

## MICROGYRIA AND CYTOMEGALIC INCLUSION DISEASE IN INFANCY

BY

L. CROME AND N. E. FRANCE

*From the Fountain Hospital and the Mothers' Hospital (Salvation Army), London*

(RECEIVED FOR PUBLICATION APRIL 13, 1959)

Many different malformations of the central nervous system can now be produced in the offspring of animals exposed during pregnancy to x rays, drugs, and dietetic deficiency or excess of vitamins, but proof of environmental causation of analogous human conditions is understandably scarce. Occasional neural, somatic, or gonadal maldevelopment has been reported after the administration to pregnant women of aminopterin (Thiersch, 1956), thiouracil (Morris, 1953), methyltestosterone (Nellhaus, 1958), and progesterone (Reilly, Hinman, Pickering, and Crane, 1958; Wilkins and Jones, 1958). Nevertheless, maternal rubella and ionizing radiation remain the only widely known extrinsic causes of human embryopathy. Hence the importance of the suggestion that another, probably infective, condition, cytomegalic inclusion disease, may be also responsible for neural malformation (Diezel, 1954).

It has been known for some time that the brain is one of the organs which may display the characteristic inclusions of cytomegalic inclusion disease. Haymaker, Girdany, Stephens, Lillie, and Fetterman (1954) collected seven such cases in the literature and added one of their own. In addition to the inclusions, some of these brains showed hydrocephalus, periventricular calcification, focal softening, haemorrhages, astrocytic proliferation, perivascular inflammatory infiltration, and ependymal granulations. These are, of course, evidence of neural disease, but not necessarily of malformation. Diezel was the first to report the association of cytomegalic inclusion disease with microgyria, or micropolygyria. Microgyria is a true cerebral malformation characterized by an excess of small abnormally formed gyri (Crome, 1952). Histological and embryological considerations suggest that the cause of this condition operates before the sixth month of foetal development.

The patient described by Diezel was an infant weighing 2,500 g. at birth who developed

neonatal jaundice, ascites with hepatomegaly, and splenomegaly, and died at 24 days. Inclusion bodies were found in the kidneys, liver, and brain. The brain weighed 130 g. and showed microgyria of the neocortical areas. Necrosis was present in the medial basal regions of both temporal lobes and in the cerebellum. The optic nerves were atrophic, there was marked hydrocephalus, and the corpus callosum was thin. On microscopy, the necrotic areas showed infiltration by lymphocytic and plasma cells with many compound granular corpuscles, macrophages, and calcified particles. The olfactory nerves were the seat of marked inflammatory change. Typical inclusions of cytomegalic inclusion disease were seen in the leptomeninges, ependymal, and glial cells, as well as in some of the undifferentiated matrix cells arranged in numerous dense periventricular foci. The only mature neurons to show inclusions were a few Purkinje cells, found after prolonged search. Architectonically, the palaeocortex and archicortex were normal and the relatively older occipital cortex was better developed than the rest of the neocortex. It appeared therefore to the author that the virus of cytomegalic inclusion disease attacked the foetus at a time when migration of nerve cells from the periventricular matrix to the periphery was complete in the older, and still afoot in the newer, parts of the brain, i.e., the beginning of the fourth month of gestation.

Diezel was able to re-examine histological preparations from the brains of the two cases of cytomegalic inclusion disease previously reported by Hartmann (1948) and by Haymaker and his co-workers. He found microgyria in one of these, whilst the other had an anomaly of gyri, possibly of microgyric nature. Microgyria was possibly present in a case of cytomegalic inclusion disease presented by Mercer, Luse, and Guyton (1953): the brain of this child had an irregular, thin-walled cyst, 1.5 cm. in diameter, in the right parietal lobe, the cortical surface around the cyst was slightly

# The Clinical Pathology Of Infancy

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## **The Clinical Pathology Of Infancy:**

