



SZABO SCANDIC

Part of Europa Biosite

Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

Weitere Information auf den folgenden Seiten!
See the following pages for more information!



Lieferung & Zahlungsart

siehe unsere [Liefer- und Versandbedingungen](#)

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

[linkedin.com/company/szaboscandic](https://www.linkedin.com/company/szaboscandic) 

ASA (m2): 293T Lysate: sc-118579

BACKGROUND

ASA (arylsulfatase A), also known as cerebrosidase-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 cerebrosidase 3-sulfate to Cerebrosidase 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.

CHROMOSOMAL LOCATION

Genetic locus: Arsa (mouse) mapping to 15 E3.

RESEARCH USE

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

PRODUCT

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

APPLICATIONS

ASA (m2): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive ASA 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.

PROTOCOLS

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