

collection of articles on some of the most recent developments and applications in molecular biology, and as such receives a firm recommendation.

DUNCAN SHAW

Congenital Metabolic Diseases

Clinical Pediatrics Volume 2. Edited by Paul A Wapnir. (Pp 429; figures + tables. \$75.00). New York: Marcel Dekker. 1985.

This book is the second of a series of publications on clinical paediatrics and is essentially a series of papers presented at a conference in 1983 to honour Garrod's description of inborn errors of metabolism.

The book is divided into eight parts and starts with some interesting historical data and discussion of Garrod's concepts. There is a limited section on new approaches to diagnosis with a chapter on the use of restriction enzymes. The third part is devoted to neonatal screening with quite a lot of duplication in the first two chapters. A review by Pang and New has a somewhat narrow approach and concentrates heavily on congenital adrenal hyperplasia. For readers outside the USA it is important to realise that this section presents a different picture of screening compared to current practice in the UK and parts of Europe. The desirability and ethical aspects of screening are not addressed, and the non-specialist reader could wrongly assume that screening for diseases such as glutathione deficiency, argininaemia, and adenosamine deficiency is justified. Computerisation of screening laboratories enabling 'megalaboratories' to screen 250 000 to 500 000 babies annually for 10 to 20 disorders certainly does not seem a reality for most countries, let alone desirable.

The section on amino acid metabolism contains an interesting selection of papers, including an excellent review of maple syrup urine disease. There is a lot of very useful dietary information which would be helpful to those directly involved in the practical management of patients with amino acid disorders.

There is a good review of the lactic and methylmalonic acidemias, and a fascinating chapter on glycerol metabolism and related disorders. Detailed discussion of enzyme measurements in the various types of glycogen storage disease are more suited to the biochemist than the paediatrician.

Part 6 deals with some aspects of the storage disorders and includes a useful update on the mucopolysaccharidoses and current status of enzyme replacement therapy. Inherited diseases of membrane transport contains an interesting and detailed chapter on leprechaunism and a useful

practical review on carbohydrate intolerance. The final section deals with aspects of purine and urea metabolism with an excellent chapter on the immunodeficiency diseases.

This book aims to be a comprehensive review of recent developments in the field of metabolic disorders for the practising paediatrician. Unfortunately, it does not fulfil this promise. Content is unbalanced, with specific texts of interest to the specialists and far too detailed for the average practising paediatrician. Although there are some very unusual and interesting chapters, not found in other available texts on this subject, there are other chapters with very little new information. For an update on this subject there are notable omissions, such as the use of chorionic villus biopsy for prenatal diagnosis and problems of maternal phenylketonuria. I felt that this inconsistency reflects that this is a publication following a conference rather than having a specific readership in mind. I was left wondering just who will buy this book.

ANNE GREEN

An Introduction to Inherited Metabolic Diseases

Edited by J B Holton. (Pp 99; figures + tables.) New York: Chapman and Hall. 1985.

This is an excellent little book. It is based on a series of undergraduate lectures about the basic principles of inherited metabolic disease. In the first chapter Garrod's concepts and some basic genetics are introduced. Then the mechanisms responsible for the defects and genetic heterogeneity are discussed. In the last two chapters the consequences and diagnosis of inborn errors are briefly covered. Examples are given throughout.

This monograph is clear, well written, and up to date since brief details of the advances as a result of studies of DNA are included. It provides a good introduction to the subject for undergraduates and others in training and I would recommend it. It also has the virtue of being short, less than 100 small pages, and, very importantly, it manages to convey some of the fascination and excitement of the subject.

J V LEONARD

Craniofacial Dysmorphology: Studies in Honor of Samuel Pruzansky

Edited by M Michael Cohen Jr and Beverly R Rollnick. (Pp x + 352; figures + tables. £58.00.) New York: Alan R Liss. 1985.

