Unilateral multicystic dysplastic kidney

EDITOR—Webb et al justify their policy of performing nephrectomy in healthy infants with prenatally detected multicystic dysplastic kidney (MCDK) on the basis of their findings in three children and their interpretation of the literature.1 Paediatricians may wish to scrutinise the methodology and conclusions of this paper more closely before referring asymptomatic infants for ‘prophylactic’ nephrectomy. On the evidence presented, the diagnosis of hypertension in their first two patients has not been conclusively proved. Furthermore, it is difficult to see how the renal pathology in their third patient can be attributed to MCDK in the absence of cysts and, more importantly, ureteric atresia.

Making a diagnosis of hypertension in children is problematic—indeed obtaining reliable reproducible blood pressure readings in fractionated infants can be very difficult. The measurement of peripheral plasma renin activity, which was performed in these children, is a relatively poor guide to the aetiology of hypertension. A credible diagnosis requires selective renal vein renin sampling. The role of echocardiography in the diagnosis and assessment of mild or moderate hypertension in childhood has yet to be validated. The authors have not identified the source of the normal ranges quoted—an important consideration since different studies have yielded different ‘normal’ values for the parameters quoted and the ‘normal’ range has often been derived from a limited number of individuals in different age groups.

In their third patient, a hypertensive girl of 14, retrograde pyelography is said to have revealed ‘remnant renal tissue in the right loin’. Atresia of the pelviureteric junction or proximal ureter is a characteristic feature of MCDK which, on retrograde pyelography, is associated with the distinctive finding of a blind ending ureter in this case. The flow of contrast material across a ureter into a remnant of the collecting system, effectively excludes the diagnosis of MCDK.

It is disappointing that the authors felt it necessary to quote the outdated findings of an informal postal survey of American paediatric urologists2 to support their arguments for nephrectomy. Published in 1978 and spanning an undefined period, this survey predates the era of prenatal ultrasound and many of the criteria demanded of a scientific study. In addition to misquoting the number of respondents to the survey (136 not 48), the authors have omitted to inform their readers that 10% of the paediatric urologists surveyed reported serious operative complications resulting from nephrectomy for MCDK including deaths, renal failure, intestinal obstruction, and removal of the wrong kidney.

The experience of other centres in relation to the risk of hypertension in MCDKs is different to that reported by Webb et al. Data on 441 children with MCDK was submitted to the Multicystic Kidney Registry3 from 49 centres in the United States and Canada. Of the 441, 260 were managed conservatively with follow up of up to five years in some cases. No cases of hypertension were reported in this series.

Rickwood et al reported a series of 44 children with prenatally detected MCDK.4 Blood pressure was measured continuously over a mean follow up of three years in 38 of these children and none was found to be hypertensive. These findings mirror our experience in Leeds where we have not encountered any case of hypertension associated with prenatally detected MCDK nor has an MCDK been identified in a child presenting with hypertension.5

Analysing 454 cases of hypertension admitted to the renal unit at the Hospital for Sick Children in Great Ormond Street, from 1975–85, Deal et al6 documented a wide spectrum of underlying renal pathology without encountering a single MCDK (J E Deal, personal communication). A retrospective study of 42 children undergoing nephrectomy for renal hypertension in Glasgow did not include a single case of MCDK. Similarly, a detailed review of 22 cases of surgically treated hypertension of renal origin in children, undertaken in Boston, did not document a single MCDK.7

It is important to distinguish clearly between other forms of renal dysplasia and MCDK, a related distinctive anomaly characterised by tense, non-functioning cysts and absent or non-functioning renal parenchyma, which is almost invariably associated with complete ureteric atresia or severe distal ureteric obstruction. The authors’ description of the radiological and pathological features of their third case clearly indicate that the renal pathology belongs in one of the former categories of dysplasia rather than representing a genuine MCDK.

Cases of hypertension conclusively linked to MCDKs have very rarely been reported but published evidence indicates that the magnitude of risk is extremely low. Menster et al,8 reviewing the published literature on the risks associated with prenatally detected MCDKs, concluded ‘It no longer seems advisable to routinely remove MCDK in young patients for either diagnostic or prophylactic reasons’.9 Paediatricians working in centres in which conservative management is undertaken can be reassured. The mounting weight of published evidence continues to favour a non-operative approach to the management of prenatally detected MCDKs in healthy infants.

