Acid sphingomyelinase Polyclonal Antibody

Catalog:	BS65948	Host:	Rabbit	Reactivity:	Human,Mouse,Rat,Dog,Pig,Cow,Rabbit,

BackGround:

Converts sphingomyelin to ceramide. Also has phospholipase C activities toward 1,2-diacylglycerolphosphocholine and 1,2-diacylglycerolphosphoglycerol. Isoform 2 and isoform 3 have lost catalytic activity.

Involvement in disease: Defects in SMPD1 are the cause of Niemann-Pick disease type A (NPDA) ; also known as Niemann-Pick disease classical infantile form. It is an early-onset lysosomal storage disorder caused by failure to hydrolyze sphingomyelin to ceramide. It results in the accumulation of sphingomyelin and other metabolically related lipids in reticuloendothelial and other cell types throughout the body, leading to cell death. Niemann-Pick disease type A is a primarily neurodegenerative disorder characterized by onset within the first year of life, mental retardation, digestive disorders, failure to thrive, major hepatosplenomegaly, and severe neurologic symptoms. The severe neurological disorders and pulmonary infections lead to an early death, often around the age of four. Clinical features are variable. A phenotypic continuum exists between type A (basic neurovisceral) and type B (purely visceral) forms of Niemann-Pick disease, and the intermediate types encompass a cluster of variants combining clinical features of both types A and B

Product:

0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

Molecular Weight:

64kDa

Swiss-Prot:

P17405

Purification&Purity:

affinity purified by Protein A

Applications:

Flow-Cyt=2ug/Test

Storage&Stability:

Store at 4℃ short term. Aliquot and store at -20℃ long term. Avoid freeze-thaw cycles.

Specificity:

Acid sphingomyelinase Polyclonal Antibody detects endogenous levels of Acid sphingomyelinase protein.

DATA:

Note:

For research use only, not for use in diagnostic procedure.

Bioworld Technology, Inc.
Add: 1660 South Highway 100, Suite 500 St. Louis Park, MN 55416,USA.
Email: info@bioworlde.com
Tel: 6123263284
Fax: 6122933841

Bioworld technology, co. Ltd.
Add: No 9, weidi road Qixia District Nanjing, 210046, P. R. China.
Email: info@biogot.com
Tel: 0086-025-68037686
Fax: 0086-025-68035151