TRMT10C:HSD17B10 (TRMT10C:SDR5C1) methylates adenosine-9 in tRNA yielding 1-methyladenosine-9

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17/05/2024

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

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

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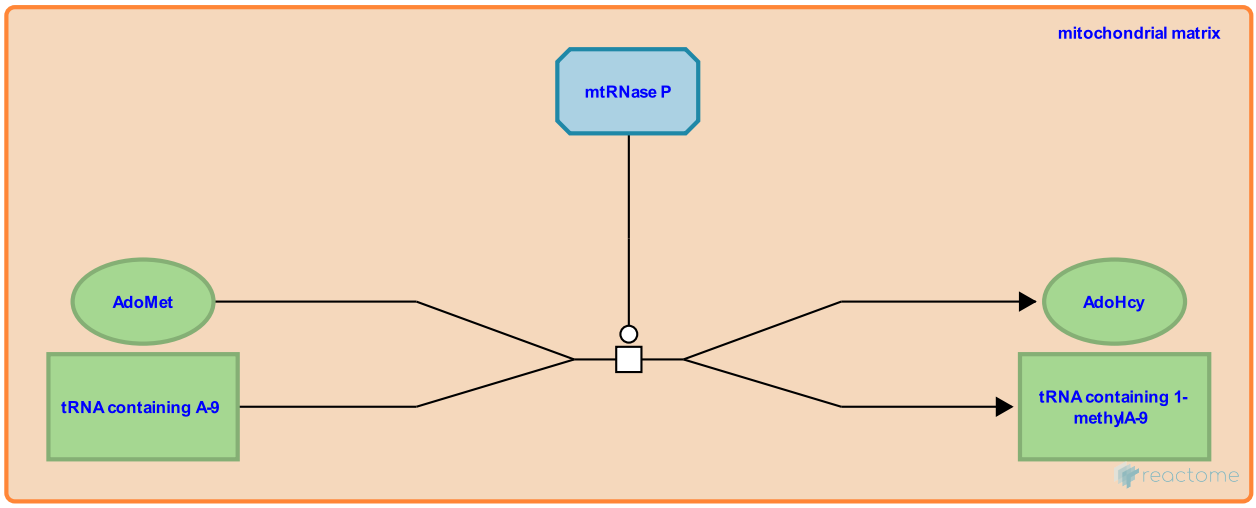
Stable identifier: R-HSA-6787594

Type: transition

Compartments: mitochondrial matrix



TRMT10C of TRMT10C:HSD17B10 (TRMT10C:SDR5C1), a subcomplex of the mitochondrial RNase P complex, methylates the 1 position of adenosine-9 in mitochondrial tRNAs (Vilardo et al. 2012). 14 of 22 mitochondrial tRNAs have an A9 residue. Methylation of A9 appears to be important for correct folding of tRNA (Helm et al. 1998). Mutations in the HSD17B10 (SDR5C1) dehydrogenase subunit of RNase P impair dehydrogenation, tRNA methylation, and tRNA processing, causing HSD10 disease, which is characterized by progressive neurodegeneration and cardiomyopathy (Vilardo and Rossmanith 2015).

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Editions

2015-07-14	Authored, Edited	May, B.
2015-08-11	Reviewed	Levinger, L.
2015-08-25	Reviewed	Motorin, Y.
2015-10-24	Reviewed	Jarrous, N.
2015-11-07	Reviewed	Powell, CA.