PostScript

LETTERS

Heterogeneity of diabetes in south Asian patients in Bradford, West Yorkshire

The incidence of type 1 diabetes in children in India and Pakistan is remarkably low.[1] However, south Asian children resident in the UK have a rising incidence of type 1 diabetes,[2] approaching that of the indigenous population.[3] There is particular concern about the emergence of type 2 diabetes in children, and the consequent morbidity and mortality.[4 5] South Asian children are particularly at risk of type 2 diabetes.

A total of 160 children aged 0–16 attend the diabetic clinic in Bradford, of which 58 (38%) are of south Asian origin. In comparison, south Asian children comprise 27% and 32% of the primary and secondary school populations, respectively. Most have type 1 diabetes, but some have more unusual conditions not seen in the indigenous children.

Case 1

A girl aged 11 years presented with diabetes without ketonuria, a body mass index of 25, and negative autoantibodies. Type 2 diabetes was diagnosed; she was treated with an oral agent, but four years later transferred to insulin because of poor glycaemic control.

Cases 2–4: DIDMOAD syndrome

One boy had a ventricular septal defect repair in infancy; he developed diabetes at the age of 3 and was started on insulin therapy. At the age of 10 he developed optic atrophy, then diabetes insipidus, chronic renal impairment, and a neuropathic bladder.

Another boy—one of three brothers with insulin treated diabetes—presented at the age of 3. He later developed optic atrophy. His two brothers have no other features of DIDMOAD syndrome (diabetes insipidus, diabetes mellitus, optic atrophy, deafness).

The third case is a girl who developed insulin dependent diabetes at the age of 3 years; she developed bilateral lens opacities two years later, treated by cataract extractions and bilateral lens implantation. Recently her visual acuity has deteriorated due to bilateral optic atrophy. She has no other features of DIDMOAD at present.

Case 5

An 11 year old boy comes from a family with a probable mitochondrial depletion syndrome. Aged 7 months, he developed diabetes which was treated with insulin. He is the fourth child from a highly consanguineous pedigree to develop diabetes in infancy. These children have a tendency to develop acute liver impairment during intercurrent illnesses. A sibling died from acute liver failure aged 22 months. Their twin cousins died aged 3½ from acute liver failure. The surviving child has intermittent deranged liver function but has never had liver failure and remains insulin dependent with poor growth and some renal impairment.

Case 6

A male infant presented at birth with insulin requiring diabetes, remaining insulin dependent thereafter. He was growth retarded at birth but had no dysmorphic features and a syndromic diagnosis was excluded. Presently aged 18 months, his growth and development are normal.

Case 7

This child was born at 34 weeks to a woman with gestational diabetes. He had hypertrophic cardiomyopathy post-delivery but this subsequently resolved. He now has visual impairment due to a retinal cone dystrophy, which is mirrored in his father and two uncles. Latterly, he developed significant obesity and acanthosis nigricans, and was diagnosed with Alström syndrome. He became hyperglycaemic without autoantibodies and is being treated successfully with metformin.

Discussion

Our clinic population includes a number of children who have diabetes in association with other unusual conditions, for whom their diabetes was not easily classified and may have been a feature of their intrinsic syndrome. The high prevalence of syndromic diabetes in this population may emanate from the high consanguinity rate in the Bradford Asian population. We suggest it is vital for children of South Asian origin presenting with new onset diabetes to be carefully assessed, with the aid of a good family history, due to the high incidence of unusual forms of diabetes in this group of children.

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Advice for new mothers

My first-born arrived just after an SHO paediatrics job in Oxford and passing my DCH and DCCH exams. I was confident in handling small patients and in fielding parents' questions but I was not confident with my own new baby. My scientific clinical self started having arguments with my unconfident maternal self, and even the slightest suspicion of criticism of my maternal skills sent me into neurotic introspection.

I went to a community clinic for my baby's six week check and the CMO asked, ''Any worries?''.

''No, none really'', I replied. ''I know in my head that he's fine. He sleeps well and he's growing right along the 50 percentile. My only problem is in confidence. Whenever anyone suggests I should be doing things differently I plunge into a turmoil of guilt and worry.''

The doctor examined my baby and on palpating his abdomen said, ''He's full of gas. The position you are feeding him in can't be quite right. He must be sucking in air as he feeds.''

As doctors I am sure we often say too much and give too much advice, but it wasn't until I left the room that I thought what an unhelpful comment this colleague had made to me. It certainly undermined my confidence—again.

