

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.

The book does have competition. The text, *Inborn Metabolic Diseases*, edited by Fernandes, Saudubray, and Tada (Springer-Verlag, Berlin, 1990) has some similarities with Holton's text. There are, however, considerable gaps in this book as the editors decided to consider only treatable disorders. Nevertheless, some will find this book preferable (if now a little dated) because of its clear approach to treatment of many inborn errors. *Clinical Biochemistry and the Sick Child*, edited by Clayton and Round (Blackwell Scientific Publications, Oxford, 1994) is another text worth browsing through before committing oneself to a purchase. This book is not strictly a text on inherited metabolic disease, 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 press, although look at the alternatives above before purchasing it.

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**Epilepsy in Children.** 2nd Ed. By Jean Aicardi. (International Review of Child Neurology.) (Pp 571; \$120 hardback.) Raven Press, 1993. ISBN 0-7817-0111-2.

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 met in Marseilles, with Aicardi a participant, to define epileptic syndromes in children. The proceedings, published in 1985, contributed to the 1989 ILAE classification of epilepsy 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 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 report to decide what seizure type is occurring? Are other techniques such as sleep or sleep 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 subdivide easily. Syndromes such 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 has enough 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 generalised tonic clonic 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 forms of epilepsy, and steroids, especially corticotrophin, are no longer the automatic choice for infantile spasms. As well as reviewing the evidence for and against current practice in each chapter, 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 authoritatively 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.

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**Neonatology and Clinical Chemistry.** By Anne Green and Imogen Morgan. (Pp 213; £15 paperback.) ABC Venture Publications, 1993. ISBN 0-902429-04-3.

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 clearly absurd (adolescent 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 glucose, fluid, and electrolytes. There is a very good chapter on neonatal screening and an excellent one on the diagnosis of inherited metabolic disorders which present 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 parenteral nutrition focuses on the metabolic complications and routine biochemical monitoring. There is a protocol for capillary blood sampling with an unfortunate photo of a baby's heel. 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 styler 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 on the newborn will be handy, 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

reference when abnormal results are 'phoned through from the laboratory.

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**Fetal and Neonatal Pathology.** 2nd Ed. Edited by Jean Keeling. (Pp 609; DM318 hardback.) Springer-Verlag, 1993. ISBN 0-387-19711-7.

The Royal College of Obstetricians and Gynaecologists and that of Pathologists have set a target rate for necropsy after perinatal death of 75%. Most regions fail to achieve this target: in Wales, for example, the rate is 57%. This is a shame because the perinatal necropsy has an important audit function: some of the best perinatal mortality meetings are those at which there is active input by a pathologist who has insight into the *faux pas* of both obstetrician and paediatrician. The perinatal necropsy also provides a most useful basis for parental counselling about future pregnancies, even when the clinicopathological classification is only changed in 12.5% of cases. It is, of course, on the histopathologist that the burden for providing a good perinatal necropsy falls and Jean Keeling's book will provide that histopathologist with first class support.

In the era of ovulation induction and in vitro fertilisation, higher order multiple pregnancies are becoming more common. The perinatal mortality among twins is five or six times that of singletons and a chapter by Geoffrey Machin has been dedicated to this topic. It is the monochorionic multiple pregnancies that carry particularly high perinatal risks. This is largely because of the presence of interfetal vascular anastomoses (IFVA) in most monochorionic placentas; IFVA can result in the development of twin transfusion syndrome and twin reversed arterial perfusion (TRAP). This last condition is associated with the congenital anomaly, holocardius acephalus. It is in this chapter of the book, ostensibly for pathologists but really remarkably clinical, that we learn that in TRAP prenatal Doppler studies have shown that the intact 'pump' twin perfuses the acardiac twin retrogradely via the umbilical artery of the latter. Arterial flow is reversed in the major part of the aorta. A proportion of the arterial blood of the pump twin, destined for the placenta, crosses instead in large placental arterioarterial anastomoses to reach the common iliac arteries of the acardiac twin. This paradoxically results in structures in the lower half of the body usually surviving better than the brain which generally does not survive the abnormal haemodynamics.

The histopathologist is a close relative of the forensic pathologist and, through him or her, the detective. Jean Keeling, in her own excellent chapter on intrapartum asphyxia and birth trauma, teaches that 'the pathologist should examine skin creases and ears for residual [meconium] staining as even a still-born fetus may have been washed before being shown to the parents and despatched to the mortuary'. She also points out that, when there are facilities for ventilation, the often under-rated factor that limits survival is the presence and extent of ischaemic damage to the myocardium. Ischaemic injury is generally restricted to the inner third of the myocardium and papillary muscle. The theme is taken up in the chapter on the cardiovascular

system by Jerry Cox and Jakob Briner. They stress that myocardial necrosis, which was once thought to be uncommon in the perinatal period, has been shown to be a relatively frequent event among those coming to necropsy after severe perinatal asphyxia. Recent necrosis is seldom identifiable macroscopically, but focal whitish scarring may identify older injury. When myocardial ischaemic damage is suspected, systematic sampling of all areas of the heart is essential and special stains are often required to identify fresh necrosis.

