



Recombinant Protein Technical Manual

Recombinant Human SMPD1/ASM Protein (aa 1-631, His Tag)(Active)
RPES1340

Product Data:

Product SKU: RPES1340

Size: 10µg

Species: Human

Expression host: Baculovirus-Insect Cells

Uniprot: NP_000534.3

Protein Information:

Molecular Mass: 65 kDa

AP Molecular Mass: 65 kDa

Tag: C-His

Bio-activity: Measured by its ability to cleave. 2-N-Hexadecanoylamino-4-nitrophenylphosphorylcholine (HNPPC). The specific activity is >1,000 pmol/min/µg.

Purity: > 94 % as determined by reducing SDS-PAGE.

Endotoxin: < 1.0 EU per µg 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: 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.

Application:

Synonyms: ASM;ASMASE;NPD

Immunogen Information:

Sequence: Met 1-Cys 631

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) 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.