Late Survival and Symptoms After Repair of Tetralogy of Fallot

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SUMMARY The long-term results of 414 patients who underwent repair of tetralogy of Fallot between 1967 and 1977 were studied and correlated with the results of others. There were nine late deaths (8-year actuarial survival 95.8%). Six of the deaths were directly related to the malformation or its treatment. Eight patients (2.4%) required reoperation. Ten patients (4.8%) had arrhythmic symptoms. Eight (3.1%) had congestive heart failure that required treatment. The risk factors associated with late events of all types, including death, were: older age at repair, a high mean ratio of peak systolic right-to-left ventricular pressures \((P_{RV/LV})\) immediately after repair, and the presence of a Potts anastomosis. Neither a transannular patch nor a previous Blalock-Taussig or Waterson anastomosis was an incremental risk factor. Bacterial endocarditis was not observed. Three hundred seven patients underwent repair primarily or after a single Blalock-Taussig or Waterston shunt and had a \(P_{RV/LV}\) of 0.85 or less after repair. Among these selected patients, the actuarial survival was 98.1%, which is still lower than that for the general population \((p = 0.12)\), and freedom from events was 95.9%. Late after repair, \(P_{RV/LV}\) was lower by 6 ± 28% \((±SD)\) than \(P_{RV/LV}\) immediately after repair \((p = 0.03)\) in the 33 restudied patients with such data. The higher the \(P_{RV/LV}\) immediately after repair, the greater the percent reduction.

MANY CENTERS have achieved very low hospital mortality rates for repair of tetralogy of Fallot, but only a few have reported the very long-term results of the methods in use to achieve good early results. To collect data concerning long-term survival and symptomatic status of patients who have had repair of tetralogy, and to identify risk factors for a poor late result, we performed a detailed follow-up study of the 414 patients who survived the operation and correlated the findings with the reports of others.

Materials and Methods

The study group consists of 398 patients with classic tetralogy of Fallot and 16 patients with tetralogy of Fallot with subpulmonary ventricular septal defect (VSD) who survived intracardiac repair and postoperative hospitalization between January 1967 and January 1977, at the University of Alabama in Birmingham Medical Center. In only four were no follow-up data obtainable. Patients were excluded from the study group if they had tetralogy of Fallot with pulmonary atresia,1 pulmonary incompetence, complete atrophicventricular canal,2 flap-valve VSD, or other major associated lesions as detailed elsewhere.3

Characteristics of the study group at the time of repair are shown in table 1. The surgical techniques have been described.4, 5 A transannular patch was used in 108 patients (26%), a nonvalved extracardiac conduit in two (0.5%), and a valved extracardiac conduit or orthotopic heterograft valve in six (1%). The mean ratio of peak systolic right-to-left ventricular pressures \((P_{RV/LV})\) \((±SD)\), measured in the operating room after repair, was 0.48 ± 0.170 for the patients without a transannular patch and 0.51 ± 0.198 for the patients with a transannular patch \((p = 0.14)\).

Data Collection

Questionnaires were sent to the patients or their parents and their local physicians in December 1977. Inquiry was made as to New York Heart Association classification of symptoms; presence and severity of activity restriction, dyspnea, peripheral edema, and cyanosis; progress in learning and behavior; heart rhythms requiring medical treatment; occurrence of bacterial endocarditis; postoperative catheterization; reoperation; and heart medications. If responses were unclear or equivocal, the physicians and patients were contacted by telephone. Data from follow-up visits in our institution and from previous correspondence were also used. Three hundred ninety-five responses were received. Data from follow-up visits in our center or from previous correspondence were available in 15 of the patients from whom no responses were obtained to this specific follow-up effort. Only four patients could not be traced.

Follow-up ranged from 2 weeks to 11.2 years (median 5.1 years). Seventy-five percent were followed for more than 3 years and 25% were followed for more than 7 years.

Of the 414 patients in the study group, 36 (8.7%) have undergone recatheterization; in 15, it was performed electively. In 33 of these 36 patients, the ratio between pulmonary and systemic blood flow \((Qp/Qs)\) was 1.6 or less, and data were available to determine the difference between \(P_{RV/LV}\) in the operating room immediately after repair and that late postoperatively. The difference was tested using the paired \(t\) test. In addition, multiple regression analysis was used to relate late \(P_{RV/LV}\) to the level observed in the operat-
ing room after repair, age at repair, the presence or absence of previous palliative surgery (and if present, its type), use of a transannular patch in the repair, and the interval between repair and recatheterization.

