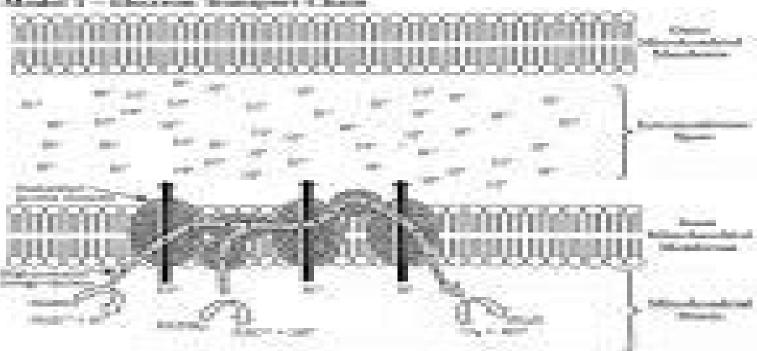


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Oxidative Phosphorylation in Health and Disease Jan A.M. Smeitink,2007-03-06 Mitochondrial diseases are often hard to diagnose From the time they were first researched without animal models patients of mitochondrial diseases were of equal interest to both clinical and basic scientists With the new research done this book includes updates on the normal structure function and molecular biology of the mitochondrial respiratory chain information on traditional diagnostical methodologies and an overview of the diagnostic promise of new technologies The hypermetabolism of Luft disease although only seen twice is also studied There are critical reviews of symptoms and signs associated with syndromes as well as updates on the genetic defects of either the mitochondrial or the nuclear genome responsible for many disorders

Biological Energy Conservation Colin William Jones, 2012-12-06 The student of biological science in his final years as an undergraduate and his first years as a graduate is expected to gain some familiarity with current research at the frontiers of his discipline New research work is published in a perplexing diversity of publications and is inevitably concerned with the minutiae of the subject The sheer number of research journals and papers also causes confusion and difficulties of assimilation Review articles usually presuppose a background know ledge of the field and are inevitably rather restricted in scope There is thus a need for short but authoritative introductions to those areas of modern biological research which are either not dealt with in standard introductory textbooks or are not dealt with in sufficient detail to enable the student to go on from them to read scholarly reviews with profit This series of books is designed to satisfy this need The authors have been asked to produce a brief outline of their subject assuming that their readers will have read and remembered much of a standard introductory textbook on biology This outline then sets out to provide by building on this basis the conceptual framework within which modern research work is progressing and aims to give the reader an indication of the problems both conceptual and practical which must be overcome if progress is to be maintained Mitochondrial Oxidative Phosphorylation Bernhard Kadenbach, 2012-06-23 This book will describe the nuclear encoded genes and their expressed proteins of mitochondrial oxidative phosphorylation Most of these genes occur in eukaryotic cells but not in bacteria or archaea The main function of mitochondria the synthesis of ATP is performed at subunits of proton pumps complexes I III IV and V which are encoded on mitochondrial DNA The nuclear encoded subunits have mostly a regulatory function However the specific physiological functions of the nuclear encoded subunits of complexes I III IV and V are mostly unknown New data indicates that they are essential for life of higher organisms which is characterized by an adult life without cell division postmeiotic stage in most tissues after the juvenile growth For complex IV cytochrome c oxidase some of these subunits occur in tissue specific subunits IV VIa VIb VIIa VIII developmental specific subunits IV VIa and VIIa as well as species specific isoforms Defective genes of some subunits were shown to induce mitochondrial diseases Mitochondrial genes and human diseases will also be covered Oxidative Phosphorylation in Health and Disease Jan A. M. Smeitink, 2004

Biological oxidation U Satyanarayana, 2014-11-07 Biological oxidation Biological oxidation Oxidative Phosphorylation Edward Charles Slater, 1958 Biogenesis of the oxidative phosphorylation machinery in plants. From gene expression to complex assembly Daniel H. Gonzalez, 2014-09-10 Mitochondrial biogenesis is an extremely complex process A hint of this complexity is clearly indicated by the many steps and factors required to assemble the respiratory complexes involved in oxidative phosphorylation These steps include the expression of genes present in both the nucleus and the organelle intricate post transcriptional RNA processing events the coordinated synthesis transport and assembly of the different subunits the synthesis and assembly of co factors and finally the formation of supercomplexes or respirasomes It can be envisaged and current knowledge supports this view that plants have evolved specific mechanisms for the biogenesis of respiratory complexes For example expression of the mitochondrial genome in plants has special features not present in other groups of eukaryotes Moreover plant mitochondrial biogenesis and function should be considered in the context of the presence of the chloroplast a second organelle involved in energetic and redox metabolism It implies the necessity to discriminate between proteins destined for each organelle and requires the establishment of functional interconnections between photosynthesis and respiration In recent years our knowledge of the mechanisms involved in these different processes in plants has considerably increased As a result the many events and factors necessary for the correct expression of proteins encoded in the mitochondrial genome the cis acting elements and factors responsible for the expression of nuclear genes encoding respiratory chain components the signals and mechanisms involved in the import of proteins synthesized in the cytosol and the many factors required for the synthesis and assembly of the different redox co factors heme groups iron sulfur clusters copper centers are beginning to be recognized at the molecular level However detailed knowledge of these processes is still not complete and especially little is known about how these processes are interconnected Questions such as how the proteins once synthesized in the mitochondrial matrix are inserted into the membrane and assembled with other components including those imported from the cytosol how the expression of both genomes is coordinated and responds to changes in mitochondrial function cellular requirements or environmental cues or which factors and conditions influence the assembly of complexes and supercomplexes are still open and will receive much attention in the near future This Research Topic is aimed at establishing a collection of articles that focus on the different processes involved in the biogenesis of respiratory complexes in plants as a means to highlight recent advances In this way it intends to help to construct a picture of the whole process and not less important to expose the existing gaps that need to be addressed to fully understand how plant cells build and modulate the complex structures involved in respiration Studies on Oxidative Phosphorylation in Mitochondrial Systems Lenola Harlene Wellman, 1952 Oxidative Phosphorylation Paul Reilly, 1998-09 Natural and Engineered Strategies that Modulate Oxidative Phosphorylation Defects Associated with Mitochondrial DNA Mutations Sarika Srivastava,2005 Oxidative Phosphorylation in Health and Disease Jan A.M.

