

ACAD-8 (A-19): sc-66706

BACKGROUND

ACAD-8 (acyl-CoA dehydrogenase family member 8), also known as isobutyryl-CoA dehydrogenase (IBD) or activator-recruited cofactor 42 kDa component (ARC42), consists of an N-terminal α -helical domain, a β -sheet domain and another α -helical domain at the C-terminal. The ACAD family of enzymes are involved in the catabolism of fatty acids and amino acids. They provide a major source of energy for the heart and skeletal muscle. ACAD-8 is a mitochondrial flavoprotein involved in valine degradation. It is responsible for converting isobutyryl-CoA to methacrylyl-CoA. ACAD-8 localizes to the mitochondrial matrix and exists as a homotetramer. Deficiency of ACAD-8 results in carnitine deficiency, dilated cardiomyopathy and formula feeding intolerance. The excretion of isobutyryl-glycine in urine is a sign of an ACAD-8 related defect.

REFERENCES

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- Zhang, J., et al. 2002. Cloning and functional characterization of ACAD-9, a novel member of human acyl-CoA dehydrogenase family. *Biochem. Biophys. Res. Commun.* 297: 1033-1042.
- Sass, J.O., et al. 2004. Isobutyryl-CoA dehydrogenase deficiency: isobutyryl-glycinuria and ACAD8 gene mutations in two infants. *J. Inher. Metab. Dis.* 27: 741-745.
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CHROMOSOMAL LOCATION

Genetic locus: ACAD8 (human) mapping to 11q25; Acad8 (mouse) mapping to 9 A4.

SOURCE

ACAD-8 (A-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of ACAD-8 of human origin.

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-66706 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

ACAD-8 (A-19) is recommended for detection of ACAD-8 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

ACAD-8 (A-19) is also recommended for detection of ACAD-8 in additional species, including equine and avian.

Suitable for use as control antibody for ACAD-8 siRNA (h): sc-61932, ACAD-8 siRNA (m): sc-61933, ACAD-8 shRNA Plasmid (h): sc-61932-SH, ACAD-8 shRNA Plasmid (m): sc-61933-SH, ACAD-8 shRNA (h) Lentiviral Particles: sc-61932-V and ACAD-8 shRNA (m) Lentiviral Particles: sc-61933-V.

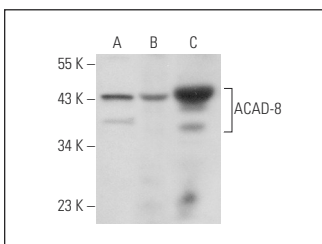
Molecular Weight of ACAD-8: 43 kDa.

Positive Controls: JAR cell lysate: sc-2276, mouse skeletal muscle extract: sc-364250 or rat heart extract: sc-2393.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



ACAD-8 (A-19): sc-66706. Western blot analysis of ACAD-8 expression in JAR whole cell lysate (A) and rat heart (B) and mouse skeletal muscle (C) tissue extracts.

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.