Evaluation of Coronary Artery Anatomy in Patients With Tetralogy of Fallot by Two-Dimensional Echocardiography

James M. Berry Jr., RDMS, Stanley Einzig, MD, PhD, Kimberly A. Krabill, MD, and John L. Bass, MD

A major coronary artery crossing the right ventricular outflow tract in patients with tetralogy of Fallot interferes with a transannular patch, and preoperative detection of this artery is important. We evaluated the ability of two-dimensional echocardiography to define noninvasively the coronary artery anatomy in 37 consecutive patients (age range, 1 day to 18 years; mean age, 40.9 months). The origin and distribution of the right anterior descending and circumflex coronary arteries, as well as any anteriorly coursing vessel, were examined from parasternal views. Complete studies were obtained in 29 (78%) of the 37 patients. Coronary artery anatomy was determined to be normal by echocardiography in 20 (69%) of the 29 patients. An anterior vessel across the right ventricular outflow tract was detected in the remaining nine patients. Six patients had an anterior descending artery from the left main coronary artery (paired anterior descending arteries in three patients, a right anterior descending artery from the left main coronary artery in two patients, and a right coronary-to-pulmonary artery fistula in one patient). Three patients had no anterior descending artery from the left main coronary artery (anterior descending artery from the right main coronary artery in two patients, and anterior descending and circumflex arteries from the right main coronary artery in one patient). Angiography, surgery, or autopsy confirmed the diagnoses in all but the final patient in whom the anterior descending artery arose from the right main coronary artery as observed at surgery, but the circumflex artery was not seen. Accurate evaluation of coronary artery anatomy is possible by echocardiography in the majority of patients with tetralogy of Fallot. Noninvasive identification of a major coronary artery coursing anteriorly can influence the timing of cardiac catheterization and surgery and the need for angiography. (Circulation 1988;78:149–156)

Surgical repair of tetralogy of Fallot may require a longitudinal ventriculotomy extending into the main pulmonary artery to relieve right ventricular outflow tract obstruction when the pulmonary annulus is small. In 5–9% of patients with tetralogy of Fallot, the anterior descending coronary artery or another major coronary artery branch courses anteriorly across the right ventricular outflow tract so that a transannular incision or patch would interrupt coronary blood flow. Relief of right ventricular outflow tract obstruction may then be difficult, particularly in younger patients. Patients with tetralogy of Fallot are routinely evaluated preoperatively with root aortography or selective coronary angiography for these coronary abnormalities. A noninvasive means of identifying patients with tetralogy of Fallot who have abnormal coronary artery anatomy could optimize timing of surgery and perhaps limit the number of cardiac catheterizations needed.

Two-dimensional echocardiography provides a noninvasive means of imaging cardiovascular anatomy and has been used to assess coronary artery branching patterns in d-transposition of the great arteries. The purpose of our study was to determine the accuracy of two-dimensional echocardiography in defining coronary artery anatomy in tetralogy of Fallot. We evaluated 37 patients at our institution by echocardiography with confirmation by angiography, surgery, or autopsy of the coronary artery branching pattern.
Patients and Methods

Between September 1984 and November 1987, 37 consecutive patients (35 with tetralogy of Fallot and two with double-outlet right ventricle and subaortic ventricular septal defect) were assessed by prospective two-dimensional echocardiography of coronary artery branching patterns in addition to angiography, surgery, or autopsy. Patients were admitted for cardiac catheterization or surgery and were not selected because of their coronary artery anatomy. Ages ranged from 1 day to 18 years, with a mean age of 40.9 months.

All echocardiograms were performed without knowledge of angiographic findings. One cardiac ultrasound technician performed the studies, often with a physician present. Ultrasound studies were performed on an Ekoline 5500D (mechanical sector scanner, Hewlett-Packard, Palo Alto, California), or a Hewlett-Packard 77020A (phased-array ultrasonoscope) with 2.5-, 3.5-, and 5-MHz transducers and recorded on videotape. A 5-MHz transducer yielded the best images of coronary anatomy in all patients. The origins of the right and left main coronary arteries were imaged from a right or left parasternal window in a plane just superior to the short axis of the aortic valve. The normal left coronary artery system was imaged from the first, second, or third left intercostal space with the transducer oriented acutely inferior so that the pulmonary valve annulus could be seen in cross section, and posterior to the annulus in longitudinal sections, in the left main, proximal left anterior descending, and circumflex coronary arteries (Figure 1A). The left anterior descending artery was further imaged from a standard left parasternal short-axis view in cross section in the anterior interventricular sulcus and traced back to its origin by scanning superiority. The right ventricular outflow tract was imaged in both long and short axes from a high left parasternal window with special attention directed toward the presence of any
antiorly coursing vascular structures. Demonstration of the circular shape of the coronary artery in cross section prevented confusion with parallel pericardial reflections (Figure 1B).

