

ACADSB (C-9): sc-398773

BACKGROUND

The acyl-CoA dehydrogenase (ACAD) family of enzymes are involved in the catabolism of fatty acids and amino acids. They provide a major source of energy for the heart and skeletal muscle. The short/branched chain specific acyl-CoA dehydrogenase (ACADSB), also designated 2-methylbutyryl-coenzyme A dehydrogenase, is a 432 amino acid protein that is ubiquitously expressed. Specifically, ACADSB forms a homotetramer within the mitochondrial matrix. ACADSB catalyzes the degradation of L-isoleucine and has the highest affinity for (s)-2-methylbutyryl-CoA, isobutyryl-CoA and 2-methylhexanoyl-CoA as substrates. Mutations in the gene encoding ACADSB result in Defects in ACADSB are the cause of short/branched-chain acyl-CoA dehydrogenase deficiency (SBCADD), an autosomal recessive disorder characterized by an increase of 2-methylbutyrylglycine and 2-methylbutyrylcarnitine in blood and urine. Patients with SBCADD have seizures and psychomotor delay as the main clinical features.

REFERENCES

1. Rozen, R., et al. 1994. Isolation and expression of a cDNA encoding the precursor for a novel member (ACADSB) of the acyl-CoA dehydrogenase gene family. *Genomics* 24: 280-287.
2. Arden, K.C., et al. 1995. Localization of short/branched chain acyl-CoA dehydrogenase (ACADSB) to human chromosome 10. *Genomics* 25: 743-745.
3. Korman, S.H., et al. 2005. 2-ethylhydracrylic aciduria in short/branched-chain acyl-CoA dehydrogenase deficiency: application to diagnosis and implications for the R-pathway of isoleucine oxidation. *Clin. Chem.* 51: 610-617.
4. Korman, S.H. 2006. Inborn errors of isoleucine degradation: a review. *Mol. Genet. Metab.* 89: 289-299.

CHROMOSOMAL LOCATION

Genetic locus: ACADSB (human) mapping to 10q26.13.

SOURCE

ACADSB (C-9) is a mouse monoclonal antibody raised against amino acids 368-411 mapping near the C-terminus of ACADSB of human origin.

PRODUCT

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

ACADSB (C-9) is available conjugated to agarose (sc-398773 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-398773 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-398773 PE), fluorescein (sc-398773 FITC), Alexa Fluor® 488 (sc-398773 AF488), Alexa Fluor® 546 (sc-398773 AF546), Alexa Fluor® 594 (sc-398773 AF594) or Alexa Fluor® 647 (sc-398773 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-398773 AF680) or Alexa Fluor® 790 (sc-398773 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

ACADSB (C-9) is recommended for detection of ACADSB of 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for ACADSB siRNA (h): sc-90519, ACADSB shRNA Plasmid (h): sc-90519-SH and ACADSB shRNA (h) Lentiviral Particles: sc-90519-V.

Molecular Weight of ACADSB: 47 kDa.

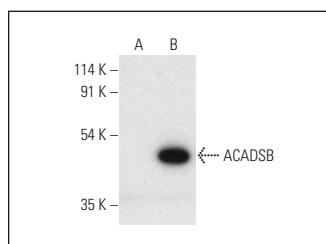
Positive Controls: ACADSB (h): 293T Lysate: sc-113801.

RECOMMENDED SUPPORT REAGENTS

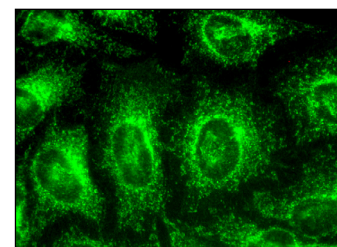
To ensure optimal results, the following support reagents are recommended:

- 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



ACADSB (C-9): sc-398773. Western blot analysis of ACADSB expression in non-transfected: sc-117752 (A) and human ACADSB transfected: sc-113801 (B) 293T whole cell lysates.



ACADSB (C-9): sc-398773. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization.

STORAGE

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

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.