# Detection of Coronary Artery Abnormalities in Tetralogy of Fallot by Two-Dimensional Echocardiography

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Patients with tetralogy of Fallot have a 5% to 19% incidence rate of abnormal distribution of coronary arteries. These abnormalities are usually detected by angiography and influence the timing and mortality rate of surgery. This study evaluates two-dimensional echocardiography as a method of assessing coronary artery distribution in tetralogy of Fallot. Forty-five consecutive patients with tetralogy of Fallot, aged 0.1 to 20.5 years (mean 5.7  $\pm$  4.3), had prospective two-dimensional echocardiographic studies to examine the branching patterns of the coronary arteries and to determine the presence or absence of a branch from the right or left coronary artery that crossed the right ventricular outflow tract. The first two patients had known coronary abnormalities and served as learning models. All other echocardiographic studies were performed without knowledge of angiographic or surgical findings.

Twenty-two studies were completed before coronary

angiography (group A) and 23 after angiography (group B). All eight patients (18%) with coronary abnormalities were correctly identified by two-dimensional echocardiography (five in group A and three in group B). Three had bilateral anterior descending coronary arteries originating from the left and right coronary arteries, two had the anterior descending artery originating from the right coronary artery, two had a large conal branch from the right coronary artery and one had origin of both left and right coronary arteries from a single left ostium. All abnormal coronary arteries were visualized crossing the right ventricular outflow tract, whereas all 21 small conal branches from the right coronary artery were not seen in the right ventricular outflow tract. Thus, two-dimensional echocardiography is a reliable technique for diagnosing coronary abnormalities in tetralogy of Fallot.

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An anomalous coronary artery coursing in the right ventricular outflow tract is a known potential complicating factor in the surgical correction of tetralogy of Fallot (1,2). Knowledge of coronary artery abnormalities influences timing, surgical technique and outcome of surgery. Performing surgery without such knowledge can lead to major complications for the patient at the time of surgery (3–5). To date, angiography remains the only method by which these coronary artery abnormalities are diagnosed before surgery (1,6,7). Two-dimensional echocardiography has been shown

to be a reliable noninvasive technique for assessing these abnormalities in Kawasaki's disease (8,9), transposition of the great arteries (10,11) and anomalous origin of the left coronary artery from the pulmonary trunk (11,12). The purpose of this study was to assess the ability and accuracy of two-dimensional echocardiography in diagnosing coronary artery abnormalities in tetralogy of Fallot.

### Methods

Study patients. From September 1987 to January 1989, 45 consecutive patients with tetralogy of Fallot, aged 0.1 to 20.5 years (mean  $5.7 \pm 4.3$ ), were seen in the pediatric cardiology clinic at Cardinal Glennon Children's Hospital. All were diagnosed as having tetralogy of Fallot by two-dimensional and Doppler echocardiography, and all had confirmatory cardiac catheterization and angiography. These patients were prospectively studied by two-dimensional echocardiography to evaluate coronary artery anatomy. At this institution, most children <3 years of age are sedated with oral

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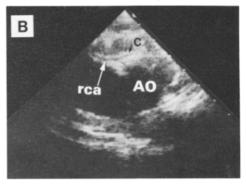
chloral hydrate (50 mg/kg body weight) before undergoing echocardiographic evaluation. Eleven patients in this study required sedation.

Echocardiography. Prospective two-dimensional echocardiographic evaluation was performed on all patients and recorded on videotape. One of two ultrasound technicians performed the studies with one of the pediatric cardiologists present. Except for the first two patients with known coronary artery abnormalities who served as learning models, all studies were performed without knowledge of the angiographic or surgical findings. Cardiac catheterization and angiography were performed after the two-dimensional echocardiographic coronary artery study in 22 patients (group A) and before echocardiography in the remaining 23 (group B). Two echocardiographic systems were used— Hewlett Packard 77020AC phased array and Acuson 128 computed sonography systems, both of which are equipped with 5 and 3.5 MHz phased array transducers. In the majority of the studies (40 of 45 [89%]), only the 5 MHz transducer was used to examine the coronary arteries.

Echocardiographic views. Six views were utilized systematically to study the origin, course and branching patterns of the coronary arteries. Particular attention was made to ascertain if a branch from the right coronary artery could be demonstrated. and then to follow the course of that branch to the right ventricular outflow tract.

