

HEXA (E-2): sc-376735

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 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 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.

REFERENCES

1. Triggs-Raine, B.L., et al. 1991. Sequence of DNA flanking the exons of the HEXA gene, and identification of mutations in Tay-Sachs disease. *Am. J. Hum. Genet.* 49: 1041-1054.
2. Yamanaka, S., et al. 1994. Targeted disruption of the Hexa gene results in mice with biochemical and pathologic features of Tay-Sachs disease. *Proc. Natl. Acad. Sci. USA* 91: 9975-9979.
3. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 606869. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
4. Sanon, A., et al. 2005. N-acetyl- β -D-hexosaminidase from *Trichomonas vaginalis*: sub and activity of inhibitors. *Biomed. Pharmacother.* 59: 245-248.

CHROMOSOMAL LOCATION

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

SOURCE

HEXA (E-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 μ g IgG_{2a} lambda light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

HEXA (E-2) is available conjugated to agarose (sc-376735 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-376735 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-376735 PE), fluorescein (sc-376735 FITC), Alexa Fluor® 488 (sc-376735 AF488), Alexa Fluor® 546 (sc-376735 AF546), Alexa Fluor® 594 (sc-376735 AF594) or Alexa Fluor® 647 (sc-376735 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-376735 AF680) or Alexa Fluor® 790 (sc-376735 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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RESEARCH USE

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

APPLICATIONS

HEXA (E-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 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 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.

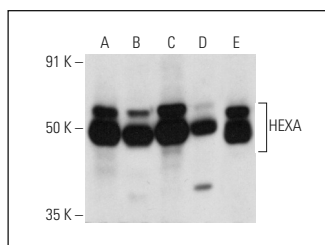
Molecular Weight of mature HEXA: 54 kDa.

Positive Controls: human liver extract: sc-363766, human stomach extract: sc-363780 or HeLa whole cell lysate: sc-2200.

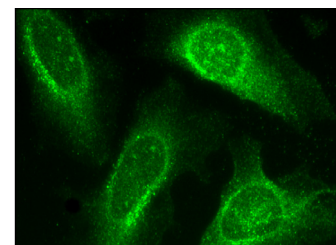
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG λ BP-HRP: sc-516132 or m-IgG λ BP-HRP (Cruz Marker): sc-516132-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-IgG λ BP-FITC: sc-516185 or m-IgG λ BP-PE: sc-516186 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



HEXA (E-2): sc-376735. Western blot analysis of HEXA expression in human liver (A), human stomach (B), human adrenal gland (C) and human brain (D) tissue extracts and HeLa whole cell lysate (E).



HEXA (E-2): sc-376735. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization.

SELECT PRODUCT CITATIONS

1. Liu, J., et al. 2021. CREG1 promotes lysosomal biogenesis and function. *Autophagy* 17: 4249-4265.

STORAGE

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