

## **Abstract**

Invaginations of the inner mitochondrial membrane originate cristae – important structural and bioenergetic mitochondrial compartments. Long-term observations of mitochondrial ultrastructure uncovered cristae dynamics, but did not identify mechanisms of cristae formation and maintenance. This thesis summarizes results of latest research on molecular mechanisms of mitochondrial cristae biogenesis, which are conserved from fungi to mammals including human. The emphasis is put on major remodeling factors:  $F_1F_0$ -ATP synthase dimers, MICOS complex, OPA1 protein and cardiolipin. Their defects lead to extensive changes on cristae level, as well as on mitochondrial, cellular and organismal levels. Various pathophysiological conditions and human mitochondrial diseases are related to these defects. More detailed research of cristae biogenesis is therefore of high significance, new findings could assist in the development of new treatments for mitochondrial disorders.