PRODUCT DATA SHEET



Bioworld Technology CO., Ltd.

Hexb (S516) Peptide

Cat No.: BS1917P

Background

(HEXB), Hexosaminidase also designated beta-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 as 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.

Swiss-Prot

P07686

Applications

Blocking

Specificity

This peptide can be used with studies using BS1917 Hexb (S516) pAb.

Purification & Purity

Synthetic peptide Hexb (S516). (Note: the amino acid sequence is proprietary). The purity is > 98%.

Product

1 mg/ml in DI water.

Storage & Stability

Store at $4\,\mathrm{C}$ short term. Aliquot and store at -20 C long term. Avoid freeze-thaw cycles.

Research Use

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