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bs-6318R-PE-Cy3

- ## Rabbit Anti-ASM/Acid sphingomyelinase Polyclonal Antibody, PE-Cy3 conjugated

Conjugated Primary Antibodies

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.

Purification: Was purified by Protein A and peptide affinity chromatography.

Storage:
Prepared as lyophilized powder or liquid and shipped on ice. Store at -20°C for one year. Protect from light.

Reconstitution:
If the antibody is in liquid form, no reconstitution needed.

Reconstitution is only required for the lyophilized antibody. Please refer to the reconstitution instruction card in the package.

For full size images and description please click HERE.

Size: 100ul or 100ug lyophilized

Concentration: 1ug/uL

Host: Rabbit

Reactivities:
Human,Mouse,Rat,Dog,Pig,Cow,Rabbit,

Application:

- IF(1:100-500)
- Not yet tested in other applications. Optimal working dilutions must be determined by the end user.

Antibody Type: Polyclonal

Isotype: IgG

Molecular Weight: 64kDa

Preservatives:
10ug/uL BSA and 0.1% NaN3.

For research use only. CAUTION: Not for human or animal therapeutic or diagnostic use.