Dr Webb and colleagues: Mr Thomas and Dr Fitzpatrick raise some important questions, to which we are grateful for the opportunity to respond to them in detail.

We agree that the diagnosis of hypertension may be difficult in small infants and children, though having detected systolic blood pressure in excess of the 95th centile for both age and height on repeated occasions using an appropriately sized cuff with the patient in the resting state, we have met widely accepted criteria for the diagnosis of hypertension in all three cases.10 The presence of end organ damage (left ventricular hypertrophy diagnosed using accepted normal ranges1) in the first two patients confirms the longstanding duration and severity of this hypertension. It is generally agreed that raised peripheral plasma renin activity, while never truly diagnostic, is a useful indicator of renally mediated hypertension; renal vein renin sampling may be potentially technically difficult from the small vein which drains a MCDK and is generally used to predict response to surgery rather than to diagnose hypertension. The resolution of hypertension and echocardiographic changes after nephrectomy provide further supportive evidence for the hypertension in these three children being renally mediated.

MCDK represents the extremes of a spectrum of obstructive cystic dysplasia; while strict pathological criteria require the presence of an atretic ureter for a diagnosis of MCDK to be made, in practice, physicians and surgeons make a clinical diagnosis on the basis of DMSA and ultrasound findings. Many children with lesions such as those seen in our third case may therefore be under followed with a clinical diagnosis of MCDK.

Our interpretation of page 215 of Bloom and Brosman’s paper10 was that 48 (35%) of the 136 urologists sent questionnaires responded to the section on clinical findings associated with MCDK; Renal vein renin, Thomas and Fitzpatrick’s interpretation is correct, 15% of 136 is a much larger number of urologists reporting hypertension than 15% of 48, and lends further weight to our argument supporting hypertension as a significant complication of the MCDK. Operative morbidity and mortality have fallen significantly since Bloom and Brosman’s publication; our previous report of a frequency of perioperative complications11 has continued over recent years.

The report of the Multicystic Kidney Registry12 on 441 children with MCDK in the United States and Canada of whom 181 underwent nephrectomy predominantly between 7 and 12 months of age, confirming that nephrectomy continues to be considered a valid treatment option in many centres. Contrary to Thomas and Fitzpatrick’s statement, four children followed up conservatively developed hypertension, though this was ‘believed to be unrelated to the multicystic kidney’. Without an alternative explanation or further investigation the authors are not prepared to state whether hypertension was unrelated, MCDK related hypertension cannot be excluded. Our paper reports three further cases of hypertension in children diagnosed as having MCDK in addition to those others previously reported in the literature.

The surgical versus conservative management of the MCDK will remain an area of great controversy. Only by collecting complete and accurate data from large, unselected series of patients treated both surgically and conservatively will the true incidence of complications of both approaches emerge, and we make a further plea for the setting up of a national registry to expedite this.

EDITOR,—Webb et al continue the debate about conservative versus surgical management of multicystic dysplastic kidney (MCDK) with case reports of three patients for whom surgery was undoubtedly indicated. We would, however, be concerned against the experience of tertiary referral centres being extrapolated to the general population.

Experience at Northwick Park Hospital, a large district general hospital, suggests that MCDK is more common than the published incidence figures of one in 4300 live births.1 We have reviewed all cases of MCDK diagnosed in infants born at Northwick Park Hospital in the 10 year period 1987–96 during which there were 35 537 live births. During that period, routine antenatal ultrasonography was carried out at 16–18 weeks’ gestation. Further scans were performed at a later stage in the pregnancy only if there was a clinical indication. Of the 14 cases of MCDK diagnosed on antenatal ultrasonography, 11 were detectable on the routine scans. In all cases, including one case in which the pregnancy was terminated at 18 weeks because of contralateral renal agenesis and multiple congenital abnormalities, the abnormality was detected by ultrasonography at 25, 26, and 39 weeks’ gestation, respectively. One further MCDK was detected by palpation of a renal mass on the routine neonatal examination after birth.

The incidence of MCDK diagnosed in an unselected district general hospital population was one in 2400 live births. As the majority of mothers were scanned on only one occasion before their pregnancy, our figures may still represent an underestimate of the true incidence of this condition.

