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

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 acidaemias, 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 fulfiil 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

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

This book, which also appears as Supplement 1 to