The Fate of Survivors of Cardiac Surgery in Infancy


SUMMARY Knowledge of the long-term effects of infant cardiac surgery is essential if further progress is to be made in not only immediate, but also long-term care. We therefore analyzed the fate of 599 infants who survived 3 weeks or more after operation in the first year of life performed over the last 25 years. Actuarial survival curves were obtained for each operation for a given condition. The rate for 3-week survival in infants was also determined for the years 1972–1976. Four risk categories were established. (1) Low initial, low late survival: pulmonary artery banding (PAB) for complete transposition of the great arteries (TGA) and ventricular septal defect (VSD)—3-week survival 83%, 5-year survival 41%; shunts for pulmonary atresia with VSD — 3-week survival 73%, 5-year survival 51%; shunts for tricuspid atresia — 3-week survival 78%, 5-year survival 70%; PAB for VSD — 3-week survival 67%, 5-year survival 81%; Blalock-Hanlon operation for TGA — 3-week survival 87%; 5-year survival 52%. (2) High initial, low late survival: Mustard’s operation for TGA — 3-week survival 94%, 5-year survival 81%; shunts for tetralogy of Fallot — 3-week survival 97%, 5-year survival 74%. (3) Low initial, high late survival: coarctation with or without persistent ductus arteriosus — 3-week survival 81%, 5-year survival 93%; total anomalous pulmonary venous drainage — 3-week survival 69%, 5-year survival 90%; pulmonary stenosis — 3-week survival 63%, 5-year survival 100%. (4) High initial, high late survival: closure of VSD — 3-week survival 97%, 5-year survival 97%.

These results demonstrate the superiority of one-stage over two-stage repair in both VSD and TGA. PAB produced unsatisfactory overall survival in all conditions studied and therefore has limited application in future surgical management. The study provides a baseline with which future developments in therapy can be compared.

THE PAST 12 years have seen a remarkable change in the pattern of open heart surgery, with more and more emphasis placed on the desirability of corrective surgery during the first year of life. This has largely been the result of a dramatic fall in immediate surgical mortality, with the result that some lesions can now be corrected in infancy with a risk of death less than 5%.

New techniques present new problems, and it has become necessary to determine (1) the effect of correction in infancy on the late mortality of the condition and (2) the place for two-stage surgery — palliative surgery in infancy followed by corrective surgery later. The latter issue can only be resolved by longitudinal studies in individual patients. Accordingly, we have ascertained the fate of 599 infants who survived cardiac surgery at the Hospital for Sick Children over the last 21 years.

Methods

The results of all operations for heart defects and coarctation of the aorta carried out in infants less than 1 year old between January 1, 1955 and January 1, 1976 were reviewed. Overseas patients with isolated persistent ductus arteriosus (PDA) were set aside because of difficulties of long term follow-up (patients with PDA are usually discharged 1–2 years after surgery). Patients dying within 3 weeks of operation were classed as immediate deaths. Potential long-term survivors — those living 3 weeks or more after operation — form the basis of this study. Their fate was determined by inspection of their subsequent case records, and correspondence with parents and the doctors responsible for their follow-up. Potential follow-up time for any patient is taken as the time from operation to October 15, 1976. Actual follow-up time is that from the operation date until the last date before October 15, 1976 about which we had information. An accurate diagnosis was established in 98% of patients by autopsy, at open heart surgery, or by cardiac catheterization and angiocardiography before and/or after surgery. In the remaining patients, who were studied early in the experience, the diagnosis was based on clinical and noninvasive investigation together with findings at extracardiac surgery. After the exclusions already mentioned, there were 699 infants operated in all. In 10 of these, the only information available was that an operation had taken place, so these were also excluded. Table 1 lists the principal diagnoses and operations performed in 599 patients.

The dates of certain types of second operation were also noted: (1) The first corrective operation after palliation in infancy. (Redirection of venous inflow with an intra-atrial baffle [Mustard’s operation] for complete transposition of the great arteries [TGA], and right atrial-to-pulmonary artery conduits [Fontan’s operation] for tricuspid atresia are classified as corrective operations.) (2) Operations for re-coarctation of the aorta. (3) Revision of Mustard’s operation to relieve obstruction of the caval or pulmonary venous pathways.