### Analytical Methods

The data were organized so that for each patient, statements could be made concerning late death, reoperation, arrhythmic symptoms and congestive heart failure. Late death from any cause was included. The data were further refined to determine whether the death was related to the tetralogy of Fallot or its repair. Reoperation included all cardiac surgical procedures undertaken after the first few postoperative days. Every patient with a symptomatic indication of need for reoperation underwent reoperation. Arrhythmic symptoms included all atrial orventricular rhythm disturbances reported by the physicians and requiring medical or surgical treatment. One patient experiencing documented but untreated episodes of symptomatic premature ventricular complexes is also included. The one patient who died suddenly of an undocumented but presumed ventricular arrhythmia is not included as having arrhythmic symptoms. For actuarial analyses, the date upon which the arrhythmic symptoms were first recognized was used. Congestive heart failure was defined as any symptom or sign of systemic or pulmonary venous hypertension for which medical therapy was initiated. For purposes of actuarial analyses, the onset of the congestive failure was taken to be that date on which medical therapy was started. The product-limit analysis method was used to describe freedom from these events. We also analyzed freedom from any event; since some patients had more than one of these events, the date of the earliest occurring event was used in the analyses.

The actuarial survival estimates were compared with the U.S. life table for 1976, matched for age, race and sex. To determine if the operation could be considered curative, we generated from this life table an expected probability of dying for each patient as if he or she were a part of the general population. From the duration of follow-up and based upon age at repair, race and sex, the total number of such expected deaths for the entire group was then calculated as the sum of the individual probabilities of dying. This was compared with the observed number of deaths by the chi-square test.

Incremental risk factors were identified and quantified by use of the proportional hazards general linear model of Cox as modified by Breslow. The factors investigated for increasing risk included age at repair, previous palliative surgery and, if done, the interval between the first palliative procedure and complete repair, as well as the type of palliative procedure, use of a transannular patch, $P_{RV/LV}$ immediately after repair, sex, race and interactions among these factors (variables formed by the multiplication of factors one with another). From this analysis, the predicted hazard of an event on the basis of one set of patient-specific characteristics compared with another was calculated from the following relationships:

$$\ln(\lambda(t)/\lambda_0) = \beta_1 x_1 + \beta_2 x_2 + \ldots + \beta_k x_k$$  \hspace{1cm} (1)

where $\lambda(t)$ is the hazard function, $\lambda_0$ is the (unknown) hazard apart from that explained by the incremental risk factors $(x_1 - x_k)$, and the contribution of each factor to hazard is determined from its statistically estimated coefficient $(\beta_1 - \beta_k)$. If $\ln(\lambda(t)/\lambda_0)$ is obtained for two sets of characteristics, and these are called $h_1$ and $h_2$, the predicted hazard for the characteristics $h_1$ relative to $h_2$ is:

$$\exp(h_1 - h_2).$$

We analyzed a selected group of 307 patients to determine late results under ideal circumstances. These patients were younger than 20 years of age at repair, underwent either primary repair or secondary repair after a single Blalock-Taussig or Waterston anastomosis, and had a $P_{RV/LV}$ of 0.85 or less immediately after repair. The selection was based on the analysis of incremental risk factors, which indicated that older age at repair, high $P_{RV/LV}$ immediately after repair, and presence of a previous Potts anastomosis were associated with an increased risk of late events.

Results are presented as the mean ± sd, and proportions (including actuarial estimates) are presented with their 70% confidence limits (CL), equivalent to ± 1 standard deviation. The actuarial estimates in the Results section refer to the 8-year value.

### Results

#### Premature Late Death

Nine of the 414 patients died during the follow-up period, so that actuarial 8-year survival was 95.8% (CL 93.9–97.1%) (fig. 1A). Four of the late deaths were from congestive heart failure and two from ventricular arrhythmias. Three deaths were considered to be unrelated to the cardiac lesions: one patient exsanguinated after a liver biopsy and two died in accidents.