During the same 10 year period, only one infant with unilateral renal agenesis was identified. Antenatal ultrasonography, compared with the published necropsy incidence in adults of one in 1070.7 Although the relatively large size of the fetal adrenal gland makes the diagnosis of unilateral renal agenesis difficult to identify on antenatal ultrasonography, our data may also be interpreted as suggesting that a large proportion of solitary kidneys identified in adult life may in fact be due to regression of MCDK. All of our infants with an antenatal diagnosis of MCDK had serial postnatal ultrasonography, a DMSA isotope scan, and micturating cystourethrogram performed postnatally. Associated renal tract abnormalities in the contralateral kidney were found in only three infants (two grade I and one grade IV vesicoureteric reflux), a similar incidence to that found in other series.8

Complication rates determined from case reports in the literature will, inevitably, be subject to reporting bias and must be interpreted in the light of data on incidence and natural history in non-referral populations if the risks of conservative management are to be put in proper perspective. Meanwhile the dilemma of management of MCDK remains controversial.

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Gastrostomy in children with Crohn’s disease

EDITOR,—We read with interest the article by Cosgrove and Jenkins, describing their experience with percutaneous endoscopic gastrostomy (PEG) in children with Crohn’s disease.1 They concluded that the advantages of gastrostomy outweigh the risks. However, we recommend that nasogastric tube feeding should be the first choice for supplemental nutrition as 25% of our patients with Crohn’s disease and growth failure used this method successfully for long time (mean duration 10.5 months).2 When this method is not possible, gastrostomy is a good and safe option. In case of previous abdominal surgery, surgical gastrostomy tube should be considered rather than PEG.

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Early micturating cystourethrograms after urinary tract infection

EDITOR,—Craig et al showed that timing of micturating cystourethrography (MCU) does not affect the detection and grading of vesicoureteric reflux (VUR) if the test is performed more than one week after diagnosis and treatment of a urinary tract infection (UTI).1 There is a view that early MCU may reveal a higher prevalence of VUR due to transient inflammatory changes at the level of the vesicoureteric junction.2 This is to some extent refuted by the work of Craig et al.1 However their study was unable to address the issue of VUR in the first week after diagnosis of UTI when the possibility of such a situation would seem greatest.

To further the issue of transient VUR, it is assumed that its presence is insignificant, therefore justifying a policy of delaying MCU (whether for one week or six weeks). The logic which supports this strategy may be questioned. An opposing view could be that if


EDITOR,—I read this paper, and Dr Bell’s commentary. Dr Bell referred to the kidneys at risk in the same manner as timewhentheurineisinfected, this may put UTI increases the likelihood of VUR at the time. Before reading this my view was rather limited to children who don’t sleep and others who sleep too much, with occasional night terrors or seizures, and wondering when I will see my first case of narcolepsy. I’ve always been haunted by Professor Illingworth’s view which blames refusal to go to sleep on parental mismanagement and advises that the child be ‘left to cry’. I prefer William Cams’s advice that ‘The devil gets into them [babies] at an early age and of course you should have the baby in your bed’. As usual the best advice is probably somewhere in between the two extremes. This is an interesting chapter and should be read. Finally, the excellent literature review. The main interest of course is ‘am I quoted?’ For me with only an occasional publication, this is unlikely, but in this case I am, slightly obliquely. Can you spot it?

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BOOK REVIEWS


The eagerly awaited 15th edition of Recent Advances in Paediatrics has just arrived. There is no disappointment. Professor David has maintained the high standard we have been led to expect. This is an excellent series which enables paediatricians to keep themselves up to date with medical progress in a relatively painless fashion. This book is well worth buying and reading carefully.

The format is unchanged. There are a dozen chapters of general paediatric interest, and the comprehensive literature review by the editor. Each topic is clearly presented with helpful tables and numerous subheadings. Each finishes with a number of key points and numerous references.

The subjects covered are wide ranging including bronchiolitis, large airways disease, thalassaemia, congenital hypothyroidism, and coeliac disease. There is a stimulating and partly speculative account of superantigen coeliac disease. There is a stimulating and partly speculative account of superantigen coeliac disease. There is a stimulating and partly speculative account of superantigen coeliac disease.