The data were punched on cards that were grouped to give a minimum of 14 patients per analysis and processed by the University of London Computer.
TABLE 1. Diagnoses, Operations and Summary of Results

<table>
<thead>
<tr>
<th>Category</th>
<th>Late survival survival ((=) SEM)</th>
<th>Early survival (1972–1976)</th>
<th>Projected overall survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n</td>
<td>5-year</td>
<td>10-year</td>
</tr>
<tr>
<td>All patients</td>
<td>599</td>
<td>76.4 (1.9)</td>
<td>67.5 (2.3)</td>
</tr>
<tr>
<td>Atrioventricular canal defects</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Banding of PA</td>
<td>14</td>
<td>92.9 (6.9)</td>
<td>76.0 (12.2)</td>
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<td>Concordant TGA</td>
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<td></td>
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<td>All Mustards</td>
<td>73</td>
<td>81.2 (5.0)</td>
<td>67.9 (6.6)</td>
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<td>Mustards with Dacron</td>
<td>31</td>
<td>80.0 (7.3)</td>
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</tr>
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<td>Mustards with pericardium</td>
<td>42</td>
<td>85.3 (5.6)</td>
<td>69.4 (6.6)</td>
</tr>
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<td>59.5 (7.9)</td>
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<tr>
<td>Coarctation of aorta</td>
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<td></td>
</tr>
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<td>84.3 (3.4)</td>
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<td>93.4 (2.9)</td>
<td>93.4 (2.9)</td>
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<td>PDA + VSD only</td>
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<td>All operations</td>
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<td>All shunts</td>
<td>17</td>
<td>51.3 (13.7)</td>
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<td>All shunts</td>
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<td>Blalock-Taussig</td>
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<td>Waterston</td>
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<td>54.2 (19.7)</td>
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<td>TAPVD</td>
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<td>Primary closure</td>
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<td>96.6 (3.3)</td>
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<tr>
<td>Miscellaneous</td>
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</table>

Late survival exceeds deaths within the first 3 weeks of surgery and refers to the entire period 1955–1975 inclusive, whereas early survival refers to 1972–1976 inclusive. The projected overall survival was obtained by multiplying 3-week survival by 5-10-15-year survival, as appropriate. Early survival has not been calculated for the group “all patients” because of the difficulty of calculating this for the “miscellaneous” group.

*98% if no VSD present.
†The same patient had a Blalock-Taussig and then a Waterston shunt.

Abbreviations: PA = pulmonary artery; TGA = transposition of the great arteries; VSD = ventricular septal defect; PDA = persistent ductus arteriosus; TAPVD = total anomalous pulmonary venous drainage.

Centre using the BMD 11S statistical package. This enabled survival curves to be compared by t test. In plotting a survival curve, account was taken of the length of observation of any patient and his or her status (alive or dead) at the end of that period. In patients who had had second operations as outlined above, a second survival curve (alive without second operation) was then plotted, taking as alternative end
points of observation (1) alive or (2) dead or undergoing a second operation. This method corresponds to that proposed for demonstrating embolism rates in patients with prosthetic valves. Patients for whom no information was available after January 1, 1975 were recorded as lost to follow-up for the purpose of the actuarial curve. In congenital heart disease, however, because the frequency of follow-up appointment is inversely proportional to the time elapsed since operation, the follow-up in patients alive when last heard of or seen was also calculated as the proportion of actual follow-up time to potential follow-up time.

In assessing the safest surgical approach to any given problem, the early death rate is obviously important, but it makes little sense to assess it for the whole of a 25-year period. Accordingly, the mortality within 3 weeks of surgery was assessed for all operations carried out in the 5-year period January 1, 1972 to January 1, 1977, including those on overseas patients.

Finally, the early and late mortalities were combined to project overall survival by multiplying the 3-week survival proportion by the survival proportion 5, 10 and 15 years after operation.

**Results**

Three-quarters of the patients alive when last seen had been followed for at least 78% of the time available for follow-up. Figure 1 summarizes a detailed analysis of the quality of follow-up.

In table 1 the 5-, 10- and 15-year survivals are summarized for each group of patients, together with the mortality within the first 3 weeks of operation, where appropriate, and the projected overall survival. In the following more detailed description, for every condition described, the number and cause (where known) of deaths between primary palliation and secondary correction are given, as is the number of deaths immediately and late after correction.

**Total Population**

Survival for the whole population of 599 patients is presented in figure 2, which shows that after an initial steep drop to 85.4 ± 1.5% survival after 1 year, the rate of fall of survival is approximately 1¼% per year.

