SANTA CRUZ BIOTECHNOLOGY, INC.

ACADSB (m): 293T Lysate: sc-126373

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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CHROMOSOMAL LOCATION

Genetic locus: Acadsb (mouse) mapping to 7 F3.

PRODUCT

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

APPLICATIONS

ACADSB (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive ACADSB 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.

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.

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.