Status of the Adult and Adolescent After Repair of Tetralogy of Fallot

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SUMMARY Because increasing numbers of patients with surgically repaired tetralogy of Fallot (TOF) are surviving to adulthood, this study was done to provide follow-up data for physicians concerned with their care. We reviewed the current status of 233 patients born before January 1962 who had repair of TOF at a median age of 9.7 years (range 11 months to 36 years). There were 26 surgical deaths (11.1%), 12 late cardiac-related deaths (5.9%) and three noncardiac deaths (1.3%). Eight of the 12 late deaths occurred suddenly and unexpectedly. In each of these eight patients we had previously documented elevated right ventricular systolic pressure as well as ventricular premature depolarizations (VPDs). Among the 95 patients who responded to a questionnaire, 84 denied symptoms, 28 were married, 44 had attended college and all were employed. Cardiac catheterization revealed an unsatisfactory hemodynamic result in 39 of 120 patients, 30 of whom were asymptomatic. The poor result was due to a large left-to-right ventricular shunt in eight patients, persistent right ventricular outflow obstruction in 25 patients, pulmonary vascular obstructive disease in five patients, and left ventricular cardiomyopathy in one patient. Among the 21 patients with VPDs, 17 underwent cardiac catheterization; 15 had a right ventricular systolic pressure > 60 mm Hg and all had elevated right ventricular end-diastolic pressure. We conclude that 1) clinical assessment alone is nonpredictive of the hemodynamic result and thus cardiac catheterization is indicated in all patients; 2) the combination of persistent elevation of right ventricular systolic pressure above 60 mm Hg and VPDs placed the patient at risk for sudden death; and 3) over 80% of adults with repaired TOF can lead a normal life without impairment of intellect, exercise tolerance or fertility.

IN THE UNITED STATES each year an estimated 24,500 newborns have a congenital heart defect.1, 2 Approximately 11%, or 2700, have tetralogy of Fallot (TOF).3-5 Based on current surgical treatment, we estimate that 89% of these patients, or 2400 per year, will survive to adulthood. The purposes of this study were: 1) to present the characteristics of these patients; 2) to determine if symptomatology, physical examination and laboratory findings are adequate predictors of the postoperative hemodynamic result; and 3) to examine the late complications and causes of late death in an effort to predict and prevent these occurrences by identifying patients at risk.

Materials and Methods

Definition

Patients were included if they had typical anatomy of TOF with a nonrestrictive ventricular septal defect and pulmonary stenosis with essentially equal peak systolic pressure in two ventricles. Patients were excluded from the study if they had preoperative right ventricular outflow or main pulmonary artery atresia ("pseudotruncus") or aortic origin of either pulmonary artery.

Clinical Material

In the files of the section of Pediatric Cardiology, Texas Children's Hospital, there were records of 253 patients with TOF born before January 1, 1962. There were 154 males and 99 females (1.56:1 ratio). The 233 patients (92%) who had had intracardiac repair (ICR) form the basis for this report. The mean age at ICR was 10.7 years (range 1.0-31.3 years). Twenty-six patients died within 30 days of operation ("early" mortality). The 207 survivors of ICR were last seen in our clinic at median age 15.8 years (range 1 month to 22 years). The follow-up period ranged from 1 month to 15.6 years (median 5.2 years). Information from the 5 years preceding the study were available for 132 patients. These patients are now 15.8-45.5 years old (median 22.2 years).

Data Collection

The chart of each patient was examined for 61 clinical variables: three identifying variables (sex, age and cause of death); 12 variables from the history and physical examination (e.g., exercise tolerance, description of outflow murmur, presence of cyanosis); eight routine laboratory variables (e.g., hemoglobin, ECG rhythm); 10 echocardiographic variables; and 28 cardiac catheterization variables. Because the chest roentgenograms were unavailable, the radiologic reports were used to determine cardiac size and pulmonary vascularity. Any degree of cardiac enlargement was classified as cardiomegaly.

In addition, a questionnaire was sent to all patients believed to be alive. This provided data on occupa-
tion, education, marital status, children and miscarriages. Ninety-five of 192 patients (49%) returned the questionnaire.

Since this was a retrospective study, not all variables could be obtained for each patient. Recent follow-up information was requested from the referring physicians. Because the follow-up process did not involve selection of patients, the results should not be biased.

