SANTA CRUZ BIOTECHNOLOGY, INC.

AASS (m): 293T Lysate: sc-118147

BACKGROUND

α-aminoadipic semialdehyde synthase (AASS), also designated lysine ketoglutarate reductase (LKR) or saccharopine dehydrogenase (SDH), is a 926 amino acid protein that exists as a homodimer in the mitochondria. AASS acts as a bifunctional enzyme containing the lysine α-ketoglutarate reductase (LKR) and saccharopine dehydrogenase activities that catalyzes the first two steps in lysine degradation. It is widely expressed with highest expression in liver and transcription of the AASS gene is induced upon starvation. Mutations in the gene encoding AASS result in various forms familial hyperlysinemias (FH), autosomal recessive disorders characterized by hyperlysinemia, lysinuria, and variable saccharopinuria. However, no adverse mental or physical effects have been found in patients with hyperlysinemia.

REFERENCES

1. Dancis, J., Hutzler, J., Woody, N.C. and Cox, R.P. 1976. Multiple enzyme defects in familial hyperlysinemia. Pediatr. Res. 10: 686-691.
2. Markovitz, P.J., Chuang, D.T. and Cox, R.P. 1984. Familial hyperlysinemias. Purification and characterization of the bifunctional aminoadipic semialdehyde synthase with lysine-ketoglutarate reductase and saccharopine dehydrogenase activities. J. Biol. Chem. 259: 11643-11646.
3. Oyanagi, K. and Nagao, M. 1998. Familial hyperlysinemia(α-aminoadipic semialdehyde synthase defect). Ryoikibetsu Shokogun Shirizu 18: 188-190.
4. Sacksteder, K.A., Biery, B.J., Morrell, J.C., Goodman, B.K., Geisbrecht, B.V., Cox, R.P., Gould, S.J. and Geraghty, M.T. 2000. Identification of the α-aminoadipic semialdehyde synthase gene, which is defective in familial hyperlysinemia. Am. J. Hum. Genet. 66: 1736-1743.
5. Praphanphoj, V., Sacksteder, K.A., Gould, S.J., Thomas, G.H. and Geraghty, M.T. 2001. Identification of the α-aminoadipic semialdehyde dehydrogenase-phosphopantetheinyl transferase gene, the human ortholog of the yeast LYS5 gene. Mol. Genet. Metab. 72: 336-342.

CHROMOSOMAL LOCATION

Genetic locus: Aass (mouse) mapping to 6 A3.1.

PRODUCT

AASS (m): 293T Lysate represents a lysate of mouse AASS transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

RESEARCH USE

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

APPLICATIONS

AASS (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive AASS antibodies. Recommended use: 10-20 µl per lane.

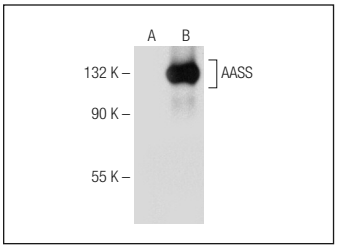
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

AASS (D-8): sc-374322 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse AASS expression in AASS transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

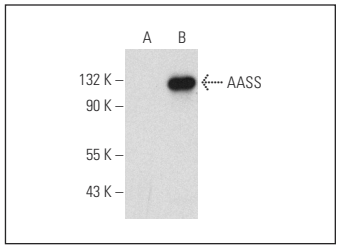
RECOMMENDED SUPPORT REAGENTS

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



AASS (D-8): sc-374322. Western blot analysis of AASS expression in non-transfected: sc-117752 (**A**) and mouse AASS transfected: sc-118147 (**B**) 293T whole cell lysates.



AASS (G-7): sc-390536. Western blot analysis of AASS expression in non-transfected: sc-117752 (**A**) and mouse AASS transfected: sc-118147 (**B**) 293T whole cell lysates.

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.

PROTOCOLS

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