SANTA CRUZ BIOTECHNOLOGY, INC.

HEXA (D-2): sc-376777

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Hexosaminidase A (HEXA), also designated β-hexosaminidase A, is a trimer composed of one α chain, one β-A chain and one β-B chain and is found in the lysosomes of cells. HEXA, along with the cofactor GM2 activator protein, catalyzes the degradation of GM2 ganglioside and other molecules containing terminal N-acetyl hexosamines in the brain and other tissues. A mutation in the α subunit of hexosaminidase is the cause of Tay-Sachs disease (TSD), also known as GM2-gangliosidosis type I. TSD is a fatal autosomal recessive lysosomal storage disease of the central nervous system (CNS) caused by insufficient activity of the HEXA enzyme that results in a failure to process GM2 gangliosides. The accumulation of GM2 ganglioside in the absence of HEXA activity causes progressive destruction of the CNS.

CHROMOSOMAL LOCATION

Genetic locus: HEXA (human) mapping to 15q23; Hexa (mouse) mapping to 9 B.

SOURCE

HEXA (D-2) is a mouse monoclonal antibody raised against amino acids 377-416 mapping within an internal region of HEXA 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.

HEXA (D-2) is available conjugated to agarose (sc-376777 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-376777 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-376777 PE), fluorescein (sc-376777 FITC), Alexa Fluor® 488 (sc-376777 AF488), Alexa Fluor® 546 (sc-376777 AF546), Alexa Fluor® 594 (sc-376777 AF594) or Alexa Fluor® 647 (sc-376777 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-376777 AF680) or Alexa Fluor® 790 (sc-376777 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

HEXA (D-2) is recommended for detection of HEXA 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), immunohistochemistry (including paraffin-embedded sections) (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 HEXA siRNA (h): sc-60783, HEXA siRNA (m): sc-60784, HEXA shRNA Plasmid (h): sc-60783-SH, HEXA shRNA Plasmid (m): sc-60784-SH, HEXA shRNA (h) Lentiviral Particles: sc-60783-V and HEXA shRNA (m) Lentiviral Particles: sc-60784-V.

Molecular Weight of HEXA precursor: 67 kDa.

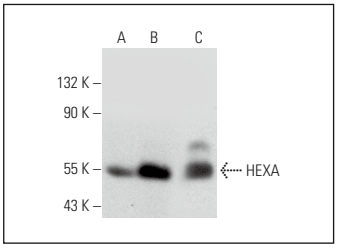
Molecular Weight of mature HEXA: 54 kDa.

Positive Controls: PANC-1 whole cell lysate: sc-364380, Hep G2 cell lysate: sc-2227 or U-937 cell lysate: sc-2239.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



HEXA (D-2): sc-376777. Western blot analysis of HEXA expression in Hep G2 (**A**), U-937 (**B**) and PANC-1 (**C**) whole cell lysates.

SELECT PRODUCT CITATIONS

1. Bedia, C., et al. 2019. GM2-GM3 gangliosides ratio is dependent on GRP94 through down-regulation of GM2-AP cofactor in brain metastasis cells. Sci. Rep. 9: 14241.
2. Brekk, O.R., et al. 2020. Upregulating β-hexosaminidase activity in rodents prevents α-synuclein lipid associations and protects dopaminergic neurons from α-synuclein-mediated neurotoxicity. Acta Neuropathol. Commun. 8: 127.
3. Davis, S.E., et al. 2023. Patients with sporadic FTLD exhibit similar increases in lysosomal proteins and storage material as patients with FTD due to GRN mutations. Acta Neuropathol. Commun. 11: 70.

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 for detailed protocols and support products.