The views were as follows: View 1. Left parasternal short-axis view to demonstrate the origin and course of the proximal left coronary artery (Fig. 1A). When the transducer was angled to the right and slightly superiorly, the right coronary artery origin and proximal course could be seen. Leftward angulation resulted in demonstration of the peripheral branches of the left coronary artery and, when present, one could visualize an abnormal coronary artery branch not originating from the left coronary artery (Fig. 2). View 2. Right parasternal short-axis view, mainly to demonstrate the course of the right coronary artery and the presence of a branch arising from the proximal right coronary artery (Fig. 3). When present, this branch could be traced toward the right ventricular outflow tract by slight superior and leftward angulation. View 3. High modified left parasternal short-axis view from the first or second intercostal space, with caudal angulation. This view was used to demonstrate the origin, but mainly the course and branches of the left coronary artery, and to demonstrate the right ventricular outflow tract by angling the transducer superiorly (Fig. 4). Special attention was paid to any arterial structure seen in the right ventricular outflow tract. The presence of a round structure with echo-bright walls prevented confusing the arterial image with other irregular muscular or epicardial reflections (Fig. 4). View 4. Modified high right short-axis view to demonstrate the origin and course of the right coronary artery (Fig. 1B). The latter two views were often successful in demonstrating the origin (ostium) of the left or the right





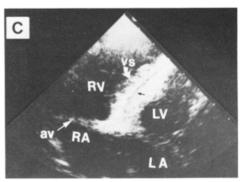


Figure 1. Echocardiographic views of normal coronary arteries. A, Left parasternal short-axis view demonstrating normal left main (lca), left anterior descending (ad) and circumflex (cir) coronary arteries. B, High modified right parasternal short-axis view demonstrating a small conal branch (c) originating from the right coronary artery (rca). This branch could not be traced to or seen in the right ventricular outflow tract. C, Modified apical four chamber view with posterior and lateral angulation and minimal clockwise rotation of the transducer demonstrating a normal left anterior descending coronary artery (small black arrow) coursing through the ventricular septum (vs). AO = aorta; av = atrioventricular groove; LA = left atrium; LV = left ventricle; PA = pulmonary artery; RA = right atrium; RV = right ventricle.

coronary arteries when the left and right parasternal views failed to do so. View 5. Apical five chamber view from a position 2 to 3 cm lateral to the left sternal border. This view was used to demonstrate the proximal course of both coronary arteries. View 6. Apical four chamber view with posterior and lateral angulation and slight clockwise rotation of the transducer. This view was used to demonstrate the

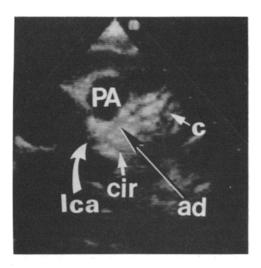
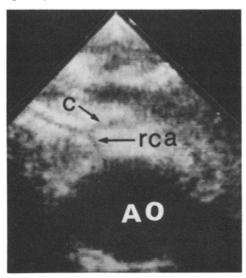
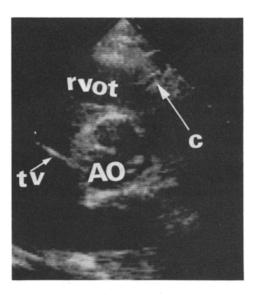


Figure 2. Modified left parasternal short-axis view with slight lateral and superior angulation demonstrating the course of the anterior descending artery (ad) and the presence of an adjacent coronary branch (large conal) (c) that does not originate from the left coronary artery (lca) (see also Fig. 8). Abbreviations in Figure 1.

longitudinal course of the anterior descending artery as it arises from either the left (Fig. 1C) or the right coronary artery, or both, in the presence of paired anterior descending arteries (Fig. 5). It could also demonstrate the superficial part of the interventricular septum and show the course of the anterior descending coronary artery (Fig. 1C and 5). This

Figure 3. A right parasternal short-axis view showing a large right coronary artery (rca) that has a large branch that could be traced and followed to the right ventricular outflow tract. This frame was obtained from a patient with origin of the anterior descending artery from the right coronary artery. Similar views were seen in patients with large conal branches and patients with paired anterior descending arteries. AO = aorta; c = conal origin of the right anterior descending artery.

