Results of Routine Preoperative Coronary Angiography in Tetralogy of Fallot

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SUMMARY
In the surgical repair of tetralogy of Fallot, morbidity and mortality are increased by certain coronary anomalies, in particular, an anterior descending branch originating from the right coronary artery or a single coronary artery in which a large coronary branch runs across the pulmonary outflow tract. In a series of 94 patients with tetralogy of Fallot who underwent cardiac catheterization, coronary artery visualization was attempted routinely, most often by flush aortography using a venous catheter. Diagnostic coronary visualization was obtained in 84 patients (89%). In these, the incidence of recognized coronary anomalies was 5%: anterior descending from the right coronary artery in four patients (4%), and single left coronary in one patient (1%). In 195 autopsy cases of tetralogy, the incidence of coronary anomalies was also 5%. Routine preoperative demonstration of the coronary artery anatomy in tetralogy patients usually can be accomplished satisfactorily and conveniently by transvenous flush aortography.

Additional Indexing Words:
Flush coronary arteriography Anomalous coronary arteries Surgical complications in tetralogy repair

IN THE SURGICAL REPAIR of tetralogy of Fallot, the size and the site of the right ventriculotomy is crucial to the ease of closure of the ventricular septal defect and relief of the infundibular stenosis. One of the determinants of the ventricular incision is the distribution of the coronary arteries over the right ventricular myocardium. In patients with tetralogy, anomalous coronary artery distribution occurs with an incidence which has been variably estimated as between 2 and 9%.1, 2

The coronary anomalies of greatest concern are origin of the anterior descending coronary artery (AD) from the right coronary artery (RCA) and certain variations of single coronary artery branching (fig. 1). Although these coronary anomalies often can be recognized at the time of ventriculotomy, postoperative morbidity and mortality may occur following the inadvertent transsection of important branches which are obscured by overlying myocardium or epicardial adhesions.1, 2 Even when recognized, an anomalous vessel may hinder the surgical approach to total repair by limiting the ventricular incision.

In order to minimize complications in tetralogy repair resulting from anomalous coronary arteries, and to allow the planning of alternative surgical approaches, an angiographic evaluation of the coronary artery distribution was undertaken in 94 patients undergoing cardiac catheterization. The objectives of the study were to determine if diagnostic coronary artery visualization could easily be made a part of the preoperative angiocardio graphic study in tetralogy of Fallot, and to discover the incidence of coronary abnormalities in a large group of tetralogy patients not selected by surgery or autopsy.

Material and Methods
Between June 1, 1972, and June 1, 1974, 94 patients with tetralogy of Fallot (criteria of Van Praagh et al.3) underwent preoperative cardiac catheterization and angiography. In all cases, informed consent was obtained prior to the procedure. In each case an attempt was made to delineate

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the coronary artery distribution angiographically, in addition to the demonstration of the intracardiac and pulmonary artery anatomy. The age range of these patients was from 1 week to 31 years. Included in the group were eight patients who had already undergone surgical correction for tetralogy and who were being considered for reoperation because of incomplete repair.

Right ventricular angiocardiology was usually performed initially. If the coronary arteries were not diagnostically visualized on this ventricular injection, an attempt was made to manipulate an NIH (side hole only) transvenous catheter through the ventricular septal defect (VSD) into the aorta, where an injection of ¾ to 1 cc per kilogram of body weight of contrast material was made at the aortic valve level (fig. 2). When a catheter could not be manipulated across the VSD, a retrograde arterial catheterization was done and flush coronary angiography performed in the aortic valve with the same amount of contrast material (fig. 3).

Results

Angiography of the Coronary Arteries in Tetralogy of Fallot

The findings are summarized in tables 1 and 2. Nondiagnostic coronary visualization occurred in ten patients; seven were older individuals (14–25 years) and flush injections in their large aortas provided only faint coronary opacification; additionally, there were three equipment failures.