Cardiac Catheterization

Cardiac catheterization after ICR of TOF has been recommended for patients at Texas Children’s Hospital since 1971. Of 207 survivors, 120 underwent cardiac catheterization 3 months to 15 years postoperatively (median 1.1 years). The hemodynamic result was considered excellent if the right ventricular systolic pressure (RVSP) was < 40 mm Hg and there was either no residual left-to-right shunt or a small shunt with a pulmonary-to-systemic flow ratio (Qp/Qs) < 1.3:1. The result was considered good if the RVSP was 40–60 mm Hg or there was a residual left-to-right shunt with a Qp/Qs 1.3–1.5:1. A result was classified as poor if the RVSP was > 60 mm Hg, or the Qp/Qs was > 1.5:1, or the distal pulmonary artery systolic pressure was > 40 mm Hg.

Statistical Methods

We used the t-test to compare arithmetic means, and the chi-square test to compare difference in sizes of groups. To determine survival curves, life table analysis was used.6 Comparison between the survival scores was made using Z scores.6

Results

Operative History and Early Mortality

Of the 233 patients who underwent ICR, 140 had a prior systemic-to-pulmonary artery anastomosis and 93 had primary ICR (fig. 1). Because there was no significant difference in comparing the types of anastomoses with respect to age at the time of palliation, the mean ages for all types are presented in figure 1. Similarly, as there was no significant difference between the palliated and nonpalliated groups with respect to age at ICR or revision of ICR, only the mean ages are given.

There were 26 early deaths (11.1%). There was no relationship between early death and either the presence of prior anastomosis or the age at ICR (table 1). However, no patient younger than 1 year old had ICR.

Early Survivors

There were 207 survivors from ICR and the postoperative course of these patients is given below.

Associated Diseases

Fatal bacterial endocarditis occurred in one patient 1 year after ICR. Nine other patients (4% of 233) recovered from bacterial endocarditis before ICR.

Three patients (1.5%) had cerebrovascular accidents (CVA) before ICR and three patients had CVAs immediately after ICR. No patient had a CVA more than 30 days after ICR. Five of these six patients with CVA were alive at a mean period of 10 years after the CVA (one died of bacterial endocarditis). Of the five, two had residual hemiparesis and three had residual seizure disorders.

Exercise Tolerance

Eighty-one percent of the patients (168 of 207) gave a history of unlimited exercise tolerance (New York Heart Association class I) and 14% (28 of 207) were class II. Ten of the 207 patients (5%) were in class III and one patient was in class IV. All of the 11 patients in class III or IV underwent cardiac catheterization,
Examination of the heart demonstrated no malformations. However, six women were infertile. Among the 95 patients who responded to a questionnaire, 44 (45%) were enrolled as students. Thirty-seven (73%) of the 51 who were no longer students had attended college; 20 of these 37 patients had graduated from college and six had earned advanced professional degrees. All who were not homemakers were employed. Occupations held by more than two patients each included: banker, construction worker, computer programmer, music instructor, and truck driver.

Eighteen men and 10 women had married. Two marriages ended in divorce. No patient reported having infertility. None of the female patients or the wives of the male patients had miscarriages. Nine men and six women have one or more children. None of the 26 progeny had congenital heart disease.

Psychosocial Data

Among the 95 patients who responded to a questionnaire, 44 (45%) were enrolled as students. Thirty-seven (73%) of the 51 who were no longer students had attended college; 20 of these 37 patients had graduated from college and six had earned advanced professional degrees. All who were not homemakers were employed. Occupations held by more than two patients each included: banker, construction worker, computer programmer, music instructor, and truck driver.

Eighteen men and 10 women had married. Two marriages ended in divorce. No patient reported having infertility. None of the female patients or the wives of the male patients had miscarriages. Nine men and six women have one or more children. None of the 26 progeny had congenital heart disease.

Physical Examination

At follow-up examination averaging 5 years after the operation, the height and weight of each patient was normal. All patients had a systolic crescendo-decrescendo murmur at the third left intercostal space. Although this murmur tended to increase in length as the right ventricular-to-main pulmonary artery gradient increased, 58% of those patients (17 of 29) with a gradient > 25 mm Hg had a short systolic murmur labeled as "insignificant." Eight patients were reported to have a pansystolic plateau murmur of a residual ventricular septal defect (VSD). These eight patients did have a VSD proven at cardiac catheterization, as did 11 other patients in whom a VSD was unsuspected on auscultation of the heart by one of the staff cardiologists. Five of these 11 patients had a Qp/Qs ≥ 1.5:1. There was a low-pitched, early diastolic decrescendo murmur of pulmonary insufficiency in 175 patients (85%).

