DIAGNOSTIC METHODS

CONGENITAL HEART DISEASE

Diagnosis of coronary artery anatomy by two-dimensional echocardiography in patients with transposition of the great arteries

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ABSTRACT With the increasing popularity of the Jatene procedure for the treatment of common or d-transposition of the great arteries (d-TGA), the preoperative definition of coronary artery anatomy in d-TGA has assumed great importance. Consequently, the reliability of two-dimensional echocardiography for determining the coronary artery anatomy was studied in 32 infants with d-TGA. Surgical observation of the coronary anatomy was used to assess the accuracy of the echocardiographic diagnosis. The coronary arteries were visualized in 29 of 32 patients (90%), predominantly with the use of parasternal and apical views. In the three remaining patients visualization of the coronary arteries was inadequate to allow determination of their anatomy. The coronary artery anatomy was correctly predicted in 25 of the 29 patients in whom the coronary arteries were visualized. The anatomic patterns included usual coronary anatomy for d-TGA (n = 16), left circumflex coronary from the right coronary artery (n = 6), single right coronary artery (n = 1), single left coronary artery (n = 1), and inverted origin of the coronary arteries (n = 1). The errors in the remaining four patients were (1) false-negative diagnosis of origin of the left circumflex coronary from the right coronary artery (n = 1); (2) false-positive diagnosis of origin of the left circumflex coronary from the right coronary artery (n = 1), and (3) diagnosis of origin of the left circumflex coronary from the right coronary artery when the correct diagnosis was single right coronary artery (n = 2). In conclusion, the coronary arteries could be visualized in 90% of patients with d-TGA and the anatomy was defined correctly in 86% of patients in whom the coronary arteries could be visualized. We anticipate that more experience will further improve the accuracy of two-dimensional echocardiography for defining coronary anatomy in patients with d-TGA.


IN 1975 Jatene reported the first successful anatomic correction of common or d-transposition of the great arteries (d-TGA) performed by switching the aorta and pulmonary artery and reimplanting the coronary arteries.1 Since then, several centers have reported excellent results using this approach for the surgical correction of d-TGA.2-5 Since coronary artery anatomy is highly variable in individuals with d-TGA,2-6-9 accurate preoperative definition of the coronary anatomy is extremely valuable in patients for whom the arterial switch operation is contemplated.2 Consequently, aortic root or selective coronary angiography is usually performed before surgery to define the coronary artery anatomy.10

Aortic root angiography may not adequately delineate the coronary anatomy and selective coronary angiography may be difficult to perform in the neonate, especially the neonate with d-TGA. Therefore, we investigated the reliability of two-dimensional echocardiography for defining the coronary artery anatomy in d-TGA.

Subjects and methods

Between August 1984 and February 1986, 61 consecutive patients underwent surgical repair of d-TGA at The Children’s Hospital, Boston. In 21 of these patients the preoperative anatomy and/or physiology was not suitable for an arterial switch operation. All of these patients underwent intra-atrial or intraventricular repair of d-TGA. The coronary artery anatomy was not specifically investigated preoperatively, nor was it described in the operative report in these 21 patients. A preoperative two-dimensional echocardiographic examination was not performed in another eight of the 61 patients.

The remaining 32 patients (age 1 day to 11 months, mean 56
days) are the subjects of this report. A two-dimensional echocardiographic examination was performed in all patients and the coronary anatomy was clearly described in the operative note or at autopsy. The diagnoses in these 32 patients were simple D-TGA (n = 24), D-TGA with ventricular septal defect (n = 5), D-TGA with ventricular septal defect and pulmonary stenosis (n = 2), and D-TGA with pulmonary stenosis (n = 1).

Echocardiography. The two-dimensional echocardiograms were obtained with an ATL Mark 600 or a Hewlett-Packard 77020 Cardiac Imager with a 5 MHz short- or medium-focus transducer. The examinations were recorded on ½ inch video-cassette tape for review in real time, slow motion, and frame by frame. Sedation with chloral hydrate was used in the patients when necessary.

