Total Correction of Tetralogy of Fallot in Infancy

Postoperative Hemodynamic Evaluation

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SUMMARY
Total intracardiac repair of symptomatic tetralogy of Fallot was accomplished in twenty-nine infants under two years of age with a mortality of six per cent. All twenty-seven survivors are asymptomatic. Postoperative hemodynamic evaluation of 17 randomly selected children was performed at least 12 months following surgery. Pulmonary regurgitation was present in 12 of the 17 children but was well tolerated, with only minimal cardiac enlargement and in no case were there symptoms or important hemodynamic consequences. Growth of the pulmonary artery with respect to the aorta occurred. Normal left ventricular performance was indicated by normal ejection fraction and normal systemic arterial pressure. Intellectual and social development were indistinguishable from a “normal” group of randomly selected children as assessed by detailed psychological testing.

Additional Indexing Words:
Cardiac catheterization       Open heart surgery       Psychological testing

TOTAL intracardiac repair of tetralogy of Fallot has been associated with increasingly satisfactory clinical and hemodynamic results and significantly decreased morbidity and mortality. During recent years earlier total intracardiac repair for symptomatic children has been recommended and survival and complication rates have improved.

The progressive nature of the right ventricular outflow tract obstruction and the frequent poor general health of the older child and adult with severe tetralogy of Fallot have led us to pursue a policy of early correction when an infant with this condition becomes symptomatic (hypoxic spells and/or cyanosis). In addition, the higher incidence in older individuals of operative and postoperative complications from collateral circulation make early total correction desirable.

Major considerations in evaluating the success of total intracardiac repair include postoperative right ventricular performance following infundibulotomy and outflow tract reconstruction, pulmonary regurgitation and its effect on cardiac performance, left heart size and performance, and the physical and intellectual development of the child.

This report concerns the postoperative hemodynamic evaluation of seventeen children who underwent primary total correction for tetralogy of Fallot in infancy.

Clinical Material
Twenty-nine infants less than two years of age, selected for their favorable cardiac anatomy, underwent primary total correction for tetralogy of Fallot at the University of Oregon Medical School between 1964 and 1972. The criterion for selection was that the diameter of the pulmonary artery be at least one-third the diameter of the aorta. These infants were between six weeks and twenty months of age when the clinical diagnosis of tetralogy of Fallot was made, based on the presence of a pulmonary stenosis murmur, single second heart sound; heart size which was normal or small; normal or diminished pulmonary vascular markings; and electrocardiogram which demonstrated right ventricular hypertrophy. Cardiac catheterization was
performed because of the onset of hypoxic spells, continuous or intermittent cyanosis or rising hematocrit. In the majority of these infants, symptoms were present either when first seen or within nine months of their first clinic visit. At cardiac catheterization, tetralogy of Fallot was considered to be present when right ventricular, left ventricular and aortic systolic pressures were equal; infundibular obstruction was demonstrated in the right ventricle, with normal or low pulmonary artery pressure; and when there was aortic overriding of a large high membranous subcrystal ventricular septal defect. Aortic oxygen saturation during the cardiac catheterization ranged from 45 percent to 91 percent with a mean for the group of 81 percent. Hematocrit at the time of cardiac catheterization ranged from 38 percent to 61 percent with a mean for the group of 47 percent. Corrective surgery was performed within two weeks of the diagnostic study and in those patients who were experiencing hypoxic spells operation was performed as an emergency procedure.

Repair was performed utilizing total cardiopulmonary bypass. The surgical technique has been previously described. Two infants died following surgery, a mortality of six percent. Major postoperative complications consisted of right ventricular failure manifested by hepatic enlargement and neck vein distention, in three patients all of whom responded to medical therapy; need for temporary tracheostomy in one patient; and a cerebrovascular accident with complete recovery in one patient. All twenty-seven survivors are asymptomatic.

We report the postoperative hemodynamic evaluation of 17 randomly selected children for whom at least 12 months had elapsed since their surgery. There was no statistically significant difference in hematocrit, aortic saturation, right ventricular pressure or age and weight at surgery between these seventeen patients and the rest of the group of twenty-nine patients. These studies were performed an average of 26.6 months (12-55 months) following surgery. All seventeen had infundibulectomy performed and eight were considered at the time of surgery to require patch graft reconstruction of the right ventricular outflow tract. In three patients, the patch was extended through the annulus and in the remaining five, the patch was ventricular only. Three patients (one with a patch and two without) had additional pulmonary valvulotomy performed.