**Atrioventricular Canal Defects**

Fourteen infants survived banding of the pulmonary artery to reduce pulmonary blood flow. In 11 of these the defect was complete, in two partial, and in one uncertain. Figure 3 shows the survival curve. By 15 years after operation, the majority of survivors had undergone debanding and repair of the intracardiac defect, but only 63.3 ± 15.4% survived. Of the four late deaths, two were at corrective surgery, one was from pneumonia and one was sudden and unexplained.

**TGA with Atrioventricular Concordance**

The first Mustard operation in an infant was carried out in this hospital in 1966, but elective repair in infants
Survival did not become our policy until 1970. Before this, the only surgical methods of palliation were closed atrial septectomy (the Blalock-Hanlon operation) and banding of the pulmonary artery for patients with an associated ventricular septal defect (VSD). These palliative operations continued beyond 1970 to an extent for patients with either failed balloon septostomy or a large VSD.

The long-term results of primary Blalock-Hanlon operation followed by secondary repair have proved disappointing (fig. 4). Although by 7 years postoperatively almost all had had Mustard operations, at 15 years only 36.4 ± 12.3% survived. Eleven of the 20 late deaths occurred more than 3 weeks after the Mustard procedure. Seven were due to presumed arrhythmias, because in each case the ECG before death showed a major abnormality, such as atrial flutter or complete atrioventricular dissociation. Other major causes of late death were the Mustard procedure itself in five, and cerebrovascular accidents before the Mustard operation in two.

The long-term results of pulmonary artery banding are even less satisfactory (fig. 5), as only 40.7 ± 12.7% were alive at 5 years. Of the 11 late deaths the principal causes were subsequent correction in four and bronchopneumonia or cardiac failure in three.

Seventy-three patients survived the Mustard procedure in infancy. Nine of these had an associated VSD that was sufficiently large to raise the left ventricular systolic pressure to at least 75% systemic in four. Survival reaches 81.2 ± 5.0% at 4 years (fig. 4). The major causes of the 13 late deaths were presumed arrhythmias in two and obstruction to caval or pulmonary venous pathways in four.

Until 1971, pericardium was used for the intracardiac baffle, but this was then changed to Dacron. When it became apparent in 1972 that this led to a high incidence of caval pathway obstruction, the use of pericardium was recommenced. With pericardium, survival reaches 83.3 ± 5.6% at 1 year, with no subsequent late deaths, whereas with Dacron, the corresponding figure at 4 years is 80.0 ± 7.3% (fig. 6). This difference is not statistically significant. However, there is a marked difference between these two groups as regards survival without surgical revision of caval or pulmonary venous obstruction (fig. 7). With Dacron, at 4 years, only 52.9 ± 9.4% have survived.
without requiring surgical revision. Thus, the difference in survival without revision in the two groups is statistically significant at the 5% level at 3 years and at the 0.5% level after 4 years. The 13 revisions in the Dacron group were achieved with only one early and one late death.

Survival after all Mustard operations was significantly better than after the Blalock-Hanlon procedure ($p < 0.025$ at 5 years; $p < 0.005$ at 8 years) (fig. 4).

Coarctation of the Aorta

Included in this group are all patients who had resection of a coarctation in infancy. Six percent of patients also underwent pulmonary artery banding, but are not classified as such. The survival curves for all groups with coarctation are all similar in that there is a certain mortality within the first 3 years from associated conditions, but thereafter there are no late deaths.

Survival in all patients reached $84.3 \pm 3.4\%$ at 4 years (fig. 8) and no late deaths occurred thereafter.

**Figure 5.** Survival in infants undergoing pulmonary artery banding for concordant transposition of the great arteries (TGA) and ventricular septal defect (VSD). The upper line represents overall survival without debanding and a "correction operation," the middle line "survival without corrective surgery," and the lowest line the number of patients on which this survival is based, as in figure 2.

The corresponding figures for isolated coarctations (with or without PDA) are $93.4\% \pm 2.9\%$ at 1.5 years and for coarctation with VSD (with or without PDA), $78.5 \pm 8.5\%$ at 1 year. In these two groups there have been no late deaths since 1966. 

**Figure 6.** Comparison of survival (to January 1, 1976) in infants undergoing Mustard's operation with the intra-atrial baffle consisting of (1) pericardium and (2) Dacron. The upper curves are survival curves, related to the left-hand ordinate. The lower curves, related to the right-hand ordinate, indicate the number of patients on whom the survival curve is based.

