SANTA CRUZ BIOTECHNOLOGY, INC.

ACADVL (N-16): sc-74900

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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.
3. Tong, M.K., et al. 2006. Very long-chain acyl-CoA dehydrogenase deficiency presenting as acute hypercapnic respiratory failure. Eur. Respir. J. 28: 447-450.
4. Gobin-Limballe, S., et al. 2007. Genetic basis for correction of very-long-chain acyl-coenzyme A dehydrogenase deficiency by bezafibrate in patient fibroblasts: toward a genotype-based therapy. Am. J. Hum. Genet. 81: 1133-1143.

CHROMOSOMAL LOCATION

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

SOURCE

ACADVL (N-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of ACADVL of human origin.

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-74900 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

ACADVL (N-16) is recommended for detection of ACADVL 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), 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).

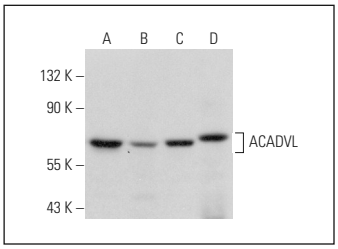
ACADVL (N-16) is also recommended for detection of ACADVL in additional species, including equine, canine, bovine and porcine.

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.

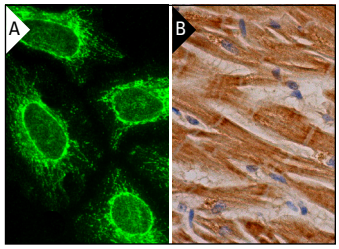
Molecular Weight of ACADVL: 66 kDa.

Positive Controls: K-562 whole cell lysate: sc-2203, HeLa whole cell lysate: sc-2200 or JAR cell lysate: sc-2276.

DATA



ACADVL (N-16): sc-74900. Western blot analysis of ACADVL expression in HeLa (**A**), K-562 (**B**) and JAR (**C**) whole cell lysates and rat skeletal muscle tissue extract (**D**).



ACADVL (N-16): sc-74900. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human heart muscle tissue showing cytoplasmic staining of myocytes (**B**).

SELECT PRODUCT CITATIONS

1. Goichon, A., et al. 2013. An enteral leucine supply modulates human duodenal mucosal proteome and decreases the expression of enzymes involved in fatty acid β-oxidation. J. Proteomics 78: 535-544.

RESEARCH USE

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