Two chapters are devoted to problems in the newborn, pulmonary hyperten- sion, and bronchiolitis. A chapter on cholestasis, not usually a clinical problem in the UK, illustrates many of the health and social problems of the undeveloped world. Spread may be rapid, the number of patients enormous, and the mortality appalling with 10 000 deaths in South America alone over three years. Good hygiene and clean water could prevent much of this, but governments lack the will to deal with poverty and the environment while population expansion and wars compound the problem.

Recent Advances in Paediatrics usually includes a chapter on recent progress in a physiological topic and its clinical applica- tion, and this is the case with nitric oxide. This is certainly a new topic which has only attracted study in the last few years, although it is certainly a new topic which has only attracted study in the last few years.

A chapter on headaches guides us through the appropriate imaging, stressing how un- common it is to find unexpected pathology with a normal clinical examination, but recognising the role of anxiety in generating investigations. The fact that headaches and a brain tumour are extremely rare, are nearly always associated with other signs and symptoms, and get progressively worse should help paediatricians to resist reaching for the imaging ‘request forms, with careful follow up being more appropriate.

The final chapter is on sleep problems. Before reading this my view was rather limited to children who don’t sleep and others who sleep too much, with occasional night terrors or seizures, and wondering when I will see my first case of narcolepsy.

I would like to add to Dr Bell’s comment in that I am sure many paediatricians out of the first flush of their youth can remember using a mixture of honey and brandy placed on a dummy as an analgesic while doing exchange transfusion. I imagine you will receive many similar letters—plus ça change... M J MAGUIRE
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recent proceedings.

The eagerly awaited 15th edition of Recent Advances in Paediatrics has just arrived. There is no disappointment. Professor David has maintained the high standard we have been led to expect. This is an excellent series which enables paediatricians to keep themselves up to date with medical progress in a relatively painless fashion. This book is well worth buying and reading carefully.

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MARTIN MONCRIEFF
Consultant paediatrician


Modest both in size and price this book stands between the grand reference book on paediatric infectious disease ‘Feigin and Cherry’ and the pocket sized manuals now available. Its challenge is to be a good, advanced reference book and yet be brief enough to be of use to the practising paediatrician who needs to look up a subject quickly. In many ways it meets the challenge. Much of the text is organised by systems rather than by infectious agents. Thus we are treated to chapters on respiratory, cardiac, gastrointestinal, and neurological infections, and those in the eyes and skeleton. Following these are sections on fever, neonatal infections, and HIV.

The text includes excellent illustrations, for example the beautiful picture of the distribution of cervical lymph nodes. The large number of x rays and computed tomograms make for particularly interesting viewing. There are a dozen pages of excellent colour plates of rashes at the start of the book. The same photographs appear later in black and white where many, such as those of scarlet fever, are of little value.

The quality of almost all the contributions is very high. Particularly impressive are the chapters on HIV and the child with recurrent infections. There is an interesting and critical discussion on urinary tract infections. It does not seem to matter that the authors, mainly from Australia, UK, and the United States, are working so far apart, except perhaps when it comes to the section on tuberculosis where the author states ‘Multiple puncture techni- ques (for example Heaf test) are no longer recommended’. Many British paediatricians are using this test, perhaps inappropriately, and it would have been useful to have had some information about it.

It is always difficult to do justice to neonatal infection in a book of this type. The section on congenital infections is comprehensive but the discussion on the neonate and young infant is disappointing. A reprint on the title page ‘management of the newborn infant 30–90 days’ confusing

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because neonatal infection is included in the text. Investigation of such infection is written without reference to important viral infections occurring in the neonate.

The index—one of the most important parts of any textbook—is usually neglected because the authors are exhausted or overworked (or both) by the time they reach the final stages. This is weak; as a typical example: the first two page entries on HIV lead to one or two lines on arthritis. Bold type should have been used to highlight the most important sections of the text.

This is an excellent book which deserves a place in the library of both the general paediatrician and specialist.

**T J DAVID**
Professor of child health and paediatrics

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This is very much an American book aimed at a home readership, and as such its applicability to UK clinical practice is limited. Neither the editors nor the contributors are likely to be familiar to British paediatricians. Although comprehensive, it does not hang together well as a coordinated textbook; it is more a series of essays on topics related to allergic disease in the widest sense. It declares itself to be aimed at primary care physicians and allergy specialists rather than paediatricians.

