

of these children, and it is for these, as well as for paediatricians, that the book has been written.

The book is divided into three parts. Part I, the general principles, is concerned with the incidence and screening for metabolic disease with basic genetics and enzymology. Part II is devoted to metabolic medicine in paediatric practice and covers the clinical manifestation of metabolic disease, hypoglycaemia, acid-base disturbances, and disorders of calcium metabolism. Part III is a description of inborn errors of metabolism based on a classification that the author has devised. He divides inborn errors into disorders of active transport, of intermediary metabolism, of synthesis, and those characterised by excessive storage or accumulation. Each aspect is covered succinctly and the descriptions of individual diseases are thorough but short. The management is usually only outlined and not discussed in detail. The advice about drug dosage is too vague and therefore potentially dangerous, for example the dose of diazoxide before pancreatectomy for islet cell tumour is given as 300 to 800 mg, and the loading dose of dihydrotachysterol for hypoparathyroidism as 3 ml, but it is not stated over what period the dose should be given, nor is the dose related to the age nor to the size of the patient.

It is a daunting task for one author to attempt to cover such a large range and inevitably this means that the book is not completely up to date. No mention is made of several recent advances such as the aetiology of familial hypercholesterolaemia and intensive feeding regimens for type I glycogen storage disease.

There is no doubt that this book will provide a useful introduction for those who work in this area of medicine and for those taking higher exams. However, it does not give sufficient detail for the clinician who has the responsibility for the care of these children. At present this book costs £27.50 and for that price has far too many misprints.

J V LEONARD

Human Genetics (Outline Studies in Biology)

By J H Edwards. (Pp 80; Figures + Tables. £1.75.) London: Chapman and Hall. 1978.

This book is one of a series of outline studies in biology which are intended to provide an interdisciplinary guide at a level between text book and research paper. For those who already have some basic knowledge of genetics, as suggested in the general editor's foreword, there is an up-to-date survey of the field of human genetics with an abundance of facts.

The style is distinctly idiosyncratic with sentences that vary from the terse to the verbose. There are remarks which do not contribute materially and which may result from the origin of the book in a course of lectures to medical students at the University of Birmingham. Such asides may make a lecture memorable but are out of place in a written form. There are, however, some striking phrases such as spontaneous mutinous or neoplastic activity and developmental punctuality. Repetition is frequent, for example for Down's syndrome.

The definition of terms is inconsistent. Multifactorial and polygenic are not regarded as acceptable, though their usefulness is admitted. Instead the word familiarity is used, though it is omitted from the useful glossary. While the reader is informed that the sex chromosomes are sometimes called allosomes, the term polymorphism is not used or defined when the variations detected by modern staining techniques are mentioned. Amniocentesis is referred to either directly or implicitly in several places but there is no single section dealing with technique, risks, and indications. The legend to figure, 2.4, which shows a cell with a normal karyotype, misleadingly refers also to the common varieties of translocation mongolism. Considerable doubt has now been thrown on aggression as the best description of the psychiatric disturbance in the minority of XYY cases.

In discussing dominant disorders, there is no mention of some which impose a heavy burden on particular families, such as hyperlipoproteinaemia (type II), whose incidence is far higher than any given in table 4.1. The important point is not made that there can be differing modes of inheritance, for example polycystic kidneys and retinitis pigmentosa. In the same table it is not clear whether the figure given refers to the incidence of haemophilia A or to both A and B, or in male births only.

In the chapter entitled Artificial Selection, the mathematics of gene frequency is given, though without reference to the section in chapter 10 where the Hardy-Weinberg equation is used without the eponym. This is unnecessary duplication, while the eponym, as well as serving as an aide memoire, honours its discoverers.

The HLA system is discussed in both the chapter on Mendelian Variation and again in that on Disorders of Defence. In the former, two barely legible diagrams of the distribution of the haplotypes for the A + B alleles in Birmingham are given showing association, but there is no explanation of the significance of W in the terminology. In the second section the association with certain diseases is stated but the fuller understanding and better delineation brought to a number of less common

diseases of the joints, such as ankylosing spondylitis, is omitted.

