Postoperative haemodynamics in tetralogy of Fallot
A study of 132 children

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SUMMARY Our study was undertaken to determine the type, incidence, and functional significance of residual anomalies in patients who have undergone corrective repair for tetralogy of Fallot. We reviewed data from cardiac catheterisations performed on 132 survivors.

A significant residual ventricular septal defect was present in only 12 patients. Resting right ventricular systolic pressure was less than 80 mmHg in 100 patients and ranged from 80 to 150 mmHg in the other 32 patients. Thirty-five patients were studied both at rest and during supine exercise. In most patients, the relation between oxygen consumption and cardiac output was normal during exercise. The stroke index and right ventricular end-diastolic pressure at rest and on exercise were compared in 34 patients. Seventeen showed a normal response to exercise. In the other 17 patients, right ventricular end-diastolic pressure rose on exercise; in 5 of these the stroke index fell during exercise, indicating abnormal myocardial response.

Our studies indicate the frequent occurrence of residual abnormalities, even in patients who appear asymptomatic, after total correction of tetralogy of Fallot.

Corrective operations for tetralogy of Fallot were first performed in 1954. Despite extensive experience at a number of centres, the postoperative course of these patients has been described principally in terms of clinical features. Until recently, little information has been available concerning postoperative haemodynamic changes.

We have performed postoperative cardiac catheterisations on 132 survivors of corrective operation for tetralogy of Fallot. This report describes the catheterisation data and the incidence and severity of residual abnormalities. Results of studies made during supine exercise are also included.

Methods

For this report, we define tetralogy of Fallot as a large ventricular septal defect with equal right and left ventricular systolic pressures, coexistent infundibular pulmonary stenosis, normal or low pulmonary arterial pressure, and a systemic arterial oxygen saturation less than 90 per cent. The aorta arose from the left ventricle in all patients, and none had double outlet right ventricle and coexistent pulmonary stenosis. In 118 patients, the pulmonary valve was abnormal, and many of these had a small pulmonary annulus. In the other 14, the pulmonary valve and annulus were normal, and the pulmonary stenosis was related to infundibular stenosis, frequently from anomalous muscle bundles.

One hundred and thirty-two patients with tetralogy of Fallot have been studied postoperatively by cardiac catheterisation. These 132 patients were from a group of 170 patients who survived corrective operation for tetralogy of Fallot. While we recommend postoperative cardiac catheterisation in all patients, 38 patients were not restudied by us, either because they have been lost to follow-up or because they are being followed by their own physician in an area distant from the University of Minnesota. Nineteen patients had had a previous Blalock-Taussig procedure, and 6 others had a Brock operation. None had had a previous total correction for tetralogy of Fallot.

The patients were catheterised in a resting, fasting state after premedication with morphine sulphate and phenobarbitone. Pressure and saturation data were obtained from the right side of the heart to

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detect the presence of a residual shunt or a pressure difference across the right ventricular outflow area. From the data, total pulmonary vascular resistance (TPVR) was calculated according to the formula: TPVR = mean pulmonary arterial pressure (mmHg) \[ \text{pulmonary blood flow (l/min)} \]

Thirty-five patients were studied both at rest and during supine submaximal exercise, using a variable resistance bicycle ergometer set at a work load designed to double the heart rate. The oxygen consumption was measured both at rest and during exercise, and cardiac output was determined by the Fick principle. Expired air was collected in a chain-driven Tissot tank and analysed by the Schonander method. Blood samples were drawn simultaneously from the brachial artery and the pulmonary artery for oxygen analysis by the method of Van Slyke. Pressure was recorded as the catheter was withdrawn through the pulmonary artery, the right ventricle, and the right atrium.

Results

Of the 132 patients, 11 were less than 4 years of age at the time of operation, 89 were 4 to 9 years of age, and the remaining 32 were between 10 and 16 years of age. Seventy-nine patients were restudied between the ages of 4 and 10 years, and 53 between 10 and 18 years of age. Most were studied one year after operation.

Resting Haemodynamics

Residual shunt

Studies showed no detectable ventricular septal defect in 106 patients; a residual communication was detected by oximetry in the other 26 (20%) patients (Table 1). A left-to-right shunt between 30 and 50 per cent was present in 14 patients, between 51 and 70 per cent in 7, and greater than 70 per cent in 2. A bidirectional shunt was present in the remaining 3 patients.

