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Produktinformation

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Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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SANTA CRUZ BIOTECHNOLOGY, INC.

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 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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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.
5. Sass, J.O., et al. 2004. Isobutyryl-CoA dehydrogenase deficiency: isobutyrylglycinuria and ACAD8 gene mutations in two infants. J. Inherit. Metab. Dis. 27: 741-745.
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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.
9. Oglesbee,D., et al. 2007. Development of a newborn screening follow-up algorithm for the diagnosis of isobutyryl-CoA dehydrogenase deficiency. Genet. Med. 9: 108-116.

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 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 µg protein in 200 µl 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 µl 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.