Book reviews


The introduction of laevodopa for the drug treatment of Parkinson's disease constituted a considerable advance in the therapeutics of that condition and there is no doubt that the lot of many patients has been markedly improved as a result. Nevertheless, the drug can cause problems. It is often necessary to use very large doses and the process of building up to the optimal amount may be time-consuming and tedious for the patient and physician alike. The high doses are needed because only a small proportion of the drug actually reaches the dopamine-deficient striatum as the rest is metabolized before it crosses the blood brain barrier. These high doses of the drug in the periphery are responsible for the undesirable side effects frequently encountered, like nausea and vomiting which either limit the amount that can be given to a particular patient or, in some instances, actually preclude the use of the drug altogether. The introduction of dopa decarboxylase inhibitors has changed things to some extent, for with these the amount of laevodopa that is metabolized in the periphery is reduced, whereas the central effects are unaltered because the inhibitors do not cross the blood brain barrier. The consequence of this is a reduction in the dose needed, a reduction in side effects due to the peripheral (but not the central) actions of the laevodopa and a greater ease of introduction and adjustment of the dose for a given patient.

In essence, this is the background to the present book which is a report of a symposium held in November 1972 at Columbia University. It contains a number of papers on the mechanisms of action of laevodopa in the disease, on the enzymology of central nervous system monoamines and on their sites of action. These are followed by several papers on the effects of combining laevodopa with various other agents and the consequences of these combinations for the patients. There is a detailed bibliography on the whole subject of extracerebral decarboxylase inhibitors.

The book has a rather narrow appeal for it is primarily of interest to neurologists, but it should contain something of interest for those who are less directly concerned with the treatment of neurological diseases but who are nevertheless interested in central nervous system neurotransmitter substances and the role they may play in the evolution of a variety of conditions. Amongst this number may be some geneticists, although it has to be conceded that to most the investment would not really be worth while. This in no way denies the general value of the book, for it is a succinct and, for the most part, readable account of an interesting and growing subject.

DAVID L. STEVENS


This book reviews developments in our knowledge of XYY males and those with Klinefelter's syndrome. It takes the story up to 1970 and presents in addition much epidemiological and clinical data based on large scale studies by the author in southern Germany.

As stated by the author, much of the data in his review is taken from the late Professor Court Brown's 1968 article on XYY males in Journal of Medical Genetics (5, 341–359). Readers wishing to read about the development of the XYY story would be better advised to refer to the latter publication.

Dr Murken's review a few steps are omitted in the historical sequence of development of our ideas and there is some over-simplification in the presentation of the results of other authors. In addition, different language idioms and some lack of clarity in the use of words such as 'weak-minded' and 'mental' would make the book confusing for those not familiar with the original publications. Lack of clarity in the language leads to loss of clear distinction between mental subnormality, mental illness, and criminality.

The main part of the book describes cytogenetic screening surveys of about 2500 individuals from a variety of groups: newborn, social deviants, and patients with congenital deformities. Very full case histories of patients with chromosome abnormality are provided; investigations include psychological studies and anthropometry with testicular histology in a few cases. Inevitably the small number of probands with chromosome abnormality and the selection of patients from within institutions must lead to caution in the interpretation of results. The predisposition of XYY males as well as XYY to socially inadequate behaviour is emphasized.

This book can be recommended to research workers interested in the relationship between criminality,
Book reviews


This book covers many aspects of the causation and management of birth defects. Although the term 'birth defects' means 'conditions of prenatal origin . . . manifest at birth or various ages later in life', the subject of this fourth international conference sponsored by the National Foundation—March of Dimes deals with malformations and diseases which are congenital, or which are apparent in infancy. This volume presents the papers of the invited speakers, and gives summaries of the workshops. Such selectiveness is good, and indeed the abstracts of the free communications are available in an earlier publication (International Congress Series No. 297). The subjects of the invited papers are wide-ranging and include cellular and molecular biology, precise analysis of chromosome structure, sex determination and differentiation, epidemiology and teratology, prenatal diagnosis and genetic counselling. In an interesting pair of papers M. M. Kaback and G. Stamatyannopoulos describe the effects of genetic screening programs in two very different communities. The most valuable workshops were those on the regulation of gene expression (J. D. Ebert), the management of spina bifida patients and their families (C. O. Carter), and the treatment of genetic disease (C. R. Scriver). The volume is completed by two chapters by the editors. A. G. Motulsky discusses the ethical aspects of the genetic advances presented. He points out that in practice the medical geneticist who considers only the best interests of a particular family will rarely have any ethical problem to face. W. Lenz provides a thoughtful summary of the conference.

Most geneticists will find one section or another particularly to their taste but an important use of the book is as a source of reference for those subjects less well known. As most papers are written as reviews, with current work explained and put into perspective, they will be useful even when further work has modified or added to some conclusions. All those who are interested in the aetiology, prevention, and treatment of birth defects will find this a helpful and stimulating book.

Sarah Bundey


Individual constitutional disorders of the skeleton are particularly difficult to define unless, like rickets and the mucopolysaccharidoses, they have consistent biochemical lesions. Many variations occur on the common themes of proportionate and disproportionate dwarfism, spinal, cranial, and limb malformations, while diagnostic overlap and mimicry are so common that the practical application of any classification, however logical, soon breaks down. Thus, no nomenclature of constitutional disorders of bone is entirely satisfactory and Dr Carter and Mr Fairbank have wisely not attempted yet another. They have chosen common sense chapter headings of several types. Thus, some relate to the site of main disorders, e.g., metaphyseal, epiphyseal, spondylo-epiphyseal, or to localized abnormalities of the limbs, skull, face, and spine. Other chapters are devoted to the muscular dystrophies, to errors of timing of development (anarchic development of bone and cartilage), to syndromes, and to inborn errors of metabolism affecting the skeleton. In view of the companion volume in the Oxford monograph series by Dr Pratt only brief reference is made to neurological conditions of skeletal significance.

This book is a complete and up-to-date summary of inherited musculoskeletal anomalies presented in a particularly clear and non-discursive manner. The clinical descriptions are brief and the atlases listed in the bibliography are necessary to define the large number of entities involved but the clinician and geneticist using 'The Genetics of Locomotor Disorders' will be able quickly to determine for any condition the probable mode of inheritance and key references for further reading. This reasonably-priced and well-produced book is a very welcome addition which will illuminate a currently rather confusing area.

Rodney Harris


The rapid exploitation of incomplete rejection of human chromosomes by some hybrid cells, especially from mice and Chinese hamsters, has led to many observations which needed to be integrated with the results of the classical methods of pedigree analysis, results which can now be supplemented by the identification of harmless chromosomal variants. The integration of these results involved difficulties in terminology, in the weighing of evidence, and in the identity of enzymes; the problem of enzyme identity can be particularly difficult since red cells and the cells established in culture often have different enzymes with similar functions, and names are based on function. Further, many laboratories specializing in hybrids do not have regular access to individuals whose heterozygosity could be checked on several tissues.

The New Haven Conference, appropriately sited at one of the major laboratories exploiting hybridization, resolved a number of problems, and identified others; this book provides a 'state of the art' review as of summer 1973. As such it is a valuable contribution to the literature on linkage. Unfortunately it is rather expensive, in part because of the excellent binding and production which are hardly justified by the rapidity of change of the subject.

M. D. Casey

Behaviour disorder, and sex chromosome abnormality for the data which it provides. It cannot be recommended as a good review of the history of this field for those not already familiar with the literature.