Postoperative Evaluation Methods

Before admission to hospital each child was seen by a medical psychologist (RGM). Depending upon age, intelligence was evaluated by means of the Cattell Infant Intelligence Scale, or the Stanford-Binet (form L-M). Social development was measured by administration of the Vineland Scale of Social Maturity to the patient's mother.

Right heart catheterization was performed under light sedation (meperidine - 25 mg, hydroxyzine - 6.25 mg, chlorpromazine - 6.25 mg/cc, at a dosage of 1 cc/20 to 30 pounds). Arterial pressure and oxygen content were obtained through a polyethylene catheter introduced into the brachial artery, or through a #19 thinwall needle introduced into the femoral artery. Cardiac output was measured by the Fick principle, using measured systemic and pulmonary artery oxygen contents and an assumed oxygen consumption of 140 ml/min/M2. Left-to-right shunting was detected by right heart oximetry, angiocardiography and the appearance times of inhaled hydrogen. Cineangiography was performed using meg-lumine diatrizoate, injecting into the right ventricle and filming in the antero-posterior, right anterior oblique, and left anterior oblique views. Left ventricle ejection fraction was measured by the one-plane cineangiographic technique as reported by Greene et al. and was assessed from the levogram phase of the right ventricular injection, filmed in the right anterior oblique view. Calculations were done on the Hewlett-Packard 9100 calculator with a 9107A digitizer.

All the pre and postoperative cineangiograms that were available (11 preoperative and 16 postoperative) were reviewed and the diameters of the pulmonary artery and aorta were measured. The diameter ratio was then squared to obtain the area ratio of the pulmonary artery to aorta. The diameter measurement was made during systole at a level approximately 0.5 cm above the annulus which passed through the sinuses to the wall of the artery. (Duplicate measurements were also made at a level 1-2 cm above the valve and these measurements were almost identical to the measurements made at the level 0.5 cm above the annulus and passing through the sinuses.) Because the level 0.5 cm above the annulus was easy to identify consistently in all cineangiograms and because it was felt that this level would be relatively uninfluenced by magnification and course of the great artery, all pulmonary artery and aorta diameter and area measurements refer to this level. In the preoperative cineangiograms, the diameter measurements were made in the AP or RAO projection from the right ventricular injection, with both pulmonary artery and aorta opacified in the same frame. In the postoperative cineangiograms the measurements were again made in the AP or RAO views, measuring the pulmonary artery from the right ventricular injection, and the aorta from the levogram phase.

Results

The age and weight distribution of the 29 infants undergoing primary total correction of tetralogy of Fallot are illustrated in figure 1. The average age was 13.5 months; the average weight 9.2 kg. The two patients who succumbed were 6 and 8 months, and 6 and 6.5 kg respectively. The 17 children undergoing postoperative hemodynamic evaluation, who constitute the basis of this report, are shown by the cross-hatched areas. They are distributed throughout the sample age and weight range.

Right Heart Measurements

Right atrial; right ventricular end-diastolic (RVEDP); right ventricular systolic; right ventricle to pulmonary artery systolic gradient and pulmonary artery pressures were the same for those with and without an outflow tract patch (table 1). There
was evidence of pulmonary regurgitation in 6 of the 9 patients without an outflow patch, compared to 6 of the 8 patients with an outflow patch. Though pulmonary regurgitation was associated with mild elevation of RVEDP, mean right atrial pressure was normal. All elevation of RVEDP greater than 5 mm Hg was associated with pulmonary regurgitation except for one patient who had an unexplained RVEDP of 7 mm Hg with RV systolic pressure of 26 mm Hg.

