

ASA (H-129): sc-68914

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 homo-octamer 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.

CHROMOSOMAL LOCATION

Genetic locus: ARSA (human) mapping to 22q13.33; Arsa (mouse) mapping to 15 E3.

SOURCE

ASA (H-129) is a rabbit polyclonal antibody raised against amino acids 145-273 mapping within an internal region of ASA of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

ASA (H-129) is recommended for detection of ASA of mouse, rat and 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)], 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).

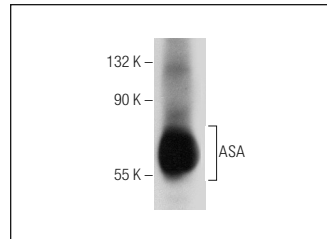
ASA (H-129) is also recommended for detection of ASA in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for ASA siRNA (h): sc-72542, ASA siRNA (m): sc-72543, ASA shRNA Plasmid (h): sc-72542-SH, ASA shRNA Plasmid (m): sc-72543-SH, ASA shRNA (h) Lentiviral Particles: sc-72542-V and ASA shRNA (m) Lentiviral Particles: sc-72543-V.

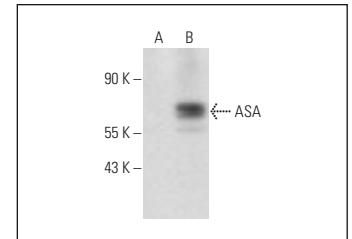
Molecular Weight of ASA: 62 kDa.

Positive Controls: mouse liver extract: sc-2256 or HeLa whole cell lysate: sc-2200.

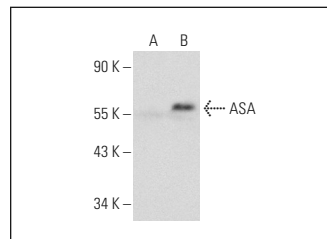
DATA



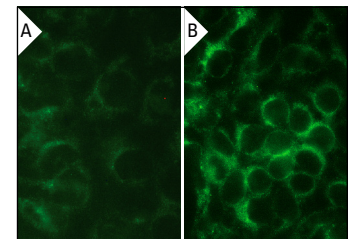
ASA (H-129): sc-68914. Western blot analysis of ASA expression in mouse liver tissue extract.



ASA (H-129): sc-68914. Western blot analysis of ASA expression in non-transfected: sc-110760 (A) and human ASA transfected: sc-158283 (B) 293 whole cell lysates.



ASA (H-129): sc-68914. Western blot analysis of ASA expression in non-transfected: sc-117752 (A) and mouse ASA transfected: sc-118578 (B) 293T whole cell lysates.



ASA (H-129): sc-68914. Immunofluorescence staining of methanol-fixed untransfected (A) and human ASA transfected HEK 293T cells (B).

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.