

ASA (h5): 293 Lysate: sc-158283

BACKGROUND

ASA (arylsulfatase A), also known as cerebroside-sulfatase, ARSA or MLD, is a 507 amino acid lysosomal protein that belongs to the sulfatase family. Functioning as a homodimer at a neutral pH and as a homooctamer at an acidic pH, ASA uses magnesium as a cofactor to catalyze the H₂O-dependent hydrolysis of cerebroside 3-sulfate to cerebroside and sulfate. Defects in the gene encoding ASA are a cause of metachromatic leukodystrophy (MLD), an intralysosomal storage disease that is characterized by ataxias, dementia, seizures, spastic tetraparesis and, ultimately, death. Additionally, defects in ASA activity are associated with multiple sulfatase deficiency (MSD), a disorder that results in decreased activity of all known sulfatases and is generally characterized by metachromatic leukodystrophy, mucopolysaccharidosis, chondrodysplasia punctata, hydrocephalus, ichthyosis, neurologic deterioration and developmental delay.

REFERENCES

1. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 607574. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
2. Jean, S., et al. 2006. Ethanol decreases rat hepatic arylsulfatase A activity levels. *Alcohol. Clin. Exp. Res.* 30: 1950-1955.
3. Jimenez, I., et al. 2006. Carbohydrate affinity chromatography indicates that arylsulfatase-A from capacitated boar sperm has mannose and N-acetylglucosamine/sialic acid residues. *Arch. Androl.* 52: 455-462.
4. Biffi, A., et al. 2006. Gene therapy of metachromatic leukodystrophy reverses neurological damage and deficits in mice. *J. Clin. Invest.* 116: 3070-3082.
5. Sevin, C., et al. 2007. Partial cure of established disease in an animal model of metachromatic leukodystrophy after intracerebral adeno-associated virus-mediated gene transfer. *Gene Ther.* 14: 405-414.
6. Consiglio, A., et al. 2007. Metabolic correction in oligodendrocytes derived from metachromatic leukodystrophy mouse model by using encapsulated recombinant myoblasts. *J. Neurol. Sci.* 255: 7-16.
7. Saravanan, K., et al. 2007. A spontaneously immortalized Schwann cell line to study the molecular aspects of metachromatic leukodystrophy. *J. Neurosci. Methods* 161: 223-233.
8. Bhattacharyya, S. and Tobacman, J.K. 2007. Steroid sulfatase, arylsulfatases A and B, galactose-6-sulfatase, and iduronate sulfatase in mammary cells and effects of sulfated and non-sulfated estrogens on sulfatase activity. *J. Steroid Biochem. Mol. Biol.* 103: 20-34.
9. Kurai, T., et al. 2007. AAV1 mediated co-expression of formylglycine-generating enzyme and arylsulfatase a efficiently corrects sulfatide storage in a mouse model of metachromatic leukodystrophy. *Mol. Ther.* 15: 38-43.

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.

CHROMOSOMAL LOCATION

Genetic locus: ARSA (human) mapping to 22q13.33.

PRODUCT

ASA (h5): 293 Lysate represents a lysate of human ASA transfected 293 cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

ASA (h5): 293 Lysate is suitable as a Western Blotting positive control for human reactive ASA antibodies. Recommended use: 10-20 µl per lane.

Control 293 Lysate: sc-110760 is available as a Western Blotting negative control lysate derived from non-transfected 293 cells.

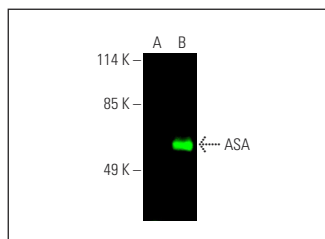
ASA (H-5): sc-365176 is recommended as a positive control antibody for Western Blot analysis of enhanced human ASA expression in ASA transfected 293 cells (starting dilution 1:100, dilution range 1:100-1:1,000).

RECOMMENDED SUPPORT REAGENTS

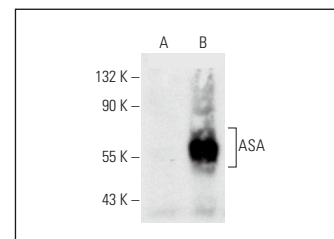
To ensure optimal results, the following support reagents are recommended:

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.

DATA



ASA (H-5): sc-365176. Near-Infrared western blot analysis of ASA expression in non-transfected: sc-110760 (A) and human ASA transfected: sc-158283 (B) 293 whole cell lysates. Blocked with UltraCruz® Blocking Reagent: sc-516214. Detection reagent used: m-IgGκ BP-CFL 680: sc-516180.



ASA (H-5): sc-365176. Western blot analysis of ASA expression in non-transfected: sc-110760 (A) and human ASA transfected: sc-158283 (B) 293 whole cell lysates.

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.