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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References

1 **Pearson GA**, Stickley J, Shann F. Calibration of the paediatric index of mortality in UK paediatric intensive care units. *Arch Dis Child* 2001;**84**:125–8.
2 **Pollack MM**, Patel KM, Ruttimann UE. PRISM III: an updated pediatric risk of mortality score. *Crit Care Med* 1996;**24**:743–52.
3 **Lemeshow S**, Teres D, Klar J, *et al*. Mortality probability models (MPM II) based on an international cohort of intensive care unit patients. *JAMA* 1993;**270**:2478–86.
4 **Knaus WA**, Wagner DP, Draper EA, *et al*. The APACHE III prognostic system: risk prediction of hospital mortality for critically ill hospitalized adults. *Chest* 1991;**100**:1619–36.
5 **Pearson G**, Shann F, Barry P, *et al*. Should paediatric intensive care be centralised? Trent versus Victoria. *Lancet* 1997;**349**:1213–7.
6 **Bennett NR**. Provision of paediatric intensive care services. *Br J Hosp Med* 1997;**58**:368–71.
7 **de Courcy-Golder K**. A strategy for development of paediatric intensive care within the United Kingdom. *Intensive Crit Care Nurs* 1996;**12**:84–9.

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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References

1 **Lucas J**, Connett GJ, Lea R, *et al*. Lung resection in cystic fibrosis patients with localised pulmonary disease. *Arch Dis Child* 1996;**74**:449–51.
2 **Chrispin AR**, Norman AP. The systematic evaluation of chest radiographs in cystic fibrosis. *Pediatr Radiol* 1974;**2**:101–5.

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.