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

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. Leaning 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 is 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 1994–95 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 to also publish it in a future paper issue. Pearson and colleagues have presented data highlighting the use of the paediatric index of mortality (PIM) score as a tool for auditing paediatric intensive care unit (PICU) performance. Whilst we would agree with the authors’ message that PIM has many advantages over other scoring systems, we feel the authors’ message that PIM has many advantages over other scoring systems, we feel the authors’ message that PIM has many advantages over other scoring systems, we feel the authors’ message that PIM has many advantages over other scoring systems, we feel the authors’ message that PIM has many advantages over other scoring systems, we feel the authors’ message that PIM has many advantages over other scoring systems, we feel the authors’ message that PIM has many advantages over other scoring systems, we feel the authors’ message that PIM has many advantages over other scoring systems, we feel the authors’ message that PIM has many advantages over other scoring systems.

Calibration of the paediatric index of mortality score for UK paediatric intensive care

Pearson and colleagues have presented data highlighting the use of the paediatric index of mortality (PIM) score as a tool for auditing paediatric intensive care unit (PICU) performance. Whilst we would agree with the authors’ message that PIM has many advantages over other scoring systems, we feel that urgent calibration is needed before this tool is adopted as a benchmark for performance in the UK. PIM variables were developed predominantly from an Australian data set (one British PICU, Birmingham participated) over 1994–95; the data used in Pearson’s validation comes from five UK PICUs, including our own over the period 1998–99. PIM continues to discriminate between death and survival reasonably well giving an area under the ROC curve of 0.840 (95% CI 0.819–0.853) marginally less than the figure of 0.90 seen in the original paper. However, from the 4 year period between development and validation the model is now calibrated poorly, as evidenced by two pieces of information from Pearson’s study. First, the overall standardised mortality ratio (SMR) is 0.87 (95% CI 0.81–0.94); this figure is remarkably concordant across 4 of the 5 PICUs. Second, from table 2, it is possible to calculate the Hosmer-Lemeshow statistic: chi-squared = 37.41, p<0.0001. This implies poor calibration, (good calibration traditionally represented by a p value >0.10). 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 highlights 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 calculation 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 PICU performance will be derived.

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 in a similar fashion 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

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References
5 MRC/DH Working Party on Intensive Care: The research needs and opportunities relevant to the NHS Medical Research Council 1997.

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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,7 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 Australia, and the new model will be recalibrating PIM using data from units in the UK (PICOS). The UK should aim for best practice (such as PIM), rather than the average of good and not-so-good units from the whole of the UK (PICOS). The UK should aim for best practice rather than being content with average practice.

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A review of surgical 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).1 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 6).

Follow up chest x rays were assessed by a consultant paediatric radiologist, using the Chrispin Norman Scoring system.2 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 or that 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

Anti-neutrophil cytoplasmic autoantibody positive glomerulonephritis in monozygotic twins

Scanty information is available concerning anti-neutrophil cytoplasmic autoantibodies (ANCAs) associated disease in children, and very few cases of familial vasculitis have been reported in the literature.4,5

We have observed two monozygotic twins developing ANCA necrotising glomerulonephritis (GN). A 7 year old boy was hospitalised for normocomplementemic acute nephritis. Pericutaaneous 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.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Lung function data: simple spirometry after bronchodilator inhalation</th>
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<tbody>
<tr>
<td>Case</td>
<td>FEV1 (% of predicted)</td>
</tr>
<tr>
<td>Preop</td>
<td>Postop</td>
</tr>
<tr>
<td>1</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>75</td>
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<td>3</td>
<td>85</td>
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<td>58</td>
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<td>5</td>
<td>60</td>
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<tr>
<td>6</td>
<td>83</td>
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<table>
<thead>
<tr>
<th>Table 2</th>
<th>Chest x ray score</th>
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<tbody>
<tr>
<td>Case</td>
<td>Operation</td>
</tr>
<tr>
<td>Preop</td>
<td>Postop</td>
</tr>
<tr>
<td>1</td>
<td>LLL</td>
</tr>
<tr>
<td>2</td>
<td>RUL</td>
</tr>
<tr>
<td>3</td>
<td>RUL</td>
</tr>
<tr>
<td>4</td>
<td>RLL and RML</td>
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Data are the Chrispin-Norman scores in the lung quadrant within which the patients had developed focal bronchiectasis and for which they underwent lobectomy (maximum score 8).
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 ANCAcs were observed at a dilution of 1/160. The stored samples of the first twin were tested and pANCAc 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, DR1; 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. An 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 pauciimmune 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 pANCAc 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 families.

In conclusion, a pANCAc test should always be performed in children with acute nephritis of unclear aetiology; a diagnosis of ANCA-GN should not preclude renal transplantation. HLA B35 may play a role in the pathogenesis of ANCA-GN.

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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% of children with hepatitis B markers. 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. The figures suggest 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 a prenatal 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 hyperhidrosis. Excessive 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; 50 mouse units) was administered intradermally using a 27G needle to the finger tips and the area over the hypothalamic and thalamic eminences of both hands. EMMA 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 thumb and the area over the hypothalamic eminence, which was reported to reduce sweating at night for up to three weeks. She is now being monitored for a further year to assess whether further treatment is required.

If you would like to buy a gentle gift for a literate medical friend, you could do worse than this lighthearted escapade, all royaltys from which might well be true. If you would like to buy a gentle gift for a literate medical friend, you could do worse than this lighthearted escapade, all royalties from which might well be true.

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 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 couples, simply because we are a 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.

Followed an overview of Islam and Muslims in Europe, it delves into issues that are important in the daily life of a Muslim family—life 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 most challenging. The chapter on birth customs is probably the most challenging. The book 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.

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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-Arafah

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 paediatricians who wish 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 set by the new course framework. It should help the teachers by providing them an agreed core curriculum. As a collaboration between Scottish departments of paediatrics, this book 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 book 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 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 current undergraduate texts to match more fundamentally the new curriculum 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, elegant, and integrated approach to core material for students undertaking their first paediatric post. It is easy to read and easy to navigate. It is not a traditional textbook, but instead, it provides a structure that gives uniformity of approach for each organ system and indeed the textbook is clearly laid out.

References


M D C Donaldson