

# HEXA (C-14): sc-48526

## 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.

## REFERENCES

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## CHROMOSOMAL LOCATION

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

## SOURCE

HEXA (C-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of HEXA of human origin.

## 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.

## PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-48526 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

HEXA (C-14) is recommended for detection of HEXA of human and, to a lesser extent, mouse and rat origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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.

## 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) 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.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.