

period of regular exercise. This is a result of an exercise-induced augmentation in the expression of protein import machinery components. During the aging process, mitochondrial content in muscle declines, and this is reflected in a reduced endurance performance. The decrease in mitochondrial content does not appear to be due to altered post-translational import mechanisms, but may be transcriptionally-related, since PGC-1 α levels are markedly reduced, particularly in slow-twitch muscle fibers. In addition, in response to a standardized acute contractile activity paradigm, signaling kinase activation is increased to a lesser degree than in muscle from young animals, which could lead to a reduced transcriptional activation with age. This likely contributes to the reduced adaptation of aged muscle to regular exercise, consisting of attenuated increases in the expression of biogenesis regulatory proteins of transcription and protein import, reduced increases in mitochondrial enzymes, and lesser improvements in endurance performance. These data suggest that the exercise-induced activation of mitochondrial biogenesis is down-regulated with age. Despite this, adaptive responses to exercise can still occur in aging muscle, leading to reduced fatigability and improved quality of life.

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13L.4 ATP-dependent proteases in biogenesis and maintenance of *Arabidopsis* mitochondria

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It is now widely accepted that tissue- and time-specific control of the quantity and quality of mitochondrial proteins is essential to cope with the challenges of changing developmental and environmental conditions. Recent studies indicate that in plant mitochondria, like in the yeast and animal ones, ATP-dependent proteases are the key components of such control. One of the best characterized plant mitochondrial ATP-dependent protease is FtsH4 from *Arabidopsis*. I will present results which imply that AtFtsH4 is involved in maintaining mitochondrial homeostasis late in rosette development under short-day photoperiod and in plant thermo-tolerance after prolonged exposure to moderately elevated temperature. I will then present some new data on the role of ATP-dependent proteases including AtFtsH4 in coordination of nuclear and mitochondrial genome expression in *Arabidopsis* mutant with impaired mitochondrial translation due to silencing of ribosomal RPS10 gene expression.

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13L.5 RNA turnover in human mitochondria

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Perturbations in functioning of mitochondrial (mt) gene expression have been linked to many human conditions including cancer, aging and neurodegenerative diseases. Therefore research on mechanisms of mt gene expression is of great importance. RNA degradation plays a very important role as it controls three major aspects of RNA metabolism: it determines the half-life of a given RNA species, it destroys the aberrant RNAs that might interfere with replication, transcription or translation in mitochondria, and finally it degrades processing intermediates. In our laboratory we are studying

the human nuclear-encoded proteins: SUV3 helicase, polynucleotide phosphorylase (PNPase) and poly(A) polymerase. A model will be presented which describes this interplay of the proteins in ensuring transcript stability and surveillance in mitochondria. In addition we shall discuss data on their participation in molecular events outside mitochondria: cell cycle control and nuclear chromatin maintenance.

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Posters

13P.1 Transfer of disulfide bonds in biogenesis of mitochondrial intermembrane space proteins

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Mitochondria play a critical role in cellular metabolism and are involved in apoptosis, ageing and a number of diseases. Biogenesis of mitochondrial proteins involves several steps including targeting to mitochondria, transport across the mitochondrial membranes, maturation and associations with partner proteins to form functional complexes ([3]). The novel MIA (Mitochondrial Intermembrane Space Assembly) pathway is essential for the biogenesis of intermembrane space proteins in the entire eukaryotic kingdom ([1,2]). A hallmark of this pathway is the regulated transfer of disulfide bonds, a process that had not been previously described in mitochondria ([1,4]). The MIA pathway represents a novel disulfide-transferring system to control the vectorial translocation of proteins into mitochondria ([7]). Mia40, one of the essential components of this pathway, acts in a receptor-like manner ([5]). It dictates precursor entry into the intermembrane space specifically selecting the proteins that possess a cysteine-containing signal MISS (Mitochondrial Intermembrane Space Signal) ([6]). Furthermore, a mode of the cooperation between Mia40 and the sulfhydryl oxidase Erv1 is unique. We propose that the simultaneous association of Mia40, Erv1 and a substrate protein in the ternary complex allows the efficient transfer of multiple disulfide bonds into substrate proteins ([8]). Our findings have important implications for the biogenesis of mitochondria, generation and transfer of disulfide bonds and their impact on protein compartmentalization and organelle functioning.

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13P.2 Biogenesis of the bacterial *cbb*₃ oxidase of *Rubrivivax gelatinosus*: Evidence for an active core-complex, composed of the catalytic subunit CcoN and the monoheme cytochrome CcoO

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