What about the medical profession? Historically it has a poor track record of awareness of the wider needs of childhood, as the history of NAWCH demonstrates. The paediatric profession will be remarkably interesting indication of whether and how much attitudes are changing.

A practical point – don’t miss the valuable notes on each chapter which (for policy makers and others) have been clearly annotated in the text. They are at the end of the book and should be searched for, as they illuminate the argument with further authority.

MICHAEL ROGERS
Senior lecturer in community child health/consultant community paediatrician


Probably most, if not all, paediatricians believe that research using child subjects is not only ethically essential but the key to fundamental improvements in the health and wellbeing of all children. This is true even in situations where there may be no direct benefit to the child subjects themselves, so-called non-therapeutic research. However, from time to time ethics advisory committees, members of parliament, and members of the public will question not only the ethics but the legality of such research. While such criticism is often based on ignorance of the facts and media over-exploitation of the circumstances, it does leave paediatricians uneasy about the possible consequences of embarking on research projects even when they may be of considerable potential value to future generations.

This American book is welcome as it gives further reassurance to paediatricians that responsible research using children is important, ethically acceptable, and unlikely to attract legal attention provided that certain conditions are met, notably a proper balance of benefits and risks; the consent of parent(s), the assent of the child where appropriate, and the approval of an ethics advisory committee. The book complements that of the US publication edited from the discussions of a group set up by the Institute of Medical Ethics ‘to encourage reflection and, perhaps, to help members of Research Ethics Committees in the discharge of their duties’ 1. The British book originated (and benefited) from distillation of the thoughtful deliberations of a large and distinguished group of scholars from many disciplines. The American book, after an interesting historical overview of paediatric experimentation (which provides ample evidence for the need for strict controls), consists of one or two author chapters on subjects grouped under sub-headings, ‘Science, ethics and the law’ and ‘Practical problems’. The core issues are well covered: the risks and benefits of research; the assessment of risks and their acceptability; the problems of consent, and the ethical and experimental use of children. British paediatricians will find the chapters ‘Ethical issues in exposing children to risks in research’ by Dan Brock, a philosopher, and ‘Vulnerable children’ by Robert Cooke, a paediatrician, of particular interest and value. In both the UK and the USA, the law has played a relatively small role to date. In the UK there is no law which specifically addresses paediatric research (and no case law), though legal provisions which protect the interests of children also circumscribe the conduct of research. In the US, only one case involving research is quoted and there is no statutory law. Nevertheless, regulations though these are relatively unrestrictive. They are recorded in a useful appendix which also contains the 1947 Nuremberg Code, the 1989 World Medical Association Declaration of Helsinki, and a list of points to consider in proposing or reviewing research involving children.

In his chapter, ‘The law of human experimentation with children’, Leonidas Glantz, a professor of health law, who ‘WITW’ has done in recent years, however, is set a general tone that indicates that children have some rights that they may exercise on their own behalf, and that parents and institutions have an obligation to respect their wishes. The role of adults to ensure that children are not subjected to unnecessary or excessive risk of discomfort. The law is a crude tool for accomplishing this goal’. Hear! Hear!

A G M CAMPBELL
Emeritus professor of child health


It is my perception that colleagues consider inherited metabolic disorders rare and esoteric, to be considered at the bottom of a list of differential diagnoses. For individual disorders this may be true, but as a group metabolic diseases are an important cause of morbidity and mortality through the neonatal period into adult life. Rapid advances in the understanding of disease mechanisms have brought with them an increased ability to diagnose disorders accurately (both by new, sensitive range of investigations before transfer to a more specialised unit for more detailed investigation and management.

The undergraduate and postgraduate training of physicians is generally poor and there has been a shortage of readable, middle sized texts on the subject. For most workers within the field, The Metabolic Basis of Inherited Disease edited by A Textbook, and currently the standard reference textbook. It is unsuitable, however, for the medical student and junior doctor who wish to have a taste of the subject rather than a feast. Holton has attempted to fill a gap in the market by producing a middle sized text which it is claimed is ‘concise, but nevertheless comprehensive and accurate’. The first edition of the book published in 1987 failed to fulfil these commendable...
ides of this was not the fault of the editor as it was produced during a time of rapid development so that it was inevitably dated on release. Some of these failings have been more than adequately redressed with this edition. The additional chapters on peroxisomal disorders and defects of the mitochondrial respiratory chain add considerable strength to the publication and are probably the best changes to the book. The other chapters, with the exception of the chapter on lysosomal storage disorders, appear to have been revised significantly to bring them very much up to date.

The content of the book is now more than adequate and it is very easy to read. I am sure that some would prefer an organ specific approach to the subject and a special section on the practical management of the sick newborn would be useful for the general paediatrician. I am uncertain how useful general paediatricians will find the detailed discussion on collagen disorders, excellent though it is. The latest edition has some important omissions. The Neurological Aspects section is non-existent. The second edition of Inborn Metabolic Diseases, edited by Clayton and Round (Blackwell Scientific Publications, Oxford, 1994) is another text well worth browsing through before committing oneself to a purchase. This book is not strictly a text on inherited metabolic disorders although many of the disorders are considered. It takes a laboratory bias, but includes interesting chapters specifically about the approach to metabolic diagnosis in the sick newborn and routine screening programmes as well as others. Its organ specific approach will appeal to many paediatricians.

In conclusion, Holton’s second edition is a great improvement over its predecessor. It will be widely read by paediatricians and deservedly so; it is probably the best text of its type currently in print, although look at the alternatives above before purchasing it.