The 8-year actuarial survival was significantly less than that of 99.7% for the age-race-sex-matched general population. Were the patients behaving as part of the general population — that is, were the

### Table 1. Characteristics of the Follow-up Study Group at the Time of Repair

<table>
<thead>
<tr>
<th></th>
<th>n</th>
<th>Median</th>
<th>Range</th>
<th>75% over</th>
<th>Range</th>
<th>25% over</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>414</td>
<td>6.5 years</td>
<td>6.5 years</td>
<td>4.2 years</td>
<td>4.2 years</td>
<td>12.8 years</td>
<td>12.8 years</td>
</tr>
<tr>
<td>Weight</td>
<td>18.4 kg</td>
<td>Range 4.63–96.6 kg</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Previous palliative surgery</td>
<td>181 patients (44%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Single Blalock-Taussig or Waterston</td>
<td>143 patients</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Previous Potts (with or without other palliative surgery)</td>
<td>20 patients</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other palliative procedures or multiple shunts</td>
<td>18 patients</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
operation "curative" — three deaths would be expected. Nine deaths were observed (p for the difference = 0.0005). Interestingly, three of the nine late deaths were unrelated to tetralogy of Fallot or its repair.

The incremental risk factors for premature late death, determined by multivariate analysis, were older age at repair (but only for deaths from all causes), high P_{RV/LV} immediately after repair, and the presence of a Potts anastomosis (table 2). A P_{RV/LV} of 0.85 after repair is predicted to give a 2.5 times greater risk of premature death within 8 years than one of 0.50. The presence of a Potts anastomosis is predicted to increase the risk of premature death 9.4 times. The use of a transannular patch or the presence of a Blalock-Taussig or Waterston anastomosis were not significant incremental risk factors for premature late death (p > 0.2).

In the case of the selected patients (n = 307, defined in the Methods section), their actuarial 8-year survival of 98.1% (CL 96.3–99.0%) was greater than that of the remaining 107 patients (p = 0.02), and it approached more closely that of the matched general population (fig. 1B). In this subgroup, three deaths were observed (two related to the tetralogy and one not related to it). If the patients were behaving as part of the general population and the operation curative, 1.2 deaths would be expected (p = 0.12).

Reoperation

Eight patients underwent reoperations, an actuarial incidence of freedom from reoperation of 97.6% (range 96.5–98.3%). All reoperations were performed within the first 5 years after repair.

Three patients, two of whom had initially had a transannular patch, were reoperated upon for aneurysm of the right ventricular outflow tract. The P_{RV/LV} in the operating room after the original repair was 0.79 in one, 0.85 in another, and 1.0 in the third. The reoperations yielded good results.

Three patients underwent reoperation for persistent right ventricular hypertension. One had a large Potts anastomosis and P_{RV/LV} after the initial repair was 1.0 despite a transannular patch. At reoperation 5 years later, stenosis of the right pulmonary artery was repaired, and an orthotopic heterograft pulmonary valve was inserted. The P_{RV/LV} after repair remained 1.0, and the patient died 8 months later of congestive heart failure. Presumably, this patient had developed severe pulmonary vascular changes from the Potts anastomosis. Another patient underwent reoperation elsewhere for a stenosis of the left pulmonary artery and one at the site of the right pulmonary artery repair incident to taking down a Waterston anastomosis; he died postoperatively. In the third patient, originally operated upon in 1972 without use of a transannular patch, placement of a transannular patch at reoperation successfully corrected the right ventricular hypertension.

Two patients had reoperation for recurrent or residual VSDs, at which time one also had anuloplasty and repair of a previously overlooked straddling mitral valve with mitral incompetence.

The incremental risk factors for reoperation were high P_{RV/LV} immediately after repair and any previous palliative operation (table 3). The incidence of reoperation was similar in the selected subgroup and in the remaining group (p = 0.23).

Late Arrhythmic Symptoms

Ten patients had arrhythmic symptoms during the follow-up period (table 4), an actuarial incidence of freedom from arrhythmic symptoms of 95.2% (CL 93.0–96.8%). The incremental risk factors for late arrhythmic symptoms were older age at repair and previous Potts anastomosis (table 5). A patient 30
TABLE 2. Cox Multivariate Analysis of Premature Late Death After Repair of Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Incremental risk factors for premature late death</th>
<th>Death from all causes</th>
<th>Tetralogy-related deaths only</th>
</tr>
</thead>
<tbody>
<tr>
<td>Older age at repair (mos)</td>
<td>0.005 ± 0.0024</td>
<td>0.004 ± 0.003</td>
</tr>
<tr>
<td>High (P_{RV/LV}) immediately after repair</td>
<td>2.7 ± 1.67</td>
<td>3.9 ± 1.95</td>
</tr>
<tr>
<td>Previous Potts anastomosis</td>
<td>2.2 ± 0.70</td>
<td>2.6 ± 0.87</td>
</tr>
</tbody>
</table>

Coefficient ± SD  
\(p\)  
\(p\)

Abbreviation: \(P_{RV/LV}\) = ratio of peak right-to-left ventricular pressure.