Table 1  Catheterisation data in 132 children after correction of tetralogy of Fallot

<table>
<thead>
<tr>
<th>Right ventricular systolic pressure (mmHg)</th>
<th>Residual shunt</th>
<th>Left-to-right (%)</th>
<th>Bi-directional Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>None</td>
<td>30-50</td>
<td>51-70</td>
</tr>
<tr>
<td>&lt; 40</td>
<td>36</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>40-59</td>
<td>36</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>60-79</td>
<td>16</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>&gt; 80</td>
<td>16</td>
<td>5</td>
<td>6</td>
</tr>
<tr>
<td>Total</td>
<td>106</td>
<td>14</td>
<td>7</td>
</tr>
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</table>
systolic pressure more than 80 mmHg, up to 150 mmHg. In only one was the right ventricular systolic pressure above systemic level. Sixteen of these patients had no residual shunt. In 14 of the 16 patients, pulmonary arterial systolic pressure was 30 mmHg or less, and in these patients the raised right ventricular systolic pressure was the result of right ventricular outflow obstruction, usually at pulmonary valve level, but in 2 patients in the infundibulum. In the other 2 patients without a residual shunt, the right ventricular systolic pressures were 85 and 90 mmHg, and pulmonary arterial systolic pressures were 40 and 90 mmHg, respectively (Table 2, cases 1 and 12). Both had a right ventricular outflow patch and pulmonary regurgitation, but in case 12 there was also peripheral pulmonary arterial stenosis.

In the other 16 patients a residual shunt was present; in 8 the shunt was greater than 50 per cent, and in 3 others it was bidirectional. In 8 patients the pulmonary arterial systolic pressure was greater than 40 mmHg (Table 2, cases 4, 5, 7, 8, 9, 10, 11, 13). Though a gradient was measured in the outflow tract, the increased pulmonary arterial pressure made an important contribution to the raised right ventricular systolic pressure. In these 8 patients, the increased pulmonary arterial systolic pressure was associated with increased right ventricular stroke volume, as a result of pulmonary regurgitation and the augmented pulmonary blood flow from a residual shunt. In 2 (cases 5 and 11) this was accentuated by an occluded left pulmonary artery, resulting from bacterial endocarditis in one, and by congenital peripheral pulmonary arterial stenosis in another (case 13).

In the remaining 10 patients with a residual shunt, the pulmonary arterial pressure was less than 30 mmHg in 5, and between 30 and 34 mmHg in the remaining 5. The gradient was in the right ventricular outflow tract in each. The raised right ventricular systolic pressure in these 10 patients was related to a combination of the obstruction and left-to-right shunt.

Thus, 33 of our catheterised patients (25%) had a shunt greater than 50 per cent, or a right ventricular systolic pressure greater than 80 mmHg. The result of the initial corrective operation in these patients was considered unsatisfactory, and a second operation has been performed in 20 to correct residual abnormalities.

### Pulmonary arterial pressure

Total pulmonary vascular resistance was normal (less than 3 units) in all but 2 patients. The peak pulmonary arterial systolic pressure was greater than 40 mmHg in 13 patients (Table 2). Pulmonary valvular regurgitation was diagnosed in each of these patients on the basis of auscultatory findings; it was associated with an outflow patch across the pulmonary annulus in 9. Of the 13 patients, 8 had a residual left-to-right shunt and 1 (case 9) a bidirectional shunt, and 4 had either an occlusion or a stenosis of a pulmonary artery.

Of the remaining 119 patients with pulmonary arterial systolic pressure less than 40 mmHg, none had an obstruction or a stenosis of a pulmonary artery. The pulmonary arterial systolic pressure was between 30 and 40 mmHg in 26 of these patients, 24 of whom had auscultatory evidence of pulmonary regurgitation. The pulmonary arterial systolic pressure was less than 30 mmHg in the remaining 96 patients; 53 had clinical pulmonary regurgitation and 43 did not.

Forty-two of the patients with a murmur of pulmonary regurgitation and 11 patients without pulmonary regurgitation had a pulmonary arterial pulse pressure greater than 20 mmHg.

We found no relation between the right ventri-
Pulmonary blood flow
At the time of cardiac catheterisation, the pulmonary flow index was less than 3.0 1/min per m² in 90 patients, between 3.0 and 4.5 1/min per m² in 15, and greater than 4.5 1/min per m² in 63. We compared the pulmonary flow index with the level of right ventricular systolic pressure. There was a tendency for patients with a higher pulmonary flow index to have a higher right ventricular systolic pressure, but there were extreme variations because of the varying relative contributions of outflow obstruction and volume of pulmonary blood flow to the height of the right ventricular systolic pressure.

Right ventricular end-diastolic pressure
In patients without pulmonary regurgitation, the right ventricular end-diastolic pressure ranged from 2 to 15 mmHg, mean 6.4 mmHg. Each patient in this group with a right ventricular end-diastolic pressure greater than 10 mmHg also had a right ventricular systolic pressure greater than 90 mmHg. In patients with pulmonary regurgitation, the right ventricular end-diastolic pressure ranged from 4 to 20 mmHg, mean 9 mmHg.

