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 recent approach to this book could be the 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 and professional reasons on how far from clearly articulated 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 acceptable, but may result in further improvements in the health and wellbeing of all children. This is true even in some 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 and media on the 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 opposition 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. In his preface to the book, John B. M. K. Addington-Hall comments on the UK 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’.

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 ethically main heads: 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 need for appropriate procedures when using 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. The basic ethical and regulatory considerations are generally the same, although the occasional differences in wording and emphasis are still there, but are not to be expected.

It is my impression 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 in 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 in utero and at birth) as well as introducing potential new therapies such as enzyme replacement or gene transfer.

We live in an exciting era!

Of course, for some the only knowledge desired or required will be the telephone number of the nearest metabolic specialist. For others a sound grounding in the basic principles of inherited metabolic disease will enable them to perform a sensitive and targeted approach and investigations before transfer to a more specialised unit for more detailed investigation and management.

The undergraduate and postgraduate texts are intended for students who are 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榈 and Long and Pediatric and Metabolic Diseases of Infancy and Childhood edited by Kliman are the standard reference textbooks. 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
ideals. 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 chapters in 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. There are, however, considerable gaps in this. There are problems, of course, such as the term ‘complex’ with one meaning when coupled with partial seizures and another with absences, certain of the syndrome categories, or a 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 the book, is to define 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 manuals that are available? Or 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 stand out clearly in the different seizure types 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.

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 a new epileptic syndrome, absence 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 syndromes and steroids, especially corticosteroids, are no longer the automatic choice for infantile spasms. As well as reviewing the evidence for and against current practice, the book includes the scientific and therapeutic data on the newer anticonvulsants, with chapters on benzodiazepines and barbiturates. A further feature is the emphasis on the practical management of the sick 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/Neonatologist


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. The combination of neonatology and clinical chemistry has not been covered before as far as I can see, and this small book is a happy exception. It includes interesting chapters on prenatal diagnosis, the newborn’s liver, the newborn’s kidney, the newborn’s GI tract, the newborn’s endocrine systems, and many others. The book is easy reading and informative. The table of contents is helpful, as is the index; it also has a useful section on realistic expectations. The book is well illustrated and includes clear, easy to understand tables and figures without excessive detail. There are a few corrigenda in this book, which is to be expected. There is a useful glossary and bibliography. The appendices are comprehensive and clear.

For the clinical biochemist, the information in this book will be handy, but what is it in it for the neonatologist? Explanations within this book are 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