

# HEXA (D-2): sc-376777

## BACKGROUND

Hexosaminidase A (HEXA), also designated  $\beta$ -hexosaminidase A, is a trimer composed of one  $\alpha$  chain, one  $\beta$ -A chain and one  $\beta$ -B chain and is found in the lysosomes of cells. HEXA, along with the cofactor CM2 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  $\alpha$  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  $\mu$ g IgG<sub>2b</sub> 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  $\mu$ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-376777 HRP), 200  $\mu$ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-376777 PE), fluorescein (sc-376777 FITC), Alexa Fluor<sup>®</sup> 488 (sc-376777 AF488), Alexa Fluor<sup>®</sup> 546 (sc-376777 AF546), Alexa Fluor<sup>®</sup> 594 (sc-376777 AF594) or Alexa Fluor<sup>®</sup> 647 (sc-376777 AF647), 200  $\mu$ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor<sup>®</sup> 680 (sc-376777 AF680) or Alexa Fluor<sup>®</sup> 790 (sc-376777 AF790), 200  $\mu$ g/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  $\mu$ g per 100-500  $\mu$ g 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.

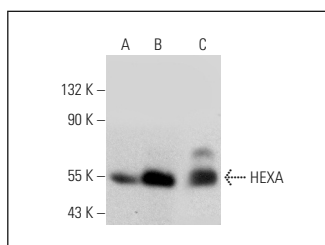
Molecular Weight of mature HEXA: 54 kDa.

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-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker<sup>™</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> 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-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz<sup>®</sup> Mounting Medium: sc-24941 or UltraCruz<sup>®</sup> Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgG $\kappa$  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

- 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.
- Brekke, O.R., et al. 2020. Upregulating  $\beta$ -hexosaminidase activity in rodents prevents  $\alpha$ -synuclein lipid associations and protects dopaminergic neurons from  $\alpha$ -synuclein-mediated neurotoxicity. *Acta Neuropathol. Commun.* 8: 127.

## 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](http://www.scbt.com) for detailed protocols and support products.

Alexa Fluor<sup>®</sup> is a trademark of Molecular Probes, Inc., Oregon, USA