Diagnostic coronary artery visualization was obtained in 84 patients; in 65 of these both coronaries were seen, and in 19, only one coronary (usually the right) was visualized. The coronary artery distribution in these 84 patients was evident after a right ventricular injection in only five, and flush aortography was required for the remaining 79 patients. In 62 of

Table 1

<table>
<thead>
<tr>
<th>Patients studied</th>
<th>94</th>
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<tbody>
<tr>
<td>Nondiagnostic coronary visualization</td>
<td>10</td>
</tr>
<tr>
<td>Diagnostic coronary visualization</td>
<td>84</td>
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<tr>
<td>By right ventriculography only</td>
<td>5</td>
</tr>
<tr>
<td>By aortography</td>
<td>79</td>
</tr>
<tr>
<td>Via antegrade venous catheter</td>
<td>62</td>
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<tr>
<td>Via retrograde arterial catheter</td>
<td>17</td>
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</table>

Table 2

<table>
<thead>
<tr>
<th>Coronary Artery Anomalies in Tetralogy of Fallot</th>
</tr>
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<tr>
<td>84 diagnostic angiocardigrams</td>
</tr>
<tr>
<td>AD from RCA</td>
</tr>
<tr>
<td>Single left coronary artery</td>
</tr>
</tbody>
</table>
CORONARY ANGIOGRAPHY IN TETRALOGY

Figure 2
Transvenous aortography showing normal coronary arteries in tetralogy of Fallot, A) antero-posterior projection and B) simultaneous lateral projection. The origin of the anterior descending coronary artery (arrow) from the left coronary artery (LCA) is obvious in the lateral view only. The right coronary artery (RCA) and its well developed conus branch are also best seen in this projection.

Figure 3
Retrograde aortography showing origin of the AD (arrow) from the right coronary artery (RCA) in a patient with tetralogy, A) and B) simultaneous antero-posterior and lateral projections respectively. In A), the course of the AD high over the infundibulum is seen, but only in the lateral projection B) is the RCA origin of the AD clearly evident. The left coronary artery (LCA) supplies a circumflex branch only. A retrograde arterial injection was necessary because of a previous, but incomplete, surgical repair.

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the 79 the ascending aorta was reached antegrade through the VSD, while in 17, eight of whom were postrepair, the venous catheter could not be manipulated through the VSD and retrograde arterial catheterization was needed.

Coronary Artery Abnormalities

Of the 84 patients with diagnostic coronary artery visualization, five had anomalies of the coronary arteries (table 2). Four patients (4%) had an anomalous origin of the AD from the RCA (the AD crossing over the RV outflow tract), and one patient (1%) had a single left coronary artery (type f in fig. 1). Although the conus and preventricular branches were enlarged in many cases, in only seven were they large enough to be of concern in the differentiation from an aberrant AD.

Surgical Follow-up

Thirty-nine patients in whom diagnostic coronary angiography was performed have undergone complete surgical correction, and in all the angiographic diagnosis was correct. Surgical verification was obtained in four of the five patients with angiographically identified anomalous coronary arteries; the fifth had palliative surgery only, and the coronaries were not visualized during the operation.

Pathology Findings

Review of the files of 195 tetralogy specimens in the Cardiac Registry, Children's Hospital Medical Center (CHMC), provided a coronary anomaly incidence of 5% (table 3); these were 3/195 (2%) AD from RCA, 3/195 (2%) single coronary artery, 1/195 (0.5%) accessory coronary artery from the noncoronary sinus, and 1/195 (0.5%) conus coronary arising from the LCA.

