

AASS (G-7): sc-390536



The Power to Question

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

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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 (human) mapping to 7q31.32; Aass (mouse) mapping to 6 A3.1.

SOURCE

AASS (G-7) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 232-253 of AASS of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-390536 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

STORAGE

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

APPLICATIONS

AASS (G-7) is recommended for detection of AASS of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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).

AASS (G-7) is also recommended for detection of AASS in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for AASS siRNA (h): sc-89857, AASS siRNA (m): sc-140738, AASS shRNA Plasmid (h): sc-89857-SH, AASS shRNA Plasmid (m): sc-140738-SH, AASS shRNA (h) Lentiviral Particles: sc-89857-V and AASS shRNA (m) Lentiviral Particles: sc-140738-V.

Molecular Weight (predicted) of AASS: 102 kDa.

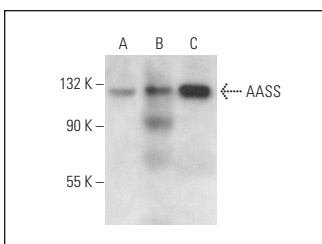
Molecular Weight (observed) of AASS: 116-128 kDa.

Positive Controls: rat liver extract: sc-2395, human liver extract: sc-363766 or AN3 CA cell lysate: sc-24662.

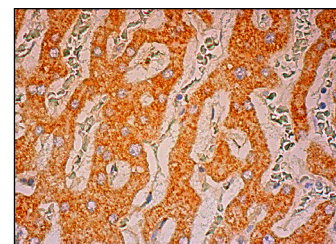
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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-IgG κ BP-FITC: sc-516140 or m-IgG κ 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-IgG κ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



AASS (G-7): sc-390536. Western blot analysis of AASS expression in AN3 CA whole cell lysate (A) and rat liver (B) and human liver (C) tissue extracts.



AASS (G-7): sc-390536. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes.

RESEARCH USE

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