SANTA CRUZ BIOTECHNOLOGY, INC.

ACADVL (D-11): sc-271225

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.

CHROMOSOMAL LOCATION

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

SOURCE

ACADVL (D-11) is a mouse monoclonal antibody raised against amino acids 472-580 mapping near the C-terminus 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.

APPLICATIONS

ACADVL (D-11) 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.

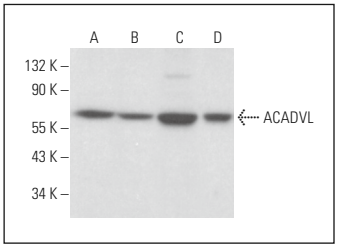
Molecular Weight of ACADVL: 66 kDa.

Positive Controls: K-562 whole cell lysate: sc-2203, HEL 92.1.7 cell lysate: sc-2270 or DU 145 cell lysate: sc-2268.

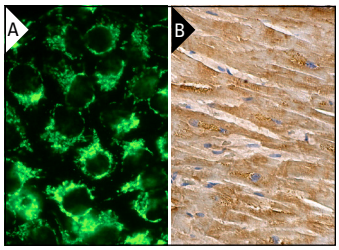
STORAGE

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

DATA



ACADVL (D-11): sc-271225. Western blot analysis of ACADVL expression in K-562 (**A**), HEL 92.1.7 (**B**) and DU 145 (**C**) whole cell lysates and HeLa nuclear extract (**D**).



ACADVL (D-11): sc-271225. Immunofluorescence staining of methanol-fixed HeLa cells showing mytochondrial localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human heart muscle tissue showing cytoplasmic staining of myocytes (**B**).

SELECT PRODUCT CITATIONS

1. Pucer, A., et al. 2013. Group X secreted phospholipase A_2 induces lipid droplet formation and prolongs breast cancer cell survival. Mol. Cancer 12: 111.
2. Obis, E., et al. 2014. Frataxin deficiency in neonatal rat ventricular myocytes targets mitochondria and lipid metabolism. Free Radic. Biol. Med. 73: 21-33.
3. Yang, X.Y., et al. 2015. Energy metabolism disorder as a contributing factor of rheumatoid arthritis: a comparative proteomic and metabolomic study. PLoS ONE 10: e0132695.
4. Dhaenens, L., et al. 2019. Endometrial stromal cell proteome mapping in repeated implantation failure and recurrent pregnancy loss cases and fertile women. Reprod. Biomed. Online 38: 442-454.
5. Xiao, C., et al. 2020. Mitochondrial energetic impairment in a patient with late-onset glutaric acidemia type 2. Am. J. Med. Genet. A 182: 2426-2431.
6. Zhou, H.L., et al. 2023. An enzyme that selectively S-nitrosylates proteins to regulate insulin signaling. Cell 186: 5812-5825.e21.

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.