BOOK REVIEWS


It is fitting that a book with this title should be published to mark the fiftieth anniversary of the appearance of Archibald Garrod's classic work bearing the same name, even if the genius and magnitude of Garrod's contributions do not receive quite the recognition they merit. To-day there are many advantages in constructing a book around this central theme. In the first place, many of the conditions that all would agree should be classified as inborn errors of metabolism have had much of their underlying biochemical mechanism elucidated; we are brought close to the primary actions of genes and can perceive the chains of events which follow. In the second place, they tend to be discrete and separable genetic entities so that their genetics in turn tend to be simple and clear-cut. All this facilitates brief and didactic treatment.

As far as is possible Professor Hsia treats each condition under the headings: clinical features, heredity, pathogenesis, diagnosis and treatment, ending with a limited number of key references. Graphically he starts with an illustrative pedigree. Then the primary gene effect (as far back as it can be traced) is stated, followed by the secondary effects, which with well studied conditions may be manifold and complex.

As long as the book deals with manifest or very probable inborn errors of metabolism there is little to criticize. The presentation is succinct yet clear. It is also authoritative and up-to-date. Students, for example those studying for the Diploma in Child Health, will find this a very useful book. It is doubtful whether any other offers so much information in so small a space. The research worker wishing to refresh his memory, or read about an unfamiliar condition, will find an excellent account, which will be a good starting point for further reading.

Yet, even in this, the main part of the book, there are a few lapses, mostly in the genetic field. On page 198 it is estimated that one person in every 16–20 carries the gene for fibrocystic disease of the pancreas. How can so high a frequency of a gene lethal in the homozygote be maintained? Dr. Hsia considers two hypotheses: (1) increased reproductive fitness of the heterozygotes, (2) a high mutation rate. (He does not mention a third possibility, namely, that there may be two or more alternative genes.) He states that as there is no evidence to support the first possibility 'we must assume that the latter is the case'. This is surely wrong. So high a mutation rate is almost unthinkable, whereas there is no evidence at all which could possibly show whether or not heterozygotes enjoy a sufficient advantage in reproductive fitness. It would not have to be large, and the onus of proof rests with those who prefer so unlikely an alternative. In dealing with classical haemophilia (Haemophilia A) it is stated: 'In most instances the defect is transmitted as a sex linked recessive character.' What can this mean? No other form of genetic transmission is known or has even been suggested.

It is when the book turns to conditions which might possibly be attributed to inborn errors of metabolism that difficulties multiply. Of course no two people would agree on the line separating what is (or might be) and what is not an error of metabolism. Probably no one else would agree in detail with Dr. Hsia. As he points out, the limits could be made so wide that everything is included, or for of course an apparently morphological defect might be due, if we but knew it, to some chemical influence during early development. The difficulty is to know where to stop. To give one example, the muscular dystrophies are included. They are so different from other conditions that not unnaturally a separate chapter has to be devoted to them, and this is where the disadvantages of the brief didactic approach are especially manifest. For the genetics are not simple, and no one could deal with them, as is attempted here for the severe generalized type, for instance, in seven lines: 'Pseudohypertrophic muscular dystrophy occurs six times as frequently in males as in females. The disease can also be transmitted by affected females. Because of this, it is generally agreed that the condition is transmitted as a sex linked recessive.' These are not the criteria for recognizing sex-linkage. The author is relying on Bell's excellent monograph of 1943. But there have been several admirable and more recent surveys; those of Stevenson and Cheeseman and of Walton, for example, which modify Bell's conclusions. The affected females are due to a mixture of sex-linked and recessive cases, and the separation can now be effected with almost complete efficiency.

