

PRODUCT DATA SHEET

Bioworld Technology,Inc.

Hexb (S516) polyclonal antibody

Catalog: BS1917 Host: Rabbit Reactivity: Human, Mouse, Rat

BackGround:

Hexosaminidase B (HEXB), also designated ta-hexosaminidase B, is a Hexosaminidase B (HEXB), also designated b-hexosaminidase B, is a tetramer of two b-A and two b-B chains and is found in the lysosomes of cells. Sandhoff disease (SD), also known GM2-gangliosidosis type II, is caused by mutations in the HEXB gene that affect the b subunit. These mutations disrupt the activity of HEXB and HEXA, which prevents the breakdown of GM2 ganglioside, a fatty material found in the brain, therby rendering both the HEXA and HEXB enzymes deficient. SD is a rare autosomal recessive disorder characterized by an accumulation of GM2 ganglioside, which causes progressive destruction of the central nervous system. Sandhoff disease is similar to Tay-Sachs disease, which is caused by mutations in the HEXA gene, although SD is more severe.

Product:

1 mg/ml in Phosphate buffered saline (PBS) with 0.05% sodium azide, approx. pH 7.2.

Molecular Weight:

~ 63 kDa

Swiss-Prot:

P07686

Purification&Purity:

The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen and the purity is > 95% (by SDS-PAGE).

Applications:

WB: 1:500~1:1000 IHC: 1:50~1:200

Storage&Stability:

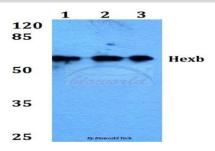
Store at 4 ${\mathbb C}$ short term. Aliquot and store at -20 ${\mathbb C}$ long

term. Avoid freeze-thaw cycles.

Specificity:

Hexb (S516) polyclonal antibody detects endogenous levels of Hexb protein.

DATA:

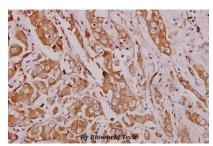


Western blot (WB) analysis of Hexb (S516) polyclonal antibody at 1:500 dilution

Lane1:Jurkat whole cell lysate

Lane2:Raw264.7 whole cell lysate

Lane3:H9C2 whole cell lysate



Immunohistochemistry (IHC) analyzes of Hexb (S516) pAb in paraf-

fin-embedded human breast carcinoma tissue at 1:100.

Note:

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

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