abcam

Product datasheet

Recombinant Human PPT1/PPT protein ab116764

1 Image

Description

Product name Recombinant Human PPT1/PPT protein

Expression system Wheat germ
Accession P50897

Protein length Full length protein

Animal free No

Nature Recombinant

Species Human

Sequence MASPGCLWLLAVALLPWTCASRALQHLDPPAPLPLVIWH

GMGDSCCNPLS

 ${\sf MGAIKKMVEKKIPGIYVLSLEIGKTLMEDVENSFFLNVNSQ}$

VTTVCQALA

KDPKLQQGYNAMGFSQGGQFLRAVAQRCPSPPMINLISV

GGQHQGVFGLP

RCPGESSHICDFIRKTLNAGAYSKVVQERLVQAEYWHDPI

KEDVYRNHSI

FLADINQERGINESYKKNLMALKKFVMVKFLNDSIVDPVDS

EWFGFYRSG

QAKETIPLQETSLYTQDRLGLKEMDNAGQLVFLATEGDHL

QLSEEWFYAH IIPFLG

Predicted molecular weight 60 kDa including tags

Amino acids 1 to 306

Specifications

Our Abpromise guarantee covers the use of ab116764 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications ELISA

SDS-PAGE Western blot

Form Liquid

Additional notes This product was previously labelled as PPT1.

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Preparation and Storage

Stability and Storage Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.3% Glutathione, 0.79% Tris HCI

General Info

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.

Involvement in 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

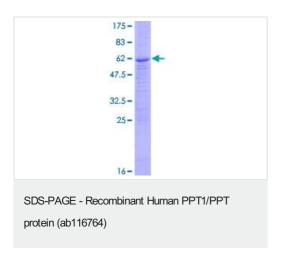
neurodegenerative, lysosomal storage diseases characterized by intracellular accumulation of autofluorescent liposomal material, and clinically by seizures, dementia, visual loss, and/or cerebral atrophy. The lipopigment pattern seen most often in CLN1 is referred to as granular

osmiophilic deposits (GROD).

Sequence similaritiesBelongs to the palmitoyl-protein thioesterase family.

Cellular localization Lysosome.

Images



12.5% SDS-PAGE analysis of ab116764 at approximately 59.73kDa, stained with Coomassie Blue.

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