Table 1
Postoperative Hemodynamic Data, Right Heart Pressure Measurements

<table>
<thead>
<tr>
<th></th>
<th>Without outflow patch</th>
<th>With outflow patch</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Average (Range)</td>
<td>Average (Range)</td>
</tr>
<tr>
<td>Whole group</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Without pulmonary regurgitation</td>
<td>2.8 (0-8) NS</td>
<td>2.8 (1-5)</td>
</tr>
<tr>
<td>With pulmonary regurgitation</td>
<td>2.3 (0-5) NS</td>
<td>4.0 (3, 5)</td>
</tr>
<tr>
<td></td>
<td>2.8 (0-8) NS</td>
<td>2.3 (1-5)</td>
</tr>
<tr>
<td>Whole group</td>
<td>5.5 (2-10) NS</td>
<td>4.8 (2-10)</td>
</tr>
<tr>
<td>Without pulmonary regurgitation</td>
<td>3 patients</td>
<td>2 patients</td>
</tr>
<tr>
<td></td>
<td>4.3 (2-6)</td>
<td>6.0 (5, 7)</td>
</tr>
<tr>
<td>With pulmonary regurgitation</td>
<td>6 patients</td>
<td>6 patients</td>
</tr>
<tr>
<td>Whole group</td>
<td>6.2 (3-10) NS</td>
<td>4.5 (3-10)</td>
</tr>
<tr>
<td>Right ventricular end-diastolic pressure mm Hg</td>
<td>38.3 (25-73) NS</td>
<td>38.6 (26-55)</td>
</tr>
<tr>
<td>Whole group</td>
<td>18.7 (8-55) NS</td>
<td>18.4 (6-34)</td>
</tr>
<tr>
<td>Right ventricular systolic pressure mm Hg</td>
<td>13 (8-15) NS</td>
<td>12 (6-20)</td>
</tr>
<tr>
<td>Pulmonary artery mean pressure mm Hg</td>
<td>20/7 (5-11) NS</td>
<td>20/6 (3-10)</td>
</tr>
<tr>
<td>Pulmonary artery pressure systolic/diastolic mm Hg</td>
<td>.56 (.32-.84) NS</td>
<td>.52 (.36-.83)</td>
</tr>
<tr>
<td>RV/Ao intraoperative</td>
<td>.39 (.27-.86) NS</td>
<td>.38 (.23-.55)</td>
</tr>
</tbody>
</table>
The single patient with an excessive right ventricular systolic pressure (75 mm Hg) had severe residual right ventricular outflow tract obstruction demonstrated by right ventricular cineangiogram. This was the only patient whose RV/Ao pressure ratio had not fallen from the intraoperative measurement. Two patients had small residual ventricular septal defects, demonstrable only by the hydrogen technique. Two patients with ventricular outflow patches had angiographic evidence of minimal tricuspid regurgitation. One of these had mild hepatic enlargement.

**Left Ventricular Performance**

The parameters of left ventricular performance considered were systemic arterial systolic pressure; cardiac index; arteriovenous oxygen difference and left ventricle ejection fraction. There was no significant difference for these measurements between those patients with and those without an outflow patch (table 2).

Systemic arterial oxygen saturation averaged ninety-five percent. Three patients had arterial oxygen saturation of 93%, two had arterial oxygen saturation of 92% and one patient had arterial oxygen saturation of 90% (attributed to alveolar hypoventilation during sedation).

**Pulmonary Regurgitation**

Pulmonary regurgitation (table 3) was thought to be present if pulmonary artery diastolic pressure was within 0 to 2 mm of that in the right ventricle. Five patients had near equalization of diastolic pressure without a murmur of pulmonary incompetence; however, no patient had a pulmonary regurgitation.
incompetence murmur without corresponding near-equalization of pressures.

Of the nine patients without an outflow patch, pulmonary regurgitation was present in six, two of whom had undergone valvulotomy in addition to the infundibulotomy.

Of the eight patients with an outflow patch, six had pulmonary regurgitation. Of these six, in three the outflow patch extended through the annulus and in three the patch was limited to the right ventricle. Only one of this latter group had had pulmonic valvulotomy. Thus, the etiology of pulmonary regurgitation in those patients in whom the valve and annulus were undisturbed (4 without a patch and 2 with a patch) is unknown.

**Chest X-ray**

A normal chest X-ray (cardiothoracic ratio less than 0.50) was found in three children without an outflow patch. Mild cardiac enlargement (cardiothoracic ratio .50 to .59) was present in twelve children, six without a patch and six with a patch. Moderate cardiac enlargement (cardiothoracic ratio .60 to .70) was present in two children, both with outflow patches (table 3). Thus, those with an outflow patch had a significantly larger cardiothoracic ratio \( P < .01 \) than those without an outflow patch. Cardiac enlargement when present, however, was only mild.

**Cineangiographic Evaluation of Right Ventricular Abnormalities (table 3)**

Residual outflow constriction, present in 4 patients without a patch, was mild in all but one (the patient whose right ventricular pressure was 75 mm Hg). Five patients with an outflow patch had mild residual outflow constriction, mainly at the suture line of the patch to the ventricle. Mild bulging of the outflow tract was present in one patient without a patch and four with a patch. Thus, except for the patient with residual severe constriction, the right ventricular abnormalities demonstrated by cineangiogram were mild.

