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Recombinant Human TPP1 / CLN2 Protein (His tag)

产品货号: TP08207

第三版

别名:CLN2;GIG1;LPIC;SCAR7;TPP-1

描述: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.

配方:PBS

分子量:60.7 kDa

序列:Met 1-Pro 563

纯度:> 95% by HPLC

浓度:

内毒素:<1.0 EU per 1 ug of protein (determined by LAL method)

存储: +4 ° C 保存 (1-2 周). 长期保存在 -20 ° C 或者 -70 ° C. 避免反复冻融.