Chest Roentgenogram

Only 85 of the patients (41%) had a normal chest roentgenogram. There was diffuse cardiomegaly in 87 patients. Cardiomegaly was noted in 20 of 39 patients (51%) with a poor hemodynamic result, but also in 31 of 81 (38%) of those with good or excellent hemodynamic results. There was no association of a pulmonary insufficiency (PI) murmur with cardiomegaly, as cardiomegaly was present in 34% of those

### Table 1. Cardiac-Related Mortality After Intracardiac Repair (ICR)

<table>
<thead>
<tr>
<th>Operative group</th>
<th>Early*</th>
<th>Late†</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Deaths/operations</td>
<td>Mortality (%)</td>
<td>Mean age (in years) at death</td>
</tr>
<tr>
<td>Previous</td>
<td>10/79</td>
<td>12.7</td>
<td>16.4</td>
</tr>
<tr>
<td>Blalock-Taussig§</td>
<td>6/60</td>
<td>10.0</td>
<td>11.5</td>
</tr>
<tr>
<td>Potts</td>
<td>5/51</td>
<td>9.8</td>
<td>10.8</td>
</tr>
</tbody>
</table>

Numbers in parentheses indicate range.
*Within 30 days of operation.
†Excludes two patients who died in accidents and one who committed suicide.
‡Includes patients who had revision of ICR. Revision includes closure of ventricular septal defect, or revision of residual infundibular, valvular or pulmonary branch stenosis.
§Blalock-Taussig group includes all patients with Blalock-Taussig except those who had Blalock-Taussig and Potts. Potts group includes all patients with Potts, including those with another anastomosis in addition to Potts.
¶Rev. ICR early mortality = 7.1%; late mortality = 23.1%.
Abbreviations: BE = bacterial endocarditis; CAVB = complete atrioventricular block; CHF = congestive heart failure; CO = cardiac output; PAH = pulmonary arterial hypertension.
Table 2. Exercise Tolerance and Cardiac Catheterization Result

<table>
<thead>
<tr>
<th>Catheterization result</th>
<th>NYHA Class III, IV</th>
<th>Class II</th>
<th>Class I</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Poor</td>
<td>6</td>
<td>4</td>
<td>29</td>
<td>39</td>
</tr>
<tr>
<td>RVP &gt; 60 mm Hg</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>and/or VSD</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(Qp/Qs &gt; 1.5:1)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>and/or PAH</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Good</td>
<td>3</td>
<td>3</td>
<td>15</td>
<td>21</td>
</tr>
<tr>
<td>RVP 40-59 mm Hg</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>and/or VSD</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(Qp/Qs &lt; 1.5:1)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No PAH</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Excellent</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RVP &lt; 40 mm Hg</td>
<td>2</td>
<td>9</td>
<td>49</td>
<td>60</td>
</tr>
<tr>
<td>No VSD or</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(Qp/Qs &lt; 1.3:1)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No PAH</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>11</td>
<td>16</td>
<td>93</td>
<td>120</td>
</tr>
</tbody>
</table>

Abbreviations: NYHA = New York Heart Association Function Classification; PAH = pulmonary arterial hypertension; Qp/Qs = pulmonary-to-systemic flow ratio; RVP = right ventricular systolic pressure; VSD = ventricular septal defect.

without a PI murmur and in 50% of those with a PI murmur. Ten patients had "increased pulmonary vascular markings." In none of these was there a left-to-right shunt or elevated left ventricular end-diastolic pressure to account for the findings.

Electrocardiogram

Conduction Disturbances. Complete right bundle branch block (CRBBB) was found in 183 of 203 patients (88%). Seventeen of the 183 patients (9.2%) had additional left-axis deviation ("left anterior hemiblock"). No patient had left-axis deviation without CRBBB. Of the total group of 203 patients, 37 had first-degree AV block; eight patients had both first-degree AV block and "left anterior hemiblock." No patient had either transient or permanent complete AV block.

