



# SZABO SCANDIC

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## Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

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### Lieferung & Zahlungsart

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### Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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# ASS1 (m): 293T Lysate: sc-126455

## BACKGROUND

ASS1, also known as argininosuccinate synthase or citrulline-aspartate ligase, belongs to the argininosuccinate synthase family. ASS1 is an urea cycle enzyme with a tetrameric structure composed of identical subunits. It is important to the urea cycle as it catalyzes the important second last step in the arginine biosynthetic pathway. A deficiency of ASS1 causes citrullinemia (CTLN1), an autosomal recessive disease which is characterized by severe vomiting spells and mental retardation.

## REFERENCES

1. Bock, H.G., et al. 1983. Sequence for human argininosuccinate synthetase cDNA. *Nucleic Acids Res.* 11: 6505-6512.
2. Freytag, S.O., et al. 1984. Molecular structures of human argininosuccinate synthetase pseudogenes. Evolutionary and mechanistic implications. *J. Biol. Chem.* 259: 3160-3166.
3. Isashiki, Y., et al. 1989. Identification of essential arginine residue(s) for Mg-ATP binding of human argininosuccinate synthetase. *Protein Seq. Data Anal.* 2: 283-287.
4. Haberle, J., et al. 2002. Structure of the human argininosuccinate synthetase gene and an improved system for molecular diagnostics in patients with classical and mild citrullinemia. *Hum. Genet.* 110: 327-333.
5. Bansal, V., et al. 2004. Citrulline can preserve proliferation and prevent the loss of CD3- $\zeta$  chain under conditions of low arginine. *JPEN J. Parenter. Enteral Nutr.* 28: 423-430.
6. Hao, G., et al. 2004. Argininosuccinate synthetase is reversibly inactivated by S-nitrosylation *in vitro* and *in vivo*. *J. Biol. Chem.* 279: 36192-36200.
7. Ito, S., et al. 2004. A pregnant patient with fulminant hepatic failure was found to carry a novel missense mutation in the argininosuccinate synthetase gene. *J. Gastroenterol.* 39: 1115-1117.
8. Lighthall, G.K., et al. 2004. Identification of salt-sensitive genes in the kidneys of Dahl rats. *J. Hypertens.* 22: 1487-1494.
9. Potter, M.A., et al. 2004. Pregnancy in a healthy woman with untreated citrullinemia. *Am. J. Med. Genet. A* 129A: 77-82.

## CHROMOSOMAL LOCATION

Genetic locus: Ass1 (mouse) mapping to 2 B.

## PRODUCT

ASS1 (m): 293T Lysate represents a lysate of mouse ASS1 transfected 293T cells and is provided as 100  $\mu$ g protein in 200  $\mu$ l SDS-PAGE buffer.

## 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](http://www.scbt.com) for detailed protocols and support products.

## APPLICATIONS

ASS1 (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive ASS1 antibodies. Recommended use: 10-20  $\mu$ l per lane.

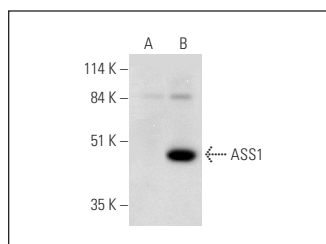
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

ASS1 (C-4): sc-514726 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse ASS1 expression in ASS1 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

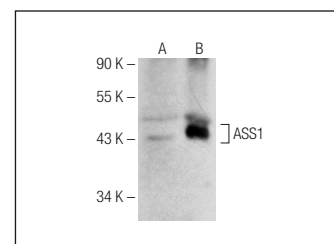
## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:  
 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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



ASS1 (C-4): sc-514726. Western blot analysis of ASS1 expression in non-transfected: sc-117752 (A) and mouse ASS1 transfected: sc-126455 (B) 293T whole cell lysates.



ASS1 (E-12): sc-365475. Western blot analysis of ASS1 expression in non-transfected: sc-117752 (A) and mouse ASS1 transfected: sc-126455 (B) 293T whole cell lysates.

## RESEARCH USE

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