**Figure 7.** Comparison of survival without revision of Mustard's operation with the intra-atrial baffle consisting of (1) pericardium and (2) Dacron. Comparison of the curve for pericardium with that in figure 5 shows that no patients in this group underwent surgical revision. The number of patients on whom this comparison is calculated is shown in figure 4.
FIGURE 8. Survival for 123 infants who underwent resection of coarctation of the aorta. The lower curve indicates the length and size of follow-up, as in figure 2.

Because none of the 123 patients died of recoarctation of the aorta, figure 9 shows the percentage who did not require operation for recoarctation of the aorta among the 104 survivors (95.1 ± 2.4% at 5 years, 79.1 ± 5.8% at 10 years, and 67.5 ± 8.0% at 15 years). Operation for recoarctation was carried out in 16 patients without mortality. Only two of these patients had been older than 3 months at their original operation.

Pulmonary Atresia with VSD

Initial palliation consisted of a Waterston19 (ascending aorta to right pulmonary artery) anastomosis in five, a Blalock-Taussig20 (subclavian to pulmonary artery) anastomosis in 10, and a Potts21 (descending aorta to left pulmonary artery) anastomosis in two. The late survival of this condition was low — 20.5 ± 16.2% at 15 years (fig. 10). There were no late deaths among patients who had had the Waterston operation. Four of the 10 late deaths were due to second operations.

Pulmonary Stenosis with Intact Ventricular Septum

There were 12 patients who survived operation, which was a closed valvotomy in nine. Open valvotomy, a Blalock-Taussig anastomosis, and a Waterston shunt were each used once. Two patients with a closed valvotomy and the shunted patients all required open valvotomy later. There were no late deaths, with survival extending to 19 years.

Tetralogy of Fallot

Initial palliation was by a Blalock-Taussig shunt in 56, a Waterston operation in 18, a Potts operation in four and a Glenn shunt (superior vena cava to right pulmonary artery)22 in one. Figure 11 shows a steady fall off in survival, to 56.1 ± 6.6% at 13 years, by which time almost all living patients had undergone correction. Of the 28 late deaths, seven were due to complete repair and five were due to shunt occlusion. Two were due to noncardiac causes (meningococcal septicemia and chicken pox) and none occurred late after total correction. The overall survival at 5 years for the Blalock-Taussig operation was 5.3% higher than for the Waterston operation, though this difference is not statistically significant (table 1). However, many of the early Blalock-Taussig shunts were done at a time when, if shunt failure occurred,
secondary correction carried a prohibitively high mortality. If survival after Blalock-Taussig shunts carried out since January 1, 1962 is compared with survival after the Waterston shunt, a statistically significant difference ($p < 0.05$) emerges 7.5 years after the shunt, by which time the majority of living patients in both groups had undergone total correction (fig. 12). Survival in the Blalock-Taussig group reaches 84.9 ± 7.1% at 7 years, with no subsequent deaths.

**Total Anomalous Pulmonary Venous Drainage**

There were only three late deaths (two in hospital and one outside) among 31 survivors of correction of this anomaly (fig. 13). Because all three patients had never recovered from the operation, the late deaths were in a rather different category from the majority. The latter was felt to be related to left ventricular hypoplasia. Survival after 1 year was 89.5 ± 7.1%.

**Tricuspid Atresia**

Twenty-seven shunts were carried out: 14 Blalock-Taussig, six Glenn, five Waterston and two Potts.23

**FIGURE 11.** Survival after all shunts for tetralogy of Fallot. The upper graph shows overall survival and survival without radical correction. The lower histogram shows the size and length of follow-up.

**FIGURE 12.** Comparison of survival after Blalock-Taussig (dotted line) and Waterston (solid line) shunts in infants with tetralogy of Fallot (upper graph). The lowest two lines of this graph represent survival without radical correction. The lower histogram indicates the length and size of follow-up in both groups.

There were 10 late deaths from a variety of causes. Survival reached 69.7 ± 9.8% at 3 years, and no late deaths occurred from 3–12 years, after which the natural history of the condition overtakes the beneficial effect of palliation (fig. 14). It is too early to say yet what influence the Fontan operation (right atrial-to-pulmonary artery valved conduit)9 will have on this survival.