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NHS Direct

The establishment of training sites for consultation skills is a worthwhile development in the service.[1] What we also now need is a fundamental change in our attitude to the educational needs of nursing staff in our daily encounters on ward rounds and in the outpatient clinics. To replace the stereotype of the nurse being merely a passive recipient of the doctor's clinical wisdom, consultants (and other medical staff) should utilise each and every clinical episode to engage the attending nurse in a clinical problem solving exercise aimed at highlighting the diagnostic issues from the nurse's perspective. To take the process one step further, nurses should be encouraged to attend the ''problem solving exercise of the week'', which has become a tradition in postgraduate centres up and down the country. Each and every nurse is

a potential future employee of NHS Direct. By optimising their educational opportunities we are making a future investment in the service.

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Cause and effect?

I read Feltblower and colleagues' article[1] with great interest, having for some time wondered whether there might be an epidemiological association between childhood leukaemia and diabetes. The observations that the authors make regarding regional distribution of disease within a country certainly warrant further investigation.

Coincidentally, Mohn and colleagues[2] have recently shown clinically significant impairment of pancreatic beta cell function in children who have been treated for acute lymphoblastic leukaemia (ALL). Although the numbers will certainly be small, is it possible that some of the observed correlation between these two conditions could stem from ''iatrogenic'' diabetes?

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BOOK REVIEWS

Nutrition in pediatrics: basic science and clinical applications, 3rd edition

W Allan Walker, John B Watkins, and Christopher Duggan. Ontario: BC Decker Inc., 2003. $198.00, hardback, pp 1075. ISBN 1 55009 226 X.

In large Victorian households, feeding the children in the nursery was traditionally left to the youngest and most inexperienced housemaid. Recognition of the importance of nutrition in children in hospital and in the community has been equally slow. Medical undergraduates continue to receive little teaching on nutrition, even though over- and under-nutrition continue to be seemingly intractable problems; a substantial proportion, maybe a third, of patients in children's hospitals exhibit evidence of malnutrition. Moreover, we are now recognising that fetal and infant nutrition are major determinants of long term health.

One reason for the glacial speed in the development of clinical nutrition has been a view that clinicians already know enough about nutrition. That view is fostered in part by nutritional knowledge being recorded in the literature in disparate sites and in different ways. Much of it is in textbooks of biochemistry and metabolism; only rarely is it easily accessible. There is no obvious course book for the Royal College of Paediatrics and Children Health Diploma Course in Nutrition (www.rcpch.ac.uk).

Having the relevant basic science pulled together in one volume is immensely helpful to specialty developments; failure to do so is enormously damaging. One of the most interesting and exciting innovations in paediatric practice in the past 50 years in the United Kingdom was the concept of community paediatrics. It has foundered, partly because the basic science underpinning the discipline was never collected, written down, and published. Clinical texts are little more than recipe books without the underpinning basic science.

Nutrition in pediatrics addresses many of these problems, and the editors are to be congratulated on a tour de force. My assumption about its scope being limited to hospital based, clinical nutrition has not been confirmed. It is useful to the paediatrician working in the community or in developing countries as well as the tertiary, hospital based specialist. The psychosocial aspects of feeding are well covered and I was pleased to see sections devoted specifically to adolescents. There are no obvious gaps.

The RCPCH is seeking to redress the lack of teaching in nutrition in undergraduate medical courses with its course in paediatric nutrition. This volume makes an ideal course book, and should also be a routine acquisition for all postgraduate and ward libraries. Buy.

I W Booth

Managing children with psychiatric problems, 2nd edition

Edited by M Elena Garralda and Caroline Hyde. London; BMJ Books, 2003. £27.50, paperback, pp 225. ISBN 0 7279 1567 3

Emerging evidence, and political pressure have changed Child and Adolescent Mental Health Services a good deal over the past 10 years. It is mystifying enough to be part of these changes. I can only imagine what it might be like to be looking on. This second edition brings a useful book up to date. It is a sort of ambassador for Child and Adolescent Psychiatry and should go a long way towards explaining the discipline to associated agencies, professionals, and commissioners. It is likely to be better at this than at its prefacing claim to help professionals ''address the psychiatric aspects of their practice''.

The overlap between editions is about 50%. There is certainly enough change to make it worth your library getting the second edition while hanging on to the first. Private purchasers might want to keep a couple of chapters that have been lost in the wash. Steinberg's chapter on Consultative Work is one. Another chapter I would want to copy and keep is McAuley's Counselling Parents in Child Behaviour Therapy. It would be an even graver loss had it not been replaced by an equally excellent chapter by Scott on Parenting Programmes. It would almost be possible to run a parenting group on the strength of a careful reading of Scott's chapter, though I would not advise anyone to try.

