
Recombinant Human SMPD1 / ASM Protein (His tag)**Cat.NO.: TP08609**

3th Edition

Synonyms:ASM;ASMASE;NPD

Description: 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.

Form:PBS**Molecular Weight:**65 kDa**Sequences:**Met 1-Cys 631**Purity:**> 95% by HPLC**Concentration:****Endotoxin Level:**<1.0 EU per 1 ug of protein (determined by LAL method)**Storage:**Can be stored at +4°C short term (1-2 weeks). For long term storage, aliquot and store at -20°C or -70°C. Avoid repeated freezing and thawing cycles.