For the practising clinician, a chapter on iatrogenic disorders (by Jean Keeling) is particularly cautionary. Here complications arising during neonatal intensive care are described: ulcerations due to endotracheal tubes, necrotising tracheobronchitis associated with high frequency jet ventilation, perforation of the lung by chest drains, oxygen toxicity and undesirable sequelae of peripheral artery sampling (for example, the slow development of a contralateral hemiplegia after temporal artery puncture, and carpal tunnel syndrome after haematomas due to radial artery sampling) are just a few that are mentioned.

George Bernard Shaw wrote in *Back to Methuselah*, 'Life is a disease; and the only difference between one man and another is the stage of the disease at which he lives'. The theme is taken up in the final section of the final chapter of this book. That section is called 'The special senses' and is by Brendan McDonald. It reminds us that at birth the taste buds are present as specialised chemoreceptors formed by modification of the oral cavity surface epithelium, with the highest density of taste receptors present on the anterior two thirds of the tongue. Unlike the olfactory chemoreceptors, the specialised taste receptor cells are continually being desquamated and replaced. The density of taste sensing cells is greatest in infancy and diminishes with advancing age.

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**Liver Disorders in Childhood.** 3rd Ed. By Alex P Mowat. (P 491; £75 hardback.) Butterworth Heinemann, 1994. ISBN 0-7506-1039-5.

The overall prognosis of paediatric liver disease has improved dramatically in the last decade owing to improved diagnostic techniques and better medical and nursing care. This has been driven to some degree by the availability of liver transplantation which has rescued many infants from certain death. However, transplantation is hazardous and there is a limited pool of donor organs so it remains a last resort. Early referral and accurate diagnoses are important in detecting children who might benefit from medical treatment or conventional surgery and thereby delay or avoid transplantation. Thus the objective of *Liver Disorders in Childhood* is to assist clinicians in identifying such children by emphasising presenting clinical features and describing associations with other diseases, for example, cystic fibrosis. The book retains the compactness and consistency of style present in previous editions. The combination of text, tables, and illustrations is easy on the eye. It is possible to dip into a section as well as indulge in a good solid read. Compared with previous editions, there are new chapters on autoimmune chronic active

hepatitis, sclerosing cholangitis, chronic hepatitis, and  $\alpha_1$ -antitrypsin deficiency as well as greatly expanded chapters on viral hepatitis and inborn errors of metabolism, which reflects the great advances in these areas. For example, hepatitis C was an unknown entity until five years ago and now details of the genome, propensity to genetic heterogeneity, epidemiology, progression to cirrhosis in infected patients, possible role in autoimmunity and treatment with interferon alfa, are available. The chapter on inborn errors of metabolism has been restructured to emphasise clinical features which alert and direct clinicians towards considering a disorder of metabolism, for example, urea cycle defects, glycogen storage diseases. This is useful as it applies some logic to the investigation and management of inborn errors of metabolism. Various disorders are discussed in more detail, of particular note are defects in bile acid synthesis and tyrosinaemia. The application of new techniques and refinements in mass spectrometry has led to the definition of at least two inborn errors of bile acid metabolism. Furthermore, new treatments for both the bile acid disorders and tyrosinaemia have been developed which may transform the management and prognosis of these disorders.

Malnutrition is common and often underestimated in chronic liver disease and is the only area not well covered in this book. As growth is one of the unique characteristics of childhood, it would have been helpful to have some discussion on detection of malnutrition and principles of nutritional support in children with liver disease.

Although liver disease in childhood is uncommon, the challenge of caring for such children is great and may involve many people in community and hospital practice. Paediatric liver disease is often chronic and requires specialist interventions from enteral nutrition to post liver transplantation monitoring. Therefore, almost all health care workers are likely to encounter children with hepatic problems at some stage and will need to refer to an authoritative and comprehensive textbook. *Liver Disorders in Childhood* fulfils that need admirably.

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**Diseases of Infection.** Edited by Norman Grist *et al.* (Pp 453; £22.50 paperback.) Oxford University Press, 1993. ISBN 0-19-262307-9.

There is a place in the market for another short text on diseases of infection. Such a book is not only for the specialist in infectious diseases but should be relevant to all branches of medicine. The difficulty faced by authors presenting a 'handbook' but attempting to cover a huge subject is one of finding the correct balance and maintaining the reader's interest, as such a format dictates that many areas of interesting controversy will be dealt with very briefly. This book succeeds in some areas, for example, very good chapters on erythematous rashes with excellent photographs, on septicemia, and on the necessity to understand how the laboratory works, and the need to supply it with the correct specimens. However, it fails on others, for example, a condensation of HIV infection to 11 pages, no definitive educational section on the present ideas on continued feeding for children with gastroenteritis and the need for an absolute