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

**LETTERS**

**Calibration of the paediatric index of mortality in UK paediatric intensive care units**

Pearson et al should be congratulated on successfully collecting the data required for calculating the PIM Score on 7253 children admitted to 5 UK paediatric intensive care units (PICUs). It is reassuring to note that the authors did not find any systematic differences between these five units in terms of their standardised mortality ratios. Leaving aside the controversies involved in cross country comparisons, it is further pleasing that they appear to conclude that mortality following admission for paediatric intensive care in 1998–99 is less than it was in 1994–95. The current results imply that 78 more children have survived following treatment in these 5 PICUs than were predicted by the 1994–99 PIM derivation model. Before this can be considered a major clinical advance, it is important to consider the health status of the additional survivors. Very different conclusions might be drawn if the additional children who survived have a very poor health status than if they have a very good health status.

The United Kingdom Paediatric Intensive Care Outcome Study (UK PICOS) was set up in response to the “Paediatric Intensive Care: A framework for the future” document and a joint United Kingdom Medical Research Council and Department of Health working paper. Both these publications recognised that, as mortality following paediatric intensive care is less than 10%, morbidity or health status may be a more important outcome of paediatric intensive care than mortality. UK PICOS is currently collecting health status measurements of children who survive following admission for paediatric intensive care in a representative sample of 21 UK PICUs. By seeking to differentiate between the survivors of paediatric intensive care, UK PICOS may lead to a risk adjustment method for health status in addition to mortality. Furthermore, UK PICOS has the potential to provide the methodology to enable cost effectiveness studies to be set up in paediatric intensive care. In the longer term this will allow organisational structures, service management, and new interventions in paediatric intensive care to be evaluated in a more rigorous manner than at present. Further details of UK PICOS are available at www.shef.ac.uk/~schart/ukpicos.

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**References**


**Authors’ reply**

Dr Tibby and Dr Murdoch note that, in our study of paediatric intensive care units (PICUs) in the UK, PIM discriminates 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, MPM and APACHE, as well as PIM. Despite Dr Tibby and Dr Murdoch’s reservations, the fact that the SMR has fallen by a similar amount 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

**PostScript**
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 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 (PICOS). 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.

Table 1 Lung function data: simple spirometry after bronchodilator inhalation

<table>
<thead>
<tr>
<th>Case</th>
<th>FEV₁ (%) of predicted</th>
<th>PVC (%) of predicted</th>
<th>Number of years followed up</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preop</td>
<td>Postop</td>
<td>Long term follow up</td>
</tr>
<tr>
<td>1</td>
<td>---</td>
<td>94 (6 y)</td>
<td>103</td>
</tr>
<tr>
<td>2</td>
<td>60</td>
<td>75</td>
<td>60</td>
</tr>
<tr>
<td>3</td>
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</tr>
<tr>
<td>6</td>
<td>83</td>
<td>83</td>
<td>60</td>
</tr>
</tbody>
</table>

Table 2 Chest x ray score

<table>
<thead>
<tr>
<th>Case</th>
<th>Operation</th>
<th>Local Chrispin-Norman scores</th>
<th>Preop</th>
<th>Postop (6 mth)</th>
<th>Long term follow up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>LLL</td>
<td>3</td>
<td>2</td>
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<tr>
<td>2</td>
<td>RUL</td>
<td>5</td>
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<tr>
<td>4</td>
<td>RUL</td>
<td>6</td>
<td>3</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>RUL and RML</td>
<td>5</td>
<td>2</td>
<td>4</td>
<td></td>
</tr>
</tbody>
</table>

Data are the Chrispin-Norman scores in the lung quadrant within which the patients had developed local bronchiectasis and for which they underwent lobectomy (maximum score 8).

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 intravenous to maintain wellbeing (case 5). Two children require anti-biotics as frequently as prior to surgery with chronic signs (cases 3 and 6). There were no predictive risk factors predictive of a less favourable outcome in these patients. Lung function has been maintained in all except one (case 6).

Follow up chest x rays were assessed by a consultant paediatric radiologist, using the Chrispin Norman Scoring system.6 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 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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Anti-neutrophil cytoplasmic 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.1,2

We have observed two monozygotic twins developing ANCA necrotising glomerulonephritis (GN). A 7 year old boy was hospitalised for normocomplementemic acute nephritis. Pericutaneous renal biopsy revealed idiopathic crescentic GN with negative immunofluorescence. Dialysis was started because of a 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.

www.archdischild.com
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. Perinephric ANCAcs were observed at a dilution of 1/160. The stored serum of the first twin was tested and pANCAcs were detected by indirect immunofluorescence. This second twin was given intravenous methylprednisolone and cyclophosphamide. The clinical picture was characterised by acute episodes resolving with repeated courses of methylprednisolone pulses.

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

The HLA antigen profiles of the two boys are A3,11; B27,35; 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.1 This emphasises the need for a precise diagnosis and aggressive treatment in such patients: CAEs 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 pauciimmune crescentic GN in a transplanted kidney in a child. Anti-rejection treatments 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.2,3

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 with some evidence suggesting a genetic pre-

Crescentic GN.

patients. They occur in about 40% of children with ANCA associated GN.

Clicking ribs—a clinical sign of rib fractures

It is well recognised in non-accidental injury that some children who have rib fractures on x-ray have no external evidence of these.