Discussion

The surgically important variations in coronary artery distribution in tetralogy of Fallot have been described previously4,5 and are summarized in figure 1. The reason for the high association of these anomalies with tetralogy is unknown. Because the coronary arteries normally form by the connection of coronary buds from the aorta with an already established myocardial vascular network, it has been suggested that anterior position (dextroposition) of the aorta in tetralogy predisposes to an anomalous origin of the AD from the RCA (the RCA artery being nearer the interventricular groove than in normal hearts).4 If this were the only explanation, the occurrence of the coronary anomaly would be expected predominantly in patients with the severest forms of tetralogy because of greater aortic anteroposition or right ventricular overriding in these cases. This is unsubstantiated, however, in the literature and in our own angiographic and autopsy experience, where an origin of the AD from the RCA has been found in mild as well as in severe forms of tetralogy. The formation of a single coronary artery apparently results from failure of one of the two coronary buds of the aorta to develop, the single remaining bud taking over the supply to the entire myocardium. This anomaly may

<table>
<thead>
<tr>
<th>Author</th>
<th>Series</th>
<th>No. of cases studied</th>
<th>LAD from RCA</th>
<th>Single coronary artery</th>
<th>Other</th>
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<tr>
<td>Present report</td>
<td>Angiographic</td>
<td>84</td>
<td>4 (4%)</td>
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<tr>
<td>White et al.3</td>
<td>Angiographic</td>
<td>2</td>
<td>1</td>
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<tr>
<td>Berry et al.1</td>
<td>Surgical</td>
<td>1400</td>
<td>23 (2%)</td>
<td>4 (0.3%)</td>
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<tr>
<td>Friedman et al.4</td>
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<td>1</td>
<td></td>
<td>1</td>
<td>None</td>
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<tr>
<td>Gadboys et al.7</td>
<td>Surgical</td>
<td>1</td>
<td></td>
<td>1</td>
<td>None</td>
</tr>
<tr>
<td>Howe et al.8</td>
<td>Surgical</td>
<td>202</td>
<td>8 (4%)</td>
<td></td>
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<tr>
<td>Kirklin et al.3</td>
<td>Surgical</td>
<td>74</td>
<td>3 (4%)</td>
<td>1 (1%)</td>
<td>None</td>
</tr>
<tr>
<td>Senning10</td>
<td>Surgical</td>
<td>27</td>
<td>4</td>
<td>1</td>
<td>None</td>
</tr>
<tr>
<td>Cardiac Registry CHMC, Boston</td>
<td>Autopsy</td>
<td>195</td>
<td>3 (2%)</td>
<td>3 (2%)</td>
<td>1—accessory cor. a. from noncor. cusp, 1—conus cor. a. from LAD</td>
</tr>
<tr>
<td>Longnecker et al.4</td>
<td>Autopsy</td>
<td>22</td>
<td>7 (7%)</td>
<td>2 (2%)</td>
<td>1—AD from LCA but passing in front of PA</td>
</tr>
<tr>
<td>Meng et al.2</td>
<td>Autopsy</td>
<td>109</td>
<td></td>
<td></td>
<td>1—RCA from PA</td>
</tr>
</tbody>
</table>

Abbreviations: LAD = left anterior descending coronary artery; RCA = right coronary artery; LCA = left coronary artery; cor. a. = coronary artery; noncor. = noncoronary; PA = pulmonary artery; AD = anterior descending coronary artery.
be no more frequent in tetralogy than in other forms of congenital heart disease.

The incidence of coronary anomalies in tetralogy of Fallot has been estimated previously from autopsy and surgical series only.1,4-10 These reports are summarized in table 3 and the frequency ranges between 2 and 9%. Because of the increased surgical mortality associated with anomalous coronary artery distribution in tetralogy patients, an obvious bias occurs when pathological material is used to determine the incidence of these anomalies. All three children with an anomalous AD from the RCA in the CHMC Cardiac Registry died because the anomalous artery had been transected during surgery. Two of the three single coronary arteries were single right coronaries giving an anterior AD over the infundibulum (type e, fig. 1), and both of these specimens were in the autopsy files because the presence of the anomalous AD branch compromised surgical exposure and led to inadequate repair. The third single coronary artery arose from the left cusp and its branching (type f, fig. 1) was of no surgical significance. The other two conus coronary anomalies were incidental autopsy findings.

