

P/4 Import and assembly of mitochondrial proteins

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Mitochondria contain about 1000 different proteins. 99% of the proteins are synthesized as precursors on cytosolic ribosomes. The precursors are imported via the translocase of the outer mitochondrial membrane (TOM complex) and are subsequently sorted into the four mitochondrial subcompartments, outer membrane, intermembrane space, inner membrane and matrix. (i) Cleavable preproteins are transported from the TOM complex to the presequence translocase of the inner membrane (TIM23 complex). The presequence translocase-associated motor (PAM) drives translocation into the matrix. (ii) Hydrophobic inner membrane proteins are transferred through the intermembrane space by a chaperone complex (small Tim proteins) and inserted into the inner membrane by the TIM22 complex. (iii) The mitochondrial import and assembly machinery (MIA) directs small proteins into the intermembrane space and promotes the formation of disulfide bonds. (iv) Beta-barrel proteins are transported from the TOM complex to the sorting and assembly machinery of the outer membrane (SAM complex).

[doi:10.1016/j.bbabbio.2008.05.016](https://doi.org/10.1016/j.bbabbio.2008.05.016)**P/5 New functions for novel mitochondrial transporters**

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A strikingly large number of mitochondrial DNA (mtDNA) mutations have been found to be the cause of respiratory chain and oxidative phosphorylation defects. These mitochondrial disorders were the first to be investigated after the small mtDNA had been sequenced in the 80's. Only recently numerous diseases resulting from mutations in nuclear genes encoding mitochondrial proteins have been characterized. Among these, nine are caused by defects of mitochondrial carriers, a family of nuclear-coded proteins that shuttle a variety of metabolites across the mitochondrial membrane. Mutations of mitochondrial carrier genes involved in mitochondrial functions other than oxidative phosphorylation are responsible for carnitine/acylcarnitine carrier deficiency, HHH syndrome, aspartate/glutamate isoform deficiency, Amish microcephaly and neonatal myoclonic epilepsy; these disorders are characterised by specific metabolic dysfunctions, depending on the physiological role of the affected carrier in intermediary metabolism. Defects of mitochondrial carriers that supply mitochondria with the substrates of oxidative phosphorylation, inorganic phosphate and ADP, are responsible for diseases characterised by defective energy production. Herein, all the mitochondrial carrier-associated diseases known to date are reviewed for the first time. Particular emphasis is given to the molecular basis and pathogenetic mechanism of these inherited disorders.

[doi:10.1016/j.bbabbio.2008.05.017](https://doi.org/10.1016/j.bbabbio.2008.05.017)**P/6 The water oxidizing enzyme**

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Photosystem II, the water oxidising enzyme of photosynthesis, put the energy (or at least a major fraction of it) into the biosphere and the oxygen into the atmosphere. It is certainly one the most influential and important enzymes on the planet. The aim of our research is to understand how this enzyme works as 1) a solar energy converter and 2) the only known thermodynamically efficient catalyst for oxidizing water. The information obtained is used in the design of artificial catalysts and photocatalysts. A chemical catalyst that has the thermodynamic efficiency of the enzyme could greatly improve the efficiency of 1) water electrolysis and photolysis for fuel (e.g. H₂) production and 2) the reverse reaction, oxygen reduction, in fuel cells. There is therefore a great interest in understanding the mechanism of this enzyme and in reproducing aspects of its function in artificial systems. I will describe our current knowledge of Photosystem II, including some recent experimental studies, as well as recent efforts in our joint Saclay/Orsay program aimed at producing bio-inspired water oxidizing catalysts.

[doi:10.1016/j.bbabbio.2008.05.018](https://doi.org/10.1016/j.bbabbio.2008.05.018)**P/7 The structure of purple bacterial antenna complexes: From single molecules to native membranes**Richard J. Cogdell^a, Alastair T. Gardiner^a, Mads Gabrielsen^a, Aleks W. Roszak^a, June Southall^a, Tatas Brotosudarmo^a, Neil W. Isaccs^a, Hideki Hashimoto^b, Juergen Baier^c, Silke Oellerich^c, Martin Richter^c, Juergen Koehler^c, Francesco Francia^d, Giovanni Venturoli^d, Dieter Oesterhelt^e^a*Division of Biochemistry and Molecular Biology, IBLS and Department of Chemistry, University of Glasgow, Glasgow G12 8QQ, UK*^b*CREST-JST and Department of Physics, Graduate School of Science, Osaka City University, 3-3-138 Sugimoto, Sumiyoshi-ku, Osaka 558-8585, Japan*^c*Experimental Physics IV, University of Bayreuth, D-95440 Bayreuth, Germany*^d*Department of Biology, University of Bologna, 40126 Bologna, Italy*^e*Department of Membrane Biochemistry, Max-Planck Institute for Biochemistry, 82152 Martinsried, Germany**E-mail: R.Cogdell@bio.gla.ac.uk*

The photosynthetic unit of purple photosynthetic bacteria typically contains two types of light-harvesting complexes, called LH1 and LH2. These antenna complexes are constructed on a modular principle. They are circular or elliptical oligomers of dimers of two low-molecular weight, hydrophobic apoproteins, called a and b, that bind bacteriochlorophylls and carotenoids non-covalently. The LH1 complex surrounds the reaction centre and, depending on the species, is either a monomer or a dimer. The LH2 complexes are arranged around the LH1-RC complexes. This plenary lecture will present the current status of structural studies on these pigment-protein complexes, based upon a combination of X-ray crystallography and single molecule spectroscopy. Then an overall view of how they are arranged in their native photosynthetic membranes will be presented.

[doi:10.1016/j.bbabbio.2008.05.019](https://doi.org/10.1016/j.bbabbio.2008.05.019)**P/8 Catalysis of substrate conversion and electron transfer by mitochondrial complex I**

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