difference probably reflects the bias of the diagnostic method used here, which favours the inclusion of patients with serious defects. Despite the fact that 35% of the patients follow up and treated medically have survived, they are in functional class III or IV of the NYHA classification, have poor quality of life, and an unfavourable outlook, thus confirming that non-surgical management of this malformation yields very poor results.

The palliative operation adds risk to the complete correction to be performed later, does not always prevent pulmonary vascular obstructive disease (as illustrated in one of our cases), and was associated, in our study, with a high surgical mortality. Our policy at present is to use the palliative procedure only in cases with left ventricular hypoplasia.17, 18

Although the overall surgical mortality of the complete repair was very high, the results for the period 1983–8 were dramatically better than those for the preceding decade (1971–82). Probable causes for this improvement are the use of surgery at an early age, increase in the experience with the technique, improved perioperative care, better selection of the patients (with exclusion of those with severe pulmonary hypertension that does not decrease in the hyperoxia and tolazoline tests), and general improvement in the care of patients with Down’s syndrome. Despite the high mortality of surgery in our study, the outlook is still better with surgery than without. The surgical mortality reported in other series ranged from 62% to 17%.19 Complete atrioventricular canal without major associated intracardiac anomalies can now be repaired in infants younger than 2 years in some of the leading centres with an acceptable mortality of 10–25%.5 7 19-21 Prospective studies, which are required to evaluate the late results of current surgical practice, need to include more objective functional assessment. Published late-non-actuarial mortality after repair of complete atrioventricular septal defect ranges from 2–19% but the duration of follow up varies considerably.8 12 21 There was a five year survival of about 47% in our children who were subjected to complete correction from 1983 to 1988, and good functional results of the operation. This compares with a poor prognosis both in terms of survival and of quality of life of the patients treated medically. This confirms the expectation, based on the results in other series, that complete repair drastically improves the natural history of this malformation. This also supports the opinion that this correction should be performed at an early age in as many patients as possible.18-22 Nevertheless, there are still doubts on the long term consequences of dysplasia of the left atrioventricular valve in the surgically corrected patient, stressing the need for monitoring of these patients over a more extended period.23 24

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Commentary

In atrioventricular septal defect and other cardiac lesions sufficient relief from the symptoms of cardiac failure, with prevention of secondary changes in the heart and lungs, may not be afforded by medical treatment alone. Early sur-gical intervention, despite the complexity of the lesion, to induce immediate improvement and minimise long term changes in both myocardial and pulmonary vasculature is the clear message in this paper.

An additional issue raised by the authors relates to the management of children with multiple handicaps. Society today expects that each handicap will be assessed in its own right, and...
alleviated so as to produce the optimum benefit to the individual. This relates to the overall quality of life and to the ability to make the fullest use of individual talents. No one would question measures to improve the hearing of the deaf child with heart disease (for example, grommets or a hearing aid) or spectacles for a myopic diabetic. But turning the question around and reducing the symptoms imposed by heart disease brings one back to the primary objectives of intervention. If the simple measures of altering a feeding pattern, judicious use of antibiotic treatment, and medical regimes to reduce cardiac failure do not produce the desired effect in the short term, the logical extension of management is to consider timely surgical intervention. The change in well being, self assurance, and independence not just for the individual but for the whole family should be the ultimate goal. It should remain a goal even if life expectancy is unaltered and could be argued if that expectancy were to be marginally curtailed. The long drawn out illness rendered by advancing pulmonary vascular disease, after a disrupted infancy and childhood, is a heavy burden for a family that timely intervention can curtail. However, in some circumstances the quality of life achieved naturally, even if less than normal, may not be capable of improvement by the most expert surgical intervention. The choice for a child with a single (cardiac) handicap, or for the child with multiple handicaps, lies in the assessment of the improved quality of life and the time that is to be sustained both with, and without, the specific type of medical and surgical intervention necessary to achieve as near perfect an anatomical repair with its consequent improved physiology.

Entering into the current decade, economic factors veiled in the term 'medical audit' will take prominence. In hospital terms the cardiac care of a multiply handicapped child may take longer for a child with a single system defect. The hospital audit, however, should be able to take into consideration the demand or otherwise on the local paediatric, community, and school medical services. The influence of the child's chronic illness or well being to the total family structure and economy merits deep reflection.

Most congenital heart lesions may be improved and some corrected surgically. The extent to which the repair leaves the heart in a normal state depends on the original lesion; many would accept that closure of a persistent ductus arteriosus is the only truly corrective operation. To this the closure of a moderate sized ventricular septal defect and some forms of total anomalous pulmonary venous drainage might also merit the accolade of 'correction'. Myocardial and pulmonary vascular changes may resolve after operation and the site of the original defect become so well endothelialised that no evidence remains.

Consideration of other lesions even as 'simple' as a secundum atrial septal defect, age at presentation, effects on the myocardium secondary to the lesion, and changes in the pulmonary vasculature, however, will determine the long term performance even after successful anatomical repair. Over and above achieving as near perfect anatomic repair and returning normal physiology the long term effects on pulmonary vascular response, myocardial performance, and the late occurrence of arrhythmias will be the indicators of how near perfection has been attained. There will be a spectrum through the varied surgical techniques needed to 'correct' simple lesions, repair the more complex ones (for example, Fallot's tetralogy, atrioventricular septal defects, coarctation of aorta, aortic stenosis, and transposition with no other defect than atrial septal defect), to essentially long term palliation—for example, persistent truncus arteriosus, pulmonary atresia with a ventricular septal defect, complex transposition, and the problems of the univentricular heart. Semilunar and atrioventricular valve replacement should also be considered palliative. Nevertheless substantial clinical improvement allowing a normal life style, though perhaps not athletic prowess, follows the majority of these procedures.

It is this improvement in well being with freedom from breathlessness and fatigue which forms the real objective in attempting to overcome the physiological impairment of congenital heart lesions. This is accompanied particularly by freedom from cyanosis and irritability, a better nutritional state, and less disturbance from intercurrent infections. An important sequel is the improved confidence to achieve a normal daily life for both the parents and child. While this improved quality of life style should enhance longevity, there are many other factors involved, and this attainment is not the sole or even prime motivation. Even with a good immediate result from surgical intervention the duration of its benefit will be influenced by the state of the myocardium after the operation (itself influenced by preoperative and perioperative factors) and also the evolution or resolution of pulmonary vascular changes. The inherent unavoidable sequelae such as the need to replace a prosthetic valve or renew a conduit (with or without a valve) will also affect ultimate prognosis. Although significant reduction in susceptibility to infective endocarditis is usually listed as a benefit, rarely is the risk eliminated and prosthetic endocarditis can be even more devastating.

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