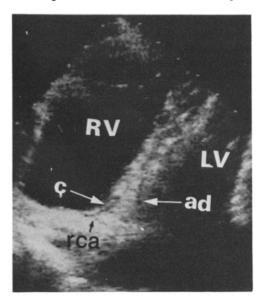


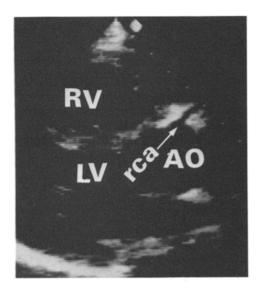


**Figure 4.** A left parasternal short-axis view with superior angulation toward the right ventricular outflow tract demonstrating an arterial structure (c) coursing in the outflow tract 1.3 cm from the pulmonary valve. AO = aorta; RVOT = right ventricular outflow tract; TV = tricuspid valve.

view utilized the vertical and the lateral resolution of the transducer to demonstrate the structures just mentioned. The subcostal views were not used in this study because they view the coronary arteries in cross section, and there-

Figure 5. Modified apical four chamber view with posterior and lateral angulation and slight clockwise rotation demonstrating two coronary arteries (c and ad) coursing on both sides of the ventricular septum (paired anterior descending arteries). One of these coronary arteries could be traced to the right coronary artery (rca) in the right atrioventricular groove. LV = left ventricle; RV = right ventricle.





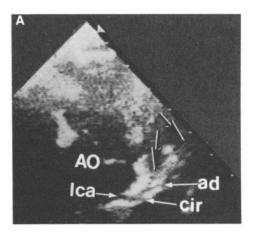
**Figure 6.** Parasternal long-axis view showing the origin of a large right coronary artery (rca) from the anterior aspect of the aorta (AO). This view was obtained from a patient with origin of the anterior descending artery from the right coronary artery. The right coronary artery (rca) is visible in this view because of clockwise rotation of the aortic cusps (viewed from below; see text). LV = left ventricle; RV = right ventricle.

fore do not provide good demonstration of the course and branching pattern of the coronary arteries (8).

Transducer parameter settings. The coronary arteries were best demonstrated by using the following adjustments in the reception focus of the echocardiographic equipment: The transmit gain was set at 35 dB. The compress was set at 40 dB. The eight time gain compensation slide pots, with a 0 to 60 dB range, were arranged in an incremental-decremental fashion as follows: the first three slide pots were set from 15 to 40 dB; the next two at 40 to 45 dB and the next three from 35 down to 10 dB. Minor alterations were necessary for each patient to obtain good contrast between the coronary artery and the surrounding myoepicardium.

Normal versus abnormal coronary arteries. The coronary artery branching pattern was considered normal in this study if the following criteria were met (Fig. 1A, B and C): 1) demonstration of the ostium and left main coronary artery originating from the left aortic sinus of Valsalva; 2) demonstration of the left main coronary artery dividing into two branches (the anterior descending artery coursing anteriorly beneath the left atrial appendage, and the circumflex artery traced to the left atrioventricular groove); 3) demonstration of the ostium and main right coronary artery arising from the right aortic sinus of Valsalva, either without branching or with a small conal branch that could not be traced to the right ventricular outflow tract (Fig. 1B); and 4) no evidence of an arterial structure in the right ventricular outflow tract.

The coronary arteries were considered to be abnormal if 1) the right coronary artery was large or seen on the



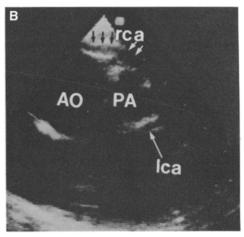
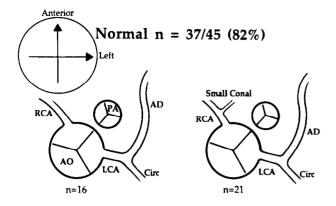


Figure 7. A, Apical five chamber view from a patient with origin of both right and left coronary arteries from a single left ostium. There is one left coronary artery (lca) that divides into three equal-sized branches. Unlabeled black arrowheads point toward the right coronary artery as it originates from the left coronary artery and to its course anteriorly. B, Modified left parasternal view from the same patient, demonstrating the course of the right coronary artery (rca) (arrows) in front of the right ventricular outflow tract (see also Fig. 8). Abbreviations as in Figure 1.

long-axis parasternal view (Fig. 6) with a large branch originating from the right main coronary artery (Fig. 3) that could be traced into the right ventricular outflow tract (Fig. 4); 2) two arteries were seen on the modified apical view along both sides of the ventricular septum, and one could be traced to the right coronary artery in the right atrioventricular groove (Fig. 5); or 3) a single large right or left coronary artery with three branches was seen (Fig. 7A and B).

