SANTA CRUZ BIOTECHNOLOGY, INC.

ACADSB (G-14): sc-104797

SANTA CRUZ BIOTECHNOLOGY
The Power to Question

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

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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.
5. Kanavin, O.J., et al. 2007. 2-methylbutyryl-CoA dehydrogenase deficiency associated with autism and mental retardation: a case report. J. Med. Case Reports 1: 98.
6. Kamide, K., et al. 2007. Association of genetic polymorphisms of ACADSB and COMT with human hypertension. J. Hypertens. 25: 103-110.
7. Sass, J.O., et al. 2008. 2-Methylbutyryl-coenzyme A dehydrogenase deficiency: functional and molecular studies on a defect in isoleucine catabolism. Mol. Genet. Metab. 93: 30-35.

CHROMOSOMAL LOCATION

Genetic locus: ACADSB (human) mapping to 10q26.13; Acadsb (mouse) mapping to 7 F3.

SOURCE

ACADSB (G-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of ACADSB of human origin.

STORAGE

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

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-104797 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

ACADSB (G-14) is recommended for detection of ACADSB of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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); non cross-reactive with other ACAD family members.

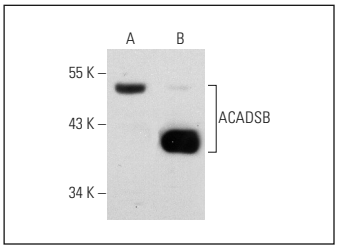
ACADSB (G-14) is also recommended for detection of ACADSB in additional species, including equine, porcine and avian.

Suitable for use as control antibody for ACADSB siRNA (h): sc-90519, ACADSB siRNA (m): sc-140793, ACADSB shRNA Plasmid (h): sc-90519-SH, ACADSB shRNA Plasmid (m): sc-140793-SH, ACADSB shRNA (h) Lentiviral Particles: sc-90519-V and ACADSB shRNA (m) Lentiviral Particles: sc-140793-V.

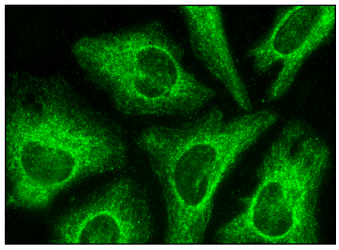
Molecular Weight of ACADSB: 47 kDa.

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

DATA



ACADSB (G-14): sc-104797. Western blot analysis of ACADSB expression in non-transfected: sc-117752 (**A**) and human ACADSB transfected: sc-113801 (**B**) 293T whole cell lysates.



ACADSB (G-14): sc-104797. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization.

RESEARCH USE

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