TABLE 3. Cox Multivariate Analysis of Late Reoperation After Repair of Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Incremental risk factors for reoperation</th>
<th>Coefficient ± SD</th>
<th>(p)</th>
</tr>
</thead>
<tbody>
<tr>
<td>High (P_{RV/LV}) immediately after repair</td>
<td>7.3 ± 1.76</td>
<td>&lt; 0.0001</td>
</tr>
<tr>
<td>Previous palliative operation</td>
<td>2.0 ± 0.90</td>
<td>0.03</td>
</tr>
</tbody>
</table>

Abbreviation: \(P_{RV/LV}\) = ratio of peak right-to-left ventricular pressure.

years old at the time of repair is predicted to have a 17 times greater risk of developing arrhythmic symptoms than a patient 5 years at repair; one 5 years of age is predicted to have a 1.4 times greater risk than a patient 2 years of age at repair. A patient with a previous Potts anastomosis is predicted to have 4.6 times increased risk for arrhythmic symptoms.

Only two patients in the selected subgroup had arrhythmias, an actuarial incidence of freedom from arrhythmias of 99.3% (CL 98.5–99.6%), higher than that in the other patients (\(p = 0.002\)).

**Congestive Heart Failure**

Eight patients had congestive heart failure requiring treatment late after operation, which represents an actuarial incidence of freedom from congestive failure of 96.9% (CL 95.4–98.0%). Four of these eight died of heart failure and are included among the late deaths. The incremental risk factors for late congestive heart failure were older age at repair, high \(P_{RV/LV}\) immediately after repair, and a previous Potts anastomosis (table 6). One patient, age 30 years at repair, is predicted to have a 4.0 times increased risk for development of congestive heart failure than one 5 years old at repair, and one 5 years old is predicted to have a 1.2 times greater risk than a patient 2 years old at repair. A \(P_{RV/LV}\) of 0.85 immediately after repair is

**TABLE 4. Arrhythmic Symptoms After Repair of Tetralogy of Fallot**

<table>
<thead>
<tr>
<th>Category</th>
<th>No. of pts</th>
<th>Age at repair (years)</th>
<th>Previous palliative operation</th>
<th>Treatment</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular arrhythmias</td>
<td>6</td>
<td>45</td>
<td>None</td>
<td>Quinidine with control</td>
<td>PVCs pre- and postoperatively</td>
</tr>
<tr>
<td></td>
<td>39</td>
<td></td>
<td>None</td>
<td></td>
<td>PVCs pre- and postoperatively</td>
</tr>
<tr>
<td></td>
<td>35</td>
<td></td>
<td>LBT Closed Brock RBT</td>
<td>Digoxin, quinidine with control</td>
<td>PVCs began 2½ yrs postoperatively</td>
</tr>
<tr>
<td></td>
<td>28</td>
<td></td>
<td>Potts</td>
<td>Propranolol with control</td>
<td>PVCs pre- and postoperatively</td>
</tr>
<tr>
<td></td>
<td>27</td>
<td></td>
<td>Potts</td>
<td>Digoxin, propranolol, disopyramide</td>
<td>Congestive heart failure; atrial fibrillation pre- and postoperatively, late death</td>
</tr>
<tr>
<td></td>
<td>1.4</td>
<td></td>
<td>None</td>
<td>Quinidine with control</td>
<td>Also paroxysmal tachycardia. (P_{RV/LV}) at recath 58/110 (0.53).</td>
</tr>
<tr>
<td>Atrial tachyarrhythmias</td>
<td>3</td>
<td>50</td>
<td>None</td>
<td>Digoxin, quinidine with control</td>
<td>Associated cerebral embolus</td>
</tr>
<tr>
<td></td>
<td>26</td>
<td></td>
<td>Potts</td>
<td>Cardioversion, digoxin with control</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>7</td>
<td></td>
<td>Waterston</td>
<td>Digoxin with control</td>
<td>—</td>
</tr>
<tr>
<td>Late variable AV block</td>
<td>1</td>
<td>36</td>
<td>Closed Brock (2)</td>
<td>Pacemaker</td>
<td>Reoperation for residual VSD 5 months postoperatively. Congestive heart failure, late death.</td>
</tr>
</tbody>
</table>