Coronary artery anatomy was judged normal when the following three criteria were met: 1) the left coronary ostium and left main coronary artery arose from the lateral aortic valve cusp, giving rise to the circumflex and anterior descending coronary arteries that coursed posterior and inferior to the pulmonary valve annulus, 2) the right coronary ostium and proximal right coronary artery arose from the right cusp as a single vessel without evidence of early branching, and 3) no vascular structure was identified anterior to the pulmonary valve or right ventricular outflow tract. Coronary artery anatomy was considered possibly abnormal whenever a vessel was identified anteriorly crossing the right ventricular outflow tract (Figures 2A and 2B), and the origin and distal distribution of any such vessel were carefully defined.

The statistical comparisons of patients’ weights and ages were performed with the Student’s unpaired t test.

Results

Echocardiographic studies adequate for determination were obtained in 29 (78%) of the 37 patients (Figure 3). Their ages ranged from 1 to 108 months, and weights ranged from 2.6 to 18.8 kg. The remaining eight patients were excluded (ages ranged from 1 day to 216 months, weights ranged from 3.35 to 59 kg) when imaging was incomplete (Table 1). In two patients, exclusion was due to large patient size (15 and 18 years old); in one patient, it was due to persistent overlying lung tissue (3 months old, with tracheostomy for bronchomalacia); and in five patients, it was due to pulmonary atresia and severe
hypoplasia of the right ventricular outflow tract and main pulmonary artery combined with inadequate patient cooperation. Two patients with tetralogy of Fallot and pulmonary atresia had adequate studies, but good cooperation, and less severe hypoplasia of the right ventricular outflow tract and pulmonary annulus area (membranous atresia). Seven of the eight technically inadequate examinations occurred during the first third of the study period. Compared with those with adequate echocardiographic studies, patients with inadequate studies were older \((p<0.005)\) and larger \((p=0.016)\). In addition to time spent on the usual evaluation of cardiac anatomy in patients with tetralogy of Fallot, up to 40 minutes of examination time was devoted to identifying and tracing the coronary arteries.

**Angiography, Surgery, and Autopsy**

Angiographic confirmation of coronary artery anatomy was possible in 28 patients, surgical confirmation in 21, and autopsy confirmation in three (Table 2). In the 20 patients in whom angiography, surgery, and autopsy were available, the angiographic diagnosis was confirmed in each. Normal coronary anatomy was present in 20 of the 29 patients. Root aortograms were performed in 19 of the 20 patients (Table 2). Small \((<\frac{3}{4}\) diameter of the right main coronary artery) conal branches from the right coronary artery that crossed the right ventricular outflow tract were present in five of the 20 patients, but it did not influence surgical management.

In the nine patients with abnormal coronary artery anatomy, root aortograms were performed in two patients, selective coronary angiograms in six patients, and no angiography in one patient (Table 2). Five different coronary abnormalities were identified. Origin of an anterior descending coronary artery from the left main coronary artery was present in six patients. Paired anterior descending coronary arteries from the right and left main arteries were seen in three of these patients (the anterior descending arteries were equal in size in one patient with the right artery supplying the upper ventricular septum, and the branch from the right artery was dominant in the other two patients); origin of the right and left coronary artery systems from a single

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**TABLE 1. Relation of Patient Size to Adequacy of Echocardiographic Study**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Weight (kg)</th>
<th>Age (mo)</th>
<th>Study problems</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>59.0</td>
<td>216</td>
<td>Inadequate penetration, large size</td>
</tr>
<tr>
<td>2</td>
<td>43.0</td>
<td>180</td>
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</tr>
<tr>
<td>3</td>
<td>24.5</td>
<td>72</td>
<td>Pulmonary atresia, uncooperative</td>
</tr>
<tr>
<td>4</td>
<td>18.6</td>
<td>60</td>
<td>Pulmonary atresia, uncooperative</td>
</tr>
<tr>
<td>5</td>
<td>12.0</td>
<td>43</td>
<td>Pulmonary atresia, uncooperative</td>
</tr>
<tr>
<td>6</td>
<td>12.2</td>
<td>30</td>
<td>Pulmonary atresia, uncooperative</td>
</tr>
<tr>
<td>7</td>
<td>3.3</td>
<td>1/30</td>
<td>Pulmonary atresia, uncooperative</td>
</tr>
<tr>
<td>8</td>
<td>4.6</td>
<td>3</td>
<td>Inadequate penetration, pulmonary disease</td>
</tr>
</tbody>
</table>

Mean \(\pm SD\)

