综述

线粒体核糖体蛋白与人类线粒体疾病

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摘要

哺乳动物线粒体核糖体(mitochondrial ribosome, mitoribosome)在漫长的进化阶段经过一系列的结构重组，rRNA比例降低，新增了部分线粒体核糖体蛋白(mitochondrial ribosomal proteins， MRPs)，成为蛋白含量最丰富的核糖体。所有MRPs均为核基因编码，在细胞质中合成，再转运到线粒体，与线粒体基因(mitochondrial DNA，mtDNA)编码的两种rRNA结合。mtDNA除编码tRNA和rRNA外，还编码组成线粒体呼吸链复合体的13种蛋白质。由于线粒体核糖体负责翻译这13种蛋白，MRPs和其他翻译工具的突变和缺陷可造成线粒体的相关疾病。

关键词： 线粒体核糖体蛋白；呼吸链复合体亚基；线粒体疾病

Mitochondrial ribosomal proteins and human mitochondrial diseases

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Abstract

Mammalian mitochondrial ribosomes (mitoribosome) have experienced a series of structure recombination during the long period of evolution. Mammalian mitochondrial ribosomes lack several major RNA stem structures of

bacterial ribosomes but they are rich in mitochondrial ribosomal proteins (MRPs). All MRPs are synthesized in cytoplasm and imported into the mitochondrial matrix, where they assemble with the two mtDNA-encoded rRNAs. In addition to tRNA and rRNA, mitochondrial DNA also encodes 13 proteins for the inner mitochondrial membrane respiratory chain complex. The mitoribosome is responsible for the synthesis of these 13 proteins. Thus, mutations or defects of MRPs or other translation tools can cause mitochondrial diseases.

Keywords: mitochondrial ribosomal proteins | respiratory complex subunits | mitochondrial diseases

全文

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引用

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