Research Interests – Prof. Dr. Peter Rehling

Mitochondria play key roles in cellular metabolism and are the main source for ATP generation by oxidative phosphorylation. Hence, it is not surprising that defects in mitochondrial functions lead to severe and often fatal neuromuscular and cardiac disorders. With our work, we aim to understand the biogenesis of mitochondria and concomitantly address how mitochondrial malfunctions lead to human disorders.

Most mitochondrial proteins are nuclear-encoded and imported into mitochondria. However, mitochondria have maintained their personal genome that encodes a small number of proteins, which are the core subunits of the oxidative phosphorylation machinery. Our projects address the molecular mechanisms by which proteins are imported into mitochondria. We analyze the regulation of mitochondrial translation specifically to understand how mitochondrial protein synthesis can adapt to cellular demands. The fact that two genomes contribute to the mitochondrial proteome raises the question as to how protein complexes derived from subunits of dual genetic origin can be assembled into functional units. This aspect of our work has led us into the analysis of model systems for mitochondrial diseases and to address why mitochondrial disorders display a surprising tissue specificity despite a systemic genetic defect.