<table>
<thead>
<tr>
<th>Adequate studies</th>
<th>10.4 (\pm 4.9)</th>
<th>30.3 (\pm 30.1)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inadequate studies</td>
<td>22.2 (\pm 19.5)*</td>
<td>75.5 (\pm 80.2)†</td>
</tr>
</tbody>
</table>

\(*p<0.005; \dagger p=0.016.\)
Table 2. Methods of Identifying Coronary Artery Anatomy

<table>
<thead>
<tr>
<th>Anatomy</th>
<th>Echocardiography</th>
<th>Angiography</th>
<th>Surgery</th>
<th>Autopsy</th>
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<tbody>
<tr>
<td>Normal</td>
<td>20</td>
<td>19</td>
<td>1</td>
<td>15*</td>
</tr>
<tr>
<td>Abnormal</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Paired anterior descendings</td>
<td>3</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Single left coronary ostium</td>
<td>2</td>
<td>0</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Coronary to pulmonary fistula</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Anterior descending from right</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Single right coronary ostium</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

Ao, root aortography; Selective, selective coronary injection.
*Does not include two patients who underwent thoracotomies that did not reveal coronary anatomy.

Ostium from the left cusp was seen in two patients; and a right coronary artery fistula that entered the anterior main pulmonary artery was seen in one patient. In the remaining nine patients, the anterior descending coronary artery did not arise from the left main coronary artery. The anterior descending artery arose from the right coronary artery in two patients. Surgery alone confirmed the origin of the anterior descending coronary artery from the right in the remaining patient thought to have a single right coronary artery. The origin of the circumflex artery was not seen; no angiography had been performed. The coronary artery anterior to the right ventricular outflow tract influenced surgical management in the nine patients.

**Echocardiography**

A normal coronary artery branching pattern was correctly identified in 20 patients (Figure 3). A small vessel identifiable as a conal branch was seen in only one patient. In the remaining nine patients, a large coronary artery was identified coursing anterior to the right ventricular outflow tract. The specific coronary abnormality was correctly predicted in each of these patients. Six patients had a normally arising left anterior descending coronary artery. A diagnosis of paired anterior descending coronary arteries was correctly made in three patients in whom the anterior coronary artery arose from the right and coursed down the interventricular sulcus (Figure 4). Origin of the right coronary artery from the left coronary system was correctly identified in two patients. No right coronary ostium was found, and the anterior coronary artery was traced from the left of the pulmonary artery anterior to the course of a normal right coronary artery (Figure 5). A right coronary artery-to-pulmonary artery fistula was correctly identified in the last of these six patients. The anterior vessel was traced from the right coronary system to an anterior communication with the main pulmonary artery (Figure 6), and a continuous negative Doppler flow signal from the fistula was recorded in the main pulmonary artery.

A normally arising left anterior descending artery was not found in the remaining three patients. The anterior descending coronary artery was correctly traced to its origin from the right coronary artery in two patients. In the final patient, echocardiography demonstrated a single coronary artery ostium on the right coronary cusp from which right anterior descending and circumflex coronary arteries arose. Although the anterior descending artery was found arising from the right coronary artery at surgery, the origin of the circumflex artery was not confirmed by angiography, surgery, or autopsy.

**Discussion**

Surgical repair of tetralogy of Fallot is being performed in progressively younger patients.11,12 Although recognized anomalies of the coronary artery system can usually be circumvented in older, larger patients with tetralogy of Fallot, repair in small patients may be significantly complicated by a major coronary artery crossing the right ventricular outflow tract. This abnormality may postpone elective surgical repair, or it may indicate that a palliative approach is needed. Noninvasive detection of such abnormalities could improve management by altering the timing of cardiac catheterization or by identifying patients in whom selective coronary angiography will be needed.

Echocardiography has been used to demonstrate coronary artery anomalies such as coronary artery fistulas.13–17 Anomalous origin of the left coronary artery from the pulmonary artery,18–20 and aneurysms of the coronary arteries in Kawasaki’s syndrome.21,22 Noninvasive evaluation of coronary artery anatomy in d-transposition of the great arteries has also been reported.10 However, we are aware of no reports with two-dimensional echocardiography to completely evaluate coronary anatomy in patients with tetralogy of Fallot.