Cardiac catheterization and angiography. We utilized standard views to demonstrate the coronary arteries during cardiac catheterization in the 45 patients (6,7). Left ventricular injection was either in the anteroposterior and lateral projections or in the left anterior oblique projection with cranial angulation and the right anterior oblique projection. Aortic root injection was performed in anteroposterior and



# Abnormal n = 8/45 (18%)

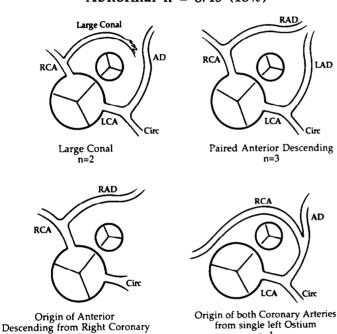


Figure 8. Schematic illustrations of normal and abnormal coronary arteries and their incidence in our 45 patients. Orientation is similar to that obtained by two-dimensional echocardiography. CIRC = circumflex; RAD = right anterior descending; other abbreviations as in Figure 1.

lateral projections if the coronary arteries were not well visualized from the left ventricular injection. Five patients had selective coronary angiography. Surgical confirmation of the coronary anatomy was available in all 26 patients who had total corrective surgery.

Statistical analysis. Visualization of the coronary arteries and their branching patterns by two-dimensional echocardiography in multiple views resulted in more confidence in the diagnosis than when only one view was successfully visualized. Therefore, we compared the weights and ages of patients who had less than two compared with two or more views using independent samples T test (p < 0.05).

Table 1. Demographic Data From 45 Patients in Relation to Echocardiographic Clarity of Coronary Anatomy

Echocardiographic Views	No.	Age (yr)	Weight (kg)
Two or more	40	4.7 ± 4	19 ± 14
Fewer than two	5	$15.7 \pm 4$	$50 \pm 10$
		p < 0.005	p < 0.005

Values are reported as mean values  $\pm 1$  SD.

# Results

Coronary artery anatomy (Fig. 8). Of the 45 patients, 8 (18%) had abnormal coronary arteries that would interfere with surgery because a large coronary artery crossed the right ventricular outflow tract. All eight of these patients (five in group A, three in group B) were correctly identified by two-dimensional echocardiography and the results confirmed by angiography. Five of the eight patients also had surgical confirmation. The following coronary artery abnormalities were found: origin of the anterior descending artery from the right coronary artery (two patients); paired anterior descending arteries originating from the left and right coronary arteries, with the artery originating from the right coronary artery coursing in the right ventricular outflow tract (three patients); a large conal artery that supplied a significant area of the right ventricular anterior wall (two patients) and origin of both the left and the right coronary artery from a single left ostium (one patient). Of the remaining 37 patients, 21 had visible small conal arteries that could not be traced to the right ventricular outflow tract (Fig. 1C). Of these, 14 had a right ventricular outflow incision and patch without sequelae.

Two patients (6%) had studies that were inadequate for determining coronary artery anatomy. This was attributed to poor penetration of the two-dimensional echocardiographic beam as a result of their older age (18 and 20 years, respectively) and larger size (52 and 65 kg). Also, three patients had only one view with clear two-dimensional echocardiographic imaging of the coronary arteries and were significantly older (p < 0.005) and larger (p < 0.005) than the other patients. The comparison of this group of patients having fewer than two views with those having more than two views is shown in Table 1. All patients with abnormal coronary arteries had clear two-dimensional echocardiographic images from multiple views, averaging four views per patient.

Potential for false positive diagnosis of abnormal coronary arteries. One of the early patients was suspected of having bilateral descending arteries on two-dimensional echocardiographic study. One year later, however, his echocardiographic study was repeated and he was found to have a large right ventricular branch from the right coronary artery. This diagnosis was confirmed later by selective coronary angiography.

