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The editors will decide, as before, whether to also publish it in a future paper issue.

The reasons for the loss of calibration are unclear. A possible, perhaps over optimistic explanation is that UK units in the latter study were all “over performing” given that individual units demonstrated an SMR of between 0.83 and 0.89. However it is unlikely that such a quantum leap in the quality of paediatric intensive care delivery has occurred over the 4 years between 1994–98, given that no major treatment breakthroughs or radical service reorganisation has occurred in this time.

More recent data from our PICU highlight the trend towards poorer calibration, where the PIM-derived SMR from 910 patients seen during the 2000 calendar year is 0.54 (95% CI 0.39–0.69). The authors acknowledge the shortcomings and state that a revised version of PIM will soon be available. However, recalibration is only worthwhile if a very broad sample of UK units participates. The UK PICOS study (paediatric intensive care outcome study) will attempt to address this, by collecting data used in the calibration of several scoring systems across the whole of the UK over a one year period commencing March 2001. From this study it is hoped that an optimal indicator of PIM performance will be derived.

Dr Tibby and Dr Murdoch point out that the reasons for the loss of calibration are unclear. A possible, perhaps over optimistic explanation is that UK units in the latter study were all “over performing” given that individual units demonstrated an SMR of between 0.83 and 0.89. However it is unlikely that such a quantum leap in the quality of paediatric intensive care delivery has occurred over the 4 years between 1994–98, given that no major treatment breakthroughs or radical service reorganisation has occurred in this time.

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Dr Tibby and Dr Murdoch note that, in our study of paediatric intensive care units (PICUs) in the UK, PIM discriminated well between children who died and children who survived, with an area under the ROC curve of 0.84. However, they are concerned that PIM had “poor calibration” because the standardised mortality rate (SMR) in the UK units was 0.87 (95% CI 0.81–0.94)—that is, the actual number of deaths was only 87% of the number predicted by PIM. In fact, this figure is almost identical to the PIM SMR for all PICUs in Australia in 1997–99, where the SMR was also 0.87 (95% CI 0.81–0.92). It is very encouraging that PIM gives such similar results in Australia and the leading PICUs in the UK, as it suggests that standards are comparable between the two groups of units and that PIM performs similarly in Australian and UK children.

It is normal for SMRs to fall with time as intensive care improves, and for mortality prediction models to need recalibration. This has happened with PRISM,3 MPM1 and APACHE,7 as well as PIM. Despite Dr Tibby and Dr Murdoch’s reservations, the fact that the SMR has fallen in parallel in both Australia and the UK suggests that standards of care have improved in PICUs in those countries in recent years.

Dr Tibby and Dr Murdoch point out that the Hosmer-Lemeshow test gives a low p value for
PIM's performance in the UK data. This test divides the sample into 10 groups, ranging from very low to very high risk of death, and compares the actual number of survivors and non-survivors in each group with the number predicted by PIM. Because PIM predicts too many deaths in the leading units in the UK, it follows that the number of actual deaths differs from the number predicted—so the Hosmer-Lemeshow p value is low. However, table 2 in our paper shows that the ratio of observed to expected deaths was similar across the 10 groups, so that the recalibrated model is likely to fit well. The fact that the Hosmer-Lemeshow test gives a low p value does not necessarily mean that a model (such as PIM) is invalid—it often means only that the standard of care in the test PICUs differs from that in the units in which the model was derived.

The PICUs that contributed the data from which the PIM score was derived were all leading units that deliver a high standard of care, so the score reflects best practice in 1994–96 when the data were collected. We are recalibrating PIM using data from units in the UK and Australia, and the new model will be available this year. Unfortunately, the quality of paediatric intensive care is not uniform in the UK, and there is evidence that some units do not perform at an optimal standard. Surely it would be preferable for the UK to use an international standard based on best practice (such as PIM), rather than the average of the whole of the UK (PICS). The UK should aim for best practice rather than being content with average practice.

