Record Nr. UNINA9910437633203321 Peroxisome biology: experimental models, peroxisomal disorders and **Titolo** neurological diseases / / edited by Gelrard Lizard Pubbl/distr/stampa Cham, Switzerland:,: Springer,, [2020] ©2020 3-030-60204-4 **ISBN** Edizione [1st ed. 2020.] 1 online resource (X, 212 p. 44 illus., 38 illus. in color.) Descrizione fisica Collana Advances in Experimental Medicine and Biology, , 0065-2598; ; 1299 Disciplina 571.655 Soggetti Peroxisomes Lingua di pubblicazione Inglese **Formato** Materiale a stampa Livello bibliografico Monografia Chapter 1. Peroxisome: metabolic functions and biogenesis -- Chapter Nota di contenuto 2. Peroxisomal Dysfunction and Oxidative Stress in Neurodegenerative Disease: A Bidirectional Crosstalk -- Chapter 3. 7-hydroxycholesterolsinduced peroxisomal disorders in glial and microglial cells: potential role in neurodegeneration -- Chapter 4. Peroxisome biogenesis disorders -- Chapter 5. Fatty Acid Oxidation in Peroxisomes: Enzymology, Metabolic Crosstalk with Other Organelles And Peroxisomal Disorders -- Chapter 6. Zellweger syndrome disorders: from severe neonatal disease to atypical adult presentation -- Chapter 7. Heimler Syndrome -- Chapter 8. Potential involvement of peroxisome in multiple sclerosis and Alzheimer's disease -- Chapter 9. Developmental and degenerative cerebellar pathologies in peroxisomal -oxidation deficiency -- Chapter 10. A mouse model system to study peroxisomal role in neurodegenerative diseases such as peroxisome biogenesis disorders -- Chapter 11. The Drosophila melanogaster as genetic model system to dissect the mechanisms of disease that lead to

neurodegeneration in adrenoleukodystrophy -- Chapter 12. Human peroxisomal 3-keto-acyl-CoA thiolase: tissue expression and metabolic regulation -- Chapter 13. Biological functions of

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Sommario/riassunto

This book provides an overview of the biology and biochemistry of peroxisomes, and discusses the contribution of these organelles to peroxisomal and neurodegenerative diseases. It begins with a detailed introduction to the biogenesis and metabolic functions of peroxisomes, and highlights their role in oxidative stress and in lipid metabolism such as fatty acid oxidation. The following chapters focus on the molecular and clinical aspects of peroxisomal disorders caused by defects in peroxisomal function. In particular, the biological aspects of peroxisomal biogenesis disorders such as Zellweger syndrome and Heimler syndrome are discussed. This includes their underlying genetic causes as well as the biochemical and metabolic defects associated with the disorders. In addition, several chapters cover recent observations suggesting an association between peroxisomal dysfunction and neurodegenerative diseases such as Alzheimer's, Multiple Sclerosis and other degenerative cerebellar pathologies. The final section of the book discusses important cell and animal models for studying the role of peroxisomes in human diseases and presents current therapeutic strategies for their treatment. This book deals with a highly topical subject that is at the heart of current research, and represents a valuable contribution for all students and researchers who want to understand the complex biology of peroxisomes and their role in human diseases.