PUS1 isoform 1 transforms uridine-27, uridine-28 yielding pseudouridine in tRNA(Lys,Ser)

Jarrous, N., Levinger, L., May, B., Motorin, Y., Powell, CA.

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

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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.

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Literature references

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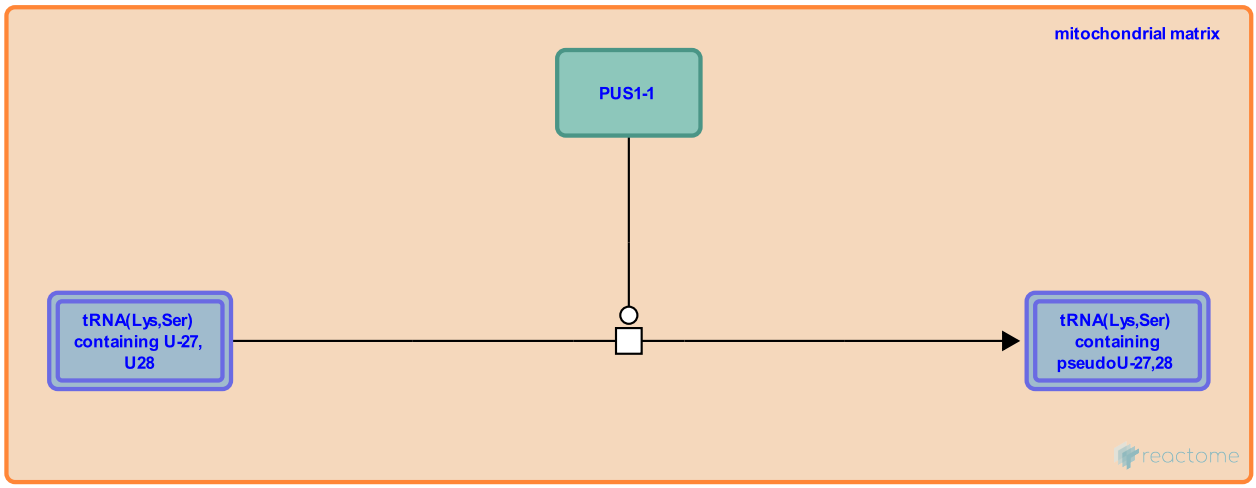
Stable identifier: R-HSA-6787566

Type: transition

Compartments: mitochondrial matrix



PUS1-1, the longer isoform of PUS1 located in the mitochondrion (Fernandez-Vizarra et al. 2007), converts uridine-27 and uridine-28 to pseudouridine residues in the anticodon stems of mitochondrial tRNA(Lys)(UUU) and tRNA(Ser)(UGA) (Patton et al. 2005, Fernandez-Vizarra et al. 2007, Sibert et al. 2008, Sibert and Patton 2012). Isomerization of uracil to pseudouridine creates an extra hydrogen bond donor and increases base stacking, acting to rigidify the RNA structure (reviewed in Charette and Gray 2000). As inferred from yeast Pus1p, PUS1 may also convert uridine to pseudouridine in other tRNAs and pre-tRNAs. Mutations in PUS1 cause mitochondrial myopathy and sideroblastic anemia (MLSA) (Bykhovskaya et al. 2004, Patton et al. 2005, Fernandez-Vizarra et al. 2007)

Literature references

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