

ASM (4H2): sc-293189

BACKGROUND

Acid sphingomyelinase (ASM) is a lysosomal protein that hydrolyzes sphingomyelin to ceramide and phosphocholine. The ASM gene encodes three proteins, ASM-1, ASM-2 and ASM-3, of which ASM-1 is the only ASM gene product that is a catalytically active enzyme. Deficiency of ASM is associated with type A and type B Niemann-Pick disease. Type A is a fatal neurodegenerative disorder seen in infancy and resulting in death by age three, whereas type B is a non-neuropathic disease that has a later onset. During monocytic cell differentiation, the expression of ASM is upregulated by the combined actions of AP-2 and Sp1 transcription factors.

REFERENCES

- Quintern, L.E., et al. 1987. Acid sphingomyelinase from human urine: purification and characterization. *Biochim. Biophys. Acta* 922: 323-336.
- Schuchman, E.H., et al. 1991. Human acid sphingomyelinase. Isolation, nucleotide sequence and expression of the full-length and alternatively spliced cDNAs. *J. Biol. Chem.* 266: 8531-8539.
- Levrn, O., et al. 1991. Niemann-Pick disease: a frequent missense mutation in the acid sphingomyelinase gene of Ashkenazi Jewish type A and B patients. *Proc. Natl. Acad. Sci. USA* 88: 3748-3752.
- Takahashi, T., et al. 1992. Identification and expression of five mutations in the human acid sphingomyelinase gene causing types A and B Niemann-Pick disease. Molecular evidence for genetic heterogeneity in the neuronopathic and non-neuronopathic forms. *J. Biol. Chem.* 267: 12552-12558.
- Langmann, T., et al. 1999. Transcription factors Sp1 and AP2 mediate induction of acid sphingomyelinase during monocytic differentiation. *J. Lipid Res.* 40: 870-880.

CHROMOSOMAL LOCATION

Genetic locus: SMPD1 (human) mapping to 11p15.4.

SOURCE

ASM (4H2) is a mouse monoclonal antibody raised against amino acids 1-364 representing partial length ASM of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain 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.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.

APPLICATIONS

ASM (4H2) is recommended for detection of ASM of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for ASM siRNA (h): sc-41650, ASM shRNA Plasmid (h): sc-41650-SH and ASM shRNA (h) Lentiviral Particles: sc-41650-V.

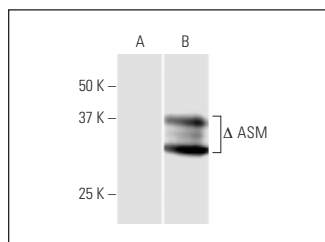
Molecular Weight of ASM: 72 kDa.

Molecular Weight of ASM cleavage product: 57 kDa.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:
 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-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).

DATA



ASM (4H2): sc-293189. Western blot analysis of ASM expression in non-transfected (A) and truncated ASM (aa 1-364) transfected (B) 293T whole cell lysates.

RESEARCH USE

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