SANTA CRUZ BIOTECHNOLOGY, INC.

ALAS-H (A-6): sc-365153

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

5-aminolevulinate 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 differentation 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-re-fractory X-linked sideroblastic anemia.

REFERENCES

1. Conboy, J.G., et al. 1992. Human erythroid 5-aminolevulinate synthase. Gene structure and species-specific differences in alternative RNA splicing. J. Biol. Chem. 267: 18753-18758.
2. Kramer, M.F., et al. 2000. Transcriptional regulation of the murine erythroid-specific 5-aminolevulinate synthase gene. Gene 247: 153-166.
3. 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.
4. Zhang, J., et al. 2002. Transient state kinetic investigation of 5-amino-levulinate synthase reaction mechanism. J. Biol. Chem. 277: 44660-44669.
5. Zheng, J., et al. 2008. Differential regulation of human ALAS1 mRNA and protein levels by heme and cobalt protoporphyrin. Mol. Cell. Biochem. 319: 153-161.
6. du Plessis, N., et al. 2009. Functional analysis of the 5' regulatory region of the 5-aminolevulinate synthase (ALAS1) gene in response to estrogen. Cell. Mol. Biol. 55: 20-30.
7. Degenhardt, T., et al. 2009. Peroxisome proliferator-activated receptor alpha controls hepatic heme biosynthesis through ALAS1. J. Mol. Biol. 388: 225-238.

CHROMOSOMAL LOCATION

Genetic locus: ALAS1 (human) mapping to 3p21.2.

SOURCE

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

STORAGE

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

APPLICATIONS

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

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

Molecular Weight of ALAS-H precursor: 71 kDa.

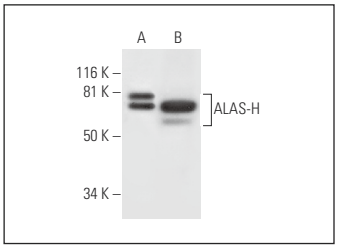
Molecular Weight of mature ALAS-H: 65 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, K-562 whole cell lysate: sc-2203 or HEL 92.1.7 cell lysate: sc-2270.

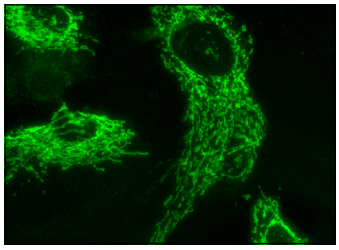
RECOMMENDED SUPPORT REAGENTS

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.

DATA



ALAS-H (A-6): sc-365153. Western blot analysis of ALAS-H expression in HEL 92.1.7 (**A**) and Hep G2 (**B**) whole cell lysates.



ALAS-H (A-6): sc-365153. Immunofluorescence staining of formalin-fixed Hep G2 cells showing mitochondria localization.

SELECT PRODUCT CITATIONS

1. Palasuberniam, P., et al. 2019. Ferrochelatase deficiency abrogated the enhancement of aminolevulinic acid-mediated protoporphyrin IX by iron chelator deferoxamine. Photochem. Photobiol. 95: 1052-1059.
2. Bruhn, C., et al. 2022. Cancer cell histone density links global histone acetylation, mitochondrial proteome and histone acetylase inhibitor sensitivity. Commun. Biol. 5: 882.

RESEARCH USE

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