ACADVL (D-11): sc-271225

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


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 IgGκ 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 lysates)], 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).


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

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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 Lumino Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ 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-IgGκ BP-BP: sc-516102 with DAB, 50X: sc-24982 and Immunohisto Mount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA

SELECT PRODUCT CITATIONS


STORAGE

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

RESEARCH USE

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