

ACADS (B-8): sc-365648



The Power to Question

BACKGROUND

ACADS (acyl-Coenzyme A dehydrogenase, C-2 to C-3 short chain), also known as SCAD or ACAD3, is a 412 amino acid homotetrameric mitochondrial flavo-protein that belongs to the acyl-CoA dehydrogenase family. ACADS catalyzes the rate-limiting step of the mitochondrial fatty acid β -oxidation pathway. Mutations of ACADS have been associated with fatty acid oxidation defects and metabolic diseases such as short-chain acyl-CoA dehydrogenase deficiency (SCAD deficiency), an autosomal recessive disorder resulting in acute acidosis and muscle weakness in infants and lipid-storage myopathy in adults. SCADs leads to the accumulation of butyrylcarnitine and ethylmalonic acid in blood and urine. ACADS contains four FAD domains.

REFERENCES

1. Corydon, M.J., et al. 1997. Structural organization of the human short-chain acyl-CoA dehydrogenase gene. *Mamm. Genome* 8: 922-926.
2. Tafti, M., et al. 2003. Deficiency in short-chain fatty acid β -oxidation affects θ oscillations during sleep. *Nat. Genet.* 34: 320-325.
3. Nasser, I., et al. 2004. Thermal unfolding of medium-chain acyl-CoA dehydrogenase and iso(3)valeryl-CoA dehydrogenase: study of the effect of genetic defects on enzyme stability. *Biochim. Biophys. Acta* 1690: 22-32.
4. Ensenauer, R., et al. 2005. Human acyl-CoA dehydrogenase-9 plays a novel role in the mitochondrial β -oxidation of unsaturated fatty acids. *J. Biol. Chem.* 280: 32309-32316.
5. Nagpal, A., et al. 2006. Crystal structures of nitroalkane oxidase: insights into the reaction mechanism from a covalent complex of the flavoenzyme trapped during turnover. *Biochemistry* 45: 1138-1150.
6. van Maldegem, B.T., et al. 2006. Clinical, biochemical, and genetic heterogeneity in short-chain acyl-coenzyme A dehydrogenase deficiency. *JAMA* 296: 943-952.
7. McAndrew, R.P., et al. 2008. Structural basis for substrate fatty acyl chain specificity: crystal structure of human very-long-chain acyl-CoA dehydrogenase. *J. Biol. Chem.* 283: 9435-9443.

CHROMOSOMAL LOCATION

Genetic locus: ACADS (human) mapping to 12q24.31; Acads (mouse) mapping to 5 F.

SOURCE

ACADS (B-8) is a mouse monoclonal antibody raised against amino acids 104-244 mapping within an internal region of ACADS of human origin.

PRODUCT

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

STORAGE

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

APPLICATIONS

ACADS (B-8) is recommended for detection of ACADS 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).

Suitable for use as control antibody for ACADS siRNA (h): sc-96082, ACADS siRNA (m): sc-140792, ACADS shRNA Plasmid (h): sc-96082-SH, ACADS shRNA Plasmid (m): sc-140792-SH, ACADS shRNA (h) Lentiviral Particles: sc-96082-V and ACADS shRNA (m) Lentiviral Particles: sc-140792-V.

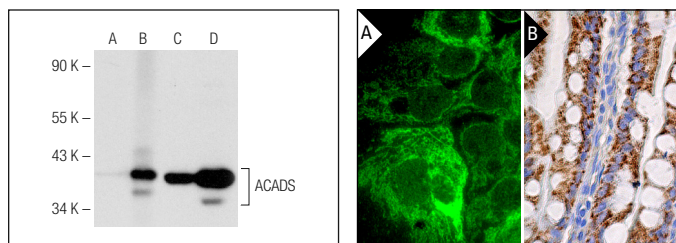
Molecular Weight of ACADS: 42 kDa.

Positive Controls: ACADS (h): 293T Lysate: sc-170175, Hep G2 cell lysate: sc-2227 or mouse kidney extract: sc-2255.

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



ACADS (B-8): sc-365648. Western blot analysis of ACADS expression in non-transfected 293T: sc-117752 (A), human ACADS transfected 293T: sc-170175 (B) and Hep G2 (C) whole cell lysates and mouse kidney tissue extract (D).

ACADS (B-8): sc-365648. Immunofluorescence staining of formalin-fixed Hep G2 cells showing cytoplasmic localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded human small intestine tissue showing cytoplasmic staining of glandular cells (B).

SELECT PRODUCT CITATIONS

1. Stomberski, C.T., et al. 2019. Molecular recognition of S-nitrosothiol substrate by its cognate protein denitrosylase. *J. Biol. Chem.* 294: 1568-1578.

RESEARCH USE

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