This book, which also appears as Supplement 1 to

the *Journal of Craniofacial Genetics and Developmental Biology*, 1985, vol 5, is dedicated to the memory of Dr Samuel Pruzansky, an internationally renowned worker in the field of craniofacial malformations until his death in 1984. Among his many achievements, Dr Pruzansky organised the first international symposium on craniofacial malformations in 1959 and founded the first Center for Craniofacial Anomalies at the University of Illinois College of Medicine in 1967.

The book contains reviews and original papers by 48 contributors from six countries and covers a wide, though by no means complete, range of clinical and experimental topics. It is divided into sections I to VI. The first includes a brief account of the life and work of Dr Pruzansky and lists his more than 190 publications. Then follows an outline of his involvement with the Center for Craniofacial Anomalies, together with reviews of techniques, such as infant cephalometric radiography, that can be used in research and as aids to diagnosis and patient management. Section III deals with growth of the cleft palate during the first five years of life, with considerations that influence the timing of surgical closure, and with velopharyngeal inadequacy in the absence of overt cleft palate. It also contains a brief but intriguing chapter reporting an unusually high frequency of non-right handedness among parents of children with right sided cleft lip (the less frequently affected side). The largest section of the book, section IV, includes accounts of both cross sectional and longitudinal cephalometric studies of various disorders, among them a detailed analysis of achondroplasia. One chapter introduces a refreshingly simple system of six measurements, taken from cephalometric radiographs, as an aid to 'quantitative dysmorphology', perhaps a useful way of side-stepping the bewildering variety of angles and dimensions that have been used in the past. Section V contains a number of clinical observations and hypotheses to explain the pathogenesis of selected syndromes, together with a description of a computerised record system for craniofacial malformations and a discussion of chromosome damage and congenital defects associated with the infamous 'Agent Orange'. The final section is devoted to experimental animal studies, the most common theme being genotype-environment interaction in the aetiology of cleft lip and palate in mice. Also included is a report of experimental fusion of the naturally cleft embryonic chick palate through surgical intervention.

The book is not a medical genetics text and only occasionally is there mention of such aspects of the various syndromes as aetiological heterogeneity and

genetic counselling. However, it does have a predominantly clinical orientation and so should find its widest readership among those, whatever their speciality, who work with patients having craniofacial malformations. It will also be of use to experimentalists striving towards a better understanding of the complexities of craniofacial development. The book is attractively produced, with reasonably clear line drawings and adequate half tone illustrations. Many of the chapters are likely to be of practical value to both clinicians and researchers, making the volume a fitting tribute to one whose professional life was devoted to helping patients with these disorders.

JEFFREY SOFAER

Sister Chromatid Exchanges. Part A. The Nature of SCEs. Part B. Genetic Toxicology and Human Studies

Edited by Raymond R Tice and Alexander Hollaender. (Pp 364; figures + tables. £58.00.) New York: Alan R Liss. 1985.

This treatise, consisting of two volumes, contains proceedings of the first ever international symposium on sister chromatid exchange (SCE), held at Brookhaven, NY in 1983, in which participants, largely through a series of research papers, provide an overview of this steadily expanding field.

Part A. The Nature of SCEs begins with a brief historical perspective. The existence of SCEs was initially postulated in 1938 by McClintock to account for the instability of ring chromosomes she had observed in maize. The subject then appears to have lain dormant until Taylor *et al* (1957) published the now famous technique of radiolabelling chromosomes with tritiated thymidine, which, due to the semiconservative nature of DNA replication, rendered SCEs visible at metaphase. This approach was given fresh impetus when Latt (1974) discovered an elegant method whereby bifiliary BrdU substituted DNA stained with a fluorochrome could be distinguished from unifiliary substituted DNA, allowing precise localisation of SCEs. Further refinement of the practicality of the technique by Perry and Wolff enabled SCEs to be readily observed in Giemsa stained preparations. The subsequent realisation that a large number of mutagens and clastogens were also inducers of SCEs has led to the method being accepted as a potentially sensitive assay for environmental genotoxic agents.

Contributors move on to consider the nature of SCE and explore the role of halogenated and natural pyrimidines, including the effects of thymidylate