

Sphingolipids, Sphingolipidoses, and Allied Disorders

Bruno W. Volk

 Springer

Sphingolipids Sphingolipidoses And Allied Disorders

M.P. Mattson

A decorative graphic consisting of a red circular shape with a white center, partially obscured by a white horizontal bar that extends from the left edge of the slide.

Sphingolipids Sphingolipidoses And Allied Disorders:

Sphingolipids, Sphingolipidoses and Allied Disorders Bruno Volk, 2013-04-17 This text contains the scientific contributions to the Fourth International Symposium on Sphingolipids Sphingo lipidoses and Allied Disorders held at the Kingsbrook Jewish Medical Center on October 25 27 1971 These meetings were conducted under the auspices of the Isaac Albert Research Institute of the Kingsbrook Jewish Medical Center and the National Tay Sachs and Allied Diseases Association Inc Four symposia held in 1958 1961 1965 and 1971 were designed to gather the most relevant and innovative of the laboratory and field studies concerned with these hereditary disorders The texts generated by these periodic meetings have mirrored the increasing absorption of the scientific community in the problems of sphingolipid metabolism The first meeting in 1958 consisted of but twelve pre sentations the majority emanating from local laboratories The current sessions contain 48 scientific presentations by scientists from nine countries and demonstrate the increas ingly diversified techniques and approaches employed in the study of these diseases Many of the authors in exploring data on the mucopolysaccharidoses and leucodystrophies as well as the sphingolipidoses have given recognition to those biochemical areas held in common by these otherwise diverse disease processes The problems of prevention and therapy of these diseases have been considered by some of the contributors Laboratory screening procedures designed to detect carriers of the va rious lipidoses are now available and the experiences of some laboratories in this area are summarized within this volume The prospective identification of heterozygotes may indeed become a powerful adjunct in genetic counseling *Sphingolipids, Sphingolipidoses and Allied Disorders* International Symposium on the Cerebral Sphingolipidoses, 4th, Kingsbrook Jewish Medical Center, 1971, 1972 *Sphingolipids, Sphingolipidoses and Allied Disorders* Bruno Volk, 2014-01-15

Sphingolipids, Sphingolipidoses and Allied Disorders Bruno W. Volk, 1972 **Sphingolipid Biochemistry** Julian N. Kanfer, Sen-itiroh Hakomori, 2012-12-06 National Library of Medicine Current Catalog National Library of Medicine (U.S.), 1972 First multi year cumulation covers six years 1965 70 **The Gangliosidoses** Bruno Volk, 2012-12-06 The history of so called storage diseases goes back to the end of the 19th and to the beginning of the 20th century when Fabry Tay Sachs Gaucher Niemann Hunter and Hurler first described the disorders which up to now are called by their eponym The clinical descriptions soon were followed by pathologic studies and within a short time the hereditary characters of these rare afflictions came to be recognized Although sporadic reports during the early part of this century dealt with biochemical analysis of the stored materials in these disorders it was actually in the late 1930s that the abnormal deposits started to attract the increasing attention of chemists S H Thannhauser brought the broad concept of lipidoses as a group of related disorders to the attention of the medical profession for the first time and in 1939 Klenk observed that the brain of a patient with Tay Sachs disease contained greatly increased amounts of a glycolipid for which he proposed the name ganglioside 20 years has thrown new light on these afflic Work carried out in the past tions and has pinpointed the enzymatic and lipid

abnormalities associated with the various storage diseases Moreover electron microscopic studies have permitted detailed investigations of the fine structure of the various organs of afflicted patients Genetic Aberrancies and Neurodegenerative Disorders M.P. Mattson,2000-01-28 Genetic Aberrancies and Neurodegenerative Disorders presents critical reviews and emerging findings concerning the roles of genetic mutations and polymorphisms in the pathogenesis of a range of neurological disorders including Alzheimer s and Parkinson s diseases trinucleotide repeat disorders stroke epilepsy and others This volume written by leading experts brings together fundamental information concerning the roles of inherited traits in the pathogenesis of different neurodegenerative disorders In addition to providing a catalogue of the known genetic alterations that are linked to specific neurodegenerative disorders the chapters detail the current state of understanding of the cellular and biochemical mechanisms whereby the genetic aberrancies lead to neuronal dysfunction and degeneration The emerging picture of each disorder painted by pathological biochemical and molecular brushes suggests that they share key mechanisms including increased levels of oxidative stress perturbed ion homeostasis mitochondrial dysfunction and apoptotic proteolytic cascades The existence of specific molecular defects provides the opportunity to design experiments that can establish the precise pathogenic mechanism of a specific mutation or genetic risk factor The value of this approach is exemplified by recent studies of how mutations in Cu Zn superoxide dismutase cause amyotrophic lateral sclerosis and how presenilin mutations result in early onset Alzheimer s disease A theme developed among the different chapters is that events that occur during aging predispose neurons to genetic aberrancies that promote degenerative cascades and that specific genetic defects exert their influence on particular populations of neurons in a disorder specific manner The chapters in this volume will stimulate readers to generate new hypotheses concerning the pathogenic mechanisms of genetic aberrancies that can be experimentally tested Biology of Brain Dysfunction Gerald E. Gaull,2012-12-06 The growth of neurochemistry molecular biology and biochemical genetics has led to a burgeoning of new information relevant to the pathogenesis of brain dysfunction This explosion of exciting new information is crying out for collation and meaningful synthesis In its totality it defies systematic summation and of course no one author can cope Thus invitations for contributions were given to various experts in areas which are under active investigation of current neurological interest and pregnant Although this project is relatively comprehensive by dint of size other topics might have been included the selection was solely my responsibility I believe systematic summation a virtual impossibility indeed hardly worth the effort The attempt to assemble all of the sections involved in a large treatise with multiple authors inevitably results in untoward delays due to the difference in the rate at which various authors work Therefore the following strategy has been adopted multiple small volumes and a relatively flexible format with publication in order of receipt and as soon as enough chapters are assembled to make publication practical and economical In this way the time lag between the ideas and their emergence in print is the shortest

Scientific Directory and Annual Bibliography National Institutes of Health (U.S.),1973

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