**Pulmonary Artery/Aorta Ratio**

Figure 2 illustrates the ratios of the diameter and the area of the pulmonary artery (PA) to the aorta (Ao) pre and postoperatively for the eight patients for whom pre and postoperative cineangiograms were available and in whom the patch, when used, did not extend through the annulus. For these eight patients pulmonary artery/aorta diameter ratio averaged 0.50 preoperatively, and had increased to an average of 0.84 postoperatively.

![Comparison of preoperative and postoperative ratios of the diameter (left) and area (right) of the pulmonary artery to the aorta.](image)

Since the cross sectional area is related to the square of the diameter, a pulmonary artery which is half the diameter of the aorta has only one quarter the cross sectional area. Hence, in figure 2 it is apparent that pre-operatively the cross sectional area of the pulmonary artery was only one quarter that of the aorta and that postoperatively it had increased to almost three-quarters.

Using all available pre and postoperative cineangiograms, there was no significant difference in the pre and postoperative PA/Ao ratios between those patients with and without outflow tract patches as listed in table 4. Increases had occurred in both groups postoperatively similar to those illustrated in figure 2.

**Intelligence and Growth**

The mean IQ was 100.93 and standard deviation ± 16.28, almost identical to the mean and sigma of

<table>
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<tr>
<th>Table 4</th>
<th>PA/Ao Diameter and Area Ratios Pre and Postoperatively</th>
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<tbody>
<tr>
<td></td>
<td>Without patch</td>
</tr>
<tr>
<td>PA/Ao diameter</td>
<td>mean</td>
</tr>
<tr>
<td></td>
<td>range (preop)</td>
</tr>
<tr>
<td>PA/Ao diameter</td>
<td>mean</td>
</tr>
<tr>
<td></td>
<td>range (postop)</td>
</tr>
<tr>
<td>PA/Ao area</td>
<td>mean</td>
</tr>
<tr>
<td></td>
<td>range (preop)</td>
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<tr>
<td>PA/Ao area</td>
<td>mean</td>
</tr>
<tr>
<td></td>
<td>range (postop)</td>
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the "normal" standardization sample of the Stanford-Binet. The range of IQ scores was from 75-125 with three individuals obtaining scores below IQ 85 (−1 standard deviation) and three above IQ 115 (+1 standard deviation). The mean score on the Vineland Scale of Social Maturity was 104.27 ± 13.28 with a range of 72-122.

Height and weight ranged from the tenth to the ninety-seventh percentile with the exception of two children whose heights were both at the third percentile while their weights were at the tenth percentile. Both these children came from families with small parents.

**Discussion**

The overall mortality of six percent in this group of symptomatic infants undergoing total repair of tetralogy of Fallot compares favorably with the combined mortality for shunt procedures and total correction in older individuals in our and other institutions.  

**Right Heart Performance**

Figure 3 compares postoperative right heart pressure measurements for the children in the current study (C), undergoing primary total correction between ages 2.5 to 23 months with those children undergoing total correction at an older age (Bristow et al. [A] and Burnell et al. [B]) in our institution. With respect to overall relief of RV outflow obstruction, RVEDP and right atrial mean pressure, the results were approximately equal for all three groups of patients.

Right atrial mean pressures for the group as a whole were low and though there was mild elevation of right ventricular end-diastolic pressure for those with pulmonary regurgitation this was not associated with elevated mean right atrial pressure.

With the exception of one patient, whose residual outflow constriction was severe, right ventricular abnormalities evaluated cineangiographically were mild.

Right heart pressure measurements and cineangiographic performance thus compare favorably with other reported postoperative, hemodynamic studies following total correction in later childhood. Furthermore, our experience also confirms the importance of adequate relief of pulmonary stenosis reported by others and the good hemodynamic result obtainable when outflow tract reconstruction is necessary.

Pulmonary regurgitation was present in 6 of the 9 patients without a patch and in 6 of the 8 patients with an outflow patch. In three children it was due to the extension of the outflow patch through the annulus, in three it was related to valvulotomy and in the remaining six it was related to infundibulectomy only. Symptomatically, hemodynamically, and by cardiothoracic ratio measurements, pulmonary regurgitation was well tolerated in all patients.

Bristow and coworkers performed serial cardiac catheterizations an average of 13 months postoperatively and again an average of 7 years postoperatively in a group of individuals who underwent total correction of tetralogy of Fallot in later childhood. They found no change in right ventricular systolic pressure and RV to PA gradient, little change in pulmonary regurgitation, and an overall fall in right atrial mean pressure between the two studies. These findings are encouraging and lead us to anticipate that the children reported in our study will continue to have satisfactory-to-excellent right heart hemodynamics.