The angiographic study reported here presents a group of tetralogy patients unselected by autopsy and surgical considerations, and therefore might be considered to yield a more accurate incidence of coronary anomalies in the general tetralogy population. The incidence of an AD from the RCA was 4% (4/84), and single coronary artery 1%, for a combined incidence of 5%.

Previous papers1-2 have commented upon the possibility of routine preoperative angiographic evaluation of the coronary arteries in tetralogy of Fallot. This report demonstrates that the coronary artery anatomy can be delineated in most patients by flush coronary aortography. Because the intravenous catheter used for right ventricular angiography usually can be manipulated into the aorta in patients with tetralogy, the aortic flush injection can be obtained in most of these patients without need of retrograde arteriography. Using this injection method, our experience indicates that coronary artery anatomy is easiest to delineate in children and preadolescent patients. The aorta becomes greatly dilated and aortic flow large in adolescent and adult tetralogy patients, making flush coronary aortography difficult. In our series of ten patients in whom flush arteriography was nondiagnostic for the coronary anatomy, seven were 14 years or older. In these patients, selective coronary arteriography merits consideration. However, selective studies may miss a coronary anomaly when multiple coronary ostia are present. Because there are often two right coronary ostia, an injection in the right sinus of Valsalva may be the best study for older individuals.

Analysis of the coronary artery anatomy in tetralogy of Fallot is best done in the lateral view (figs. 2 and 3) because in this projection the origins of the right and left coronary arteries are most separated. The counterclockwise aortic rotation (viewed from above) in tetralogy associated with right ventricular overriding results in the right and left coronary cusps having an almost direct anterior-posterior relationship (fig. 1), and frontal viewing of the coronaries often simulates a single coronary artery because of the superimposed coronary origins. Therefore, lateral or steep oblique filming is recommended, the AD being identified by its characteristic course in the interventricular groove.

Because the incidence of coronary artery anomalies in tetralogy of Fallot may be as low as 5% and because the surgeon often can define the coronary anatomy at the time of surgery, it could be argued that preoperative angiographic demonstration of the coronaries is unnecessary. Our study suggests that routine, preoperative transvenous coronary arteriography in tetralogy is easily performed and should be considered for the following reasons:

1) Injury to an anomalous vessel is associated with a greatly increased surgical risk.1-2,4-10 Preoperative demonstration of such a vessel allows for planning of the best type of operation, or even deferral of total repair in favor of palliative surgery. This is particularly pertinent when infant total repair is being considered.

2) The coronary artery distribution is not always definable by surgical observation. The anomalous vessel can have either an intramyocardial course, or in the case of previous surgery involving opening of the pericardium, may be obscured by epicardial adhesions.1

3) The coronary aortography described in this report adds little extra time and no increased morbidity to the preoperative catheterization when performed correctly with a side-hole only, transvenous catheter passing through the VSD into the aorta.

Routine, retrograde arterial coronary angiography in tetralogy patients is probably unwarranted.1,2 However, there are two clinical instances in which a retrograde study would seem indicated preoperatively if a transvenous catheter cannot be manipulated into the aorta: 1) patients who have had previous intrapericardial surgery, where epicardial adhesions may obscure an anomalous coronary artery, and 2) infants in whom total repair is being contemplated. The demonstration of an anomalous coronary artery coursing over the infundibulum in these babies may be an indication for a palliative procedure rather than a corrective one.
When an anomalous coronary artery is recognized either preoperatively by angiography or at the time of thoracotomy, several surgical approaches to the repair are possible, and these include: 1) a transverse or tailored incision paralleling the anomalous vessels, 2) an incision caudal to the abnormal artery with an additional pulmonary artery incision above it, 3) a valved conduit graft from the right ventricle to the pulmonary artery, bypassing the anomalous coronary and underlying infundibular stenosis, and 4) an incision tunnelled underneath the aberrant artery.

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