SANTA CRUZ BIOTECHNOLOGY, INC.

HEXB (M-40): sc-134581

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

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.

REFERENCES

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3. Bikker, H., et al. 1989. Demonstration of a Sandhoff disease-associated autosomal 50-kb deletion by field inversion gel electrophoresis. Hum. Genet. 81: 287-288.
4. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 606873. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
5. Yamamoto, N. and Urade, M. 2005. Pathogenic significance of α-N-acetylgalactosaminidase activity found in the hemagglutinin of influenza virus. Microbes Infect. 7: 674-681.
6. Sanon, A., et al. 2005. N-acetyl-β-D-hexosaminidase from *Trichomonas vaginalis*: substrate specificity and activity of inhibitors. Biomed. Pharmacother. 59: 245-248.
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CHROMOSOMAL LOCATION

Genetic locus: Hexb (mouse) mapping to 13 D1.

SOURCE

HEXB (M-40) is a rabbit polyclonal antibody raised against amino acids 387-426 mapping within an internal region of HEXB of mouse origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

APPLICATIONS

HEXB (M-40) is recommended for detection of HEXB of mouse and rat origin by Western Blotting (starting dilution 1:200, 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).

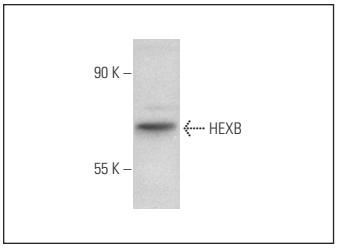
Suitable for use as control antibody for HEXB siRNA (m): sc-60786, HEXB shRNA Plasmid (m): sc-60786-SH and HEXB shRNA (m) Lentiviral Particles: sc-60786-V.

Molecular Weight of HEXB: 63 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



HEXB (M-40): sc-134581. Western blot analysis of HEXB expression in Hep G2 whole cell lysate.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.