

ASL (C-15): sc-68249

BACKGROUND

ASL (argininosuccinate lyase), also known as ASAL or arginosuccinase, is a member of the lyase 1 family of proteins and is predominantly expressed in the liver. Localizing to the cytoplasm and existing as a homotetramer, ASL catalyzes the hydrolytic cleavage of argininosuccinic acid (ASA) to fumarate and arginine, an essential step of the urea cycle which is crucial for the detoxification of ammonia. This reaction is also involved in the biosynthesis of arginine. In addition, ASL shares high sequence homology with the avian and reptilian eye lens protein, δ -crystallin. Mutations in the gene encoding ASL lead to an accumulation of ASA in body fluids and result in arginosuccinic aciduria (ASAuria), an autosomal recessive disorder that is characterized by hyperammonemia, liver enlargement, convulsions, physical and mental retardation, episodic unconsciousness and dry and brittle hair showing trichorrhexis nodosa (weak points or nodes in the hair shaft).

REFERENCES

1. Turner, M.A., et al. 1997. Human argininosuccinate lyase: a structural basis for intragenic complementation. *Proc. Natl. Acad. Sci. USA* 94: 9063-9068.
2. Yu, B. and Howell, P.L. 2000. Intragenic complementation and the structure and function of argininosuccinate lyase. *Cell. Mol. Life Sci.* 57: 1637-1651.
3. Sampaleanu, L.M., et al. 2001. Three-dimensional structure of the argininosuccinate lyase frequently complementing allele Q286R. *Biochemistry* 40: 15570-15580.
4. Yu, B., et al. 2001. Mechanisms for intragenic complementation at the human argininosuccinate lyase locus. *Biochemistry* 40: 15581-15590.

CHROMOSOMAL LOCATION

Genetic locus: ASL (human) mapping to 7q11.21; Asl (mouse) mapping to 5 G1.3.

SOURCE

ASL (C-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of ASL of human origin.

PRODUCT

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

Blocking peptide available for competition studies, sc-68249 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

PROTOCOLS

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

APPLICATIONS

ASL (C-15) is recommended for detection of ASL of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

ASL (C-15) is also recommended for detection of ASL in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for ASL siRNA (h): sc-61998, ASL siRNA (m): sc-61999, ASL shRNA Plasmid (h): sc-61998-SH, ASL shRNA Plasmid (m): sc-61999-SH, ASL shRNA (h) Lentiviral Particles: sc-61998-V and ASL shRNA (m) Lentiviral Particles: sc-61999-V.

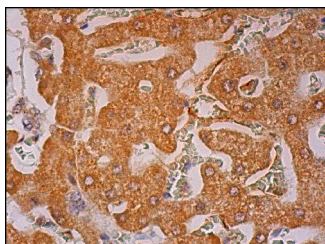
Molecular Weight of ASL: 51 kDa.

Positive Controls: mouse liver extract: sc-2256 or Hep G2 cell lysate: sc-2227.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



ASL (C-15): sc-68249. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes.

RESEARCH USE

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



Try **ASL (B-1): sc-166787** or **ASL (E-5): sc-374353**, our highly recommended monoclonal alternatives to ASL (C-15).