Laboratory: Bioenergetics

Speaker: Tomáš Mráček

Title: How to study mitochondrial energy production

Annotation:

In our laboratory we study mitochondrial complexes involved in the cellular ATP provision. We are interested in the assembly of those multisubunit protein complexes, detailed aspects of their molecular function and how the cell/organism adapts their dysfunction. In addition to basic research in this area we also have clinical collaborations on inborn mitochondrial disorders and on heart failure. Currently, we can offer projects focussed on ATP synthase and cytochrome c oxidase.

Speaker: Tomáš Mráček, Petr Pecina

Topic: Molecular mechanisms of pathogenicity in ATP synthase disorders, Regulation of mitochondrial oxidative phosphorylation by tissue-specific isoforms of cytochrome *c* oxidase

Annotation:

Mutations in mitochondrial FoF1 ATP synthase responsible for severe inborn errors of metabolism. One of the striking features is the tissue specificity of symptoms associated with mutations in individual subunits. Recently, we have developed animal models for two genes causing ATP synthase disorder – Tmem70 and Usmg5. The aim of this project is to explore differences in tissue presentation as well as compensatory or regulatory mechanisms involved to mitigate pathogenic phenotype.

Expression of tissue-specific isoforms of mitochondrial cytochrome c oxidase (COX) represents a crucial mechanism of regulation of oxidative phosphorylation. COX6B subunit occurs as ubiquitous isoform (COX6B1) or as a protein with exclusive testicular expression (COX6B2). We propose to construct COX6B knock-out/knock-in models in HEK293 cells to characterize their functional features. The role COX6B2 will also be explored in the physiological context of sperm maturation and capacitation.