

ASS1 (C-4): sc-514726

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

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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- ζ 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 (human) mapping to 9q34.11; Ass1 (mouse) mapping to 2 B.

SOURCE

ASS1 (C-4) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 373-400 at the C-terminus of ASS1 of human origin.

PRODUCT

Each vial contains 200 μ g IgM kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-514726 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

ASS1 (C-4) is recommended for detection of ASS1 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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).

Suitable for use as control antibody for ASS1 siRNA (h): sc-45810, ASS1 siRNA (m): sc-45811, ASS1 shRNA Plasmid (h): sc-45810-SH, ASS1 shRNA Plasmid (m): sc-45811-SH, ASS1 shRNA (h) Lentiviral Particles: sc-45810-V and ASS1 shRNA (m) Lentiviral Particles: sc-45811-V.

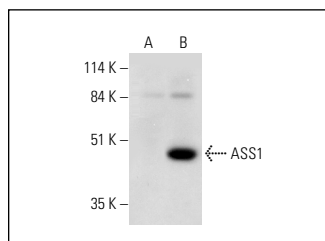
Molecular Weight of ASS1: 47 kDa.

Positive Controls: ASS1 (m): 293T Lysate: sc-126455 or IB4 whole cell lysate: sc-364780.

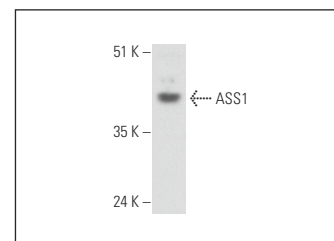
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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. 2) Immunoprecipitation: use Protein L-Agarose: sc-2336 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

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 (C-4): sc-514726. Western blot analysis of ASS1 expression in IB4 whole cell lysate.

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.

PROTOCOLS

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