## Recombinant Human TPP1/CLN2 Protein (His Tag)

#### Catalog No. PKSH030613

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

Description	
Synonyms	Tripeptidyl-Peptidase 1;TPP-1;Cell Growth-Inhibiting Gene 1 Protein;Lysosomal Pepstatin-Insensitive Protease;LPIC;Tripeptidyl Aminopeptidase;TPP1;CLN2;GIG1;LPIC;SCAR7;TPP-1
Species	Human
Expression Host	Baculovirus-Insect Cells
Sequence	Met 1-Pro 563
Accession	AAH14863.1
Calculated Molecular Weight	60.7 kDa
Observed molecular weight	60 kDa
Tag	C-His
Bioactivity	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 $\mu$ g of the protein as determined by the LAL method.
Storage	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80°C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of reconstituted samples are stable at < -20°C for 3 months.
Shipping	This product is provided as lyophilized powder which is shipped with ice packs.
Formulation	Lyophilized from sterile 20mM Tris, 500mM NaCl, pH 7.4, 10% glycerol Normally 5 % - 8 % trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Please refer to the specific buffer information in the printed manual.
Reconstitution	Please refer to the printed manual for detailed information.
Data	



> 95 % as determined by reducing SDS-PAGE.

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### 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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