Right ventricular end-diastolic pressure was higher in patients in whom a pulmonary valvotomy was done or an outflow patch was placed (mean 8 mmHg and 9.8 mmHg, respectively) than in those patients who had neither an outflow patch nor a pulmonary valvotomy (mean of 5.4 mmHg), but in part this difference may be related to anatomical differences between those patients with an abnormal pulmonary valve and annulus and those with infundibular stenosis and a normal pulmonary valve.

Exercise haemodynamics
Thirty-five patients were studied both at rest and during submaximal supine exercise. Cardiac output was measured by the Fick principle. The 35 patients ranged in age from 6 to 16 years. Twenty-six were studied one year after operation, and the other 9 at intervals up to 4 years. Their heart rates ranged from 60 to 105/minute at rest and from 110 to 190/minute on exercise. No patients developed aarrhythmia during exercise.

The cardiac index in each of the 35 patients increased with exercise. The cardiac index during exercise was compared with the corresponding oxygen consumption (Fig. 1). Thirty-one patients showed a normal relation between these two measurements. The cardiac index of the other 4 patients was low in relation to oxygen consumption, indicating impaired cardiac function. Each of these 4 patients had clinical evidence of pulmonary regurgitation and a wide pulmonary artery pulse pressure. Severe residual right ventricular outflow obstruction was also present in 2 of the 4 patients. The third had a small residual ventricular septal defect only. The remaining patient had no residual defect, and his right ventricular systolic pressure was 45 mmHg.

The peak right ventricular systolic pressure increased on exercise in 34 patients (Fig. 2). At rest the right ventricular systolic pressure ranged from 34 to 105 mmHg; with exercise, the range increased to 38 to 140 mmHg. In the other patient, there was essentially no change.

On exercise, both the peak systolic and the mean systemic arterial pressures increased. In only one patient did right ventricular systolic pressure ris...
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Fig. 2 Simultaneous change in cardiac index and peak right ventricular systolic pressure from rest to exercise in 35 children after repair of tetralogy of Fallot. Observations on individual patients connected. ○ = rest; ● = exercise.

above systemic on exercise, and in this patient the peak systolic systemic arterial pressure increased from 110 to 130 mmHg while the right ventricular systolic pressure increased from 100 to 140 mmHg.

Both stroke index and right ventricular end-diastolic pressure were studied at rest and during exercise in 34 patients (Fig. 3). Seventeen patients showed a normal response to exercise. Their right ventricular end-diastolic pressure either stayed the same or decreased, while their stroke index showed either an increase or a slight decrease. Pulmonary regurgitation was present in only 4 of these patients. Only 2 had right ventricular systolic pressures greater than 50 mmHg.

The other 17 patients showed a major increase in right ventricular end-diastolic pressure. In 12 of these patients, stroke index either remained the same or increased. Stroke index fell in the remaining 5 patients. Pulmonary regurgitation was present in 15 of the patients in whom right ventricular end-diastolic pressure rose, and the right ventricular systolic pressure was greater than 50 mmHg in 9 of these patients. It is difficult to determine if abnormal responses to exercise were related to abnormalities of myocardial function or to the presence of residual structural abnormalities.

Discussion

Kirklin et al. (1965), Malm et al. (1966), Burnell et al. (1969), and Gotsman et al. (1969) have shown that repair of tetralogy of Fallot is feasible with a low operative mortality and excellent clinical results. Many, however, have failed to corroborate the clinical improvement with haemodynamic data. Our early experience indicated that residual abnormalities were frequently present after correction of tetralogy of Fallot. These anomalies were often observed in patients who were asymptomatic, and their presence could not have been detected without cardiac catheterisation.

In our haemodynamic studies and in those of others (Bristow et al., 1962; Lillehei et al., 1964; Kirklin et al., 1965; Malm et al., 1966; Burnell et al., 1969; Gotsman et al., 1969; Bristow et al., 1970; Jarmakani et al., 1972; Epstein et al., 1973; Ruzyllo et al., 1974; Finnegan et al., 1976), ventricular septal defect and right ventricular outflow obstruction have been identified as the major residual abnormalities. Residual ventricular septal defect has been found in 15 to 20 per cent of patients after corrective operation. In most patients, the shunts have been small and the pulmonary/systemic flow ratios less than 2:1 (Malm et al., 1966; Gotsman et al., 1969). In large series of patients, the incidence of residual
left-to-right shunts greater than 2:1 was 1 to 7 per cent (Malm et al., 1966; Gotsman et al., 1969; Ruzyllo et al., 1974). The incidence of residual shunts in our study was 20 per cent. Nine per cent of our patients had a large residual defect leading to either a left-to-right shunt greater than 2:1 or a bidirectional shunt. The usual location of the residual defect was along the superior-posterior margin of the patch. The incidence of the occurrence of residual ventricular septal defect in our patients has not changed from our early to our most recent surgical experience.

