

## Recombinant Mouse SMPD1/ASM Protein (His Tag)

**Catalog Number:** PKSM040504

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

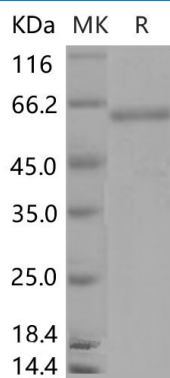
### Description

<b>Species</b>	Mouse
<b>Source</b>	Baculovirus-Insect Cells-derived Mouse SMPD1/ASM protein Met 1-Leu 626, with an C-terminal His
<b>Calculated MW</b>	66.3 kDa
<b>Observed MW</b>	63 kDa
<b>Accession</b>	Q04519
<b>Bio-activity</b>	Measured by its ability to cleave 2-N-Hexadecanoylamino-4-nitrophenylphosphorylcholine(HNPPC). The specific activity is > 1, 500 pmoles/min/μg.

### Properties

<b>Purity</b>	> 85 % as determined by reducing SDS-PAGE.
<b>Endotoxin</b>	< 1.0 EU per μg of the protein as determined by the LAL method.
<b>Storage</b>	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.
<b>Shipping</b>	This product is provided as lyophilized powder which is shipped with ice packs.
<b>Formulation</b>	Lyophilized from sterile 20mM Tris, 500mM NaCl, 10% glycerol, pH 8.0, 0.1% Tween20 Normally 5% - 8% trehalose, mannitol and 0.01% Tween 80 are added as protectants before lyophilization. Please refer to the specific buffer information in the printed manual.
<b>Reconstitution</b>	Please refer to the printed manual for detailed information.

### Data



> 85 % as determined by reducing SDS-PAGE.

### Background

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Toll-free: 1-888-852-8623  
Web: [www.elabscience.com](http://www.elabscience.com)

Tel: 1-832-243-6086  
Email: [techsupport@elabscience.com](mailto:techsupport@elabscience.com)

Fax: 1-832-243-6017

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) 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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