Recombinant Human SMPD1/ASM Protein (His Tag)

Catalog No. PKSH030434

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

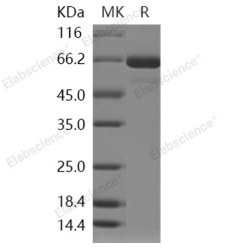
Description

Synonyms	ASM;ASMASE;NPD
Species	Human
Expression Host	Baculovirus-Insect Cells
Sequence	Met 1-Pro628
Accession	NP_000534.3
Calculated Molecular Weight	66.3 kDa
Tag	C-His
Bioactivity	Measured by its ability to cleave 2-N-Hexadecanoylamino-4-nitrophenylphosphorylcholine (HNPPC). The specific activity is > 1000 pmol/min/μg.

Properties

Purity	> 90 % 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 20 mM Tris, 500 mM NaCl, 25 % glycerol, pH 7.5.
Reconstitution	Not Applicable

Data



> 90 % as determined by reducing SDS-PAGE.

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 to mouse SMPD1. SMPD1 is a monomeric lysosomal enzyme that converts sphingomyelin (a plasma membrane lipid)

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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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