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 paediatrician interested in this book will be of the most 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 reasons on publishers’ behalf) have not been clearly annotated in the text. They are at the end of the book and should be searched for, as they illustrate the argument with further authority.

MICHAEL ROGERS
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Probably most, if not all, paediatricians believe that research using child subjects is not only desirable but essential to 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 of the facts and media and 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. The book is the result of a symposium and will be published in book form in the near future. It has been the result of an initiative by the Child Health Ethics Committee, of the Royal College of Physicians of London, in the light of the recent controversy concerning the use of children in research.

The main problems that children face in research are twofold. First, they have a different capacity to understand what they are being asked to do from adults. Second, there are problems with controlling for the effects of parents and the way that parents have of understanding the research. The American book is a useful guide for those who are interested in the field, but it is likely to be more directly relevant to the work of those who are involved in research and in the legal and ethical issues of research on children.


It is my perception that colleagues who consider inherited metabolic disorders rare and esoteric, should 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 mortality and morbidity, particularly 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 at a molecular level 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 sensible range of investigations before transfer to a more specialised unit for more detailed investigation and management.

The undergraduate and postgraduate teaching of inherited metabolic disease is currently 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 Zuckier and Cotlove (Pp 1340 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

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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 changes 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 has been great emphasis on the classification of the syndromes. There has been a (partial into extensive) additions and amendments Arciardi. There will be currently by alternatives sick Aicardi. It was clearly redressed about the text on Biochemistry Clinical Biochemistry (if adequate and is an exception of the chain add considerable 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 has been great emphasis on the classification of the syndromes. There has been a (partial into extensive) additions and amendments Arciardi. There will be currently by alternatives sick Aicardi. It was clearly redressed about the text on Biochemistry Clinical Biochemistry (if adequate and is an exception of the chain add considerable


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 combinations are bring 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 blood and electrolytes. There is a very good chapter on neonatal screening and an excellent one on the diagnosis of inherited metabolic disorders prevalent 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 example of a picture of the newborn infant. The chemical 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 laborious spring loaded heel lance. The biochemical reference ranges in the appendix are comprehensive and clear.

For the clinical biochemist, the information is useful, but what is in it for the neonatologist? Everything 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


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 was that of the International League Against Epilepsy (ILAE). In 1983 a workshop conference, with Arciardi as Chair, was held 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. 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 this book, is an advantage. Each seizure type is characterized, 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 recognized, 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 medical journals? Is the seizure type occurring? Are other techniques such as sleep or sleep deprived EEGs used as often as they should be? Here Arciardi'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 strongly resemble the syndromes such as the Lennox-Gastaut and Doose mean different conditions to different people: Arciardi 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 succeeds in making up the gap 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 Arciardi treats with cautious optimism. The use of older drugs is also discussed: for example, if an adolescent presents with epilepsy, the best treatment 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 manage epilepsy and steroids, especially corticosteroids, are no longer the automatic choice for infantile spasms. As well as reviewing the evidence for and against current "practice" in epilepsy research 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: Arciardi's second edition is one of the best ways to find out more.
The Inherited Metabolic Diseases

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