## Discussion

Echocardiography. This study highlights the importance of two-dimensional echocardiography in assessing the coronary artery anatomy in tetralogy of Fallot. There is now increasing evidence that two-dimensional echocardiography is very reliable in assessing coronary artery anatomy in a variety of pathologic conditions such as Kawasaki's disease (8,9), anomalous origin of the left coronary artery from the pulmonary trunk (11,12), dextro-transposition of the great arteries (10,11) and coronary artery fistula (13). Also, we have used two-dimensional echocardiography to successfully differentiate between right coronary artery fistula and aortico-right ventricular tunnel, based on the presence of normal coronary artery size in the latter condition (14).

There are clear benefits from using two-dimensional echocardiography for the assessment of the coronary arteries in tetralogy of Fallot. 1) When a coronary artery abnormality is demonstrated by two-dimensional echocardiography, the cardiologist will be prepared to perform selective coronary angiography if the coronary artery abnormality is not well seen on routine left ventricular angiography or aortography. This is particularly true when there is a systemic to pulmonary shunt, which results in simultaneous filling of the pulmonary arteries that overshadow the coronary arteries and makes identification of the correct coronary anatomy almost impossible. Coronary artery abnormality in one of our patients in group B with origin of the anterior descending artery from the right coronary artery was missed when only a left ventricular injection was used. The coronary artery abnormality was discovered at surgery and because of her age (2 years), a palliative procedure was done. Subsequent selective right coronary artery injection demonstrated the anomaly well, and she had total correction with a conduit connecting the right ventricle and pulmonary artery at the age of 6 years. Her coronary artery abnormality was clearly seen by two-dimensional echocardiography at that time. One patient in group A who had paired anterior descending arteries from both the left and right coronary arteries underwent right sinus of Valsalva cusp injection, which demonstrated the anomaly well. This anomaly could have been easily missed if only a left ventricular angiogram was performed because it was not readily seen in that injection. Other investigators (1,3,5) have reported coronary artery abnormalities that have been missed at the time of cardiac catheterization and later discovered at surgery, autopsy or repeat angiography.

Influence of detection of coronary abnormalities by echocardiography on patient management. Because the diagnosis of tetralogy of Fallot in infancy can be accurately made by two-dimensional echocardiography, early cardiac catheterization and angiography are often unnecessary for the initial diagnosis and management. Thus, these invasive tests should be performed only before the planned total repair. At present, there is an increasing trend toward performing total repair of tetralogy of Fallot at a progressively younger age (15). At our institution, elective surgical repair is performed between the 2nd and 3rd year of life because of evidence that there is more fibrosis in the right ventricle (16) and a higher incidence of arrhythmias (17) in those with later repair. It is technically difficult to place a conduit in the outflow tract or perform other surgical techniques to preserve the abnormal coronary artery in patients <3 years of age. Therefore, when a coronary artery abnormality that could interfere with a right ventriculotomy is found by two-dimensional echocardiography, cardiac catheterization or definitative surgery should be postponed until the patient attains an appropriate age and size (2).

Sedation is very important in successful performance of a diagnostic echocardiographic study in constantly moving or anxious children. To date, we have used sedation with chloral hydrate in >400 patients without adverse reaction. The time spent in assessing coronary artery anatomy using the six views ranged between 10 to 40 min. At the beginning of the study, 35 to 40 min was required for most patients but with increasing experience, this range decreased to 10 to 20 min. We believe that this is time well spent and that the benefits outweigh the costs.

Clues to echocardiographic diagnosis. There are a few clues that should alert the echocardiographer to the presence of abnormal coronary arteries. 1) The presence of a large right coronary artery, which could be seen on the parasternal long- (Fig. 6) or short- (Fig. 3) axis view is indicative of abnormal coronary anatomy. This appearance was demonstrated in all seven patients with abnormal coronary arteries originating from the right coronary artery, which is larger because it supplies more areas of the heart than does the left coronary artery in these patients (1). When seen on the parasternal long-axis view, a large right coronary artery indicates that the aortic cusps are rotated clockwise (when viewed from below), a condition associated with abnormal coronary arteries (mainly origin of the anterior descending artery from the right coronary artery) (1,18).

