

BOEKBESPREKINGS : BOOK REVIEWS

PORPHYRIA IN SOUTH AFRICA

The Porphyrrias. A story of inheritance and environment. By G. Dean, M.D., M.R.C.P. Pp. xi + 118. Illustrated. R2.50. London: Pitman Medical Publishing Co. 1963.

This monograph of just over 100 pages records Dr. Dean's investigation of the genetics of porphyria in South Africa. As he himself says in his foreword, it is really in the nature of a detective story. He recounts how he unravelled the fascinating story of the inheritance of porphyria variegata or South African genetic porphyria. Although he had the help of many able people in this investigation, it was due to his energy and pertinacity that the inheritance of the gene was traced to a couple who married at the Cape in 1688.

The book commences with a brief account of the historical background and the chemistry of the porphyrins and their precursors. Thereafter the book has a highly personal flavour, commencing with a description of his first encounter with porphyria while engaged in consultant practice. Illustrative case reports follow. He also compares the South African form with the Swedish form which he was able to investigate in Sweden and points out the major differences between these two conditions. This is followed by a short chapter on the erythropoietic porphyrias and a brief account of symptomatic porphyria. During a visit to Turkey he was able to study cases of hexachlorobenzene-induced porphyria due to the ingestion of treated seed wheat. There are also chapters on the acute

attack and the diagnosis of porphyria in which he stresses the use of the faecal screening test.

Perhaps the most interesting chapter is that in which the master family tree is reproduced showing that the gene was derived from a couple who came from Holland and were married in Cape Town in 1688. Their marriage lines are reproduced on Plate 8.

The book is well produced and has few defects. There are a few typographical errors, e.g. 'metabolytes' (p. 6), 'oxydase' (p. 10) and occasional statements are open to question, e.g. on p. 10, 'increased urinary porphobilinogen is frequently found in lead poisoning', and on p. 69, 'in the quiescent phase of porphyria variegata this test (i.e. the Watson-Schwartz test) is always negative'. The section on acute porphyria is somewhat disappointing. Uraemia and alkalosis do not feature, nor is there any discussion of the possible role of inappropriate secretion of the anti-diuretic hormone and the importance of fluid restriction in the treatment of this hyponatraemic syndrome.

A surprising omission in the chapter on symptomatic porphyria is the lack of reference to the work of other investigators in South Africa, notably the excellent study from Durban—the best documented account available.

This book, which records a fine piece of painstaking genetic research, is a welcome addition to the porphyric literature and is one that every doctor in this country should read.

L.E.

SURVEY OF PERINATAL MORTALITY

Perinatal Mortality. 1st report of the 1958 British Perinatal Mortality Survey under the auspices of the National Birthday Trust Fund. By N. R. Butler, M.D., M.R.C.P., D.C.H. and D. G. Bonham, M.A., F.R.C.S., M.R.C.O.G. Pp. xvi + 304. R3.00. Edinburgh and London: E. & S. Livingstone Ltd. 1963.

Stillbirths and deaths up to 7 days after birth are included in this review of 17,000 births, and over 7,000 stillbirths and neonatal deaths. Of these 60% were stillbirths and 34% first-

week deaths. The perinatal mortality rate was 33/1,000. Intra-partum anoxia and/or birth trauma was responsible for almost one-third of the mortality. Late neonatal deaths were dominated by congenital malformations (45%) and infection (26% pulmonary and 10% extrapulmonary). Everything is correlated with everything else and the whole production is masterly and most informative. F.J.F.

PARTIAL GASTRECTOMY

Partial Gastrectomy. Complications and metabolic consequences. Ed. by F. A. R. Stammers, C.B.E., T.D., B.Sc., Ch.M., F.R.C.S. and J. A. Williams, M.B., Ch.M., F.R.C.S. Pp. ix + 309. Illustrated. R8.30. London and Durban: Butterworths. 1963.

This is a book which should be possessed and valued by every surgeon who ever performs gastrectomy. Every trainee surgeon should know it and understand it. Above all, every physician who refers patients to surgical colleagues with a view to gastrectomy should read and evaluate this book in terms of his own practice. 'Now we see through a glass darkly, then face to face.' Questions are answered and doubts are resolved in a most stimulating and authentic manner. In the next edition I hope the authors will devote an additional chapter to vagotomy and so-called drainage procedures.

R.D.H.B.

NEUROLOGY IN CHILDHOOD

Neurometabolic Disorders in Childhood. Proceedings of a Symposium held at Sheffield, England, in May 1963. Ed. by K. S. Holt and J. Milner. Pp. vii + 112. Illustrated. R1.50. Edinburgh and London: E. & S. Livingstone Ltd. 1964.

The principles of adult neurology have to be applied with caution to the changing and developing neurological situation in childhood. Biochemical factors are among the most important of those affecting the developing nervous system, and it was with these that the symposium was concerned.

The chapters of the book cover the effects of oxygen, CO₂, glucose, copper, and various enzyme defects. They review the situation as known in 1963; valuable to all paediatricians, neurologists, biochemists and physiologists. F.J.F.