It would most certainly not be safe to set out with a prescription pad having read only Prendergast's manic romp through Drug Treatments. This is where it is most true to say that any textbook is out of date by the time it is published. Not mentioning recent problems with paroxetine and venlafaxine, or the rapid increase in the use of controlled release stimulants in hyperactivity, can be forgiven, but this chapter has a number of idiosyncrasies which I would not expect in an overview directed at non-specialists.

A great deal has been done to create consistency, while preserving the styles of the individual authors. Each chapter starts with a boxed overview and most include sections on indications and contraindications, techniques, and evidence base. This book comes out of the process well armed with chapters on the important therapeutic modalities, most areas of interagency working, liaison, and legal aspects. There are significant omissions, however. Paediatricians based both in hospitals and in the community are increasingly exposed to deliberate self harm and drug and alcohol misuse. We are encouraged by the new children's National Service Framework to work together and we are increasingly doing so with, for example, autistic spectrum disorders and chronic fatigue syndrome. I would have thought that more on these areas would have been welcome The other gap, given current political and funding trends, is a chapter discussing changes in the working relationship with social services. As there is one chapter on child behaviour therapy and another on child behaviour therapy in groups, space could have been made for some of these important areas.

If the book deserves to sell well, it will not be on the strength of its illustrations, one of which greets us from the front cover. My son thought that some of them were drawn by an adult pretending to be a child which, if it were true, would not be an example of good psychiatric practice. They are not needed to break up the text, and the fact that pictures have migrated from one chapter to another between editions suggests that their positions are almost arbitrary. Somehow they contrive to be at once both condescending and perplexing.

Having got over this minor irritation, the more I looked into this book, the more I liked it. It is easy to read, is well referenced, and punches above its weight.

A West

Pediatric urology

Edited by John P Gearhart. New Jersey: Humana Press, 2003, $125 (hardback), pp 319. ISBN 1 58829 110 3

Paediatric urology has come of age over the past few years, with it now being formally recognised as a distinct subspecialty of both urology and paediatric surgery within the European Union. In parallel with this development there has been a rapid expansion in the number of paediatric urologists practising in the United Kingdom. With this increasing specialisation there is a risk of isolation, and developments in the field of urology may go unrecognised in the broader areas of paediatrics, paediatric surgery, and urology. Therefore there is a need for publications to highlight recent developments. The non-specialist rarely reads major reference textbooks (of which there were two published in

2002) outside their own field, and smaller texts providing a general overview for associated specialities are required. I am uncertain what audience this book has been written for but I believe it goes some way to meet the requirement of an update for the generalist.

For a multiple author book it is well edited and there is little repetition. It is relatively inexpensive and generally a very enjoyable read. It is well illustrated and well referenced. It should not be regarded as a reference text but rather as one that can easily be read from start to finish over a short period, and I believe it will update the reader with most of what is happening in paediatric urology. I do not think the specialist paediatric urologist will find much new here and the book will be of far more interest for the general paediatrician, paediatric surgeon, urologist, and trainees in those areas.

The book covers most of the major areas (but not all) of urinary tract pathology, including: prenatal diagnosis, vesicoureteral reflux, duplex systems, voiding dysfunction, neuropathic bladder, bladder exstrophy, stones, oncology, hypospadias, undescended testis, and urological emergencies.

One chapter deserves special mention: ''Developmental perspectives of children with genitourinary anomalies''. Conditions such as incontinence and genital abnormalities can be particularly worrying for children and adolescents, and the point is clearly made that ''identity becomes entwined with anomaly'' and as a result adolescents may become developmentally blocked. The importance of identifying abnormal development to prevent problems and to treat psychological conditions that have already developed, and the need for every paediatric urology service to have formal psychological and psychiatric support is clearly made. It is encouraging to see such a holistic approach to patient care.

The chapters on vesicoureteral reflux provide an excellent overview, and it is refreshing to find an American text adhering to evidence based principles on the role of surgery. Although the topic of urinary tract infection is alluded to in a number of chapters, it is disappointing that it was not covered in a separate chapter dealing with presentation, investigation, and treatment. I also think it would have been useful to include a general chapter on urinary tract obstruction rather than have it dealt with under prenatal diagnosis.

The chapters on hyospadias and bladder exstrophy are excellent, but for the general reader there is perhaps a little too much surgical detail. It is as if the authors were undecided about which audience they were aiming at. I also think it would have been useful if the chapter on exstrophy addressed long term management rather than concentrating on the initial closure of the bladder in infancy. This is of particular importance in the United Kingdom setting where the management of exstrophy is now limited to two national centres, and there is a need for referring paediatricians to know what will happen to their patients.

Despite these shortcomings, overall I think this book is a valuable contribution, and most paediatric and urology departments would benefit from having it on the shelves of their libraries.

