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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).[1] 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.[2 3] 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 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.[4 5] 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/~scharr/ukpicos.

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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.[1] 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 indication 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.[1] 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),[1] marginally less than the figure of 0.90 seen in the original paper.[2] However, from the 4 year period between development and validation the model is now calibrating poorly, as evidenced by two pieces of information from Pearson's study.[1]

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,[1] 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 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 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.

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Authors' reply

Dr Tibby and Dr Murdoch note that, in our study of paediatric intensive care units (PICUs) in the UK,[1] 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,[2] MPM[3] and APACHE,[4] 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

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,[1] 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.[5–7] 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 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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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).[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 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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Table 1 Lung function data: simple spirometry after bronchodilator inhalation

Case	FEV_1 (% of predicted)			FVC (% of predicted)			Number of years followed up
	Preop	Postop (6 mth)	Long term follow up	Preop	Postop (6 mth)	Long term follow up	
1	—	94 (6 y)	103	—	91	106	10
2	60	75	60	76	87	81	4
3	85	76	76	103	94	91	5
4	58	59	66	66	66	71	5
5	46	46	58	74	71	84	4
6	83	83	60	77	83	58	9

Table 2 Chest *x* ray score

Case	Operation	Local Chrispin–Norman scores		
		Preop	Postop (6 mth)	Long term follow up
1	LLL	3	2	5
2	RUL	5	3	4
3	RUL	5	2	4
4	RUL	5	4	5
5	RUL	6	3	5
6	RLL and RML	5	2	4

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).

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.[1–3]

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 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.