

HEXB A chain (H-40): sc-134580

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

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CHROMOSOMAL LOCATION

Genetic locus: HEXB (human) mapping to 5q13.3.

SOURCE

HEXB A chain (H-40) is a rabbit polyclonal antibody raised against amino acids 408-447 mapping within an internal region of HEXB of human origin.

PRODUCT

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

APPLICATIONS

HEXB A chain (H-40) is recommended for detection of HEXB A chain of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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).

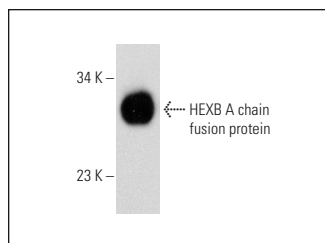
Suitable for use as control antibody for HEXB siRNA (h): sc-60785, HEXB shRNA Plasmid (h): sc-60785-SH and HEXB shRNA (h) Lentiviral Particles: sc-60785-V.

Molecular Weight of HEXB A chain: 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 A chain (H-40): sc-134580. Western blot analysis of human recombinant HEXB A chain fusion protein.

STORAGE

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

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.