

HEXB (R-15): sc-48535

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, thereby 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

1. Beutler, E., Kuhl, W. and Comings, D. 1975. Hexosaminidase isozyme in type O GM2 gangliosidosis (sandhoff-Jatzkewitz disease). *Am. J. Hum. Genet.* 27: 628-638.
2. O'Dowd, B.F., Quan, F., Willard, H.F., Lamhonwah, A.M., Korneluk, R.G., Lowden, J.A., Gravel, R.A. and Mahuran, D.J. 1985. Isolation of cDNA clones coding for the β subunit of human β -hexosaminidase. *Proc. Natl. Acad. Sci. USA* 82: 1184-1188.
3. Bikker, H., van den Berg, F.M., Wolterman, R.A., de Vijlder, J.J. and Bolhuis, P.A. 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-acetyl-galactosaminidase activity found in the hemagglutinin of influenza virus. *Microbes Infect.* 7: 674-681.
6. Sanon, A., Tournaire-Arellano, C., El Hage, S.Y., Bories, C., Caujolle, R. and Loiseau, P.M. 2005. N-acetyl- β -D-hexosaminidase from *Trichomonas vaginalis*: substrate specificity and activity of inhibitors. *Biomed. Pharmacother.* 59: 245-248.
7. Casal, J.A., Cano, E. and Tutor, J.C. 2005. β -hexosaminidase isoenzyme profiles in serum, plasma, platelets and mononuclear, polymorphonuclear and unfractionated total leukocytes. *Clin. Biochem.* 38: 938-942.

CHROMOSOMAL LOCATION

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

SOURCE

HEXB (R-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of HEXB of human origin.

STORAGE

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

PRODUCT

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

Blocking peptide available for competition studies, sc-48535 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

HEXB (R-15) is recommended for detection of HEXB of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

HEXB (R-15) is also recommended for detection of HEXB in additional species, including canine, bovine, porcine and avian.

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: 63 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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.