Mitochondrial RNase P (mtRNase P) cleaves the 5' ends of pre-tRNAs and ELAC2 (RNase Z) cleaves the 3' ends of pre-tRNAs in the L strand transcript

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28/04/2024

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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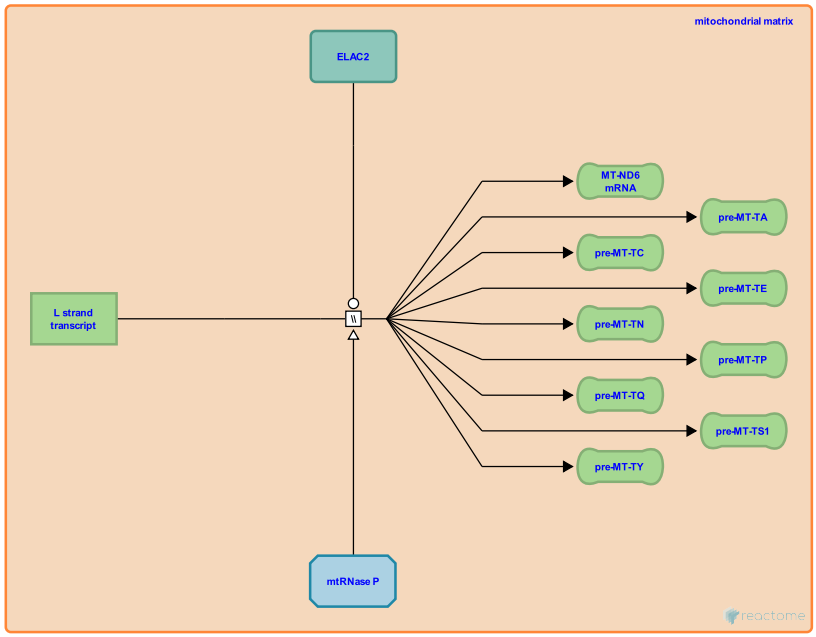
Stable identifier: R-HSA-6786854

Type: omitted

Compartments: mitochondrial matrix



RNase P, ELAC2, and additional unknown nucleases cleave L strand transcripts to release the tRNAs and an mRNA contained in the long polycistronic transcripts.
Mitochondrial RNase P, comprising 3 protein subunits and no RNA moiety (Holzmann et al. 2008), endonucleolytically cleaves polycistronic mitochondrial transcripts at the 5' ends of the tRNA sequences (Sanchez et al. 2011, Howard et al. 2012, Vilardo et al. 2012, Li et al. 2015, Reinhard et al. 2015, Vilardo and Rossmanith 2015). A subcomplex of RNase P also functions as a tRNA methyltransferase and the SDR5C1 subunit is an amino acid and fatty acid dehydrogenase. Mutations in the SDR5C1 subunit of RNase P cause HSD10 disease, which is characterized by progressive neurodegeneration and cardiomyopathy (Vilardo and Rossmanith 2015)
ELAC2 cleaves polycistronic mitochondrial transcripts at the 3' ends of the tRNA sequences (Brzezniak et al. 2011, Sanchez et al. 2011). Different isoforms of ELAC2 are present in the nucleus and mitochondria (Rossmanith 2011). Mutations in ELAC2 cause cardiac hypertrophy (Haack et al. 2013).
Unknown nucleases also cleave the L strand transcript at a site 3' to MT-ND6 (reviewed in Van Haute et al. 2015).

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Editions

2015-07-09	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.
2015-11-18	Reviewed	Suzuki, T.