

# GM2-AP (F-14): sc-68088

## BACKGROUND

GM2-AP (GM2 ganglioside activator), also known as GM2A or SAP-3 (shingolipid activator protein 3), is a 193 amino acid protein that localizes to the lysosome. Existing as a small glycolipid transport protein, GM2-AP acts as a ganglioside-specific co-factor that, together with  $\beta$ -hexosaminidase A (HEXA), stimulates the breakdown of glycolipid GA2 and ganglioside GM2 and is important for the degradation of proteins containing terminal N-acetyl hexosamines. Mutations in the gene encoding GM2-AP are the cause of GM2-gangliosidosis type AB (GM2GAB), which is also known as tay-sachs disease AB variant, and is an autosomal recessive disease that is characterized by ganglioside GM2 accumulation in the presence of both hexosaminidase A and B.

## REFERENCES

- Schröder, M., et al. 1989. Isolation of a cDNA encoding the human GM2 activator protein. *FEBS Lett.* 251: 197-200.
- Xie, B., et al. 1991. Isolation and expression of a full-length cDNA encoding the human GM2 activator protein. *Biochem. Biophys. Res. Commun.* 177: 1217-1223.
- Klima, H., et al. 1991. Characterization of full-length cDNAs and the gene coding for the human GM2 activator protein. *FEBS Lett.* 289: 260-264.
- Schröder, M., et al. 1991. A mutation in the gene of a glycolipid-binding protein (GM2 activator) that causes GM2-gangliosidosis variant AB. *FEBS Lett.* 290: 1-3.
- Nagarajan, S., et al. 1992. Evidence for two cDNA clones encoding human GM2-activator protein. *Biochem. J.* 282: 807-813.
- Xie, B., et al. 1992. Identification of a processed pseudogene related to the functional gene encoding the GM2 activator protein: localization of the pseudogene to human chromosome 3 and the functional gene to human chromosome 5. *Genomics* 14: 796-798.
- Schröder, M., et al. 1993. Molecular genetics of GM2-gangliosidosis AB variant: a novel mutation and expression in BHK cells. *Hum. Genet.* 92: 437-440.
- Chen, B., et al. 1999. Structure of the GM2A gene: identification of an exon 2 nonsense mutation and a naturally occurring transcript with an in-frame deletion of exon 2. *Am. J. Hum. Genet.* 65: 77-87.

## CHROMOSOMAL LOCATION

Genetic locus: GM2A (human) mapping to 5q33.1; Gm2a (mouse) mapping to 11 B1.3.

## SOURCE

GM2-AP (F-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of GM2-AP 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.

## 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-68088 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

GM2-AP (F-14) is recommended for detection of GM2-AP of mouse, rat and human 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).

GM2-AP (F-14) is also recommended for detection of GM2-AP in additional species, including equine, bovine and porcine.

Suitable for use as control antibody for GM2-AP siRNA (h): sc-62385, GM2-AP siRNA (m): sc-62386, GM2-AP shRNA Plasmid (h): sc-62385-SH, GM2-AP shRNA Plasmid (m): sc-62386-SH, GM2-AP shRNA (h) Lentiviral Particles: sc-62385-V and GM2-AP shRNA (m) Lentiviral Particles: sc-62386-V.

Molecular Weight of GM2-AP: 21 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.

## 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) or our catalog for detailed protocols and support products.



Try **GM2-AP (E-7): sc-514437**, our highly recommended monoclonal alternative to GM2-AP (F-14).