- 2) The presence of a coronary artery in the right ventricular outflow tract (1 to 2 cm below the pulmonary valve) is a clear indication of the presence of abnormal coronary anatomy (Fig. 2C). This abnormality was seen in all eight patients with abnormal coronary arteries. We recommend that the echocardiographer examine this area very carefully and perform several scans of the margin of the right ventricular outflow tract to ensure the absence of an abnormal coronary artery in this area.
- 3) The modified apical view with posterior and lateral angulation and slight clockwise rotation enabled the assessment of the presence of paired anterior descending arteries and differentiation of this condition from a large conal branch; in the case of paired anterior descending arteries, there was evidence of two coronary arteries coursing along

both sides of the ventricular septum (Fig. 5). We were able to demonstrate this in all three patients with paired anterior descending arteries that were confirmed angiographically. In the case of a large conal coronary artery, only one anterior descending artery was seen. This view can demonstrate the normal anterior descending artery clearly in >50% of our patients. To our knowledge, use of this view has not been reported before (8–10). The differentiation between a large and surgically significant conal artery from a small insignificant one was successfully based on whether it could be traced to and seen in the right ventricular outflow tract.

4) Demonstration of only one coronary ostium with a single large coronary artery that has three equal branches should indicate a single origin of both coronary arteries. In the patient with a single left coronary ostium, the three branches were well visualized (Fig. 7A) and the right coronary artery course could be followed in front of the right ventricular outflow tract (Fig. 7B) and later in front of the aorta. This patient's diagnosis was confirmed at surgery. He did not require a conduit placement because the right coronary artery descended down away from the pulmonary valve after its initial course in the right ventricular outflow tract. A transannular incision and patch procedure were performed in the area free of coronary arteries without sequelae.

Potential for false positive coronary abnormalities on twodimensional echocardiography. One of the early patients was suspected to have bilateral anterior descending arteries because he had a visible branch from the right coronary artery that crossed the right ventricle 3 cm below the pulmonary valve. However, 1 year later when his initial echocardiographic study was reviewed before cardiac catheterization, it was clear that the diagnosis was incorrect because there was only one anterior descending artery seen on the modified four chamber view, and the distance of the coronary artery from the pulmonary valve was too great. The correct diagnosis (right ventricular branch) was made on repeat echocardiographic study and subsequently confirmed by cardiac catheterization and selective coronary angiography. In contrast, all abnormal coronary arteries crossed the right ventricular outflow tract 1 to 2 cm from the pulmonary valve. To our knowledge, there is only one report (5) describing a death associated with inadvertent division of a right ventricular branch; however, the right anterior descending coronary artery was additionally divided in that patient, and could have been primarily responsible for the death. We did not include our early patient among the abnormal cases because the surgical team believed that the right ventricular branch can be avoided easily during surgery. We did not have false negative results in this study.

Incidence of abnormal coronary abnormalities. The 18% incidence rate of significant coronary abnormalities in this group of patients is higher than the 5% to 10% rate found in other studies (1,5-7), but is equal to Senning's reported incidence rate (19) of 19%. In our study group, we did not

encounter all coronary artery abnormalities reported for tetralogy of Fallot (5). However, if this systematic approach to assess coronary artery anatomy is followed, it is unlikely that a major abnormality would be missed. Our findings clearly indicate that the older the patient, the more difficult it is to demonstrate coronary artery anatomy by echocardiography (Table 1). Therefore, we recommend that coronary artery anatomy be studied as early as possible by two-dimensional echocardiography, preferably at the time the diagnosis of tetralogy of Fallot is first made.

Conclusions. Two-dimensional echocardiography can be reliably used to diagnose coronary artery anatomy in tetralogy of Fallot. The presence of an abnormal coronary artery should alter the plan of management and preparation so that selective coronary angiography is performed before corrective surgery if the abnormality is not well seen on left ventricular or aortic root angiograms. In the future, we may be able to avoid the additional risks of angiography and rely solely on echocardiography to manage patients with tetralogy of Fallot.

#### Addendum

Since the original submission of this manuscript for publication, Berry et al. (20) reported their experience with two-dimensional echocardiographic assessment of coronary anatomy in tetralogy of Fallot. Our study and theirs agree on the incidence of coronary artery abnormalities and the importance of two-dimensional echocardiographic evaluation of the coronary anatomy before angiography. Important additions in our study include the ability to differentiate between a large conal branch originating from the right coronary artery and paired anterior descending arteries originating from both right and left coronary arteries. The differentiation is made possible by utilizing the previously not described modified apical view. Also, we were able to demonstrate small conal arteries and to differentiate between small and large conal arteries.

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