SANTA CRUZ BIOTECHNOLOGY, INC.

ACADVL (H-7): sc-376239

BACKGROUND

ACADVL (acyl-coenzyme A dehydrogenase, very long chain), also known as VLCAD, LCACD or ACAD6, is an inner mitochondrial membrane protein that belongs to the family of acyl-CoA dehydrogenases. Involved in lipid metabolism, ACADVL has catalytic activity toward esters of long chain and very long chain fatty acids and functions in the first step of the fatty acid β-oxidation pathway. Defects in the gene encoding ACADVL are the cause of very long chain acyl-CoA dehydrogenase deficiency (VLCAD deficiency), an autosomal recessive disease that leads to impaired long-chain fatty acid oxidation. VLCAD deficiency can give rise to three different phenotypes: a severe childhood form with a high incidence of mortality and cardiomyopathy; a mild childhood form with hypoketotic hypoglycemia and low mortality rates; and an adult form characterized by myoglobinuria and rhabdomyolysis. Two isoforms of ACADVL exist due to alternative splicing events.

REFERENCES

1. Mathur, A., et al. 1999. Molecular heterogeneity in very-long-chain acyl-CoA dehydrogenase deficiency causing pediatric cardiomyopathy and sudden death. Circulation 99: 1337-1343.
2. Andresen, B.S., et al. 1999. Clear correlation of genotype with disease phenotype in very-long-chain acyl-CoA dehydrogenase deficiency. Am. J. Hum. Genet. 64: 479-494.

CHROMOSOMAL LOCATION

Genetic locus: ACADVL (human) mapping to 17p13.1; Acadvl (mouse) mapping to 11 B3.

SOURCE

ACADVL (H-7) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 505-541 within an internal region of ACADVL of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

ACADVL (H-7) is available conjugated to agarose (sc-376239 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-376239 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-376239 PE), fluorescein (sc-376239 FITC), Alexa Fluor® 488 (sc-376239 AF488), Alexa Fluor® 546 (sc-376239 AF546), Alexa Fluor® 594 (sc-376239 AF594) or Alexa Fluor® 647 (sc-376239 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-376239 AF680) or Alexa Fluor® 790 (sc-376239 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-376239 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

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RESEARCH USE

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

APPLICATIONS

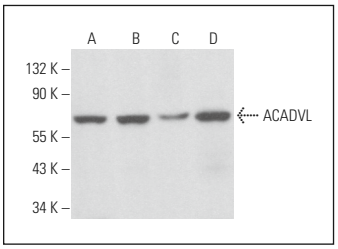
ACADVL (H-7) is recommended for detection of ACADVL of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 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).

Suitable for use as control antibody for ACADVL siRNA (h): sc-72427, ACADVL siRNA (m): sc-72428, ACADVL shRNA Plasmid (h): sc-72427-SH, ACADVL shRNA Plasmid (m): sc-72428-SH, ACADVL shRNA (h) Lentiviral Particles: sc-72427-V and ACADVL shRNA (m) Lentiviral Particles: sc-72428-V.

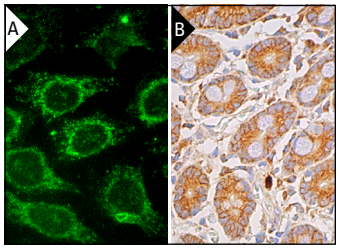
Molecular Weight of ACADVL: 66 kDa.

Positive Controls: K-562 whole cell lysate: sc-2203, Jurkat whole cell lysate: sc-2204 or 3T3-L1 cell lysate: sc-2243.

DATA



ACADVL (H-7): sc-376239. Western blot analysis of ACADVL expression in K-562 (**A**), Jurkat (**B**), 3T3-L1 (**C**) and IB4 (**D**) whole cell lysates.



ACADVL (H-7): sc-376239. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human duodenum tissue showing cytoplasmic staining of glandular cells (**B**).

SELECT PRODUCT CITATIONS

1. Benatti, R.O., et al. 2014. Maternal high-fat diet consumption modulates hepatic lipid metabolism and microRNA-122 (miR-122) and microRNA-370 (miR-370) expression in offspring. Br. J. Nutr. 111: 2112-2122.
2. Hu, G., et al. 2020. Chronic exercise provides renal protective effects with upregulation of fatty acid oxidation in the kidney of high fructose-fed rats. Am. J. Physiol. Renal Physiol. 318: F826-F834.
3. Lu, X., et al. 2023. UBE2M-mediated neddylation of TRIM21 regulates obesity-induced inflammation and metabolic disorders. Cell Metab. 35: 1390-1405.e8.
4. Schmitt, L., et al. 2024. Targeting mitochondrial metabolism by the mito-toxin bromoxib in leukemia and lymphoma cells. Cell Commun. Signal. 22: 541.

STORAGE

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