Total 10

Abbreviations: LBT = left Blalock-Taussig; RBT = right Blalock-Taussig; PVC = premature ventricular complex; AV = atrioventricular; \(P_{RV/LV}\) = ratio of peak pressure left-to-right ventricular pressure.
TABLE 5. Cox Multivariate Analysis of Late Arrhythmic Symptoms After Repair of Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Incremental risk factors for late arrhythmic symptoms</th>
<th>Coefficient ± SD</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Older age at repair</td>
<td>0.0095 ± 0.00198</td>
<td>&lt; 0.0001</td>
</tr>
<tr>
<td>Previous Potts anastomosis</td>
<td>1.5 ± 0.71</td>
<td>0.03</td>
</tr>
</tbody>
</table>

TABLE 6. Cox Multivariate Analysis of Congestive Heart Failure After Repair of Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Incremental risk factors for congestive heart failure</th>
<th>Coefficient ± SD</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Older age at repair</td>
<td>0.005 ± 0.0028</td>
<td>0.10</td>
</tr>
<tr>
<td>High PRV/LV immediately after repair</td>
<td>4.3 ± 1.69</td>
<td>0.01</td>
</tr>
<tr>
<td>Previous Potts anastomosis</td>
<td>3.4 ± 0.77</td>
<td>&lt; 0.0001</td>
</tr>
</tbody>
</table>

Abbreviation: PRV/LV = ratio of peak left-to-right ventricular pressure.

predicted to increase the risk for congestive heart failure 4.4 times more than one of 0.50. The presence of Potts anastomosis is predicted to increase the risk of congestive heart failure 29 times.

By way of comparison, two patients in the selected subgroup were treated for congestive heart failure within the first 2 months after leaving the hospital. The actuarial incidence of freedom from congestive heart failure among these patients was 99.3% (CL 98.7-99.7%), which is higher than that in the other patients (p = 0.01).

Incremental Risk Factors for Any Late Event

The 35 events described above (late death, reoperation, arrhythmic symptoms and congestive heart failure) occurred in 24 of the 414 patients. Thus, 91.2% (CL 88.9-93.1%) of patients were event-free at 8 years (fig. 2a). The incremental risk factors for occurrence of any event were older age at repair, high PRV/LV immediately after repair, and a previous Potts anastomosis (table 7). A transannular patch and a single Blalock-Taussig or Waterston anastomosis were not incremental risk factors in this analysis. A patient age 30 years at the time of repair is predicted to have a 4.4 times greater risk for any late event than a patient 5 years of age at repair; one 5 years of age is predicted to have 1.2 times greater risk than a patient 2 years of age at repair. A PRV/LV of 0.85 immediately after repair is predicted to increase the risk for any late event by 3.1 times compared with a PRV/LV of 0.50. A Potts anastomosis is predicted to increase the risk for any event 5.4 times.

For the selected subgroup, the actuarial freedom from any event was 95.9% (CL 94.1-97.2%) (fig. 2B), higher than that for the other patients (p = 0.0002).

Other Events

No instances of endocarditis were reported by patients, parents or physicians.

Besides the activity restrictions due to late congestive heart failure or arrhythmias, one patient reported limitation of physical activity for reasons we could not determine. All other patients are active without apparent limitation.

Late Postoperative Cardiac Catheterization

Two of the 36 patients who were recatheterized (6%, CL 2-13%) had large recurrent or residual VSDs (Qp/Qs > 2.0), and both were recatheterized because of unfavorable signs of symptoms. They were 27 and 36 years old at the time of operation. Three (8%, CL 4-16%) had small VSDs (Qp/Qs > 1.3 but ≤ 1.6), four (11%, CL 6-19%) had trivial VSDs (Qp/Qs ≤ 0.8).
1.3), and 27 (75%, CL 65–83%) had no shunt at the ventricular level. Two patients had residual shunts at reopened Potts anastomoses, and one had a residual shunt at the recanalized subclavian artery of a ligated Blalock-Taussig shunt.

