

# ACAD-8 (m): 293T Lysate: sc-118182

## 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  $\alpha$ -helical domain, a  $\beta$ -sheet domain and another  $\alpha$ -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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4. 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.
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6. Battaile, K.P., et al. 2004. Structures of isobutyryl-CoA dehydrogenase and enzyme-product complex: comparison with isovaleryl- and short-chain acyl-CoA dehydrogenases. *J. Biol. Chem.* 279: 16526-16534.
7. Goetzman, E.S., et al. 2005. Convergent evolution of a 2-methylbutyryl-CoA dehydrogenase from isovaleryl-CoA dehydrogenase in *Solanum tuberosum*. *J. Biol. Chem.* 280: 4873-4879.
8. Pedersen, C.B., et al. 2006. Variations in IBD (ACAD8) in children with elevated C4-carnitine detected by tandem mass spectrometry newborn screening. *Pediatr. Res.* 60: 315-320.
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## STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.

## CHROMOSOMAL LOCATION

Genetic locus: *Acad8* (mouse) mapping to 9 A4.

## PRODUCT

ACAD-8 (m): 293T Lysate represents a lysate of mouse ACAD-8 transfected 293T cells and is provided as 100  $\mu$ g protein in 200  $\mu$ l SDS-PAGE buffer.

## APPLICATIONS

ACAD-8 (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive ACAD-8 antibodies. Recommended use: 10-20  $\mu$ l per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

## RESEARCH USE

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