The book is not tightly edited, and as well as some sloppy typographical errors, there is also a great deal of repetition. For example, most of the chapter on diagnostic tests is repeated elsewhere, and the chapters on asthma in children and adults respectively have much in common. There is almost no cross referencing between chapters. With better editing the book could be considerably shorter.

Good points include liberal use of boxes containing key points, a generally readable style, and a healthy scepticism towards unproved and unscientific tests and treatments. There are helpful explanations of underlying theories of allergic disease, good descriptions of diagnostic tests including skin testing, and a quite comprehensive description of childhood asthma, which goes beyond what one might expect in a book confined to allergic disease.

There are thorough, although rather repetitive, chapters on allergic disorders of the upper respiratory tract, eyes, and skin.

Weak points include paucity of illustrations, poor quality black and white photographs and, in the chapters related to drug treatment, there tends to be too much pharmacology and not enough on trials of clinical efficacy. Surprisingly, in spite of the comprehensive account of asthma, there is very little on inhaler devices and nothing on comparison of new treatments.

However, the main factor limiting the usefulness of this book to British doctors is its North American parochialism. Thus the thorough account of environmental allergens refers only to pollen species found in the USA, and the insect bites that may cause anaphylaxis are inflicted by species unfamilial to us. The clinical management of asthma suggested differs quite considerably from standard UK practice. For example, chest radiography for every patient, different bronchodilator drugs, and much later use of inhaled steroids), and there is much greater emphasis on the use of immunotherapy and tolerance regimens for all types of allergic disease.

The best chapter is probably the last, 'Controversies in Allergy', which commendably debunks most of the alternative and unproved methods of diagnosis and treatment that seem to be finding increasing favour on both sides of the Atlantic.

In summary, this book will not find a ready market in the UK, and readers wanting accounts of the various allergic diseases and their treatments would probably be better served by this particular book as their first option. There are better, more internationally orientated texts available. That said, there are some excellent chapters and the subject is covered most comprehensively.

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Julian Shield and David Baum have brought together an impressive group of authors from various parts of the globe for this latest edition to the monograph series of Clinical Paediatrics on childhood diabetes.

It makes an enjoyable introduction for anyone interested in this complex childhood endocrine disease but it is not a practical management manual.

Only two of the chapters deal with any practical aspects of care, those on diabetic ketoacidosis and neonatal diabetes. The chapter on diabetic ketoacidosis discusses the physiology of the metabolic derangement in depth and includes the recent recommendations for its management from the British Society for Paediatric Endocrinology and Diabetes. The chapter on neonatal diabetes includes some useful pointers on management of this rare condition and an insight into its relationship to glucose intolerance in later life.

The majority of the book is thus a series of ‘state of the art’ dissertations from renowned authors with an emphasis on science and potential clinical applications rather than current practice.

There is an excellent series of three review chapters on epidemiology, genetics, and possible preventive interventions. The paediatric aspects of the Diabetes Control and Complications Trial are crystallised in a multiauthor chapter headed by William Tamborlane, which also poses some interesting questions on how to achieve optimal management. The issue of pathogenesis and screening for microvascular complications is taken up in a chapter by Henrik Mortensen with, as might be expected, the emphasis on microalbuminuria.

There are also excellent chapters on maturity onset diabetes of the young, associated immunological diseases, and a final chapter on growth hormone and insulin-like growth factor-I and their role in the difficulties of adolescent management and possible therapeutic role in the future from Zvi Laron.

This book should be essential and stimulating reading for all those paediatricians...
involved in the care of children with diabetes. Not that it will greatly improve our care of them in its own right but it will go a long way to explaining much of the frustration in management and put the all important science involved in the care of children with diabetes. I am looking forward to looking through the whole book and will certainly complement it. PENELOPE J DISON

