



## Mouse Anti-PPT1 antibody (SLM51262M)

SLM51262M

<b>Product Name:</b>	PPT1
<b>Chinese Name:</b>	小鼠抗棕榈酰蛋白硫酯酶1单克隆抗体
<b>Alias:</b>	CLN1; INCL; Palmitoyl protein hydrolase 1; Palmitoyl protein thioesterase 1; Palmitoyl-protein hydrolase 1; Palmitoyl-protein thioesterase 1; PPT; PPT-1; PPT1; PPT1_HUMAN.
<b>Organism Species:</b>	Mouse
<b>Clonality:</b>	Monoclonal
<b>克隆号:</b>	10G4
<b>React Species:</b>	Human,
<b>Applications:</b>	WB=1:500-2000ELISA=1:500-1000IHC-P=1:20-100IHC-F=1:20-100 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
<b>Molecular weight:</b>	31kDa
<b>Cellular localization:</b>	cytoplasmic
<b>Form:</b>	Lyophilized or Liquid
<b>Concentration:</b>	1mg/ml
<b>immunogen:</b>	KLH conjugated synthetic peptide derived from human PPT1:
<b>Lsotype:</b>	IgG
<b>Purification:</b>	affinity purified by Protein A
<b>Storage Buffer:</b>	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
<b>Storage:</b>	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.
<b>PubMed:</b>	<a href="#">PubMed</a>
<b>Product Detail:</b>	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]

**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 (PubMed:8816748)

**Subcellular Location:**

Lysosome.

**DISEASE:**

Defects in PPT1 are the cause of neuronal ceroid lipofuscinosis type 1 (CLN1) [MIM:256730]. A form of neuronal ceroid lipofuscinosis with variable age at onset. Infantile, late-infantile, juvenile, and adult onset have been reported. 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 pattern seen most often in CLN1 is referred to as granular osmiophilic deposits (GROD).

**Similarity:**

Belongs to the palmitoyl-protein thioesterase family.

**SWISS:**

P50897

**Gene ID:**

5538

**Database links:**

[Entrez Gene: 281421](#)Cow

[Entrez Gene: 5538](#)Human

[Entrez Gene: 19063](#)Mouse

[Entrez Gene: 29411](#)Rat

[Omim: 600722](#)Human

[SwissProt: P45478](#)Cow

[SwissProt: P50897](#)Human

[SwissProt: O88531](#)Mouse

[SwissProt: P45479](#)Rat

[Unigene: 3873](#)Human

[Unigene: 277719](#)Mouse

[Unigene: 1574](#)Rat

**Important Note:**

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.