The coronary artery anatomy was correctly defined by two-dimensional echocardiography in all 29 patients with adequate studies. A major coronary artery across the right ventricular outflow tract was identified in the nine patients in whom the coronary arteries were abnormal. The specific anomaly present was correctly recognized in eight patients. In the ninth patient, an anterior descending coronary artery crossing the right ventricular outflow tract
was correctly diagnosed. Circumflex anatomy was not defined at surgery. We were able to detect a variety of different abnormalities including paired anterior descending coronary arteries and the origin and course of a single coronary artery crossing the right ventricular outflow tract. The types of anomalies were similar to those reported in other series.²⁻⁷

High left parasternal images proved to be the most sensitive for detection of an abnormal coronary artery crossing the right ventricular outflow tract. Apical and subcostal windows allowed visualization of the coronary arteries in some patients, but relations were more difficult to ascertain. Demonstration of a circular cross-sectional shape, as well as the origin and distal distribution of a suspected vessel across the right ventricular outflow tract, eliminated confusion with parallel pericardial reflections. It is not possible to visualize the entire course of a coronary artery in a single plane, and multiple imaging planes from both right and left parasternal windows with patients in right and left lateral decubitus or supine positions are usually necessary. There can be no single uniform approach to evaluation of coronary anatomy because of the multiple factors affecting acoustic penetration and the variation in anatomy relative to the chest wall.

High-quality echocardiographic images of the right ventricular outflow tract and main pulmonary artery are an absolute necessity for critical evaluation of coronary anatomy. No conclusions can be drawn about the presence or absence of coronary anomalies if these images are not obtained. Although we cannot establish a size limitation from our experience, obtaining images of older and larger patients may be more difficult. It is the younger patient in whom the best studies are most likely to be obtained and in whom the detection of an aberrant coronary artery across the right ventricular outflow tract is most important. As with all cardiac ultrasound procedures, adequate patient cooperation is essen-

**FIGURE 4.** Echocardiograms of parasternal short-axis views through the aortic root (Ao) in a patient with paired anterior descending coronary arteries. Panel A: Origin of a large anterior descending coronary artery (AD) from the right coronary artery (RCA) that then courses across the right ventricular outflow tract. Panel B: In a slightly inferior scan, a smaller left anterior descending coronary artery (LAD) courses posterior to the right ventricular outflow tract. PA, pulmonary artery.
tial. In five patients, the difficulty of these procedures was compounded by poor patient cooperation and by pulmonary atresia and severe hypoplasia or atresia of the right ventricular outflow tract and main pulmonary artery, where the relation of the coronary arteries and a strand of tissue representing the right ventricular outflow tract/pulmonary annulus were difficult to define. Confidence in the ability to define coronary anatomy developed as this study progressed. Failure of reexamination in three patients whose initial studies were inadequate suggests that a learning curve will not overcome technical limitations in the ability to obtain good images.

Evaluation of coronary anatomy in these patients requires a considerable dedication of resources. Both technicians and physicians must invest time in learning views and techniques for tracing coronary artery distribution. The patients themselves undergo longer ultrasound studies, although the amount of time varies according to the technical difficulty of the study and the anatomic abnormality present. Nevertheless, we have found that the results of these efforts are beneficial in preoperative evaluation.

Up to 19.3% of patients with tetralogy of Fallot have been reported to have a conal branch crossing the right ventricular outflow tract that is at least as large as the right coronary artery. We did not observe a large conal branch in any of the study group either by angiography or echocardiography. A large conal branch could be distinguished from an aberrant anterior descending artery by tracing its termination to the right ventricular infundibulum rather than down the interventricular sulcus. An anomalous coronary artery may also run between the aorta and pulmonary artery rather than across the right ventricular outflow tract. Although the ability of echocardiography to detect either of these abnormalities could not be assessed in the patients that we examined, these abnormalities are of less surgical importance.

**Figure 5.** Echocardiogram of the parasternal short-axis view in a patient with the origin of the right coronary artery (RCA) from the left coronary system. RCA (→) courses from the left across the right ventricular outflow tract toward its normal distribution. Ao, aortic root; PA, pulmonary artery.

**Figure 6.** Echocardiogram of the parasternal short-axis view through the pulmonary artery (PA). Large fistula (F) arises from the right coronary artery (RCA) and courses to the left across the right ventricular outflow tract communicating with the main pulmonary artery. Ao, aortic root.
The incidence of coronary artery anomalies in the patients with adequate studies (nine of 29, or 31%) is in considerable excess of the 5–9% reported in the angiographic and autopsy series.2–7 These patients were not selected because of coronary artery abnormalities. This represents the incidence among patients admitted during the study period, not the true overall incidence of coronary anomalies in patients with tetralogy of Fallot at our institution.

Accurate evaluation of coronary artery anatomy by two-dimensional echocardiography is possible in the majority of patients with tetralogy of Fallot. The technique holds considerable promise for directing cardiac catheterization in patients for whom selective coronary or cusp injections or specially angulated root aortograms may be needed to adequately define coronary anatomy. It may also be possible to identify children with tetralogy of Fallot and an aberrant coronary artery in whom cardiac catheterization before elective surgery can be postponed until the patient reaches a size at which surgical repair can be more safely performed. With additional experience, it may even become possible to eliminate the need for angiography to define coronary artery anatomy in some of these patients, reducing the risk of cardiac catheterization.

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References


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