Mitochondrion is an important organelle maintaining energy metabolism of the cell and participating in signalization, cell cycle and apoptosis. Its pathology causes several diseases. Replication, transcription and translation take place in mitochondria, similarly like in nucleus, though there are only 13 protein coding genes. However, these processes, as well as mitochondrial DNA and RNA, vary significantly from those present in nucleus. Mitochondrial DNA is circular and both strands are replicated separately. Mitochondria form polycistronic transcripts, which are subsequently processed by tRNA.

Mitochondrial ribosome evolved from prokaryotic one, but contains only half as much rRNA. Missing rRNAs are replaced by ribosomal proteins. These ribosomes contain even more proteins compared to much larger cytoplasmatic eukaryotic ones. This work is focused on current topic of mitochondrial molecular genetics: mitochondrial rRNA and ribosome, especially ribosomal assembly. In this process mitochondrial rRNAs interact with nuclear encoded proteins. The whole process probably takes place on the inner mitochondrial membrane close to the nucleoid. Our understanding to whole mechanism can help us to find a way how to cure mitochondrial pathologies.