



ALAS-E (h): 293T Lysate: sc-114245

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 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

1. Conboy, J.G., Cox, T.C., Bottomley, S.S., Bawden, M.J. and May, B.K. 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., Gunaratne, P. and Ferreira, G.C. 2000. Transcriptional regulation of the murine erythroid-specific 5-aminolevulinate synthase gene. *Gene* 247: 153-166.
3. Furuyama, K. and Sassa, S. 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. and Ferreira, G.C. 2002. Transient state kinetic investigation of 5-aminolevulinate synthase reaction mechanism. *J. Biol. Chem.* 277: 44660-44669.

CHROMOSOMAL LOCATION

Genetic locus: ALAS2 (human) mapping to Xp11.21.

PRODUCT

ALAS-E (h): 293T Lysate represents a lysate of human ALAS-E transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

APPLICATIONS

ALAS-E (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive ALAS-E antibodies. Recommended use: 10-20 µl per lane.

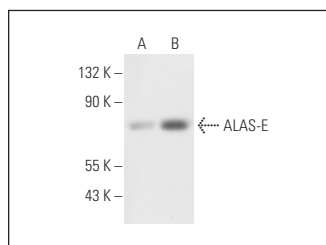
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

ALAS-E (D-4): sc-166739 is recommended as a positive control antibody for Western Blot analysis of enhanced human ALAS-E expression in ALAS-E transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

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.

DATA



ALAS-E (D-4): sc-166739. Western blot analysis of ALAS-E expression in non-transfected: sc-117752 (A) and human ALAS-E transfected: sc-114245 (B) 293T whole cell lysates.

RESEARCH USE

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

PROTOCOLS

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