Over the years, several considerations on the radiology in non-accidental injury I, and sev-

eral of my colleagues, have come across a small number of children who have been pre-

sented to health care professionals with a parental complaint of feeling a “clicking” sensation or “grating” feeling in the rib cage or, indeed, even hearing an odd “click”. This has been ignored as a sign of rib fractures. The children have often been sent home only to return with further injury.

I write to draw the attention of paediatricians to this sign in the hope that it will assist in recognising this for what it is.

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Lipid and glucose metabolism in 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. However, until now all studies have been done in adults. The aim of this study was to evaluate the influence of highly active anti-retroviral therapy (HAART) on serum levels of 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 HDL after PI use in HIV-1-infected children, as was previously observed in adults.1 However, in contrast with adults, a marked increase in HDL and normal glucose levels was observed.2 The total cholesterol/HDL ratio, fasting triglyceride and FFA levels remained stable over time.

To date, it has not been revealed whether these metabolic changes are the result of HAART or if HIV-1 infection itself is responsi-

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% 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 (4.7%) was a carrier.

The total cholesterol/HDL ratio, fasting triglyceride and FFA levels remained stable over time.

To date, it has not been revealed whether these metabolic changes are the result of HAART or if HIV-1 infection itself is responsi-

ble. Hyperglycaemia 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.3 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 also be partially 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.1 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 we unfortunately have to conclude that HAART also affects the lipid and glucose metabolism in children. We would like to thank I. Zwang and M A. C. van Fessel for performing the laboratory analyses, W. C. J Hop for statistical advice, G. J. Brunning, F. Pistor, H. J. Scherpber, and T. F. W. Wolfs for their co-operation.

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children is not undertaken, this may result in unnecessary exposure of these children to hepatitis B infection.

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Reference

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 treating childhood hyperhidrosis.1,2 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 excessive hyperhidrosis. Excess palmar sweating caused difficulty with school work (difficulty holding a pen, with the ink smudging the paper because of sweating) and social embarrassment. Botulinum toxin type A (Dysport; 20 mouse units) was administered intradermally using a 27G needle 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 hydroxide 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

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Caring for Muslim Patients


Islam is the religion of one-fifth of humanity and, with an estimated population of 1.6 million Muslims form 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 which is what this publication is trying to do. It is divided into nine chapters, peppered with anecdotes and examples, with a summary box at the end of chapters, and concludes with useful appendices on Islam and the Internet, Muslim organisations, and a glossary.

Following an overview of Islam and Muslims in Europe, it delves into issues that are important in the daily life of a Muslim family—like birth, marriage, and death as well as health matters at times like the fasting month of Ramadan and pilgrimage to Mecca. Chapters on Fasting and Pilgrimage may not be of direct interest to the paediatrician but remembering the holy days in the Islamic calendar may improve, among other things, clinical attendance.

The chapter on birth customs is probably the one that will interest paediatricians most. We read about practices following the birth of a new baby, like whispering the name of God in their ear, shaving the hair on the seventh day, and remembering the holy days in the Islamic calendar. These practices may have contradicted the teachings of Islam. As a Muslim myself, I may not agree with some of the 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.

H Marcovitch

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, pharmacological 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
high expectation for a strong potential in children. The two factors drove the research into childhood migraine many steps forwards. Unfortunately despite the huge amount of new knowledge on the subject and, possibly, the increased prevalence of headache and migraine in children, there is more need than ever for an up-to-date publication on the subject. Until now, only two books on childhood headache and migraine are available on the paediatric bookshelf. The Classical books of Charles Barlow (Headache and migraine in childhood, Oxford: Blackwell Scientific, 1988) and of Judith Hackaday (Migraine in childhood, London: Butterworth, 1988) remained the most recent sources of information and advice for practising paediatricians and general practitioners. Therefore, this book comes at an appropriate time to fill some of the gaps in the paediatric literature.

The book deals mainly with the diagnostic issues, differential diagnosis, and the management of childhood headache in a simple and practical way. Complex concepts and mechanisms were introduced and discussed with simplicity that made the reading of the book flow easily. Headache was introduced as a pain syndrome that has its own methods of measurement and management in the early part of the book. The general direction the book was determined, therefore, by the fact that 7 out of the 10 contributing authors been pain scientists, clinical psychologists, or child psychiatrists. Such an influence towards the psychology of pain has enhanced the quality of the book and enriched its value and contents. Therefore, the book provides the researcher on the subject of pain and headache a valuable reference to understand difficult issues in relation to pain measurement, impact of pain and headache on child's life and also the management of headache including behavioural modification.

From the point of view of the practising general paediatricians who deal with children with headache in busy medical paediatric clinics, the book provides a good brief overview of the causes of headache, diagnostic assessment, and treatment. The use of simple data collection sheet would be very useful to assist the attending physician in establishing the diagnosis of the type of headache and also in identifying both the trigger and relieving factors. The editors propose, in two appendices, lengthy interviews of the child and the parents that may defy the practicality of the consultation. It would be more appropriate to the clinician if those interviews were short and direct. Also, diaries would be a useful tool to help understand the child's headache by recording symptoms as they occur.

There is no doubt that this book will prove to be an important and useful reference 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-Arefeh

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 book 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 their book has adopted a “problem oriented approach”. This does not match other books that start with clinical signs and symptoms; such a true problem orientated 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 reference. 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 of the course. They could help the teachers by providing them an agreed core curriculum. As a collaboration between Scottish departments of paediatrics, this should not 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 examination of 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 one could level criticism at other similar textbooks. This could be seen as an argument for radical redesign of all undergraduate texts to match more fundamental 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 fresh, clearly structured text for early professional training. 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.

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