Late \( P_{RV/LV} \) in 33 restudied patients without important residual shunts was 0.55 ± 0.205, significantly lower than the 0.61 ± 0.234 in the operating room in these patients (\( p = 0.03 \)) (fig. 3). The change in \( P_{RV/LV} \) was related to its level immediately after repair (\( r = -0.50, p = 0.003 \)): If \( P_{RV/LV} \) immediately after repair was less than about 0.4, the change tended to be small but upward, whereas if it was above this value, the change tended to be downward. The change was not related to presence of a previous palliative operation, age at operation, presence or absence of a transannular patch, or the interval between operation and restudy (\( p > 0.2 \) for all). The relationship between \( P_{RV/LV} \) immediately after repair and at restudy is expressed by the regression equation

\[
\text{Late } P_{RV/LV} = 0.14 \pm 0.065 + 0.68 \pm 0.100 \cdot \text{OR } P_{RV/LV}
\]

(3)

where \( \text{OR} = \text{in the operating room}, p(\text{OR } P_{RV/LV}) < 0.0001 \), and the standard deviation for prediction of individual late \( P_{RV/LV} = 0.13 \). Late \( P_{RV/LV} \) less than 0.7 is associated with a \( P_{RV/LV} \) less than 0.83 immediately after repair.

Discussion

Methods and Limitations

This is a study of survival and symptoms late after repair of tetralogy of Fallot. We believe our data are reliable because of the care taken in collecting and analyzing the follow-up data. A state that seems asymptomatic to the patient, family and doctor may be associated with less-than-normal exercise capacity and undetected arrhythmias. The catheterization data presented are selective, but we believe they are of value when combined with data from the operating room, because of the surgically important relation of \( P_{RV/LV} \) immediately after repair to that late postoperatively.

Survival

Overall late survival is excellent and better than life expectancy without surgery. However, survival is not quite as good as for the age-race-sex-matched general population, suggesting that some factors have prevented the operation from being curative (providing a normal life expectancy) in all instances. Our results, though of a shorter follow-up, are similar to those reported by Fuster and associates for patients operated upon at the Mayo Clinic between 1955 and 1965 and followed for up to 18 years. Our own analysis of the basic Mayo Clinic data, supplied by the authors, using the Cox proportional hazards regression model, indicates that the incremental risk factors for late death were high \( P_{RV/LV} \) immediately after repair and the presence of a Potts anastomosis, findings similar to those in this study. As in our study, no effect of transannular patching or of a previous Blalock-Taussig or Waterston shunt was evident in the Mayo Clinic data.

The finding that the selected subgroup of 307 patients had a survival rate very close to that of the age-, sex- and race-matched general population emphasizes the importance of the incremental risk factors for premature death.

Actuarial survival rates beyond 20 postoperative years are not available, and the true “cure rate” is therefore not known.

Reoperation

Only a few patients in this series have undergone reoperation. Since routine postoperative catheterization has not been considered necessary, a few asymptomatic patients may have important residual shunts or residual right ventricular hypertrophy necessitating reoperation in the future. This is unlikely, we believe, because of routine measurements of \( P_{RV/LV} \) and shunts perioperatively, and restudy of those selected patients with unsatisfactory early results. Since all reoperations were in the first 5 years postoperatively, it is unlikely that longer follow-up will increase the incidence of this event, except possibly in patients with transannular patches. In such patients the finding of increasing right ventricular enlargement suggests that some may ultimately require the insertion of a valve beneath the patch. This procedure has resulted in good palliation.

Arrhythmic Symptoms

Although we have not performed an analysis of the postoperative ECGs for this study, we do know by history the incidence of symptomatic arrhythmias. Also, only one of the 414 patients in this series died suddenly.

In an analysis of their late results, Garson and col-

![Figure 3. Relationship of late ratio of peak right-to-left systolic pressure \( (P_{RV/LV}) \) to that immediately after repair. The solid line is the line of identity, and the dashed lines identify the 0.4 level.](http://circ.ahajournals.org/doi/fig/10.1161/01.CIR.65.2.408)
leagues noted that of eight patients who died suddenly, all had premature ventricular complexes (PVCs) postoperatively. None had left anterior hemiblock, first-degree atrioventricular block, or complete atrioventricular block. There was a significant (p < 0.001) relationship in their study between the occurrence of PVCs and right ventricular pressure of 60 mm Hg or greater. There was no association between the occurrence of PVCs and pulmonary incompetence during their follow-up study.

Our study emphasizes that older age at repair and a Potts anastomosis are also incremental risk factors for arrhythmic symptoms.

