### SANTA CRUZ BIOTECHNOLOGY, INC.

# ACAD-8 (H-165): sc-135147



#### 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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- 3. Nguyen, T.V., Andresen, B.S., Corydon, T.J., Ghisla, S., Abd-El Razik, N., Mohsen, A.W., Cederbaum, S.D., Roe, D.S., Roe, C.R., Lench, N.J. and Vockley, J. 2002. Identification of isobutyryl-CoA dehydrogenase and its deficiency in humans. Mol. Genet. Metab. 77: 68-79.
- 4. Zhang, J., Zhang, W., Zou, D., Chen, G., Wan, T., Zhang, M. and Cao, X. 2002. Cloning and functional characterization of ACAD-9, a novel member of human acyl-CoA dehydrogenase family. Biochem. Biophys. Res. Commun. 297: 1033-1042.
- 5. Sass, J.O., Sander, S. and Zschocke, J. 2004. Isobutyryl-CoA dehydrogenase deficiency: isobutyrylglycinuria and ACAD8 gene mutations in two infants. J. Inherit. 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 (H-165) is a rabbit polyclonal antibody raised against amino acids 251-415 mapping at the C-terminus of ACAD-8 of human origin.

#### PRODUCT

Each vial contains 200 µg lgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

#### **APPLICATIONS**

ACAD-8 (H-165) 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 µg per 100-500 µg 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 (H-165) is also recommended for detection of ACAD-8 in additional species, including equine, canine, bovine and porcine.

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.

#### **RECOMMENDED SECONDARY REAGENTS**

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat antirabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

#### **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.

#### **PROTOCOLS**

See our web site at www.scbt.com or our catalog for detailed protocols and support products.

## MONOS Satisfation Guaranteed

Try ACAD-8 (D-5): sc-390038, our highly recommended monoclonal alternative to ACAD-8 (H-165).