isovaleryl-CoA + FAD => beta-methylcrotonyl-CoA + FADH2

European Bioinformatics Institute, New York University Langone Medical Center, Ontario Institute for Cancer Research, Oregon Health and Science University.

18/01/2021

Introduction

Reactome is open-source, open access, manually curated and peer-reviewed pathway database. Pathway annotations are authored by expert biologists, in collaboration with Reactome editorial staff and cross-referenced to many bioinformatics databases. A system of evidence tracking ensures that all assertions are backed up by the primary literature. Reactome is used by clinicians, geneticists, genomics researchers, and molecular biologists to interpret the results of high-throughput experimental studies, by bioinformaticians seeking to develop novel algorithms for mining knowledge from genomic studies, and by systems biologists building predictive models of normal and disease variant pathways.

The development of Reactome is supported by grants from the US National Institutes of Health (P41 HG003751), University of Toronto (CFREF Medicine by Design), European Union (EU STRP, EMI-CD), and the European Molecular Biology Laboratory (EBI Industry program).

Literature references

Fabregat, A., Sidiropoulos, K., Viteri, G., Forner, O., Marin-Garcia, P., Arnau, V. et al. (2017). Reactome pathway analysis: a high-performance in-memory approach. *BMC bioinformatics, 18*, 142. ↗

Sidiropoulos, K., Viteri, G., Sevilla, C., Jupe, S., Webber, M., Orlic-Milacic, M. et al. (2017). Reactome enhanced pathway visualization. *Bioinformatics, 33*, 3461-3467. ↗

Fabregat, A., Jupe, S., Matthews, L., Sidiropoulos, K., Gillespie, M., Garapati, P. et al. (2018). The Reactome Pathway Knowledgebase. *Nucleic Acids Res, 46*, D649-D655. ↗

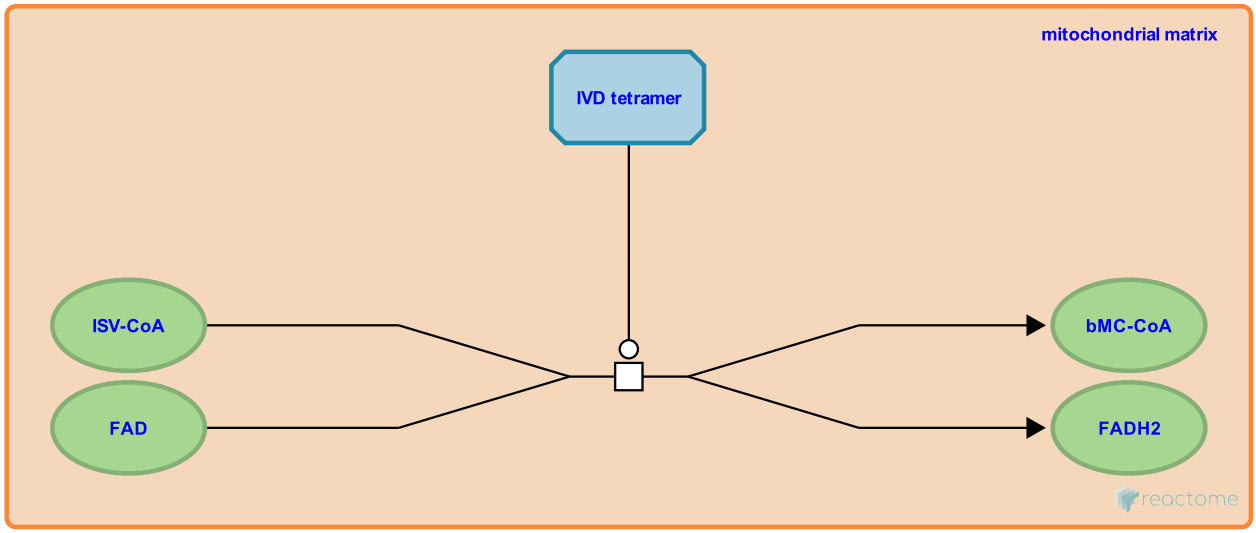
Fabregat, A., Korninger, F., Viteri, G., Sidiropoulos, K., Marin-Garcia, P., Ping, P. et al. (2018). Reactome graph database: Efficient access to complex pathway data. *PLoS computational biology, 14*, e1005968. ↗

isovaleryl-CoA + FAD => beta-methylcrotonyl-CoA + FADH2 ↗

Stable identifier: R-HSA-70745

Type: transition

Compartments: mitochondrial matrix



Mitochondrial isovaleryl dehydrogenase (IVD) catalyzes the reaction of isovaleryl-CoA and FAD to form beta-methylcrotonyl-CoA and FADH2 (Finocchiaro et al. 1978; Rhead and Tanaka 1980). Crystallographic studies demonstrated the existene of a tetramer of IVD polypeptides lacking an aminoterminal mitochondrial targeting sequence (Tiffany et al. 1997).

Literature references

Tiffany, KA., Roberts, DL., Wang, M., Paschke, R., Mohsen, AW., Vockley, J. et al. (1997). Structure of human isovaleryl-CoA dehydrogenase at 2.6 A resolution: structural basis for substrate specificity. *Biochemistry, 36*, 8455-64. ↗

Finocchiaro, G., Ito, M., Tanaka, K. (1987). Purification and properties of short chain acyl-CoA, medium chain acyl-CoA, and isovaleryl-CoA dehydrogenases from human liver. *J Biol Chem, 262*, 7982-9. ↗

Rhead, WJ., Tanaka, K. (1980). Demonstration of a specific mitochondrial isovaleryl-CoA dehydrogenase deficiency in fibroblasts from patients with isovaleric acidemia. *Proc Natl Acad Sci U S A, 77*, 580-3. ↗