Congestive Heart Failure

Paralleling our experience, Rocchini and colleagues found that older age at repair and a previous Potts anastomosis increased the risk for late congestive heart failure, as did Zerbini. As in our series, Poirier and associates found PRV/LV after repair to be considerably higher in their patients with late congestive heart failure.

The finding that a Potts anastomosis is an incremental risk factor for congestive failure is not unexpected. The tendency for the development of pulmonary vascular disease after a Potts anastomosis is well known. Poirier and colleagues reported that three of the 13 late deaths in their series were due to severe pulmonary vascular disease. Each of these three had had a Potts anastomosis performed several years before complete repair. In Rocchini and associates' study of congestive heart failure after repair of tetralogy, pulmonary artery systolic pressure was elevated both preoperatively and postoperatively in seven of 17 patients in whom it was measured at both times. All seven patients had had a Potts or Waterston shunt. The similarity between Potts and Waterston anastomoses, that is, the critical size of the aortotomy and the late distortions of the pulmonary artery related to each, is noteworthy.

In our patients, a large residual VSD was present in two of the six patients catheterized late because of congestive heart failure. Rocchini and colleagues reported that a large residual VSD was the most prevalent abnormality leading to congestive heart failure. It was present in 31 of 36 patients with that condition. They noted that all postoperative patients with Qp/Qs ratios greater than 2:1 had congestive heart failure.

Activity and Lifestyle

The high proportion of patients without apparent limitation of activity indicates the generally good results from the operation. However, testing by formal exercise studies provides more complete information. Wessel and colleagues in such a study, found determinants of late functional status similar to ours. They found no significant difference in exercise performance (duration of exercise) according to peak right ventricular pressure (PRV ≤ 50 vs PRV > 50) in patients without postrepair residual VSD or pulmonary valve incompetence. Pulmonary incompetence was associated with a significant decrease in exercise capacity. A transannular patch was also associated with a significant decrease (p < 0.01) in exercise performance, different from our findings because of the greater sensitivity of their methods. They also found a significant reduction in duration of exercise in patients with residual VSD, large cardiothoracic ratio on chest film, or a Potts anastomosis.

PRV/LV Late After Repair

We found that important residual right ventricular hypertension increased the risk of an unfavorable late event, as did Garson et al. We also found that PRV/LV late after repair is predictable from measurements made in the operating room. We, in collaboration with Bertranou, have made a similar analysis of the data from other centers involving 324 patients (data and analysis available upon request), and the equation relating PRV/LV immediately after repair to that late postoperatively is:

\[
\text{Late PRV/LV} = 0.15 \pm 0.027 + 0.56 \pm 0.046 \cdot \text{OR PRV/LV} \quad (4)
\]

(p for postrepair PRV/LV < 0.0001).

The similarity of this equation to equation 3 supports the conclusion that late postrepair PRV/LV, and thus to some extent the patient's prognosis, can be predicted from the PRV/LV measured in the operating room immediately after repair.

Relevance to Current Surgical Practices

Our study indicates no difference in late results whether a low perioperative mortality is achieved by routine primary repair in infancy or by using, when necessary, an initial Blalock-Taussig shunt and definitive repair while still young (2-3 years old). However, this matter has not been fully resolved. Borow et al. suggested that left ventricular function is more normal late postoperatively when primary repair is done in infancy.

Our study indicates that the obsolescence of the Potts anastomosis is appropriate. The study emphasizes that the VSDs should be completely closed. Our data and reports of others do not resolve completely the issue of the indications for the use of transannular patching in relieving pulmonary stenosis. Transannular patching should probably be avoided when possible because of its deleterious effects on exercise capacity in some patients, and the possibility that 10-30 years later a pulmonary valve insertion may be needed. Moderate residual pulmonary stenosis (reasonably defined as a late postrepair PRV/LV ≤ 0.7) is very well tolerated as long as the pulmonary valve is competent; therefore, avoidance of transannular patching seems possible when such late PRV/LV ratios can be obtained without it. According to equations 3 and 4, this means a postrepair PRV/LV of less than about 0.80-0.85. When necessary, simple transannular patching rather than valve insertion seems adequate, as our data and the Mayo study do not

*Data were also kindly supplied by Drs. J. Aigueperse, J. Aubert, F. Fontan and A. Choussat.
show an unfavorable effect on survival and symptoms 10–20 years postoperatively. As indicated earlier in the discussion, very late postoperative pulmonary valve insertion may be necessary.

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