Recombinant Human TPP1/CLN2 Protein (His Tag)

Catalog Number: PKSH030613

Note: *Centrifuge before opening to ensure complete recovery of vial contents.*

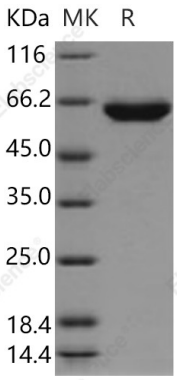
Description

Species	Human
Source	Baculovirus-Insect Cells-derived Human TPP1/CLN2 protein Met 1-Pro 563, with an C-terminal His
Calculated MW	60.7 kDa
Observed MW	60 kDa
Accession	AAH14863.1
Bio-activity	Measured by the cleavage of AlaAlaPheAMC. The specific activity is > 850 pmoles/min/μg.

Properties

Purity	> 95 % as determined by reducing SDS-PAGE.
Endotoxin	< 1.0 EU per μg of the protein as determined by the LAL method.
Storage	Store at < -20°C, stable for 6 months. Please minimize freeze-thaw cycles.
Shipping	This product is provided as liquid. It is shipped at frozen temperature with blue ice/ gel packs. Upon receipt, store it immediately at < - 20°C.
Formulation	Supplied as sterile 20mM Tris, 500mM NaCl, pH 7.4, 10% gly

Data



> 95 % as determined by reducing SDS-PAGE.

Background

Tripeptidyl-peptidase 1 (TPP1 / CLN2) is a member of the sedolisin family of serine proteases. The protease functions in the lysosome to cleave N-terminal tripeptides from substrates, and has weaker endopeptidase activity. It is synthesized as a catalytically-inactive enzyme which is activated and auto-proteolyzed upon acidification. TPP1 / CLN2 May act as a non-specific lysosomal peptidase which generates tripeptides from the breakdown products produced by lysosomal proteinases. Defects in TPP1 / CLN2 are the cause of neuronal ceroid lipofuscinosis type 2 (CLN2), a form of neuronal ceroid lipofuscinosis which is associated with the failure to degrade specific neuropeptides and a subunit of ATP synthase in the lysosome. 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.

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