试剂盒
	RA10021	4h 快速 WB 即用型全流程试剂盒
	RA10042	5h 畅享版 WB 全流程试剂盒
	RA10037	校准级彩色预染蛋白 Marker (8-180kDa)
	RA10038	校准级彩色预染蛋白 Marker (10-250kDa)
	RA10039	校准级高分子彩色预染蛋白 Marker (25-400kDa)
TSA 多重荧光染色试剂盒	RA10008	TSA 双标三色多重荧光染色试剂盒 (mIHC)
	RA10009	TSA 三标四色多重荧光染色试剂盒 (mIHC)
	RA10010	TSA 四标五色多重荧光染色试剂盒 (mIHC)
	RA10011	TSA 五标六色多重荧光染色试剂盒 (mIHC)
	RA10012	TSA 六标七色多重荧光染色试剂盒 (mIHC)
IHC 检测试剂盒	RA10006	HRP Anti-Mouse/Rabbit IHC Detection System
	RA10007	Polymer-HRP Anti-Mouse/Rabbit IHC Detection System
抗体标记试剂盒	RE80004p	辣根过氧化物酶(HRP)抗体标记试剂盒
	RE80002q	Sulfo-NHS-生物素标记试剂盒
	RE80007p	Cy3 荧光素标记试剂盒
	RE80011p	Fluor488 荧光素标记试剂盒
	RE80017p	Fluor750 荧光素标记试剂盒
	RE80005p	藻红蛋白(R-PE) 抗体快速标记试剂盒
	RE80040	PE-Cy7 串联染料抗体快速标记试剂盒
稳转细胞系构建服务 (免费赠送全膜 WB 验证)	TS-0001	过表达稳转细胞系构建
	TS-0002	敲低稳转细胞系构建
	TS-0003	敲除细胞系构建

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