SANTA CRUZ BIOTECHNOLOGY, INC.

AASS (H-5): sc-390511

SANTA CRUZ BIOTECHNOLOGY

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

1. Dancis, J., et al. 1976. Multiple enzyme defects in familial hyperlysinemia. Pediatr. Res. 10: 686-691.
2. Markovitz, P.J., et al. 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.

CHROMOSOMAL LOCATION

Genetic locus: AASS (human) mapping to 7q31.32; Aass (mouse) mapping to 6 A3.1.

SOURCE

AASS (H-5) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 231-253 of AASS of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

AASS (H-5) is available conjugated to agarose (sc-390511 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-390511 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-390511 PE), fluorescein (sc-390511 FITC), Alexa Fluor® 488 (sc-390511 AF488), Alexa Fluor® 546 (sc-390511 AF546), Alexa Fluor® 594 (sc-390511 AF594) or Alexa Fluor® 647 (sc-390511 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-390511 AF680) or Alexa Fluor® 790 (sc-390511 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-390511 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

RESEARCH USE

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

APPLICATIONS

AASS (H-5) 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 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).

AASS (H-5) 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.

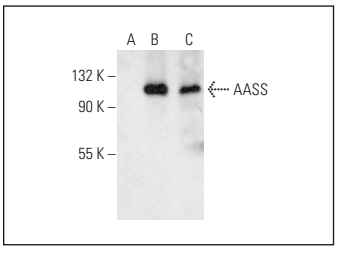
Molecular Weight (observed) of AASS: 116-128 kDa.

Positive Controls: AASS (m): 293T Lysate: sc-118147 or rat liver extract: sc-2395.

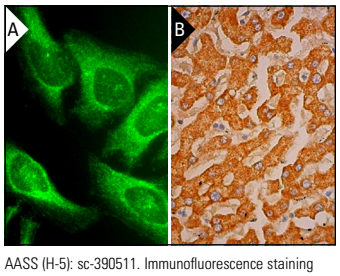
RECOMMENDED SUPPORT REAGENTS

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



AASS (H-5): sc-390511. Western blot analysis of AASS expression in non-transfected: sc-117752 (**A**) and mouse AASS transfected: sc-118147 (**B**) 293T whole cell lysates and rat liver tissue extract (**C**).



AASS (H-5): sc-390511. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes (**B**).

STORAGE

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