**Ventricular Septal Defect**

Until the end of 1971, the management of infants with intractable heart failure resistant to medical treatment was banding of the pulmonary artery,14 but thereafter the policy was changed to primary closure.14 Figure 15 compares survival after these two methods of management. By 2 years, primary closure is significantly superior ($p < 0.05$) and by 4 years the differ-
Figure 13. *Survival after correction of total anomalous pulmonary venous drainage (TAPVD) in infancy, arranged as in figure 2.*

Survival is even more marked \( (p < 0.01) \). Survival at this stage is 96.6 ± 3.3% for primary closure, and 80.7 ± 4.7% for banding. The only cause of late death after primary closure was noncardiac (thoracic dystrophy); thus, there have been no late deaths related to VSD closure in either group. In two patients in the banded group, spontaneous closure of the VSD had occurred by the time debanding was performed. Seven patients died during debanding and closure of the VSD, if this was necessary. Reoperation was the major cause of late death in this group. One patient died from a ruptured aneurysm of the pulmonary trunk.

**Discussion**

Heart disease causes more than half the deaths from congenital malformations in the first year of life.\(^{25, 26}\) Infancy is by far the most perilous time for patients with congenital heart disease. The evidence for this assertion has been admirably reviewed by Hoffman.\(^{27}\) In brief, there is wide divergence between estimates of survival based on intensive population studies, such as that by Carlgren\(^{28}\) in Gothenburg (1941–1950), and those based on autopsy studies alone, such as that by Menashe and colleagues\(^{22}\) in Portland (1957–1961). Carlgren found that 662 of 1000 children born with congenital heart disease would survive until their first birthday. Menashe estimated that 722 of 1000 deaths from congenital heart disease would be in the first year of life. If birth rate is increasing during an autopsy study, the number of infants at risk of dying will increase, and infant mortality will appear disproportionately large,\(^{27}\) but this can hardly explain the wide divergence. The difference is probably due to mild congenital heart disease, such as mild pulmonary stenosis, which would be more likely to be recorded in a population study involving live patients than in an

![Figure 14. Survival after all shunts for tricuspid atresia in infancy, arranged as in figure 4.](image)

![Figure 15. Comparison of survival after primary closure and debanding of the pulmonary artery (PA) in 106 infants with ventricular septal defect (VSD). The lower two histograms relate to the size of population studied, and the steeply sloping line relates to survival after banding without debanding and VSD closure.](image)
Quantification of Late Survival

Late results of surgery are most commonly presented as percentage late mortality: late deaths divided by number of operations times 100. This method is precise only in the unlikely event of there being no late deaths or infinite time available for follow-up. The percentage of late mortality depends not only on the true prognosis but also on the total time of follow-up and the distribution of follow-up within that time. Thus it is impossible to accurately compare the percentage of late mortality between one surgical report and another unless the mean follow-up times are identical. The most precise method is cohort analysis, as carried out by Taussig and colleagues, on all shunts carried out by Blalock between 1944 and 1951, very few of them in infancy. This method is extremely slow in producing results, because so much time must elapse before analysis can begin. The actuarial method enables late survival to be estimated while operations are actually going on.

Percentage late mortality consistently overestimates late survival, because it is only concerned with what has happened, whereas actuarial survival also considers what will happen to patients whose follow-up has been relatively short on the basis of deaths in those whose follow-up has been longer.

If assessing late mortality from a single operation presents problems, the assessment of two-stage corrections is more daunting yet. Even if there is no late mortality from the first operation, the early mortality from the first operation means that there are fewer patients at risk of dying from the second. Thus, direct addition of mortalities from the two operations is only accurate if both approach zero. Any significant late mortality between stages means that overall survival can only really be assessed actuarially. The disadvantage of the method is that it is only meaningful when the vast majority of living patients has had secondary correction. For this reason “survival without correction” curves have been plotted. These curves do not indicate what would happen to a population if secondary correction were not used; they merely indicate the proportion of patients living without correction.

Because of the very great improvement since 1951 in immediate mortality of all operations, early mortality has been assessed over the past 5 years, and projected overall survival after surgery for each category calculated accordingly. The extent to which this reflects survival in the entire population depends upon what lesion is being considered. In conditions that present as critical emergencies in the first month of life, such as complete TGA, total anomalous pulmonary venous drainage with pulmonary venous obstruction or severe coarctation of the aorta, an unquantifiable number of deaths will occur before the patients reached the hospital and more deaths will occur in hospital before operation. Deaths before surgery are less likely to occur in VSD or tetralogy of Fallot.