P S J Malone

Tuberous sclerosis complex: from basic science to clinical phenotypes

Edited by Paolo Curatolo. Cambridge: Cambridge University Press, 2003. £55.00 (hardback), pp 301. ISBN 1 898 68339 5

Tuberous sclerosis has always had the capacity to confuse clinicians. Friedrich von Recklinghausen confused it with neurofibromatosis, when on 25 March 1862 he presented a case to the Obstetrical Society of Berlin. The case he described was of a young infant who had died soon after childbirth and who was discovered on postmortem examination to have multiple cardiac myomata in the ventricular walls and a ''great number of scleroses'' in the brain. It was a French physician, Desire Magloire Bourneville, a pupil of Charcot, who has won the plaudits for correctly appreciating that tuberous sclerosis was a separate disease. In 1879 he described the case of a 15 year old girl who died at the Salpetriere in Paris. She had suffered from epilepsy and severe learning difficulties for most of her life and was afflicted with a disfiguring vesicular-papular eruption on her face. An autopsy revealed that she had many hard sclerotic lesions in the cerebral cortex and white nodular lesions protruding into the lateral ventricles. Bourneville coined the term ''tuberous sclerosis of the cerebral convolutions'' for this unique pathology. The term tuberous sclerosis has stuck, although the French still patriotically talk of Bourneville's disease.

Part of the reason that tuberous sclerosis has confused us, and still does, is that there has been a relative lack of population based research of the disease. In 1908 Vogt proposed a triad of clinical features that he felt characterised the disease, namely seizures, mental handicap, and ''adenoma sebaceum''. No doubt the description accurately reflected his anecdotal clinical experience. It is still quoted in medical textbooks to this day. However, it grossly oversimplifies the disease. Epidemiological studies of the disease have revealed that more than half of those with tuberous sclerosis complex (TSC) will have a normal intellect and that complications related to the disease may occur in any organ of the body. The plea for more epidemiological research is not just an academic point. Patients may, and probably do, suffer if clinical decisions about their medical care are based on the results of hospital case series that are distorted by selection biases.

Professor Curatolo's book goes a long way to demystifying this complex disease. Occasionally the authors fall into the trap of uncritically quoting many of the aforementioned misleading case series. However, this is more than compensated for by some excellent chapters within the book on the neurological and neuropsychiatric complications of the disease. The piece on the cognitive and intellectual impairments of the disease is the most balanced and comprehensive summary of the subject to date. Bolton clearly points out that the majority of TSC individuals fall within the normal range of IQ, but also suggests that many of those with high intelligence may also have specific neuropsychological impairments. He also explores the question of what causes the intellectual impairments in TSC in some detail and concludes that it is likely to be related to not just the extent of the structural brain abnormality but also the age of onset and type of seizure disorder as well as the nature of the underlying genetic mutation.

Curatolo and Seri's chapter on seizures is also strong. Particularly interesting is the discussion of how epileptogenesis in TSC may be related to an impairment of GABA mediated neuro-inhibition. It is startling how well vigabatrin, a specific inhibitor of gamma amino butyric acid aminotransferase, is supposed to work in the treatment of infantile spasms in tuberous sclerosis. In Hancock and Osborne's Cochrane Review of the treatment of infantile spasms, they found that there was a 95% response rate in those individuals with TSC who were treated with vigabatrin. The acid test of this therapy, of course, will be to see whether the apparent remarkable success rate in stopping spasms is matched by an improved intellectual outcome in these patients later in life.

The genetics of TSC are fascinating, and Kwiatkowski and colleagues carefully and clearly take the reader through the complexities of the subject. The disease is caused by alterations in two genes, TSC1 and TSC2, on chromosomes 9 and 16 respectively. Both are tumour suppressor genes and mutations in either can be responsible for all the complications seen in the disease. The gene products of the two genes, hamartin and tuberin, appear to act together in the regulation of cell growth by inhibiting a substance known as mTOR (target of rapamycin). In those individuals with the disease this inhibition is thought to be reduced and therefore there is the formation of the hamartomas that are characteristic of the disease. It is exciting that rapamycin, a clinically available compound, also selectively inhibits mTOR and therefore there is now the possibility that rapamycin may provide some benefit in reducing the clinical manifestations of TSC. Fittingly, Professor Curatolo's book finishes with the intriguing suggestion that at last we may be on the threshold of being able to do something to ameliorate this complex and potentially devastating disease.

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CORRECTION

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Toni S, Reali M F, Fasulo A, *et al*. The use of insulin pump improves the metabolic control in children and adolescents with type 1 diabetes *Arch Dis Child* 2004;**89**:796–7. The initial of Dr Festini was incorrectly published as an F. The correct initial is P.