

# ALAS-H (C-6): sc-137094

## BACKGROUND

5-aminolevulinic acid synthase 1 (ALAS-H) and 2 (ALAS-E) are two isoforms of ALAS, an enzyme catalyzing the first step of the heme biosynthetic pathway in mammals. The erythroid-specific isoenzyme, ALAS-E, regulates the first step of hematopoietic cell differentiation and iron metabolism in the liver. ALAS-H is a housekeeping protein which mediates synthesis of early heme in the mitochondria of most cells. Succinyl CoA associates with ALAS-E in protein conformation change and translocation of ALAS-E into the mitochondria and does not interact with ALAS-H. The ALAS-E 5'-flanking region contains binding sites for nuclear activators such as GATA-1, NF-E2 and EKLf. Since the ALAS gene maps to the X chromosome, mutation of the gene leads to the pyridoxine-refractory X-linked sideroblastic anemia.

## REFERENCES

- Conboy, J.G., et al. 1992. Human erythroid 5-aminolevulinic acid synthase. Gene structure and species-specific differences in alternative RNA splicing. *J. Biol. Chem.* 267: 18753-18758.
- Furuyama, K., et al. 2000. Interaction between succinyl CoA synthetase and the heme-biosynthetic enzyme ALAS-E is disrupted in sideroblastic anemia. *J. Clin. Invest.* 105: 757-764.

## CHROMOSOMAL LOCATION

Genetic locus: ALAS1 (human) mapping to 3p21.2; Alas1 (mouse) mapping to 9 F1.

## SOURCE

ALAS-H (C-6) is a mouse monoclonal antibody raised against amino acids 57-210 mapping near the N-terminus of ALAS-H of mouse origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>1</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

ALAS-H (C-6) is recommended for detection of precursor and mature ALAS-H 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).

Suitable for use as control antibody for ALAS-H siRNA (h): sc-44728, ALAS-H siRNA (m): sc-44729, ALAS-H shRNA Plasmid (h): sc-44728-SH, ALAS-H shRNA Plasmid (m): sc-44729-SH, ALAS-H shRNA (h) Lentiviral Particles: sc-44728-V and ALAS-H shRNA (m) Lentiviral Particles: sc-44729-V.

Molecular Weight of ALAS-H precursor: 71 kDa.

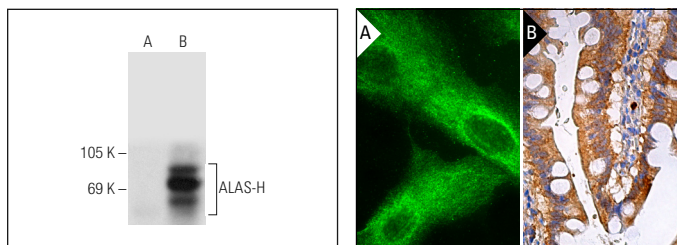
Molecular Weight of mature ALAS-H: 65 kDa.

Positive Controls: JAR cell lysate: sc-2276, K-562 whole cell lysate: sc-2203 or ALAS-H (m): 293T Lysate: sc-118326.

## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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 A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

## DATA



ALAS-H (C-6): sc-137094. Western blot analysis of ALAS-H expression in non-transfected: sc-117752 (A) and mouse ALAS-H transfected: sc-118326 (B) 293T whole cell lysates.

ALAS-H (C-6): sc-137094. Immunofluorescence staining of formalin-fixed Hep G2 cells showing cytoplasmic localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded human small intestine tissue showing cytoplasmic staining of glandular cells (B).

## SELECT PRODUCT CITATIONS

- Bakthavatsalam, D., et al. 2014. Chaperonin-containing TCP-1 complex directly binds to the cytoplasmic domain of the LOX-1 receptor. *FEBS Lett.* 588: 2133-2140.

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