Book reviews


In this monograph a detailed comparison is made between 34 patients with Klinefelter's syndrome (47, XXY) and 16 patients in whom hypogonadism is due to causes other than an abnormal chromosome constitution. The purpose of the investigation was to study the effects of the chromosome constitution 47, XXY on intelligence, personality, and mental illness. The patients were referred to the hypogonadal study section associated with the medical out-patients' clinic at Copenhagen University Hospital on account of gynaecomastia, infertility, or small or undescended testes. Thus the patients were derived from a more representative population than in most previous studies which have usually depended on screening hospitals for the mentally ill or the mentally subnormal. Some selection inevitably did occur and the authors attribute the relatively high mean IQs of both groups to the exclusion of institutionalized patients and the predominantly urban population which was studied. The patients underwent careful psychiatric, psychological and anthropological, physical, and endocrine examinations which were conducted independently of each other and before disclosure of the results of the chromosome studies. In addition, urinary gonadotrophins and 17-ketosteroids were assayed.

The authors predicted that they would be able to distinguish between the Klinefelter groups of patients and the cytogenetically normal hypogonadal patients on the basis of the psychiatric examinations and a psychiatric weighted score: the Klinefelter patients were expected to display more psychiatric abnormalities. They were correct in their expectations and found that the Klinefelter patients fell into two groups—a psychiatrically more disabled group of 19 patients who were older and less intelligent, and a more stable group of 15 patients. In addition to a greater proneness to mental illness, Klinefelter patients were found to show more immaturity, feelings of insecurity, and boastful or self-assertive behaviour. Their intelligence was lower (mean IQ 102-8) than the other hypogonadal patients (mean IQ 115-3). Criminal behaviour, including sexual offences against children, was also relatively frequent. The psychiatric morbidity in Klinefelter patients is thus frequent (41%) and serious and increases with advancing age. The authors say that they are uncertain of the pathogenesis of these mental abnormalities, but attribute their principal causation to the abnormal sex chromosome constitution XXY. Environmental factors may also play a part in view of their findings that the more unstable group of Klinefelter patients had come from more disturbed homes. Hypogonadism as such did not appear to be responsible for psychiatric abnormalities.

This study therefore throws light on the frequency and character of psychiatric disturbances in the Klinefelter syndrome. The presence of these disturbances, especially when found in patients with gynaecomastia or with extremely small testes allows a diagnosis to be made from other causes of hypogonadism unassociated with an abnormal sex chromosome structure.

The authors came to a number of conclusions of practical importance. They advise against drawing the patient's notice to the smallness of his testes or his sterility. Gynaecomastia, more commonly seen in the Klinefelter patients, is often a source of serious distress and a mastectomy in patients suitably handled by psychotherapy is often beneficial. Testosterone treatment judiciously administered is useful in relieving impotence and increasing libido, and may improve the mental state, but should preferably be begun before the age of 25.

The work discussed in this monograph follows the best Scandinavian traditions of medical research with a scrupulous attention to detail, careful case reporting, and sophisticated statistical treatment of the results. The only apparent flaw in the study is the possibility that the psychiatrists in the course of their examination might have been influenced by noticing the physical appearance of patients with severe hypogonadism who, as a rule, fall into the Klinefelter group. In theory this might have biased their psychiatric assessment; in practice this objection is probably unwarranted. Some of the terms used to describe personality traits such as 'immaturity' or psychological defects such as 'word blindness' would have meant more if accompanied by operational definitions. These main criticisms do not, however, detract significantly from a serious and valuable contribution to the understanding of the psychological and psychiatric sequelae of chromosomal aberrations.

