

ASM (m2): 293T Lysate: sc-126454

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

1. Quintern, L.E., Weitz, G., Nehrkorn, H., Tager, J.M., Schram, A.W. and Sandhoff, K. 1987. Acid sphingomyelinase from human urine: purification and characterization. *Biochim. Biophys. Acta* 922: 323-336.
2. Schuchman, E.H., Suchi, M., Takahashi, T., Sandhoff, K. and Desnick, R.J. 1991. Human acid sphingomyelinase. Isolation, nucleotide sequence and expression of the full-length and alternatively spliced cDNAs. *J. Biol. Chem.* 266: 8531-8539.
3. Levran, O., Desnick, R.J. and Schuchman, E.H. 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.
4. Takahashi, T., Suchi, M., Desnick, R.J., Takada, G. and Schuchman, E.H. 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.
5. Langmann, T., Buechler, C., Ries, S., Schaeffler, A., Aslanidis, C., Schuierer, M., Weiler, M., Sandhoff, K., de Jong, P.J. and Schmitz, G. 1999. Transcription factors Sp1 and AP-2 mediate induction of acid sphingomyelinase during monocytic differentiation. *J. Lipid Res.* 40: 870-880.

CHROMOSOMAL LOCATION

Genetic locus: Smpd1 (mouse) mapping to 7 E3.

PRODUCT

ASM (m2): 293T Lysate represents a lysate of mouse ASM transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

ASM (m2): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive ASM antibodies. Recommended use: 10-20 µl per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

RESEARCH USE

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

PROTOCOLS

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