Consultant paediatrician


There is no substitute for direct experience in learning practical techniques, but a logical and reasoned approach backed up by clear diagrams is an invaluable aid in turning the apprentice into a confident professional. Pediatric and Neonatal Tests and Procedures was written to instruct on common, non-specialist procedures and to form an account about specialised ones. It aims at a wide audience from medical students to paediatric specialists. There are 59 contributors to 48 chapters on subjects ranging from venepuncture to ECMO, and bandaging to developmental assessment. Four chapters appealed particularly to me: a clear account of ECG interpretation, a brief but useful description of cardiac ultrasonography, a well illustrated piece on diagnostic ultrasound screening, and a reasonably up to date section on high frequency oscillation ventilation. Most chapters are well referenced up to 1993, although references are not always cited in the text. There are clear differences between UK practice and some of the assertions in this book. The American Academy of Pediatrics recommends screening at surprisingly frequent intervals, eight visits in the first two years alone, and there is sometimes an aggressively interventionist attitude: ‘In the newborn child with difficulty breastfeeding, a frenulectomy can be performed in the office setting’. Presumably these examples reflect different financial arrangements in the American and British health care systems. There is an attitude towards children’s distress which would be unusual here: ‘A papoose board is used to securely restrain toddlers’ (for bronchoscopy). Some advice is potentially dangerous: the risk of infection from blood is no greater here than in the USA, yet the chapter on venepuncture does not mention gloves, illustrates un gloved hands, and talks without adverse comment about using mouth pipettes to collect capillary blood samples. The British paediatrician may also find the preference for butterfly needles rather than plastic cannulas for intravenous infusions quaintly old fashioned.

For practical techniques I know of no other book with such extensive coverage, but for investigations I would read Addy’s book. It seems to me that £65 is a lot to pay for the four chapters which I found useful.

MARY WHEATER
Consultant paediatrician


The prognosis for infants and children with inborn errors of metabolism is often directly related to the speed of diagnosis and institution of appropriate treatment. Attempts to shorten the period of diagnostic uncertainty are to be applauded. Most short textbooks on metabolic disorders emphasise clinical presentation and treatment, but this publication takes a different approach and concentrates on the diagnostic process. It is almost entirely biochemically based and little information about the molecular tests available are presented. This is a weakness especially in a field where molecular biology is beginning to make a major impact. I suspect this would be corrected in a second edition.

I like the way in which the chapters followed a standard format and all contributions are written by a researcher who worked in their field which gave the text an authoritative feel. It is unfortunate that the end result is not attractive. Indeed one’s initial impression is of a sea of tables, algorithms, and biochemical pathways with little intervening text. It is clear on closer inspection that an enormous amount of work must have gone into the coordination and publication of the book, but I fear, however, that it will miss its intended audience in my experience many general paediatricians when faced with a possible metabolic problem want clear advice on what tests to do and in addition want the result of the tests to be interpreted for them. The pages of reference data on various values and various biological fluids will not be of interest to them and I suspect most will read only the summaries at the end of each chapter, which indeed are excellent. Where access to a specialist opinion is limited the tables and algorithms are a helpful guide to diagnosis and for non-specialist laboratories the tables of data should help in interpreting test results. For workers within the field of inherited metabolic disease there is little new in the text, but I am sure all would agree how useful it is to have such a comprehensive set of data within a single short text.

In conclusion I am uncertain whether ‘do it yourself’ books such as this are the correct response to our expanding knowledge of inherited metabolic disease. Although it is true that the number of patients with inherited metabolic disease there is little new in the text, but I am sure all would agree how useful it is to have such a comprehensive set of data within a single short text.


Unlike its subject, this slim volume, in familiar red, is not deficient in energy nor micro-nutrients. Feeding the Disabled Child is a wide based review of a rapidly developing subject, the fundamental human activity of feeding, which in the severely disabled child can be perverted by disability from a pleasurable activity to an ordeal for the child and a chore for the parent. Normal development of feeding is considered; an interesting minute of neural development, coordination, learned skill, and opportunity. Then the causes of feeding difficulty in the disabled child are reviewed, the effects of inadequate intake of basically adequate food, ineffective feeding,
and the effects of reflux. The interlocking nature of problems is demonstrated by chapters on the consequences of malnutrition on neurodevelopment and the effects on the respiratory system of neurological deficit. Then the authors move to the assessment of a child with disability and feeding difficulties, a process focusing on strengths as well as weaknesses, clinically, nutritionally and by investigation. Two problems which greatly worry parents, drooling and constipation, receive a chapter each. Therapeutic options in the disabled child are considered over two chapters and then the authors round off with personal views on the ethics and implications of feeding programmes for the child with the disability.

