SANTA CRUZ BIOTECHNOLOGY, INC.

ASL (E-5): sc-374353

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 leads to an accumulation of ASA in body fluids and results 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., et al. 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 (E-5) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 1-29 at the N-terminus of ASL of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

ASL (E-5) is available conjugated to agarose (sc-374353 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-374353 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-374353 PE), fluorescein (sc-374353 FITC), Alexa Fluor® 488 (sc-374353 AF488), Alexa Fluor® 546 (sc-374353 AF546), Alexa Fluor® 594 (sc-374353 AF594) or Alexa Fluor® 647 (sc-374353 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-374353 AF680) or Alexa Fluor® 790 (sc-374353 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-374353 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

ASL (E-5) is recommended for detection of ASL of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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), 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).

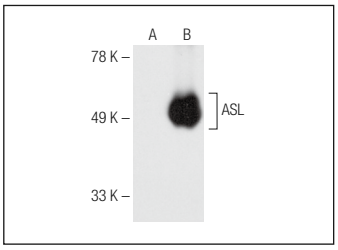
ASL (E-5) is also recommended for detection of ASL in additional species, including canine and porcine.

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.

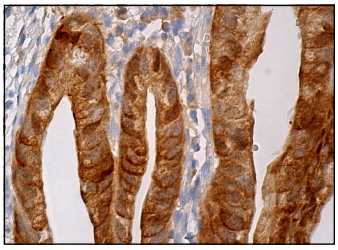
Molecular Weight of ASL: 51 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, mouse liver extract: sc-2256 or ASL (h): 293 Lysate: sc-110465.

DATA



ASL (E-5): sc-374353. Western blot analysis of ASL expression in non-transfected: sc-110760 (**A**) and human ASL transfected: sc-110465 (**B**) 293 whole cell lysates.



ASL (E-5): sc-374353. Immunoperoxidase staining of formalin fixed, paraffin-embedded human gall bladder tissue showing cytoplasmic and nuclear staining of glandular cells.

SELECT PRODUCT CITATIONS

1. Pankowicz, F.P., et al. 2018. Rapid disruption of genes specifically in livers of mice using multiplex CRISPR/Cas9 editing. Gastroenterology 155: 1967-1970.e6.
2. Poillet-Perez, L., et al. 2018. Autophagy maintains tumour growth through circulating arginine. Nature 563: 569-573.
3. Diez-Fernandez, C., et al. 2019. Argininosuccinate neurotoxicity and prevention by creatine in argininosuccinate lyase deficiency: an *in vitro* study in rat 3D organotypic brain cell cultures. J. Inherit. Metab. Dis. 42: 1077-1087.

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.