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Long term results of lung resection in cystic fibrosis patients with localised lung disease

We have previously reported favourable short term outcomes following lobectomy in six children with cystic fibrosis and severe localised bronchiectasis (range 6 months to 6 years post-operation). Prior to surgery all had significant respiratory symptoms despite aggressive conventional treatment, including frequent courses of intravenous antibiotics. Computerised tomography and ventilation scans showed severe localised disease with little or no evidence for bronchiectasis elsewhere. Lung function was maintained or improved in all but one case from six months post-surgery, and all had improved symptoms.

All children have now been reassessed at least four years postoperatively (table 1). Three remain much improved, with few symptoms and minimal need for intravenous antibiotic therapy. One child remains better than prior to surgery, but has recently required increased intervention to maintain wellbeing (case 5). Two children require antibiotics as frequently as prior to surgery with chronic signs (cases 3 and 6). There were no preoperative risk factors predictive of a less favourable outcome in these patients. Lung function has been maintained in all except one case (case 6).

Follow up chest X-rays were assessed by a consultant paediatric radiologist, using the Chrispin Norman Scoring system. New radiological changes have tended to occur in the zones previously occupied by the resected lobe (table 2). One of the patients has had a bronchoscopy following right upper lobectomy (case 3). Upwards displacement of the right middle lobe bronchus appeared to be causing airway narrowing. Such distortion of the lung anatomy may predispose to bronchiectasis in lobes that have shifted so as to occupy the spaces previously occupied by the resected lobe.

Our long term results suggest that surgical resection is a worthwhile option in selected children with severe localised symptomatic bronchiectasis. Detailed preoperative assessment is essential to exclude patients with more extensive lung damage. While there is a good long term improvement of symptoms and preservation of lung function in the majority of patients, there is a tendency for new radiological abnormalities to occur in the zones previously occupied by resected lobes.

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### References


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### Anti-neutrophil cytoplasmic antibody autoantibody positive glomerulonephritis in monozygotic twins

Scanty information is available concerning anti-neutrophil cytoplasmic autoantibodies (ANCA) associated disease in children, and very few cases of familial vasculitis have been reported in the literature.

We have observed two monozygotic twins developing ANCA necrotising glomerulonephritis (GN).

A 7 year old boy was hospitalised for normocomplementemic acute nephritis. Percutaneous renal biopsy revealed idiopathic crescentic GN with negative immunofluorescence. Dialysis was started because of worsening in renal insufficiency. Despite several courses of daily plasma exchanges combined with intravenous methylprednisolone and cyclophosphamide, there was no improvement; one year later, the boy received a cadaveric renal transplant.

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### Table 1

<table>
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<tr>
<th>Case</th>
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<th>FVC (% of predicted)</th>
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### Table 2

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Persistent proteinuria appeared four years after transplantation, when a renal biopsy revealed focal necrotising GN.

At the age of 10 years, the identical male twin was found to have microscopic haematuria and proteinuria of >1 g/24 h with normal renal function. Renal biopsy showed focal necrotising GN with 20% cellular and segmental crescents. Perinuclear ANCs were observed at a dilution of 1/160. The stored samples of the first twin were tested and pANCs were detected by indirect immunofluorescence.

This second twin was given intravenous methylprednisolone and cyclophosphamide. The clinical picture was characterised by acute episodes resolved with repeated courses of methylprednisolone pulses.

ANCA positivity in the second twin (also found retrospectively in the first twin's veins) allowed us to classify the disease as a renal limited vasculitis expressed by necrotising and crescentic GN.

The HLA antigens profiles of the two boys are A3,11, B7,27,35, DR12, DQ1.

Acute nephritis or urinary abnormalities were the initial onset symptoms in our patients. They occur in about 40% of children with ANCA-associated GN. This emphasises the need for a precise diagnosis and aggressive treatment in such patients. ANCA should be sought in the presence of acute nephritis or persistent urinary abnormalities of unclear aetiology, and not only in children with frank vasculitis or rapidly progressive GN.

We believe this to be the first report of the recurrence of pauci-immune crescentic GN in a transplanted kidney in a child. Anti-rejection treatment with steroids and cyclosporine A seems to be a useful means of controlling disease flare ups.

Furthermore, as far as we are aware, this is the first report of pANCA GN in HLA-identical twins. The pathogenesis of ANCA-GN is unknown but likely implicates genetic and/or environmental influences.

