Recombinant Human SMPD1/ASM Protein (aa 1-631, His Tag)(Active)

Catalog No. PKSH030372

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

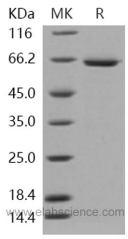
Description

Synonyms	ASM;ASMASE;NPD
Species	Human
Expression Host	Baculovirus-Insect Cells
Sequence	Met 1-Cys 631
Accession	NP_000534.3
Calculated Molecular Weight	65 kDa
Observed molecular weight	65 kDa
Tag	C-His
Bioactivity	Measured by its ability to cleave. 2-N-Hexadecanoylamino-4-nitrophenylphosphorylcholine (HNPPC). The specific activity is >1,000 pmol/min/μg.

Properties

Purity	> 94 % as determined by reducing SDS-PAGE.
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	Lyophilized from sterile 50mM Tris, 100mM NaCl, pH 8.0, 0.1% OGP, 10% glycerol
Reconstitution	Please refer to the printed manual for detailed information.

Data



Background

Sphingomyelin phosphodiesterase 1 (SMPD1) , also known as ASM (acid sphingomyelinase), is a member of the acid sphingomyelinase family of enzymes. Three isoforms have been identified, isoform 1 is 631 amino acids (aa) in length as the pro form, while Isoform 2 and isoform 3 have lost catalytic activity. The active SMPD1 isoform 1 contains one saposin B-type domain that likely interacts with sphingomyelin, and a catalytic region. Human SMPD1 is 86% aa identical

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to mouse SMPD1. SMPD1 is a monomeric lysosomal enzyme that converts sphingomyelin (a plasma membrane lipid) into ceramide through the removal of phosphorylcholine. This generates second messenger components that participate in signal transduction. Defects in SMPD1 are the cause of Niemann-Pick disease type A (NPA) and type B (NPB), also known as Niemann-Pick disease classical infantile form and Niemann-Pick disease visceral form. Niemann-Pick disease is a clinically and genetically heterogeneous recessive disorder. NPB has little if any neurologic involvement and patients may survive into adulthood.

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