Further, because actuarial late mortality decreases, as time goes on (this was clearly shown for the Blalock-Taussig operation for tetralogy), the projected overall survival figures present an overly pessimistic view of the prospects for an infant undergoing surgery today. Actuarial projection as we have used it overstates mortality rate, particularly where improvement of management has occurred recently. This is especially so where such improvement has taken place in the immediate results of secondary corrective operations, because these are usually the main cause of late death after primary palliation. In theory, this problem might have been overcome by separating the late results of primary palliation from the early results of secondary correction, much as we have separated the early results of palliation from the late results. Using more recent results from secondary correction would enable one to obtain a more up-to-date estimate of the probability of surviving secondary correction, and that probability could then be multiplied by the probability of surviving until the time of secondary correction. However, on closer examination, certain problems emerge. In particular, this approach demands that the group withdrawn alive from follow-up after primary palliation be matched in all important respects with the group undergoing secondary repair. To give an extreme example of the problems that might arise, suppose that secondary correction in patients forming the basis of the study of survival after palliation was only carried out as an emergency in moribund patients. These dying patients would be regarded as “withdrawn alive” from palliation. Suppose further that the immediate risks of secondary correction were assessed in a group of patients who had had previous palliation, but who were in excellent condition at the time of secondary correction. These techniques of selection would give an unduly optimistic picture of both palliative and corrective surgery taken separately; combining the two would magnify the errors. Objective measurements of the well-being of patients have varied so much over the period of this long study that we felt it unjustifiable retrospectively to match severity in these two groups of patients in the necessary fashion. We elected instead to incorporate the early results of corrective surgery in the late results of palliative surgery, accepting that the results would appear pessimistic, but knowing at least that they would be based upon facts rather than on inspired guesses. Actuarial analysis of surgical results in congenital heart disease has until recently been rare, and concerned mainly with older children. More recently a number of excellent reports have documented long-term survival in individual lesions both actuarially and parametrically.

Early and Late Mortality

If 90% survival is defined as high, examination of table 1 leads to identification of four groups with respect to survival.
The most disheartening group is that in which early and late survival was low. This includes all groups of patients who underwent banding of the pulmonary artery, the Blalock-Hanlon operation, resection of coarctation in the presence of VSD (though there have been no late deaths since 1966), and shunts for tricuspid and pulmonary atresia.

The next group is that in which immediate survival is gratifyingly high, but late results are not so satisfactory. This group includes Mustard’s operation, and shunts for tetralogy of Fallot.

Arguably more satisfactory for everyone concerned is the group in which the initial risks are high, but the late results are satisfactory. This includes coarctation with or without PDA, total anomalous pulmonary venous drainage and critical pulmonary stenosis.

The only operation for which early and late survival exceeds 90% is primary closure of VSD. The median projected overall 5-year survival rate for all groups is 58%.

Primary vs Two-stage Correction

In no case were management policies randomized. Even so, the superiority of primary closure of VSD over banding followed by debanding seems clear. The same is true for Mustard’s operation as a primary procedure rather than after a Blalock-Hanlon procedure, certainly for simple TGA. The management of TGA with VSD is more complex. A Blalock-Hanlon operation alone is not enough. However, there were not enough patients in the primary Mustard group to compare with banding of the pulmonary artery. The results of banding were so poor that almost any different approach is likely to be an improvement. The arterial switch operation**5**8 59 may be appropriate for this group. Overall survival in simple TGA can more probably be improved by modifying surgical technique to reduce venous obstruction and arrhythmias after Mustard’s operation than by pulmonary artery banding followed by an arterial switch.

Insufficient data were obtained to assess the relative merits of primary as opposed to two-stage repair of tetralogy of Fallot. The comparison between palliation with the Waterston and Blalock-Taussig shunts was between patients in similar age ranges, but perhaps not with similar degrees of severity. Nevertheless, the result suggests that primary total correction should be compared with two-stage correction with the Blalock-Taussig rather than the Waterston shunt as the first stage. The 85% long-term survival after the Blalock-Taussig shunt since 1961 was achieved largely at a time when the immediate mortality after secondary correction was much higher than it is today.