Rhythm Disturbances. We found ventricular premature depolarizations (VPDs) on routine 15-lead ECGs taken at rest in 21 of 207 patients (11.2%). The VPDs were unifocal in 19 patients and multifocal in two. Two patients age 26 and 30 years had VPDs before ICR. In the other 19, the VPDs were noted 2 months to 10 years after ICR. Only five patients had VPDs noted on ECGs recorded immediately postoperatively. None of these patients had VPDs recorded subsequently. Dynamic 24-hour ECGs were not done on these patients.

Postoperative treadmill examination is an important provocative test. Because this was not routine, only 17 of 207 had a treadmill exercise test. Two of these 17 patients were among the group who had VPDs before the test. With exercise, the VPD frequency increased in one patient and decreased in the other.

In none of the patients were the VPDs induced by exercise.

The most important association with VPDs on the resting ECG was an elevated right ventricular systolic or end-diastolic pressure. Seventeen of 21 patients with VPDs had cardiac catheterization and 15 of 17 had a right ventricular systolic pressure &gt; 60 mm Hg, as well as a right ventricular end-diastolic pressure &gt; 7 mm Hg.

The relationship of VPDs to right ventricular systolic pressure &gt; 60 mm Hg was statistically significant (chi square = 35.0, p &lt; 0.001), as well as the relationship of VPDs to right ventricular end-diastolic pressure &gt; 7 mm Hg (chi square = 17.3, p &lt; 0.001). Each of the two patients with a normal right ventricular systolic pressure had an elevated end-diastolic pressure of 8 mm Hg. VPDs were found in 10 of 30 patients who had pulmonary stenosis causing a right ventricular systolic pressure &gt; 60 mm Hg. VPDs were also found in four of five patients with pulmonary vascular obstructive disease (distal pulmonary systolic pressure &gt; 40 mm Hg without a left-to-right shunt or increased left atrial pressure).

There was no association between VPDs and the presence or severity of pulmonary insufficiency, or with cardiomegaly on chest roentgenogram in patients whose right ventricular pressure was &lt; 60 mm Hg.

Echocardiogram

Echocardiograms were performed on 31 patients. We compared the measurements with the normal measurements for our laboratory. A measurement more than 2 standard deviations from the mean was considered abnormal. Abnormalities included an enlarged aorta in 25 patients, an enlarged left atrium in 11, a small left ventricular end-diastolic dimension in eight, and an enlarged right ventricular end-diastolic dimension in 22; 10 patients had reduced septal excursion or paradoxical septal motion and 16 had mitral valve prolapse. The ratio of left ventricular pre-ejection period to ejection time was abnormally prolonged in six of 12 patients in whom the measurement was possible. The left ventricular shortening fraction was decreased in seven of the 31. Each of these seven patients had decreased septal excursion. Because only 20 of 31 patients had cardiac catheterization, we did not attempt to correlate these findings.

The echocardiograms of this small group of patients did reveal many abnormalities; the most serious were the abnormalities of left ventricular size and function. Longer follow-up will be required to evaluate their significance.

Cardiac Catheterization

We found that 60 of 120 patients (50%) who had cardiac catheterization had an excellent result (see Methods and table 3), 21 patients (18%) had a good result, and 39 patients (32%) had a poor result. The age at ICR was not significantly different between those with a good or excellent result (10.4 years) and those with a poor result (11.8 years). The presence of a
prior anastomosis did not significantly affect the hemodynamic result (table 3).

**Residual Right Ventricular Outflow Gradient.** Forty percent of patients had a right ventricular systolic pressure ≥ 40 mm Hg with an outflow tract gradient such that the distal main pulmonary artery pressure was normal (figs. 2 and 3). In 47 of 50 patients the gradient was situated proximal to the midportion of the main pulmonary artery (at the infundibulum, valve, or proximal pulmonary outflow patch insertion). Twenty-two of the 50 patients had a gradient distal to the mid-main pulmonary artery. It was impossible to ascertain by angiographic appearance or pressure withdrawal recording if the gradient was caused by unrelieved congenital proximal pulmonary branch stenosis or by obstruction at the site of the pulmonary artery outflow patch insertion into the pulmonary artery confluence.

Of the 38 patients with prior Potts anastomosis, only one had a kink at the anastomosis site causing obstruction of the left pulmonary artery. None of the 40 patients with a prior Blalock-Taussig anastomosis had pulmonary branch stenosis resulting from the anastomosis.