**Left Ventricular Performance**

Recent reports of Jarmakani and coworkers indicate that left ventricular performance as
essed by a reduced ejection fraction in the presence of decreased afterload is depressed in children with tetralogy of Fallot who are cyanotic and remains depressed following total correction in childhood, with and without previous shunt procedures. In the children we report, who were corrected in infancy, the normal ejection fractions, in the presence of normal systemic arterial pressure, suggest normal left ventricular performance. Cardiac index and arteriovenous oxygen difference were also normal for the group. Our studies suggest the possibility that abnormalities of left ventricular performance that have been demonstrated in the older cyanotic child with tetralogy of Fallot and which continue to be present following shunting procedures and total correction\textsuperscript{14} may be avoided, halted or reversed when total correction is performed in infancy.

**Pulmonary Artery/Aorta Size**

Small pulmonary artery size relative to that of the aorta is an anatomic and probably a developmental aspect of tetralogy of Fallot. By comparing the diameter and area of the pulmonary artery to that of the aorta in the pre and postoperative cineangio-
grams, we have been able to demonstrate a postoperative increase in the size of the pulmonary artery with respect to the aorta.

Since in our hospital symptomatic tetralogy of Fallot is operated upon, we are not able to compare unoperated with operated children. Because of the not infrequent progressive development of collateral circulation\textsuperscript{10} and the occasional progression of outflow tract narrowing,\textsuperscript{6} it is probable that pulmonary artery growth in the older unoperated child with tetralogy of Fallot is not of the same magnitude as in these children who underwent total correction in infancy.

**Intelligence and Growth**

Intelligence may be difficult to assess accurately in small children with congenital heart disease. A recent report by Linde, Rasof and Dunn\textsuperscript{18} emphasizes that in developmental and intelligence tests, cyanotic children score lower than acyanotic children with congenital heart disease and children without congenital heart disease, and that this performance deficit persists throughout all age periods.

For the 17 patients in the current study, cyanosis when present was generally mild since total correction was commonly performed as the treatment for hypoxic “spells” or cyanosis. Uncorrected, it might be expected that some of these children would have developed intellectual deficit and growth failure or that they as a group would be distinguishable from a “normal” group by the occurrence of a lower mean score and a greater number of low scores (<1 standard deviation below the mean). For the whole group, the mean IQ, range and distribution of IQ scores are nearly identical to the “normal” standardization group. In regard to self-help, communication, locomotion, occupation, self-direction and socialization, these children have been shown to be developing at a rate comparable to that of a “normal” group of children. This would suggest that the parents perceive their children as sufficiently normal that they do not overprotect or overindulge them.

Height and weight were also representative of a group of normal children, with two children at the third percentile for height and one child above the ninety-seventh percentile for height. No child was below the tenth percentile for weight.

**Summary**

Twenty-nine symptomatic infants less than two years of age underwent primary total correction of tetralogy of Fallot, utilizing total cardiopulmonary bypass, between 1964 and 1972. The only criterion for selection was that the diameter of the pulmonary artery be at least one-third that of the aorta. Two infants (6\%) died as a result of surgery. The twenty-seven survivors are asymptomatic.

Seventeen randomly selected children underwent hemodynamic and angiographic study 12 to 55 months postoperatively. Eight of the 17 had patch graft reconstruction of the right ventricular outflow tract.

Two children had small residual ventricular septal defects, detectable only by the hydrogen inhalation technique; two had minor tricuspid insufficiency. One patient had important residual obstruction (RV systolic pressure 75 mm Hg).

Right heart measurements indicated an average RV pressure of 39 mm Hg with an average RV to PA peak systolic gradient of 19 mm Hg.

Pulmonary regurgitation was present in 6 of 8 with an outflow tract patch and in 6 of 9 with no outflow tract patch. In no case did pulmonary regurgitation produce symptomatic or important hemodynamic consequences.

In 8 patients without pulmonary annulus patching and in whom pre and postoperative angiograms were available for review, there was a change in the ratio of the area of the pulmonary artery to the area
of the aorta from .26 preoperatively to .71 postoperatively.

Ejection fraction and systemic arterial pressure were normal suggesting normal left ventricular performance.

Physical growth and intellectual and social development were indistinguishable from a randomly selected group of "normal" children.

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