The Clinical Pathology of Infancy Frederick William Sunderman, 1967      **The Clinical Pathology of Pregnancy and the Newborn Infant** Maurice N. Cauchi, James B. Brown, Gwendolyn L. Gilbert, 1984      **Current Catalog** National Library of Medicine (U.S.), 1992-10      *Sudden Death in Infancy, Childhood and Adolescence* Roger W. Byard, 2004-01-08 This unique comprehensive survey of virtually all aspects of sudden death in infants and childhood will be an essential source of reference for pathologists clinicians and lawyers who deal with such cases Individual sections deal in detail with deaths due to inflicted and non inflicted injuries and to natural diseases This new edition includes 1200 new references 300 new illustrations and an extensively revised chapter on sudden infant death syndrome The intentional injury chapter has additional material on head trauma the biomechanics of injury neonaticide suicide and subtle and unusual trauma The chapter on non intentional injury has also been expanded to more accurately reflect its importance as a cause of death Deaths in the first week of life are also covered This new edition also covers the full range of natural causes of death and their pathological investigation undertaken in light of advances in our understanding of genetic susceptibility and pathophysiology      *Potter's Pathology of the Fetus and Infant E-Book* Enid Gilbert-Barness, Raj P. Kapur, Luc Laurier Oigny, Joseph R. Siebert, 2007-05-01 This comprehensive reference addresses all aspects of fetal and neonatal pathology including complicated pregnancies multiple pregnancies abortion placental pathology and disorders affecting the full term neonate A consistent organization allows for quick access to specific guidance and nearly 2 500 illustrations 2 350 in full color depict conditions and abnormalities as they present in practice facilitating diagnosis An Image Bank on CD ROM new to this edition features all of the illustrations from the 2 volume set downloadable for presentations Offers comprehensive coverage of all common and rare embryonic fetal and infant disorders in one source Correlates clinical pathologic and genetic findings for each systemic disease Emphasizes the genetic and molecular basis of birth defects Features nearly 2 500 illustrations 2 350 in full color which depict each abnormality or condition as they present in practice Presents practical information on autopsy techniques and protocols Provides the latest guidance on molecular pathology immunohistochemistry DNA technology and more Offers an expanded discussion of developmental biology related to the pathogenesis of birth defects Features user friendly summary tables and diagnostic flow charts making information quick and easy to find Includes a CD ROM featuring all of the illustrations from the 2 volume set      *Pathology of Childhood and Adolescence* Consolato M. Sergi, 2020-10-28 This book covers the full range of pathologic conditions encountered during childhood and youth including tumors and tumor like conditions of all organ systems with direct links to developmental biology pathways and genetics It provides a user friendly road map to the main diagnostic criteria and combines an organ related approach with an explanation of the diagnostic approaches to various specific diseases and syndromes including sequential segmental analysis of congenital heart disease More than 500 new full color macro and microphotographs using more than 500 multi

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*Brain Damage in the Preterm Infant* Nigel Paneth, Raoul Rudelli, Elias Kazam, William Monte, 1994-01-10 A detailed survey allowing the formulation and testing of hypotheses about the causes and consequences of haemorrhage **Summary of Proceedings, Second International Conference on Causes of Sudden Infant Death, Seattle, 1969** National Institute of Child Health and Human Development (U.S.), 1971 **Sudden Death in the Young** Roger W. Byard, 2010-09-16 Determining the cause of death in children and young adults can pose considerable challenges Professor Byard provides for the first time a complete overview of pathological aspects of sudden death in the young from before birth to middle adult life Highly illustrated with more than 800 colour figures this third edition contains new sections on sexual abuse pregnancy related deaths and rare natural diseases as well as expanded coverage of unexpected death in young adults up to the age of 30 years Chapters are organised by systems and cover all aspects of natural death as well as accidents suicides and homicides Supported by extensive referencing and numerous tables the book can also be used as a practical autopsy manual An encyclopaedic overview and analysis of sudden death in the young this is a key text for pediatric and forensic pathologists pediatricians and lawyers and physicians involved in medicolegal cases *Pathology of Infancy and Childhood* John M. Kissane, 1975 Medical Record George Frederick Shrady, Thomas Lathrop Stedman, 1901 Handbook of Laboratory Animal Science, Volume III, Third Edition Jann Hau, Steven J. Schapiro, 2013-12-26 While replacing and reducing the use of laboratory animals are integral parts of the 3Rs replace reduce refine which form the cornerstones of laboratory animal science biomedical research involving animals remains absolutely essential for the advancement of the medical veterinary agricultural and biological sciences Building upon the bestselling previous edition the Handbook of Laboratory Animal Science Volume III Third Edition Animal Models complements volumes I and II of the third edition by completing the task of providing a comprehensive overview of animal models in all biomedical disciplines The three Rs have been integrated throughout this handbook to promote efficient and humane experimental work with animals Written by international experts each chapter focuses on an important subdiscipline of laboratory animal science and can be used as a stand alone text This volume contains new chapters for six additional disease animal models spinal cord injury cardiovascular diseases sudden infant death syndrome developmental disorders eye diseases and human cancer It also presents a new chapter on applying reduction and refinement to animal models This handbook can be used for undergraduate and postgraduate laboratory animal science courses and as a handbook for scientists who work with animals in their research for university veterinarians for regulators and for other specialists in laboratory animal science **Pathology of Heart Disease in the Fetus, Infant**

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