**Residual Left-to-Right Shunt.** There was a residual VSD in 19 of 120 patients; in nine (5.9%) the Qp/Qs was ≥ 1.5:1. In each of these patients, the right ventricular systolic pressure was > 40 mm Hg, but in only two patients was the distal pulmonary artery pressure elevated. (The remaining six patients had coexistent anatomic obstruction to right ventricular outflow.)

A residual insignificant left-to-right shunt through the anastomosis (Qp/Qs < 1.5:1) was present in five of the 37 patients (13.5%) who had a prior Potts procedure, and in three of 40 (7.5%) with a prior Blalock-Taussig anastomosis.

**Distal Pulmonary Arterial Hypertension.** Eight of the 120 patients (6.7%) had pulmonary arterial hypertension with a distal pulmonary artery branch systolic pressure ≥ 40 mm Hg (fig. 3). Two of these patients had a VSD with a large residual left-to-right shunt. One had left ventricular failure with a left ventricular end-diastolic pressure of 30 mm Hg and the other five had pulmonary vascular obstructive disease. Each of these five had had a surgical anastomosis more than 10 years before ICR; four had had a Potts and one a Blalock-Taussig anastomosis.

**Late Deaths**

The total number of late deaths is uncertain since 60 of 207 patients have been lost to follow-up. Therefore, the 6% (12 of 203) late cardiac-related death figure is a minimum. These deaths occurred in four ways:

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### Table 3. Results of Cardiac Catheterization After Repair of Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Surgical group</th>
<th>Criteria for operative result</th>
<th>Operative result</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Excellent</td>
<td>Good</td>
<td>Poor</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>RVP &lt; 40 mm Hg</td>
<td>RVP 40-59 mm Hg and/or VSD</td>
<td>RVP ≥ 60 mm Hg and/or VSD</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>No VSD or</td>
<td>(Qp/Qs &lt; 1.5:1)</td>
<td>(Qp/Qs ≥ 1.5:1)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>No PAH</td>
<td>No PAH</td>
<td>No PAH</td>
<td></td>
</tr>
<tr>
<td>Blalock-Taussig before ICR</td>
<td>RVP</td>
<td>2</td>
<td>1</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td></td>
<td>VSD</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>PAH</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>RVP + VSD</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>n = 40†</td>
<td>Total</td>
<td>23/40 = 58%</td>
<td>4/40 = 10%</td>
<td>13/40 = 32%</td>
<td></td>
</tr>
<tr>
<td>Potts before ICR</td>
<td>RVP</td>
<td>6</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>VSD</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>PAH</td>
<td>0</td>
<td>4</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>RVP + VSD</td>
<td>0</td>
<td>3</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>n = 38</td>
<td>Total</td>
<td>16/38 = 43%</td>
<td>7/38 = 18%</td>
<td>15/38 = 39%</td>
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</tr>
<tr>
<td>Primary ICR</td>
<td>RVP</td>
<td>10</td>
<td>8</td>
<td>1</td>
<td></td>
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<tr>
<td></td>
<td>VSD</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>PAH</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>RVP + VSD</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>n = 42</td>
<td>Total</td>
<td>21/42 = 50%</td>
<td>10/42 = 24%</td>
<td>11/42 = 26%</td>
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<tr>
<td>All ICR</td>
<td>RVP</td>
<td>18</td>
<td>25</td>
<td>1</td>
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<td></td>
<td>VSD</td>
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<td></td>
<td>PAH</td>
<td>0</td>
<td>6</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td></td>
<td>RVP + VSD</td>
<td>1</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>n = 120</td>
<td>Total</td>
<td>60/120 = 50%</td>
<td>21/120 = 18%</td>
<td>39/120 = 32%</td>
<td></td>
</tr>
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</table>

*Criteria for operative result (good and poor result): RVP = right ventricular systolic pressure; VSD = ventricular septal defect with pulmonary-to-systemic flow ratio as indicated; PAH = pulmonary arterial hypertension (distal pulmonary artery pressure ≥ 40 mm Hg); RVP + VSD = elevated right ventricular pressure and ventricular septal defect (designated as poor result group if either RVP or VSD qualified for that group).†Numbers in columns represent number of patients in each class.*

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**TABLE 3. Results of Cardiac Catheterization After Repair of Tetralogy of Fallot**
Number of Patients (total N = 120)

FIGURE 2. Histogram of number of patients at each level of right ventricular systolic pressure in our 120 patients who underwent cardiac catheterization after intracardiac repair of tetralogy of Fallot. In three of the eight patients with a hemodynamically significant ventricular septal defect (VSD), the elevated right ventricular (RV) pressure was caused by the VSD rather than by RV outflow obstruction. Qp/Qs = pulmonary-systemic flow ratio; PAH = pulmonary arterial hypertension.