The onset of disease at different times in two identical twins seems to suggest a genetically determined susceptibility rather than environmental triggers. Review of the literature revealed few reports of familial vasculitis, with some evidence suggesting a genetic predisposition of the HLA class I antigen present in our twins (A11, B35), and antigen B35 alone have also been found in two identical twins seems to suggest a genetic predisposition.

In conclusion, a pANCA test should always be performed in children with acute nephritis of unclear aetiology; a diagnosis of ANCA-GN should not preclude renal transplantation.

Hepatitis B prevalence among Somali households in Liverpool

A cross sectional descriptive study was undertaken in the Liverpool Somali population in order to determine the prevalence of hepatitis B markers. Sessions were held at two health centres providing care for Somali households. A total of 439 subjects were screened, of whom 194 (43.3%) were children aged less than 15 years. It was found that 5.7 per cent of the study population were carriers of HBsAg.


References


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Lipid and glucose metabolism in HIV-1-infected children treated with protease inhibitors


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References


HIV-1-infected children treated with protease inhibitors

The use of protease inhibitors (PIs) in patients with HIV-1/AIDS has been associated with peripheral lipodystrophy, hyperlipidaemia and insulin resistance. Most studies have been done in adults. The aim of this study was to evaluate the influence of highly active antiretroviral therapy (HAART) on serum levels of fasting triglyceride, total cholesterol, high density lipoprotein cholesterol (HDL), low density lipoprotein cholesterol (LDL), free fatty acids (FFAs) and glucose in twenty HIV-1-infected children treated during a minimum period of 18 months with an indinavir (HIV) or nelfinavir (NFV) containing regimen of HAART.

The lipid values were evaluated at two time points: within the first month of HAART (“baseline values”) and after 18 months or more (range 18–24 months). Serum levels of fasting glucose was only evaluated at follow up.

In summary, we found an increase in serum levels of total cholesterol and LDL after PI use in HIV-1-infected children, as was previously observed in adults. However, in contrast with adults, a marked increase in HDL and normal glucose was observed. The total cholesterol/HDL ratio, fasting triglyceride and FFA levels remained stable over this period.

To date, it has not been revealed whether these metabolic changes are the result of HAART or if HIV-1 infection itself is responsible. Hypertriglyceridemia and low levels of total cholesterol, HDL and LDL, have been detected in HIV-1-infected patients without prior antiretroviral therapy, especially in the late phase of the disease. Thus, the significant rise of total cholesterol, HDL and LDL in HIV-1-infected children may not only be attributed to the effects of HAART, but may be also partially be the result of a normalisation of pre-existing lipid abnormalities.

It is difficult to discriminate the metabolic effects of PIs from those of other antiretroviral drugs in this study. Most children received a combination of a PI, zidovudine, and lamivudine, which are also reported to cause lipodystrophy and lipid abnormalities. Eleven children were pretreated with zidovudine before the start of HAART. These children had significantly lower levels of total cholesterol and LDL at baseline than naive children, suggesting that zidovudine itself may have an effect on the lipid metabolism.

After these results were obtained we have to conclude that HAART also effects the lipid and glucose metabolism in children.

We would like to thank L.Zwag and M A C van Fessum for performing the laboratory analyses, W C J Hop for statistical advice, G J Bruining, F Pistor, HJ Schepfer, and T F W Wolfs for their co-operation.

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Hepatitis B prevalence among Somali households in Liverpool

A cross sectional descriptive study was undertaken in the Liverpool Somali population in order to determine the prevalence of hepatitis B markers. Sessions were held at two health centres providing care for Somali households. A total of 439 subjects were screened, of whom 194 (43.3%) were children aged less than 15 years. It was found that 5.7 per cent of the study population were carriers of HBsAg.

Seven of 80 (8.7%) children born in the UK and aged 5 years or less had evidence of exposure to hepatitis B. Of their mothers only one was a carrier, one had anti-HBc antibody, and five were non-immune. These data suggests that horizontal HBV transmission continues at an early age among Somali immigrants.