Smeitink, 2008-11-01 Mitochondrial diseases are often hard to diagnose From the time they were first researched without animal models patients of mitochondrial diseases were of equal interest to both clinical and basic scientists With the new research done this book includes updates on the normal structure function and molecular biology of the mitochondrial respiratory chain information on traditional diagnostical methodologies and an overview of the diagnostic promise of new technologies The hypermetabolism of Luft disease although only seen twice is also studied There are critical reviews of symptoms and signs associated with syndromes as well as updates on the genetic defects of either the mitochondrial or the nuclear genome responsible for many disorders Biological Energy Conservation: Oxidative Phosphorylation; 2nd Ed Colin William Jones, 1981 Oxidative Phosphorylation in Liver Mitochondria Sylvia Panagos Lianides, 1959 **Phosphorylation by Pea Seedling Mitochondria** Nestor Rosa, 1960 **Evidence for a Soluble Intermediate of** Oxidative Phosphorylation Isolated from Cabbage Mitochondria Morris Alfred Johnson, 1966 An intermediate of oxidative phosphorylation was apparently solubilized from cabbage mitochondria. The method of solubilization used primarily was extraction of a mitochondrial acetone powder with 0 1 M glycylglycine pH 7 4 at 4 C The intermediate betrayed its presence by causing the transfer of Pi32 to ADP with the resultant formation of ATP32 or G 6 P32 when the hexokinase trap was used The formation of ATP32 or G 6 P32 was determined as N E P32 non extractable P32 by the isobutanol benzene extraction procedure The transfer reaction used for assay took place in a completely soluble system and both ADP and extract were required. The historical development of an isolation procedure and of a suitable assay system were described Initial attempts to detect an intermediate were directed at introducing the radioactive label of Pi32 while the mitochondria were still intact or during the extraction of the acetone powder At these stages of the work the assay was also conducted during the extraction of the acetone powder by inclusion of the acceptor system ADP hexokinase and glucose and Mg in the extraction medium Most of the significant facts were obtained through the use of a clarified extract which had not been exposed to Pi32 previously Although it was possible to centrifuge the crude extract at 105 000 X g for 90 min without sedimenting the component necessary for the transfer reaction this high speed centrifugation was not employed on a routine basis The alleged intermediate was apparently proteinaceous as indicated by its response to heat aging dialysis inhibitors and trichloroacetic acid TCA The transfer activity of the extract was lost upon incubation at 50 C for five min upon aging at 4 C upon the addition of TCA and in the presence of 2 X 10 5 M p chloromercuribenzoic acid PCMB Although non dialyzable the extract was inactivated by dialysis probably due to the loss of an essential small molecule such as NAD or NADP Both NAD and NADP stimulated the transfer activity in crude extracts and in extracts which had undergone the centrifugation at 105 000 X g The action of these coenzymes was attributed to their possible role in promoting the labeling of the intermediate from Pi32 In addition to these two activators the rate of the transfer reaction was markedly dependent upon the Pi concentration up to 1 X 10 3 M Pi The yield of intermediate from slightly less than two kilo grams of cabbage was usually 50

100 millimicromoles under favorable assay conditions The probability that this represents less than the maximum yield was considered The transfer activity was rather severely inhibited by 1 X 10 3 M arsenate and partially inhibited by 1 X 10 4 M dinitrophenol DNP it was not sensitive to oligomycin ATP AMP and GDP did not substitute for ADP nor did NADH substitute for NAD The intermediate appeared to be labile when kept in an aqueous system in the presence of Mg Problems posed by such items as the impurity of the Pi32 and by side reactions were discussed The extract also contained succinic thiokinase exchange activity Most of the available evidence including a preliminary ammonium sulfate fractionation would indicate that this exchange was due to a separate protein On the basis of experimental evidence obtained in this study a sequence for the operation of the transfer reaction in the assay system was proposed It was suggested that both the non phosphorylated AH and the phosphorylated XH forms of an intermediate of oxidative phosphorylation were present in the extract It was further speculated that NAD and or NADP functioned as the A of AH and were active in promoting the labeling of the XH form with Pi32 In view of no ready alternate explanation for the transfer activity of the extract it was suggested that this activity merits consideration as the expression of an intermediate of oxidative phosphorylation **Oxidative Phosphorylation in** Mitochondria During Embryonic Development Anna Miriam Henson, 1967 Effect of Fluorescamine and Its Derivatives on Mitochondrial Oxidative Phosphorylation David Da-Wei Shiuan, 1981 Studies on the Efficiency of Oxidative Phosphorylation in Eschericha Coli B Craig Warren Rice, 1980 On the Biological Transfer of Energy. A Theoretical Study of Oxidative Phosphorylation and High Energy Phosphates Barbro Grabe, 1960 **Evidence for a Soluble** High Energy Intermediate of Oxidative Phosphorylation in Mitochondria from Blowflies Albert Yung-Kwang Sun,1967

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