If the reader wishes to know how many nerves are involved in swallowing or about feeding teams, about Sandler's 'head cocking' syndrome or the management of reflux, then this volume will help. The authors have made their 'object' improving the quality of life of the individual and the carer. This book will, with humanity and erudition, assist others in that aim.

A D KINDLEY
Consultant paediatrician


This is No 30 of Oxford Monographs on Medical Genetics. It discusses various demographic features of Arabs, selected disease entity among Arabs, genetic disorders in Arab countries and geographic regions, as well as cultural and religious attitudes to genetic issues. It provides an explanation of the important observation that there are many reports of genetic diseases in Arab populations. This could be explained by the large families and high level of consanguinity which increase the frequency of autosomal recessive conditions. In addition, with control of infectious diseases, genetic disorders will become a more prevalent problem.

Until now, the exact size of the problem of genetic diseases was not known because of lack of demographic and epidemiological population-based studies. However, there are several case reports and hospital based studies that indicate an increasing number of genetic diseases. Some of these were described first in Arab populations, for example, limb/pelvis, hypoplasia/aplasia syndrome described in a Palestinian family from Kuwait; subsequently reports came from other countries including Brazil and Italy.

No source of information has collected these reports in a systematic way, hence the importance this book which helps researchers in getting information on genetic diseases among Arab populations.

We would expect more description of genetic diseases in different Arab countries with advanced health care and improved laboratory technology. Therefore, revision and updating of this book are expected in the future because the subject is changing rapidly.

As a paediatric neurologist, I found the book very useful, in particular the chapter on new syndromes first reported among Arabs. More than 100 new single gene syndromes are described. Chapters on genetic disorders among the bedouin and cultural aspects are important for all doctors treating patients with genetic disorders. The chapters are based on the geography of Arab countries resulting in some repetition.

The editors have tried to cover the subject as broadly as possible but I think that coverage for certain diseases was not optimum, for example, endocrine and kidney disorders, congenital adrenal hyperplasia and congenital nephrosis, which are observed in certain Arab tribes. Similarly, the coverage of genetic and metabolic disorders in Saudi Arabia was also not complete, since Saudi Arabia is the origin of all of the migratory populations to other Arab countries. Many inborn errors of metabolism have been described from Saudi Arabia, for example in a special supplement of Developmental Medicine and Child Neurology in April 1991.

Despite these limitations the book provides a good source of information regarding genetic diseases in Arab countries. It is a good reference book for paediatricians, geneticists, neurologists, and other medical specialists in Arab countries as well as physicians from other countries who may face a medical problem in Arab patients and who may not be aware that these rare genetic diseases do occur in Arab populations frequently.

AHMED AL Jarallah
Paediatric neurologist


There are few, if any, books related to paediatric endocrinology that can be recommended unequivocally. Given the pace at which paediatric endocrinology has expanded, notably because of very rapid progress in molecular biology, textbooks involving this subject become quickly obsolescent, therefore posing a greater challenge to the authors. This Colour Atlas of Paediatric Endocrinology and Growth tackles the subject courageously and with a different approach, mainly focusing on clinical aspects supported by lavish photographs. Unexpectedly, the text is thorough covering in detail the presenting signs, symptoms, differential diagnosis, and investigations while being very practical and avoiding superficial information. It is written in a way which is easy to understand. Accompanying tables, clear growth charts, and appendices are simple to use and the most successful aspect of the book. They will certainly help unravel the subject and will act as a useful tool to memorise what ought to be retained by the reader. The illustrations act as the main lure for the buyers who have tried them effective visualisation of diseases and concepts described.

The paragraphs regarding treatment end pathophysiology are deliberately brief but clear. The book's intended audience is junior doctors as well as the general paediatrician who can easily extract practical information during daily practice. It is unfortunate, and yet understandable with the quality and the quantity of the illustrations that this small book is expensive, which has not been helped if the authors had provided a few key scientific references at the end of each chapter. To sum up, in spite of minor misgivings, this book is a publication of great distinction; practical, ambitious, but simple and without a hint of self importance.

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Warning: do not take this title literally. Not only is this obviously not a paediatrician, but it is unlikely to fit in your pocket. However take the title figuratively, and it aptly describes this manual of paediatrics: complete but succinct.