J E WARRAITH
Consultant paediatrician


There has been an explosion of new ideas and methods for the diagnosis and management of epilepsy. The pace of change is reflected in extensive additions and amendments to the second edition of Epilepsy in Children. In 1981 the current classification of epileptic seizures, into partial (focal), generalised, or unclassified types was published by the International League Against Epilepsy (ILAE). In 1983 a workshop with Aicardi was held in Venice, Italy, to participate, to define epileptic syndromes in children. The proceedings, published in 1985, contributed to the 1989 ILAE classification of epileptic syndromes and required a second edition in 1992. That is a problem, of course, such as the term ‘complex’ with one meaning when coupled with partial seizures and another with absence, certain of the syndrome categories, or sometimes clumsy terminology. There is also a sizeable minority of children whose epilepsy defies classification. However, the result, helped by better ways of imaging the structure and function of the brain, has brought considerable clarity and understanding, and a common language, to what had seemed a confused subject.

A practical approach to diagnosis, demonstrated in Aicardi’s book, is to judge the seizure types a child experiences, with other clinical features if present, and then consider the underlying syndromes. Although each seizure type has presumably occurred since mankind first had epilepsy, it is fascinating how many new clinical features are still being recognised, particularly for those of frontal origin. It is surprising that some clinicians, and textbooks for that matter, still seem to believe that all focal seizures are of temporal lobe origin or still seem happy with the vague and obsolete terms ‘grand mal’ or ‘petit mal’. Ictal and interictal EEG appearances are essential to diagnosis, for example distinguishing typical and atypical absences, and illustrate the inappropriateness of separating neurophysiology from clinical practice: how can a sensible opinion be given if only scanty clinical details are available and do all clinicians read the technical details? For example, does this seizure type is occurring? Are other techniques such as sleep or deprived EEGs used as often as they should be? Here Aicardi’s book is a definitive guide.

There are a host of syndromes, some less clear cut and some whose natural history is still being studied, and yet more which are poorly classified, such as the myoclonic seizure disorders of early childhood, which do not suit the classification of disorders of seizure type. As the Lennox-Gastaut and Doose mean different conditions to different people: Aicardi accepts the existence of the former but not the latter. Special situations, such as seizures in neonates, after head injury or with tumours, need separate consideration, as does the increased interest in all forms of status, particularly non-convulsive. Again Epilepsy in Children is clear, concise, and very up to date. It also provides a thorough overlap between different chapters, such as febrile convulsions and complex partial seizures, to let each be read alone without a need to constantly cross reference, which is very useful when wanting an opinion on individual patients.

Increased diagnostic and prognostic accuracy has been complemented by the introduction of new ‘designer drugs’ and by the resurgence of epilepsy surgery, exciting developments that Aicardi treats with cautious optimism. The use of older drugs is also discussed: for example, if an adolescent presents with epilepsy and a single tonic seizures due to juvenile myoclonic epilepsy, carbamazepine does not help and may make matters worse; withdrawing treatment after two years’ seizure free existence is not appropriate to many epilepsies, and steroids, especially corticosteroids, are no longer the automatic choice for infantile spasms. As well as reviewing the evidence for and against current practice, recent advances in investigation and treatment are further covered in the last part of the book, expanded from three to five chapters, which also deal with the differential diagnosis, prognosis, and overall management. This again reflects increased emphasis on treating the whole child and not just fits. Clearly an international text cannot go into specifics which vary in different countries, such as the value of self help groups – especially in social problems such as schools, jobs and driving – but the only area now not covered in depth is the origin and management of the behaviour disorders which can be associated with epilepsy.

The first edition received rave reviews: this edition is even better. It is difficult to review a book by a major figure without hagiography, particularly when it is authoritative based on both extensive personal experience and over 2500 references. My copy already has a queue of colleagues waiting to borrow it. There are plenty of reasons for a new optimism in epilepsy: Aicardi’s second edition is one of the best ways to find out more.

PETER BAXTER
Consultant paediatrician/paediatric neurologist


If all the clinical specialties and all the service specialties are listed in separate columns and one is chosen from each, the numerous combinations provide opportunities for books. Some combinations are more exciting (lymphocytic immunology), while others are of doubtful importance (gynaecological haematology). Some combinations of specialties though are relevant and a book can distil information that might be difficult to extract from major textbooks in either specialty. This book, bringing together neonatology and clinical biochemistry, is such an example. It is a smallish (not quite pocket sized) paperback written by a neonatologist and clinical biochemist from Birmingham who work together. It is one of a series of books commissioned by the Association of Clinical Biochemists.

The first two chapters, on neonatal care and newborn physiology, are aimed at biochemists. There are then chapters on the biochemistry of the term and the preterm infant. Jaundice is comprehensively discussed and there are good sections on measurements and electrolytes. There is a very good chapter on neonatal screening and an excellent one on the diagnosis of inherited metabolic disorders in the newborn period. Both have clear diagrams of the relevant metabolic pathways that show the site of the enzyme deficiency and its consequences. The chapter on drugs is very brief and apart from a table on therapeutic drug monitoring seems out of place. The chapter on parental nutrition focuses on the metabolic complications and routine biochemical monitoring. There is a protocol for capillary blood sampling with an attached photo of a newborn finger. The impression given is that the heel has been squeezed to the point of gangrene! It was sad to see the description of the use of a manual heel stylet rather than the equally effective but less painful spring loaded heel lance. The biochemical reference ranges in the appendix are comprehensive and clear.

For the clinical biochemist, the information is about newborn care. The book will be handy, but what is in it is for the neonatologist. All the information within this book is contained in the large textbooks of neonatology and paediatrics, and much will be found in the standard working manuals of neonatal intensive care. It is very convenient though to have the biochemistry of neonatal care concentrated into one small volume. This is the sort of book to keep on the newborn intensive care unit, for quick
The Inherited Metabolic Diseases

J E Wraith

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