

HEXA (H-40): sc-134577

BACKGROUND

Hexosaminidase A (HEXA), also designated β -Hexosaminidase A, a trimer composed of one α chain, one β -A chain and one β -B chain, 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

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2. Yamanaka, S., Johnson, M.D., Grinberg, A., Westphal, H., Crawley, J.N., Taniike, M., Suzuki, K. and Proia, R.L. 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/>
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5. 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.
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CHROMOSOMAL LOCATION

Genetic locus: HEXA (human) mapping to 15q23.

SOURCE

HEXA (H-40) is a rabbit polyclonal 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 in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

APPLICATIONS

HEXA (H-40) is recommended for detection of HEXA 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), 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 shRNA Plasmid (h): sc-60783-SH and HEXA shRNA (h) Lentiviral Particles: sc-60783-V.

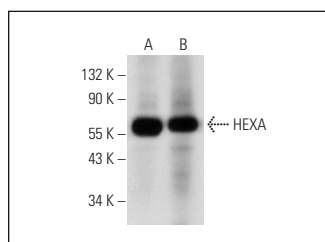
Molecular Weight of HEXA precursor: 67 kDa.

Molecular Weight of mature HEXA: 54 kDa.

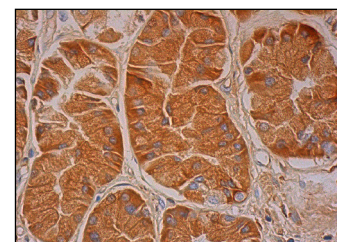
RECOMMENDED SECONDARY REAGENTS

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. 4) Immunohistochemistry: use ImmunoCruz[™]: sc-2051 or ABC: sc-2018 rabbit IgG Staining Systems.

DATA



HEXA (H-40): sc-134577. Western blot analysis of HEXA expression in rat liver (A) and mouse liver (B) tissue extracts.



HEXA (H-40): sc-134577. Immunoperoxidase staining of formalin fixed, paraffin-embedded human upper stomach tissue showing cytoplasmic staining of glandular cells.

RESEARCH USE

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

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Try **HEXA (D-2): sc-376777** or **HEXA (E-2): sc-376735**, our highly recommended monoclonal alternatives to HEXA (H-40).