SANTA CRUZ BIOTECHNOLOGY, INC.

ASL (H-300): sc-98619

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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

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4. Yu, B., et al. 2001. Mechanisms for intragenic complementation at the human argininosuccinate lyase locus. Biochemistry 40: 15581-15590.
5. Linnebank, M., et al. 2002. Argininosuccinate lyase (ASL) deficiency: mutation analysis in 27 patients and a completed structure of the human ASL gene. Hum. Genet. 111: 350-359.
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7. Christodoulou, J., et al. 2006. Deletion hotspot in the argininosuccinate lyase gene: association with topoisomerase II and DNA polymerase α sites. Hum. Mutat. 27: 1065-1071.
8. Lee, H.J., et al. 2006. Critical role of tryptophanyl residues in the conformational stability of goose δ-crystallin. Exp. Eye Res. 83: 658-666.
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CHROMOSOMAL LOCATION

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

SOURCE

ASL (H-300) is a rabbit polyclonal antibody raised against amino acids 1-300 mapping at the N-terminus of ASL 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

ASL (H-300) 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), 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).

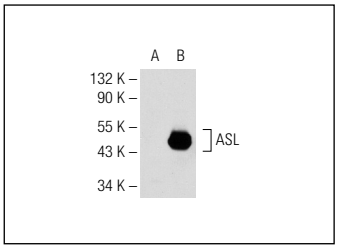
ASL (H-300) is also recommended for detection of ASL in additional species, including equine, canine, bovine and porcine.

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, ASL (h): 293 Lysate: sc-110465 or Hep G2 cell lysate: sc-2227.

DATA



ASL (H-300): sc-98619. Western blot analysis of ASL expression in non-transfected: sc-110760 (**A**) and human ASL transfected: sc-110465 (**B**) 293 whole cell lysates.

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.