

Quality control is vital for the energy production of cells

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Mitochondria generate most of the energy cells need through a respiratory chain for which they must produce their own proteins. The research group of Associate Professor Henna Tyynismaa, University of Helsinki, Finland, has discovered a "quality control" mechanism in the mitochondria, which is necessary for the construction of a functional respiratory chain.

Tyynismaa's group studied a mitochondrial enzyme that attaches the correct amino acid to the transfer RNA, which directs protein synthesis. Working together with researchers from the Chinese Academy of Sciences and the University of Turku, the group established that this enzyme makes mistakes and can attach the wrong amino acids to the transfer RNA. This means that the [amino acid sequence](#) in the resulting [respiratory chain](#) proteins would be incorrect, unless the enzyme had an additional mechanism to correct its mistakes.

"We found that the error-correction mechanism of this [mitochondrial enzyme](#) is vital for the cell—even a slight decrease in the correction mechanism had an adverse effect on the function of the cell. The significance of this mechanism for the mitochondria was previously unknown," explains Tyynismaa.

Deviations in the mitochondrial protein synthesis can result in many different hereditary diseases. The enzyme, which is the focus of this research, is also known to harbour disorders caused by genetic anomalies, which may result either in a severe neonatal heart disease or a

brain disease with an onset in early adulthood.

"We do not currently know whether these diseases are the result of the weakened ability of the enzyme to attach the correct amino acids to the transfer RNA or problems in its error-correction mechanism. A better understanding of the enzyme's fundamental mechanisms may help us determine the impact of the faulty genes in the future," says Tyynismaa.

The research was published in the *Nucleic Acids Research* series.

More information: Taru Hilander et al, Editing activity for eliminating mischarged tRNAs is essential in mammalian mitochondria, *Nucleic Acids Research* (2017). [DOI: 10.1093/nar/gkx1231](https://doi.org/10.1093/nar/gkx1231)

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