1) suddenly and unexpectedly (eight of 12), 2) from congestive heart failure associated with either a residual VSD or pulmonary arterial hypertension (two of 12), 3) from bacterial endocarditis (one of 12), and 4) at reoperation for residual defects (one of 12). Two additional patients died in automobile accidents and a third committed suicide. Except for the patient who died from bacterial endocarditis, we found that 10 of the other 11 late cardiac-related deaths had residual hemodynamic abnormality. Among the eight patients who died suddenly, seven had had cardiac catheterization. In each of the seven, right ventricular systolic pressure was $\geq$ 70 mm Hg. Three of these patients had pulmonary vascular obstructive disease and four of

Number of Patients (total N = 120)

FIGURE 3. Histogram of patients at each level of main pulmonary artery (PA) systolic pressure. Eight patients had increased pulmonary artery systolic pressure ($> 40$ mm Hg) without demonstrable pulmonary artery branch stenosis: Five had pulmonary vascular obstructive disease, two had a large ventricular septal defect (VSD) and one had left ventricular (LV) failure. PVOD = pulmonary vascular obstructive disease.
seven had anatomic pulmonary outflow obstruction. One-third (seven of 21) of those patients with a right ventricular systolic pressure $\geq 70$ mm Hg died suddenly.

Two additional patients who had not had postoperative cardiac catheterization died from congestive heart failure. Each had clinical evidence of a poor result. One had the clinical and radiographic findings of a large residual VSD and the other had had a prior Potts anastomosis with a main pulmonary artery systolic pressure of 80 mm Hg before ICR. The occurrence of late death was not statistically related either to age at operation or prior anastomosis.

One further important association with late death was the presence of VPDs. Eight of 21 patients (38%) with VPDs died 3 months to 8 years (mean 3.4 years) after ICR. All eight patients who died suddenly had VPDs before their death. None of these patients had either left anterior hemiblock, first-degree AV block, or complete AV block. Of the patients who experienced sudden death, half died during strenuous exercise. In addition, seven of the eight had had cardiac catheterization and in each, the right ventricular systolic pressure was $\geq 70$ mm Hg and the right ventricular end-diastolic pressure was $> 7$ mm Hg (table 4).

Longevity

According to our actuarial data, approximately 90% of patients who had ICR of TOF lived to be 15 years of age, and we estimate that 70% will live to be 30 years old. We have not gathered data on enough patients to make accurate statements about longevity beyond 30 years of age.

Although it is possible that even patients with excellent surgical results will have decreased survival, we found that if a patient survives ICR, his or her longevity depends upon the surgical result (fig. 4). Of 120 patients who had cardiac catheterization after ICR, 97% of those who had a good or excellent hemodynamic result were alive both 5 and 10 years after the operation, while 91% of those with a poor result lived 5 years and only 74% of those with a poor result lived 10 years. Using estimates from all our patients (fig. 5), approximately 1% of survivors of ICR died each year for the first 10 years. If a patient survived longer than 10 years after ICR, the mortality decreased. All 97 of our patients who survived at least 10 years after ICR are still alive.

Discussion

In this study of patients with TOF born more than 15 years ago, we estimate that well over half will survive to the fourth decade. If a patient with TOF is born today, he will most likely not have the same management, or the same prognosis as the patients in this study. Current reports of early mortality for ICR have been as low as 0–5%, significantly lower than our early mortality of 11.1%. In addition, as the operation is performed on younger infants, the hemodynamic results are improving; we have recently performed cardiac catheterization on 16 children, each of whom weighed less than 10 kg at ICR of TOF. Only one of these infants (6%) had a poor result. In the present study of patients who were older at the time of ICR (10.7 years), 32% of the patients had a poor result.

For the next 5–10 years, however, patients such as those in this study will be seen by the internist. Most of the patients are asymptomatic, but 8–34% may have a poor operative result.

Prediction of a Poor Surgical Result

Because a poor operative result increases a patient's risk of dying, clinical identification of these patients would be helpful.