Overall survival with two-stage repair of atrioventricular canal was, with two exceptions, the lowest recorded. This has been a strong incentive toward primary repair in infancy, and the initial results of this approach have been encouraging, with an 83% hospital survival in two reports.61 62

Pulmonary atresia with VSD can be a correctable lesion.63 The poor survival recorded here may be partly the result of inadequate preoperative evaluation in patients operated on before 1975. Selective angiography with injection into major aortopulmonary collateral arteries can now be performed at any age, showing the presence or absence of central pulmonary arteries and the nature of pulmonary blood supply. In pulmonary atresia the outlook may be improved by better preoperative investigation.64 65

Corrective Operation — Fact or Fiction?

The word corrective has been used throughout this article to indicate definitive repair. However, no primary operation described has been entirely without long-term complications or mortality. Even an apparently straightforward operation such as resection of coarctation is not corrective in all cases, as evidenced by the rate of operations for recoarctation. However, if no operation with any long-term complications were to be classed as corrective there would be no corrective surgical operation, so we retain the adjective corrective. In this case, as in all others, the results of surgical treatment must be set against the results of medical treatment or no treatment for the condition. The mortality in infants with coarctation, with or without medical treatment, was reported as 41%,66 and 100% in infants with coarctation and any associated lesion.67 Though the overall prognosis for VSD is excellent,68 the mortality in infants presenting with congestive heart failure was recently reported as 11 of 77 (14.3%) on medical treatment.69 Between 1957 and 1964 in California, 89.3% of patients with TGA were dead by their first birthday.70 Of 25 patients with total anomalous pulmonary venous drainage and pulmonary venous obstruction treated medically, only two (8%) survived their first year.71 Without treatment, two-thirds of infants with tricuspid atresia die in the first year of life.72

So, while the management of congenital heart disease in infancy has made great strides over the past 25 years, the need for detailed and prolonged follow-up, not just with regard to mortality, but also with regard to function, is clear. Only by such studies can the need for further improvements in management be identified and acted upon.

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Fontan Procedure for Tricuspid Atresia

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SUMMARY A Fontan procedure has been performed on 29 patients for tricuspid valvar atresia. The age range was 8 months to 33 years (median 10 years), the pulmonary vascular resistance ranged from 1.8–6.1 units • m² (mean 3.3 units • m²), and the mean pulmonary arterial pressure ranged from 13–45 mm Hg (mean 21 mm Hg). Twenty-nine previous operations had been performed in 23 patients. Fourteen other associated cardiopulmonary anomalies were present in 12 patients. There were four hospital deaths (13.8%), all in patients with complicating features. Among the last 22 consecutive patients who have undergone operation, one died (4.5%). Complete atrioventricular block necessitated pacemaker implantation in one patient. No late deaths occurred. Of the 19 patients followed 3 months or more from the time of operation, eight have no restriction of exercise capacity, nine have only mild restriction, and two have a poor result. The Fontan approach to tricuspid atresia has several theoretical advantages over previously used shunts or pulmonary artery banding, the operative mortality in patients who have suitable anatomy and hemodynamics is low, and the results have been good.

TRICUSPID VALVULAR ATRESIA, a congenital cardiac defect often occurring in association with other complex cardiovascular malformations, carries a very poor prognosis, although certain types of tricuspid atresia have benefited from various shunting and banding procedures. Recently, a more physiologic repair — anastomosis of the right atrium to either the rudimentary right ventricle or the pulmonary arteries (Fontan procedure) — has been reported from various centers. This report describes the entire Mayo Clinic experience with the Fontan procedure for tricuspid atresia, involving 29 patients.

The term “tricuspid atresia” can be confusing unless it is further defined, because the term has been used to describe hearts in which either the right or the left atrioventricular valve may be atretic. In this report, therefore, the term is used to describe the congenital cardiac malformation in which there is situs solitus of the atria and absence of the right atrioventricular connection, with a main ventricle of left ventricular type and a rudimentary ventricle of right ventricular type (outlet chamber). The ventriculoarterial connections may be of any type. We agree with the observation that most cases of tricuspid atresia are examples of univentricular heart with outlet chamber and atresia of the right atrioventricular valve. In this manuscript, the term “rudimentary right ventricle” will be used to describe this outlet chamber.

Patient Profile

The first Fontan procedure for tricuspid atresia at our institution was performed in October 1973, and this consecutive series of 29 patients extends from then through March 1979. The patients were classified ac-
The fate of survivors of cardiac surgery in infancy.
F J Macartney, J F Taylor, G R Graham, M De Leval and J Stark

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