The UK is one of the few western European countries which has chosen not to comply with the WHO recommendations for universal hepatitis B vaccination. This position has recently been defended, although no reference was made for the need to immunise high risk ethnic groups, outside an antenatal screening programme. Evidence of previous hepatitis B infection in children is not uncommon among the Somali population in Liverpool. This has implications for screening of children who may benefit from immunisation. If screening of high risk groups and vaccination of susceptible...
TREATING CHILDHOOD HYPERHIDROSIS WITH BOTULINUM TOXIN TYPE A

Recently there have been a number of published studies on the use of botulinum toxin type A for hyperhidrosis.1 These studies focus on its use in adults and we would like to highlight that it can also be useful in treating childhood hyperhidrosis. As in adults, hyperhidrosis can have considerable impact on quality of life in children. This is illustrated by a 13 year old healthy girl referred for treatment of axillary hyperhidrosis. Excessive palmar sweating caused difficulty with school work (difficult holding a pen, with the ink smudging the paper because of sweating) and social embarrassment. Botulinum toxin type A (Dysport; 30 mouse units) was administered intradermally using a 27G needle to the fingertips and the area over the hypothalamic and thenar eminences of both hands. EMLA cream was used for topical anaesthesia. She reported sufficient reduction in palmar sweating within one week to improve her school work. She noticed grip strength reduction that lasted three weeks but did not affect hand function significantly. The beneficial effect of botulinum toxin lasted four months after which she requested further treatment. Repeat injections were given to the fingertips only. No adverse effect on grip strength was reported despite some functional benefit from reduced sweating. To date she has had four courses of treatment over a period of two years with good effect. Although treatments such as aluminium chloride and iontophoresis can be effective and may be preferred in children, we suggest that botulinum toxin should be considered for children with refractory hyperhidrosis who do not want surgery.1

If you would like to buy a gentle gift for a literary medical friend, you could do worse than this lighthearted escapist, all royalties from which are paid to the Royal Medical Benevolent Fund.

The editor, Dr Saleem Goolamali, has persuaded various of the great, good (and Lord Archer) to provide an anecdote or two, often with a medical reference. Sir Edward Heath professes his love of American football, Rabbi Lionel Blue provides a recipe which will please encopretologists everywhere and a recently retired Regius Professor reminisces on how fate might have led him to be a ballet dancer instead. Would Covent Garden's gain have balanced Oxford's loss? Lord Archer relates a childhood memory which might have helped him.

You won't split your sides but there are plenty of chuckles—and the RMBF is well worth supporting in its aim to provide help for doctors or their families who have fallen on hard times. You can order your copy free from MedLem, 25 Highfield Road, Northwood, HA6 1EU, UK.

H Marcovitch

CARING FOR MUSLIM PATIENTS


Islam is the religion of one-fifth of humanity and, with an estimated population of 1.6 million Muslims forming Britain's largest religious minority group. There is, therefore, a need for a book that gives advice and guidance to non-Muslim healthcare professionals when dealing with Muslim patients and their families. The authors believe is exacerbated by our own reluctance as health advisors to discuss abortion with Muslim couples, simply because we assume that they would always refuse it.

More modern issues like adoption, fostering, and organ transplants are discussed. Do you know that the Muslim Law Council, a UK based organisation, strongly supports Muslims donating organs? Not surprisingly, the majority of Muslims reject the idea of their dead undergoing postmortem examination, but the authors issue a call to Muslim jurists to study this issue and give believers clear guidelines.

This is an interesting book which I very much enjoyed reading. If its aim was to provide information, it has succeeded. I feel, however, that it should not be used as a religious reference by health professionals to make decisions or give advice, as it is contradicting the teachings of Islam. As a Muslim myself, I may not agree with some statements in the book nor wish my patients to believe them to be a religious command. Telling asthmatics not to use their inhalers during fasting (in case part of the inhaled medication enters the oesophagus!) is an example. Some statements made by the authors are based on cultural practices rather than religious facts. These practices may have developed over centuries, specific to a particular Islamic society and as an Arab, some customs described in the book are as unfamiliar to me as they would be to a non-Muslim. This book tells us a lot about customs and practices in Muslims of Asian roots and in that respect, would be an invaluable reference.

