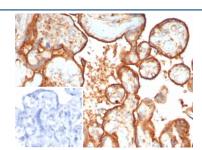


# Hexosaminidase B Antibody / HEXB [clone HEXB/7762] (V5089)

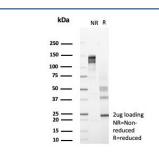
Catalog No.	Formulation	Size
V5089-100UG	0.2~mg/ml in 1X PBS with 0.1 mg/ml BSA (US sourced), 0.05% sodium azide	100 ug
V5089-20UG	0.2 mg/ml in 1X PBS with 0.1 mg/ml BSA (US sourced), 0.05% sodium azide	20 ug
V5089SAF-100UG	1 mg/ml in 1X PBS; BSA free, sodium azide free	100 ug

## **Bulk quote request**

Availability	1-3 business days
Species Reactivity	Human
Format	Purified
Clonality	Monoclonal (mouse origin)
Isotype	Mouse IgG2, kappa
Clone Name	HEXB/7762
Purity	Protein A/G affinity
UniProt	P07686
Localization	Lysosome
Applications	Immunohistochemistry (FFPE): 1-2ug/ml for 30 min at RT
Limitations	This Hexosaminidase B antibody is available for research use only.



IHC staining of FFPE human placental tissue with Hexosaminidase B antibody (clone HEXB/7762). Inset: PBS used in place of primary Ab (secondary Ab negative control). HIER: boil tissue sections in pH 9 10mM Tris with 1mM EDTA for 20 min and allow to cool before testing.



SDS-PAGE analysis of purified, BSA-free Hexosaminidase B antibody (clone HEXB/7762) as confirmation of integrity and purity.

#### **Description**

Hexosaminidase B (HEXB), also designated beta-hexosaminidase B, is a hexosaminidase B (HEXB), also designated beta-hexosaminidase B, is a tetramer of two beta-A and two beta-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 beta 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.

## **Application Notes**

Optimal dilution of the Hexosaminidase B antibody should be determined by the researcher.

#### **Immunogen**

A recombinant fragment of the human protein was used as the immunogen for the Hexosaminidase B antibody.

## **Storage**

Aliquot the Hexosaminidase B antibody and store frozen at -20oC or colder. Avoid repeated freeze-thaw cycles.