Residual right ventricular outflow tract obstruction occurs often in patients after repair of tetralogy of Fallot (Kirklin et al., 1965; Malm et al., 1966; Shah and Kidd, 1966). Residual stenosis is considered significant when the peak right ventricular systolic pressure is greater than 80 mmHg or the right ventricular/pulmonary arterial gradient is greater than 50 mmHg. The percentage of patients with severe residual stenosis varies from zero in small series (Burnell et al., 1969; Jarmakani et al., 1972; Ruzyllo et al., 1974) to 4 to 5 per cent in larger series (Kirklin et al., 1965; Gotsman et al., 1969; Ruzyllo et al., 1974). Resting peak right ventricular systolic pressure was greater than 80 mmHg in 24 per cent of our patients. The most common site of outflow tract stenosis was at the level of the pulmonary annulus. No difference in the level of right ventricular systolic pressure was found between the patients with a patch across the pulmonary annulus and those without such a patch. This fact may be related to a difference in the anatomy of the right ventricular outflow tract in patients requiring a patch. The presence of a residual shunt with increased pulmonary blood flow contributes to the raised right ventricular systolic pressure in some patients.

Pulmonary regurgitation frequently occurs after the reconstruction of the right ventricular outflow tract in patients with tetralogy of Fallot. The degree of pulmonary regurgitation is difficult to evaluate, but our patients seem to have tolerated it quite well, even the 4 patients with an obstructed pulmonary artery and raised pulmonary arterial pressure. Pulmonary regurgitation was present in over half of our patients and, as expected, was associated with a widened pulmonary arterial pulse pressure. These patients frequently had high pulmonary arterial systolic pressure that may have contributed to the raised right ventricular systolic pressure.

Malm et al. (1966), Shah and Kidd (1966), Bristow et al. (1970), Epstein et al. (1973), and Finnegan et al. (1976) have shown the resting and exercise cardiac output to be normal in patients after repair of tetralogy of Fallot. Shah and Kidd (1966) showed that an increase in cardiac output with exercise is related to normal changes in heart rate and stroke volume. Bristow et al. (1966) reported a normal relation between oxygen consumption and cardiac output. Epstein et al. (1973) studied patients in an upright position doing intensive exercise. The cardiac indices of these patients were low (6·1 ± 0·3 l/min per m²) compared with normal individuals (8·9 ± 0·3 l/min per m²). This occurred though these patients had no residual septal defect and only minimal outflow tract gradient. In 4 of our 35 patients studied during supine exercise, the cardiac index was abnormally low in relation to oxygen consumption.

Half of our 34 patients studied showed an abnormal response to exercise. Pulmonary regurgitation was more common in the 17 patients with an abnormal response, and the right ventricular systolic pressure was more frequently raised. However, there was no significant correlation between behaviour of right ventricular end-diastolic pressure and stroke index, and the presence of residual ventricular septal defect or outflow tract obstruction.

It is uncertain whether abnormal response of right ventricular end-diastolic pressure and stroke index reflects abnormal ventricular compliance or abnormal ventricular contractility. Studies of right ventricular volume would assist determination of right ventricular function. We noted, as did Bristow et al. (1970), that abnormalities in right ventricular function at rest were magnified during exercise, and that right ventricular end-diastolic pressure was raised to abnormal levels in several patients.

We did not routinely study left ventricular function in our patients, but intend to do so in the future, since abnormalities of left ventricular function were shown in other studies. Sunderland et al. (1972) have shown normal left ventricular ejection fraction in children who were operated upon before the age of 2 years, but, in contrast, Jarmakani et al. (1972) have shown that left ventricular ejection fraction is depressed in patients operated upon at an older age. Thus, postoperative cardiac function may be dependent on the age at which operation is done.

Because of the significant incidence and severity of residual abnormalities occurring in children after tetralogy of Fallot repair, many of whom are asymptomatic, we believe that every child should undergo postoperative cardiac catheterisation. The studies should include exercise haemodynamics and assessment of left ventricular function. Understanding of the postoperative results in tetralogy of Fallot will be improved with the development of methods to measure right ventricular volume.

We wish to acknowledge the various paediatric...
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cardiology staff and fellows who assisted and performed many of the cardiac catheterisations, and our cardiac surgeons who recognised the need for postoperative cardiac catheterisation and stressed the importance of such procedures to the parents of our patients.

References


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