Another handbook. Is there any need for it? And if so, is it any good? The answer to the first question depends on what the book aims to do and whether it has been done before. Written by the paediatricians of the British Columbia Children's Hospital in Vancouver this book is based on the handbook given to their trainees and residents, and it aims to give practical advice to junior doctors, medical students, and nursing staff. The key word here is 'practical': a basic knowledge of the common entities encountered in paediatrics is assumed, allowing it to focus on diagnosis and specific management. To the best of my knowledge this handbook is unique in this aspect, and it is exactly what junior doctors require. Other handbooks either are restricted to major emergencies or a short version of a textbook, with non-specific or no advice on management.

However, for the same reasons I would question the usefulness of this book for medical students, at least in this country. The average medical student would be more likely to want to know what the haemolytic uraemic syndrome is, than how to manage it. That apart, the answer to the first question of whether there is any need for this book, is yes.

To answer the second question I put the book to the ultimate test and took it on call with me for a whole weekend. It was very useful. The chapters are clear and concise, with emergency management separated from background information and further management. Common conditions are given priority, and they are approached as they would be clinically, without having a final diagnosis. For example the cardiology chapter is divided into broad headings of cardiac failure, cyanosis and murmurs, rather than into individual structural cardiac conditions. There is a comprehensive pharmacology section which includes common drugs, their major side effects and interactions of each drug, as well as a good chapter on surgical problems. The answer to the second question of whether the book is good, is also yes.

One final area which sets this book apart from its competitors is that it attempts to tackle some of the ethical and practical problems of being and surviving as a junior doctor. The authors obviously considered
these areas as important as how to manage a severe case of croup—important enough to include them in a pocket book. This is an attitude which is more prevalent on the other side of the Atlantic than it is here, and it is refreshing to see the issue being dealt with head-on, rather than relying solely on hard learned experience.

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TRAINING PACKAGE

Equal Rights/Equal Access—Improving the Care of Minority Ethnic Children with Disability or Chronic Illness. A Training Package. Produced by the Department of Child Health, University of Wales College of Medicine, Cardiff, 1996. ISBN 1-899717-05-6. The package is available from Joanne Plummer, Department of Child Health, University of Wales College of Medicine, Heath Park, Cardiff CF4 4XN, price £105.75. Anyone wishing to buy the package must attend the ‘Equal Rights/Equal Access’ course; for further information contact Joanne Plummer at the above address or on phone 01222 743375.

As paediatricians consider making services more child and family friendly, we come up against political issues such as inequality of care, the needs of minorities, and different cultural views. Having trained and worked in South Africa, I am only too aware of the impact such issues have on health care provision. The authors’ viewpoint is that we live in a multicultural society and need to be aware of people’s cultural background, at the same time recognising that cultures are mixing and changing and that all individuals are different in what has shaped their lives. We have a responsibility to take account of these factors in our work.

This manual was produced under the auspices of the Access to Black and Minority Ethnic Children with Disability Project in South Glamorgan. It details a six hour training course designed to be run by two trainers who between them have experience in training, race awareness, and disability in childhood. The emphasis is on promoting the understanding of multicultural practice, the impact of racism on service delivery, and the development of sensitive and non-judgmental attitudes. It is designed for professionals from various disciplines working with children with disability.

The authors have creatively and sensitively put together a format on how to run the course as well as the subject matter, allowing some flexibility to take account of local circumstances. Such a package of planning details, handouts, and overheads would be most welcome in other teaching that I do. The shifts in the course from discussion to group activities and video clips with suitably ethnic music, kept my attention and interest.

I particularly liked some of the examples relating to my own experience including the doctor asking for the child’s ‘Christian’ name; the father who does not believe that a woman paediatrician can be the ‘real doctor’; exercises highlighting our prejudices or expectations when we think we are taking ethnic issues into account and the trap of cultural stereotyping. The handouts raise issues such as poverty, and particularly pleasing is that they offer solutions, such as a comprehensive strategy for health service personnel including recommendations for managers and commissioners.

Reviewing this manual from the material provided without having participated in the training, is a bit like deciding to buy a car based on the manufacturers’ sales pitch, without having had the chance to test drive the car. These manufacturers may highlight the background research and the de-lux features but the vital factor is to know whether the performance matches the specifications. I do however feel strongly tempted to buy.

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Archives of Disease in Childhood—http://www.archdischild.com

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Unilateral multicystic dysplastic kidney

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