R Tawfik

THE CHILD WITH HEADACHE: DIAGNOSIS AND TREATMENT


Over the past 10–15 years there has been a large volume of research into headache, in general, and childhood headache in particular. Research interest and publications have covered vast areas of previously neglected aspects of childhood headache including epidemiology, pathogenesis, clinical features, classification, impact on child's life and education, management, psychological adjustment, and medical treatment. Two major developments have helped to drive research into childhood headache and migraine. Firstly, the publication of the classification and diagnostic criteria for headache disorders, cranial neuralgias, and facial pain by the International Headache Society in 1988 triggered better understanding, research interest and debate into headache. Secondly, the introduction of a new generation of specific anti-migraine medications in the early 1990s has started a huge wave of research into migraine. Sumatriptan was the first of many 5HT1 agonists to show effective relief of migraine headache in adults associated with...
There is no doubt that this book will prove to be an important and useful resource for paediatricians treating children with headache. Other publications dealing with the practical issues and the organisation of headache services for children are also needed.

I Abu-Arafeh

Core Paediatrics and Child Health


Another textbook of paediatrics finds its way to market, to take its place alongside those already in print. In their introduction, Haddad et al write that they have written this for undergraduates and junior doctors undertaking their first paediatric post. The underlying concepts arise from prior collaborative work undertaken by departments of Child Health in Scottish Universities in response to the GMC guidelines contained in “Tomorrow’s Doctor”. This work, reported in Medical Education, provides a structure that gives uniformity of approach for each organ system and indeed the textbook is clearly and consistently laid out.

As with many other authors of textbooks, the authors start with an assumption that the layout of texts will influence learning. It is difficult to find any supportive evidence in educational literature and any research suggests that it is assessment rather than course material that drives acquisition of knowledge and reasoning skills. Nevertheless it seems reasonable to assume that those learning paediatrics should be able to choose from a selection of texts written and laid out differently. As such, it could be commended to students if they are considering the purchase of a textbook to support their learning, and I feel sure it will take its place in the “top five” of UK paediatric textbooks.

Although system based, the authors claim that their book is “a problem oriented approach”. This does not match other books that start with clinical signs and symptoms; such a true problem oriented approach can be seen in Field et al’s book. This difference highlights the difficulty of writing a text for both students and practising doctors. Anecdotally, students, who seem to prefer topic based teaching while SHOs, may find a true problem based approach more suited to their needs. They do, nevertheless, include “Key problems”, and have useful sections that review underpinning science, such as “Essential background”. For the enthusiastic student who wishes to pursue any topic further, they have included “Beyond core” material and sections entitled “Highlights and hypotheses”.

At over 300 pages, it probably contains more than is needed at undergraduate level but could be seen as core and a suitable text for SHOs. SHOs might find its system based layout less helpful in their learning how to practice paediatrics, but it would be a useful starting point for revision for postgraduate exams.

Teachers need to look at evaluation from a different perspective. How should they evaluate material for students undertaking their course? Fundamentally, any text should support and NOT divert student effort from the learning objectives and the other resources that help the teachers by providing them an agreed core curriculum. As a collaboration between Scottish departments of paediatrics, this book should present a problem north of the border, but others will need to analyse it mindful of their own course objectives. As a tutor at Imperial College School of Medicine this would raise problems. Our main course objectives are that:

1. Students should acquire understanding of families, their structure and how children are supported within this.
2. Students should acquire the skills of history taking and examining children along with the necessary communication skills.
3. Students should acquire a basic knowledge of common and important childhood diseases.

This textbook clearly supports the last objective but neither 1 nor 2, although it is only fair to say that this criticism could be levelled against other similar textbooks. This could be seen as an argument for radical redesign of all undergraduate texts to match more fundamentally the course aims rather than a “topic based” core curriculum, but such discussion is outside the remit of a book review such as this.

My one major criticism is that it divides up history taking and examination according to body systems. Development of these clinical skills must be the cornerstone of undergraduate education, and dissection of history taking and examination makes it a difficult text from which to teach these essential practical skills. Having said that, this book offers a clearly structured text for early professional education, and it will be interesting to see how it is received by the consumer, the medical student or doctor undertaking general professional training.

M D C Donaldson

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