

Table 5 Group C: where necropsy showed clinically important extracardiac pathology

Polysplenia	3
Cerebral arteriovenous malformation	1
Pulmonary hypoplasia	1
di George's syndrome	1
Hepatoblastoma	1
Asplenia	1
Pulmonary haemangiomas	1
Abnormal karyotype	1
Lymphocytic thyroiditis	1

in table 5. In only one case was the extracardiac pathology the cause of death (cerebral arteriovenous malformation with associated infarction). The other anomalies, however, sometimes helped in the attribution of a "syndrome" to the case. In 15 cases no additional information was obtained by necropsy (group D).

Discussion

The claim that necropsy has been rendered obsolete by the improved techniques of modern clinical diagnosis is refuted by this study. Admittedly, in only one of our cases did postmortem examination show clinical cardiac diagnosis to be completely wrong. The criteria for assessing the value of necropsy, however, must be broader than merely the complete disagreement of clinical and necropsy diagnosis. In our series 38% of cases had undiagnosed additional cardiac anomalies or surgical flaws which may have contributed to death in 13 cases. Porter and Keeling's study found a high rate of significant discrepancy between necropsy and clinical diagnoses.³ Forty four percent of postmortem examinations on neonatal deaths in that series yielded clinically important information. In our series of more highly investigated children 80% of necropsies yielded additional information, although the precise clinical importance of some of the findings has yet to be evaluated. Intensive clinical investigation in life increases the need for detailed post mortem examination to ensure that the basis for the adopted treatment was sound.

A disproportionate number of the major clinico-pathological discrepancies arose in children who had only had echocardiographic assessment (four out of seven cases). This supports the observation of Goldman,⁴ who found that discrepancies at necropsy were much more common in cases where diagnosis was based solely on the result of a newer technique such as ultrasound, computed tomography scanning, or nuclear medicine than those based on other investigations. In our study aortopulmonary window caused particular diagnostic problems on echocardiography as this region is hard to visualise. It was missed twice in children with interrupted aortic arch, a

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well recognised though rare association and diagnostic pitfall.⁵ Aortopulmonary window was also erroneously diagnosed in a child with a complete atrioventriculoseptal defect. Minor unsuspected cardiac lesions were found in 28%, illustrating the incomplete resolution of these forms of cardiac imaging. These lesions would not have influenced the management or outcome had they been detected. They may assume greater importance, however, as methods of treating the severe concomitant abnormalities improve.

Scrutiny of highly skilled, complex surgery on tiny hearts at necropsy is essential for the continued improvement in surgical technique. Flaws in surgical technique were uncommon in our series but when present they provided valuable lessons for the management of future cases.

Myocardial necrosis is increasingly recognised in children dying in the perinatal period. Less well recognised is the association of necrosis with cardiac malformation when the distribution of dead tissue may reflect the site of an obstructive cardiac lesion.⁶ The necroses in our cases were presumed to be related to recent cardiac surgery in most cases. Techniques of paediatric myocardial preservation during bypass operations are less well determined than for adult procedures.

It is difficult to be certain of the clinical importance of the foreign body embolism in the lungs of these cases. The ease of their histological demonstration, on random lung sections, suggest that they were present in large numbers. Filter material, plastic tubing, and antifoaming agents are well documented sources of embolism following cardiac bypass procedures but these usually have systemic distribution.⁷ Other sources also have to be excluded as not all the cases occurred following surgery. Although we have no evidence that these emboli caused problems in the children, the long term consequences may be important in these patients who often already have impaired pulmonary circulation.

Both these complications of myocardial necrosis and foreign body embolism were shown by histological examination. This highlights the essential role of routine histological sampling even in the absence of macroscopic lesions.

Clinically important incidental abnormalities were uncommon but when present sometimes led to a unifying diagnosis for the case. No additional information was obtained from 20% of cases. As paediatric pathologists we are aware of the value of negative findings in counselling bereaved parents. Although unsatisfactory in explaining the outcome of the case, the negative findings may still be used to comfort parents that all appropriate measures had been taken.

Pathologists must share some of the blame for the

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