

HEXA (N-19): sc-48530

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.

CHROMOSOMAL LOCATION

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

SOURCE

HEXA (N-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus 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.

Blocking peptide available for competition studies, sc-48530 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

HEXA (N-19) is recommended for detection of HEXA of mouse, rat and 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).

HEXA (N-19) is also recommended for detection of HEXA in additional species, including equine.

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.

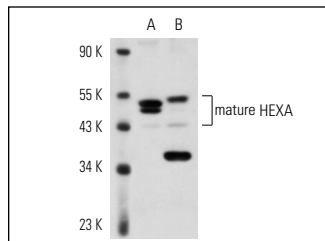
Molecular Weight of mature HEXA: 54 kDa.

Positive Controls: rat liver extract: sc-2395, mouse liver extract: sc-2256 or 3611-RF whole cell lysate: sc-2215.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



HEXA (N-19): sc-48530. Western blot analysis of mature HEXA expression in rat liver (A) and mouse liver (B) tissue extracts.

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.



Try **HEXA (D-2): sc-376777** or **HEXA (E-2): sc-376735**, our highly recommended monoclonal alternatives to HEXA (N-19).