A final chapter of four pages is entitled 'The Future of Biochemical Genetics'. Two tables are given. The first is a list of 67 conditions said to be genetic, but for which there is little or no evidence for a biochemical cause. But it includes some conditions which are only very partially genetic, for example, harelip and cleft palate and clubfoot, and at least one which is hardly genetic at all, namely, disseminated sclerosis. The other table is baffling. It purports to give a list of 35 conditions stated to be biochemical, but which 'cannot be classified as among the hereditary diseases in the classical sense', though 'they may be genetic'. Yet the list includes the Laurence-Moon-Biedl syndrome and Milroy's disease, which are genetically determined, and, astonishingly, secretion and non-secretion of the blood group substances, and ability to taste phenylthio- carbamidc, which are 100% genetic in their determina-
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Professor Illingworth has collected a team of 14 collaborators to review recent thought on cerebral palsy and, though he makes no claim to have edited a textbook on the subject, between them the authors cover an impressive amount of ground. There are three chapters on classification, pathology and diagnosis; four on the physical, psychological and intellectual handicaps of palsied children; three on education and the statutory and voluntary services available in this country; a chapter each on therapy with drugs, physical medicine and speech therapy; and finally two surgical chapters, by an orthopaedic and neuro-surgeon respectively. Professor Illingworth has chosen his team well, and it is obvious that he has briefed them carefully, so that the book achieves marked unity of purpose even though the styles vary considerably from chapter to chapter. The quality is high throughout; not only is a tremendous amount of information available, but to anyone working with a special interest in this subject the book provides an outlook of informed and intelligent sympathy which work with these children often brings out. At the same time it is free of that kind of uncritical optimism which colours much of the writing on handicapped children. It would be difficult to pick out any chapter for special comment, but Professor Illingworth's opening chapter is a good summary of the scope of the subject, the aetiology and classification, and the chapters on diagnosis, education, treatment of deafness in these children and on drug therapy are perhaps the most immediately useful.

The authors are drawn from England, Australia and America. The English and even the Australian is simple enough to understand, but some of the American is difficult for the English reader, and unfortunately good translations are unobtainable. For instance, 'Far from envisioning static, fixed mechanisms, the neurohumoral orientation views organismic mechanisms as essentially dynamic, ever in flux and, within wide limits, capable of compensatory activity such that noxious influences can be tolerated to a considerable degree without resulting in clinically manifest derangements. Above all, it considers neurohumoral circuits as expressive of relationships that obtain among component parts—relationships not apprehendable by scrutiny limited to the anatomical elements themselves'. The reviewer thinks he understands but is relieved that the author's next sentence begins: 'This rubric appears to be one of the hardest for the student to grasp.' This neo-American medical lingo is tedious reading and spoils an otherwise thoughtful, almost philosophical chapter on neurosurgery.

As usual, the binding and the paper is first-class. Some of the photographs, particularly those of abnormal brains, are helpful, and I would say this book is well worth its 50s. as a book of reference for a practising paediatrician.


Griffiths-Mitchell, Mitchell-Nelson to Nelson in seven unfolding editions. A book which a quarter of a century ago set out to be an encyclopedia of paediatrics in two volumes has now become the accepted text throughout the English-speaking world.

The present volume performs the exceptional feat of including much that is new but at the same time being somewhat shorter than its predecessor. There are new sections on: 'Prenatal Factors in Diseases of Children' by Josef Warkany and F. Clarke Fraser, on 'Diseases of Mesenchymal Tissues', by Ralph J. P. Wedgwood, on 'Behaviour Problems Associated with Organic Brain Damage' by John B. Bartram and an excellent chapter by Clement Smith and C. Davenport Cook on 'Pulmonary Ventilation in Health and Disease'.

Like the aphorism 'which says too much in trying to be too small' the excessive condensation which has enabled so much paediatric information to be distilled into such a small compass extracts its penalties. The book is an impressive compendium of facts and data but it is on the whole humourless and impersonal. One can feel the pressure of the Editor's pen erasing the anecdotes, the historical sides and the individual bigotry which make some smaller text-books memorable. Again the accepted viewpoints are well expounded but the reasoning which has led to their acceptance is frequently lacking. Amid the welter of facts there is little space for opinion and doubt. This may be admirable in a book of reference but in an undergraduate text-book it is liable to breed the student who is too pre-occupied in absorbing facts to have time to think. For this reason the seventh edition is more suited for a place in the practitioner's cupboard than on the student's desk.

In a book with no less than 81 contributors the uniform excellence of each section is remarkable. Since all the writers work in the United States there is in some sections a strong transatlantic emphasis. Certain views on dysplasia of the hip joint, or on allergy to cow's milk may be intriguing rather than acceptable in this country. Perhaps it is more surprising in a book of this magnitude that there is so much agreement with accepted British practice.

The illustrations and production are of the extremely high standard one expects in American text-books. The diagrams are easy to understand and the short selection of references at the end of each chapter provides a useful entrée to the literature of each topic. This edition will do much to maintain and enhance the reputation its predecessors have gained.