The normal exercise tolerance displayed by 80–95% of patients after ICR does not necessarily imply a good hemodynamic result. However, exercise intolerance after ICR is associated with a poor result. Eighty-six percent of Rocchini's patients with symptoms of congestive heart failure had a residual VSD. Nineteen of our 11 patients with exercise intolerance after ICR had a poor result.

The data from our physical examination was less helpful in predicting the postoperative result. Fifty-eight percent of patients with significant right ventricular outflow obstruction had a murmur judged to be benign. The pansystolic murmur of residual VSD was helpful when present, but it was not detected as a separate murmur in 63% of the patients with large residual VSDs.

The chest roentgenogram was the least helpful. The frequently observed residual cardiomegaly did not

| Table 4. Mortality Related to Presence of Ventricular Premature Depolarizations (VPDs) and to Right Ventricular Pressure |
|---------------------------------------------------------------|---------------------------------------------------------------|
| Condition                                      | Mortality                  | Condition                                      | Mortality                  |
|----------------------------------------------------------------------------------------------------------------|
| VPD                                             | 8/21 (38%)                 | No VPD                                       | 4/179 (2%)                 |
| RVSP $\geq 60$ mm Hg                            | 8/37 (22%)                 | RVSP $< 60$ mm Hg                            | 1/83 (1%)                  |
| RVSP $\geq 70$ mm Hg                            | 7/22 (32%)                 | RVSP $< 70$ mm Hg                            | 2/98 (2%)                  |
| VPD + RVSP $\geq 60$ mm Hg                      | 8/15 (53%)                 | No VPD + RVSP $> 60$ mm Hg                   | 0/22 (0%)                  |
| VPD + RVSP $\geq 70$ mm Hg                      | 7/11 (64%)                 | No VPD + RVSP $> 70$ mm Hg                   | 0/11 (0%)                  |

Abbreviation: RVSP = right ventricular systolic pressure.
correlate with hemodynamic abnormality. There is no known way to predict accurately by clinical means the hemodynamic result found at cardiac catheterization.

Relationship of VPDs and Right Ventricular Pressure to Late Deaths

The incidence of VPDs on routine ECG varied from 5–8% in patients after ICR of TOF.16-18 With exercise, especially in the postexercise recovery period, the incidence of VPDs has been reported to increase; in one study 23% of postoperative patients had VPDs.16 We have found that patients who have VPDs are likely to have associated high right ventricular systolic or end-diastolic pressure. VPDs in our patients indicated a poor surgical result. In several other reports, VPDs were observed in patients who later died suddenly and unexpectedly.16-18 Thirty-eight percent of our patients who had VPDs later died suddenly. VPDs were especially ominous in patients with pulmonary vascular obstructive disease. Because of the reported association of CRBBB-left anterior hemiblock pattern (with or
without first-degree AV block) with sudden death.\textsuperscript{17, 19} we examined all ECGs for these findings. None of these conduction disturbances was found in the patients who died.

In our series, therefore, it seems to be the presence of VPDs and resultant ventricular dysrythmias, rather than AV block, which are responsible for sudden death after ICR of TOF.

Recommendations

Since assessment based on clinical variables is unreliable in estimating the success of operation for TOF, cardiac catheterization should be performed postoperatively on all patients within 6–8 months of operation.

The patient with a good or excellent hemodynamic result (see above) may be followed by a cardiologist infrequently, and routine later catheterization may not be necessary.\textsuperscript{9}

If the hemodynamic result is poor and a residual surgically accessible defect is found, reoperation should be considered. If surgery is not attempted, the patient should be monitored frequently for dysrythmias, especially VPDs.

All postoperative patients should have, at the time of cardiac catheterization, a resting ECG, treadmill exercise test and ambulatory 24-hour electrocardiographic monitoring. If VPDs are not detected, an annual physical examination (not necessarily by a cardiologist) should suffice for screening purposes. If VPDs are detected at rest, the dysrythmias should be treated.\textsuperscript{20} It is not yet known if exercise-induced or frequent VPDs found on tape monitoring need to be treated.

Patients with a good or excellent hemodynamic result after intracardiac repair of tetralogy of Fallot are essentially normal. They have normal exercise tolerance, normal intelligence and normal fertility. Their eventual longevity must await results of continuing follow-up.

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