G. F. M. RUSSELL


Most of this short monograph is in agreement with one's own experience. The emphasis on the important role of emotional disturbance in precipitating myasthenic crisis is not sufficiently well known. The vicious cycle of ventilatory failure and cerebral hypoxia following upper respiratory infection is well described. In other ways the book is less satisfactory. I believe that the serious deterioration which may occur in the first 48 hours after thymectomy is almost entirely cholinergic. The validity of this opinion is supported by the fact that I have not lost a single patient post-operatively in the last 14 years with management based on this view. (The 8% mortality
Book reviews

NEUROSURGERY OF INFANCY AND CHILDHOOD 2nd edition.
It is 15 years since 'Ingraham and Matson' was welcomed as the first comprehensive publication in the field of paediatric neurosurgery. Dr. Ingraham died in 1965 and this second edition has been radically rewritten by Dr. Matson; alas, a few months before publication, Dr. Matson died at the height of his powers. This book, therefore, stands as a testimonial to his life's work, which makes the reviewer's task difficult—is he writing a review or an obituary? With such an important volume, already established as a major reference work in the field, and further editions certain to be demanded, some comments for the ears of future editors may be judged in order without detracting from the magnificent achievement of the author.

The book as it stands is a personal one, reporting the experience and methods of the Children's Hospital Medical Center in Boston. Indeed, in his preface Dr. Matson offers it as a monograph, and insists that it is not an encyclopaedic reference text. Yet that is what most readers will expect of a book of this price, running to 1,000 pages and 700 references. In fact, no important aspect of paediatric neurosurgery is omitted, this second edition having added chapters on the surgery of epilepsy and pain, on benign intracranial hypertension and on Sturge Weber disease. The reports of Dr. Matson's uniquely large series of various conditions, particularly of intracranial tumours, are fascinating and invaluable. He explicitly states in the preface that he has deliberately decided not to engage in critical comparison with the experience or methods of other clinics, which the reader is encouraged to do for himself from the original sources. But the material here is seldom presented with as full an analysis as in a scientific paper, and this makes it less easy than it sounds to carry out such comparisons. As a result the book leaves many intriguing questions unanswered, particularly the reasons which led Dr. Matson to reject the several alternative approaches that are available for many of the problems discussed.

It might be thought that to expand the book to include such critical comparisons would make an already over-weight book unmanageable. However, there is little doubt that much could be done to condense the information in the present edition by effecting economies of style and illustration. There are over 600 illustrations, but the time has come in this as in many other books to question the real value to the reader of black and white reproductions of operative fields or of histological sections, or of pictures of excised tumours lying by the inevitable statutory scale. The many photographs of happy young adults with a legend indicating how ill the individuals were as children, were appropriate in the days when neurosurgery still had to justify itself, but seem a little outdated in the '70s. These are, of course, minor criticisms. This is an invaluable tome, of interest not only to surgeons but to all doctors who are concerned to know how one world famous clinic deals with the wide variety of problems now coming within the scope of paediatric neurosurgery.

CYTOLOGY OF TUMOURS AFFECTING THE NERVOUS SYSTEM
This book is, to the best of my knowledge, the first to be devoted entirely to the use of smears in the diagnosis of cerebral tumours. Before the various types of tumour are illustrated, there is a short chapter on the appearances of normal, gliotic, and non-neoplastic brain tissue.

The book is essentially an atlas, as the text amounts to little more than legends for the illustrations. The latter, in general, are of fairly good quality. One might question some of the diagnoses, but the book should be welcomed as a bold attempt on the part of two neurosurgeons to provide an atlas that will be of considerable interest to clinicians and trainee neuropathologists.

My major criticism is that the subject is dealt with too superficially. The difficulties often encountered in attaching a precise diagnosis to a particular tumour are insufficiently stressed, with the result that many might be led to assume that the technique is simple and foolproof.

The inclusion of smears that presented difficulties in diagnosis, supported by information about the ultimate diagnosis in sections would have greatly enhanced the value of the book. As all the illustrations occupy a full page, twice the number of illustrations could have been incorporated without increasing the size of the book.

Finally, it is unfortunate that there is no mention of the value of smears in establishing the presence of some other types of brain pathology such as encephalitis. Nevertheless, until a better text is forthcoming, this book should be used frequently in laboratories and clinical units using the smear technique in the diagnosis of brain tumours.

J. HUME ADAMS

It may be that this useful monograph will be the last one concerning us in Britain, as the Animal Health Division of the Ministry of Agriculture has already started on its plan to eradicate brucellosis entirely.
CRISES IN MYASTHENIA GRAVIS

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