SANTA CRUZ BIOTECHNOLOGY, INC.

HEXB B chain (D-9): sc-376781

BACKGROUND

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

CHROMOSOMAL LOCATION

Genetic locus: HEXB (human) mapping to 5q13.3; Hexb (mouse) mapping to 13 D1.

SOURCE

HEXB B chain (D-9) is a mouse monoclonal antibody raised against amino acids 122-166 mapping within an internal region of HEXB of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

HEXB B chain (D-9) is available conjugated to agarose (sc-376781 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-376781 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-376781 PE), fluorescein (sc-376781 FITC), Alexa Fluor® 488 (sc-376781 AF488), Alexa Fluor® 546 (sc-376781 AF546), Alexa Fluor® 594 (sc-376781 AF594) or Alexa Fluor® 647 (sc-376781 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-376781 AF680) or Alexa Fluor® 790 (sc-376781 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

HEXB B chain (D-9) is recommended for detection of HEXB B chain of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for HEXB siRNA (h): sc-60785, HEXB siRNA (m): sc-60786, HEXB shRNA Plasmid (h): sc-60785-SH, HEXB shRNA Plasmid (m): sc-60786-SH, HEXB shRNA (h) Lentiviral Particles: sc-60785-V and HEXB shRNA (m) Lentiviral Particles: sc-60786-V.

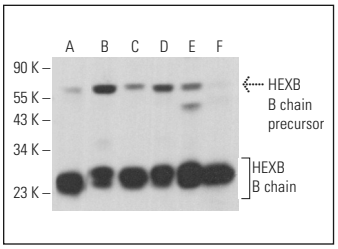
Molecular Weight of HEXB B chain: 63 kDa.

Positive Controls: HEK293 whole cell lysate: sc-45136, Hep G2 cell lysate: sc-2227 or HeLa whole cell lysate: sc-2200.

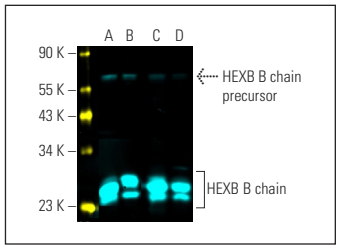
STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

DATA



HEXB B chain (D-9): sc-376781. Western blot analysis of HEXB B chain expression in Hep G2 (**A**), Caco-2 (**B**), NCI-H1299 (**C**), HEK293 (**D**) and WI-38 (**E**) whole cell lysates and human small intestine tissue extract (**F**).



HEXB B chain (D-9) Alexa Fluor® 647: sc-376781 AF647. Direct fluorescent western blot analysis of HEXB B chain expression in Hep G2 (**A**), HeLa (**B**), U-87 MG (**C**) and MCF7 (**D**) whole cell lysates. Blocked with UltraCruz® Blocking Reagent: sc-516214. Cruz Marker™ Molecular Weight Standards detected with Cruz Marker™ MW Tag-Alexa Fluor® 488: sc-516790.

SELECT PRODUCT CITATIONS

1. Mazzulli, J.R., et al. 2016. Activation of β-glucocerebrosidase reduces pathological α-synuclein and restores lysosomal function in Parkinson's patient midbrain neurons. J. Neurosci. 36: 7693-7706.
2. Kim, M.J., et al. 2017. The Parkinson's disease-linked protein TMEM230 is required for Rab8a-mediated secretory vesicle trafficking and retromer trafficking. Hum. Mol. Genet. 26: 729-741.
3. Kim, M.J., et al. 2018. Acid ceramidase inhibition ameliorates α-synuclein accumulation upon loss of GBA1 function. Hum. Mol. Genet. 27: 1972-1988.
4. Pellegrini, D., et al. 2019. Quantitative microproteomics based characterization of the central and peripheral nervous system of a mouse model of Krabbe disease. Mol. Cell. Proteomics 18: 1227-1241.
5. Greaney, A.M., et al. 2020. Platform effects on regeneration by pulmonary basal cells as evaluated by single-cell RNA sequencing. Cell Rep. 30: 4250-4265.e6.
6. Han, J., et al. 2021. Involvement of CASP9 (caspase 9) in IGF2R/CI-MPR endosomal transport. Autophagy 17: 1393-1409.
7. Park, J.H., et al. 2021. Disruption of nucleocytoplasmic trafficking as a cellular senescence driver. Exp. Mol. Med. 53: 1092-1108.
8. Stojkovska, I., et al. 2022. Rescue of α-synuclein aggregation in Parkinson's patient neurons by synergistic enhancement of ER proteostasis and protein trafficking. Neuron 110: 436-451.e11.
9. Kim, M.J., et al. 2022. Lysosomal ceramides regulate cathepsin B-mediated processing of saposin C and glucocerebrosidase activity. Hum. Mol. Genet. 31: 2424-2437.

RESEARCH USE

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