Chapter 10 consists of a series of paragraphs on different topics, otherwise unmentioned, which are arranged alphabetically rather than in a related sequence. One important point made under the heading of paternity is awareness that non-paternity is higher in a child conceived before marriage, and that care is needed therefore before giving the poor prognosis for an autosomal recessive disorder found in a first born child.

There are quite a number of misprints including two on p 31 and the index is not always accurate. Amniocentesis does not appear on p 68 but there are important sections on p 69. No references are given by design in the text but there are a number of monographs or reviews given under suggestions for further reading, together with the author's useful comments on their value.

Although the intention of this book is commendable, in that of starting from the basic set of components, it has not succeeded. Whether this analytical approach would appeal to many medical students is open to question, but before it could be recommended, considerable rewriting would be required. Furthermore, there are other volumes of similar size which meet the needs of the medical student. Whether it would be more useful to science graduates with some knowledge of genetics is also uncertain because of their unfamiliarity with many of the medical diagnoses mentioned but not described.

A W JOHNSTON

Genetics and Cancer in Man

By R N Schimke. (Pp viii + 108; Figures + Tables. £7·00.) Edinburgh, London, New York: Churchill Livingstone. 1978.

Malignancy is a major cause of human morbidity and mortality. The most common forms of neoplasia occur in later adult life, and this makes both prospective genetic studies and the retrospective collection of adequate family data more difficult. These, added to the many other difficulties of investigation of the inheritance of human disorders, have tended to turn many away from the study of cancer genetics in man. Yet there is increasing evidence that among those patients with the common neoplasms there is a group of families in whom cancer predisposition behaves for practical purposes as a simply inherited trait. For example, among the wide and apparently random distribution of carcinoma of the colon in the general population, it is seen regularly as a development in the hereditary

polyposis syndromes where it behaves like a straightforward dominant, although the bulk of colonic cancer cases may not be genetic at all. But even in those families in which the genetic risk of malignancy is high, there remain some disturbing features. Monozygotic twin concordance is frequently far less than expected, even when allowance is made for differences in expression and lack of penetrance.

The volume by Professor Schimke on genetics and cancer in man is very welcome. It is part of the series published by Churchill Livingstone under the general editorship of Professor Alan Emery designed to provide trainee and practising doctors with easily accessible and authoritative information about genetic aspects of their specialities. There are nine chapters. The first is introductory, setting out the difficulties of cancer genetics, the origin of heritable tumours in man, pitfalls in the analysis of site-specific neoplasia, malignancy in recessive disorders, the association with malignancy of the HLA system and cytogenetics, and the clonal origin of tumour cells.

Chapter 2 concerns embryonal tumours, uncommon except for haemangiomas, lymphangiomas, and various types of pigmented naevi. It is tacitly assumed that they originate in maldeveloped fetal tissue, although they may not become clinically evident until childhood, the incidence declining rapidly with advancing age, so that most develop before the age of 5. The usually solid embryonal tumours contain histologically mixed types of tissue or are sarcomatous and usually are not considered heritable. But nephroblastoma, retinoblastoma, neuroblastoma, hepatoblastoma, the sarcomas, and teratoma show quite distinct hereditary predispositions. In the first two approximately one-third show clear evidence of inheritance, the remainder a much lower familial tendency.

The next chapter deals with the hamartomas, solid or cystic tumours formed by excessive aggregation of either single or multiple tissue elements, and the hamartoma syndromes where different organs throughout the body contain these conglomerates. Here neurofibromatosis, tuberous sclerosis, von Hippel-Lindau syndrome, and the basal cell naevus syndrome are discussed, all of which show clear evidence of different forms of Mendelian inheritance, while in the syndromes too, for example the Gardner syndrome, the Peutz-Jegher syndrome, the Turcot syndrome, the evidence for Mendelian inheritance is strong.

In the next chapter on cancers of the skin, the summary table of skin abnormalities as indicators of potential non-cutaneous malignancy is particularly useful. So too in chapter 5 on endocrine gland