The echocardiographic views that were most useful for imaging the coronary arteries included the parasternal short-axis view,11,12 the apical four-chamber view,12,13 a left oblique subxiphoid view obtained by rotating the transducer approximately 30 degrees clockwise from the long-axis view of the left ventricle, and a parasternal long-axis view angled to the left. The parasternal short-axis view at the level of the aortic root usually displayed the origins of the coronary arteries from the

![FIGURE 1. Parasternal views of the origins of the coronary arteries. Top and Middle left, Short-axis views through the aortic root showing the origin of the left and right coronary artery, respectively. Bottom left, Long-axis view angled to the right through the aortic root, an alternate method for visualizing the origin of the right coronary artery. The scale for all three panels is as shown in the top panel. Top right, Line drawing illustrating the short-axis imaging plane for displaying the origin of the left (A) and right (B) coronary artery. Bottom right, Imaging plane of alternate long-axis view for the right coronary artery. Ao = aorta; LCA = left coronary artery; PA = pulmonary artery; RA = right atrium; RCA = right coronary artery; a = anterior; l = left; r = right; s = superior.](image-url)
right and left posterior sinuses of Valsalva (figure 1). However, the origin of the right coronary artery was often best seen in a parasternal long-axis view through the aortic root (figure 1). Counterclockwise rotation from the parasternal short-axis view allowed the right coronary artery to be followed to the right atrioventricular groove.

The course of the left coronary artery could be followed by angling leftward from a parasternal long-axis view through the aortic root. When the scan plane was tangential to the left side of the aortic root, the origin and proximal portion of the left coronary artery were displayed in cross section. Further leftward angulation tracked the left coronary artery to its bifurcation into the left anterior descending and left circumflex coronary arteries (figure 2).

The apical four-chamber and left oblique subxiphoid views were useful for location of the left circumflex coronary when it arose from the right coronary artery or for location of the left coronary artery in most cases of single right coronary artery.

In 26 patients the echocardiographic description of the coronary artery anatomy was derived from the initial prospective interpretation. In the other six patients, the coronary artery anatomy was not described in the echocardiographic report. These six echocardiograms were reviewed blindly by one of the authors (S.P.S.) for description of coronary anatomy.

Results

Coronary artery anatomy. The coronary anatomy observed in the 32 patients with D-TGA is shown in figure 3.

FIGURE 2. Leftwardly angled parasternal long-axis views for imaging the left coronary artery and branches in usual coronary artery anatomy for D-TGA. Top left, A view through the anterior interventricular groove showing the left anterior descending coronary artery arising from the left coronary artery. Bottom left, Further leftward angulation demonstrates the left circumflex coronary artery in the left atrioventricular groove. Note that the anterior descending and circumflex coronary arteries are nearly perpendicular to each other. The scale for both panels is as shown in the top panel. Right, Line drawing illustrating the scan plane for displaying the left anterior descending (top) and left circumflex (bottom) coronary arteries. LAD = left anterior descending coronary artery; LCCA = left circumflex coronary artery; other abbreviations as in figure 1.
The orientation of the figures is that of a parasternal short-axis echocardiographic view. LAD = left anterior descending; LCCA = left circumflex coronary artery; RCA = right coronary artery.

**Echocardiographic diagnoses.** The coronary arteries could be imaged in 29 of the 32 patients (90%) by two-dimensional echocardiography. In the three remaining infants the coronary arteries were not imaged adequately. At surgery, all three were found to have the usual coronary anatomy accompanying D-TGA.

The coronary artery anatomy was diagnosed correctly by two-dimensional echocardiography in 25 of the 29 patients (86%) in whom the coronary arteries could be visualized.

Usual coronary anatomy for D-TGA was correctly diagnosed in 16 of 17 patients (11/12, prospective; 5/5, blinded review). In the remaining patient, origin of the left circumflex coronary artery from the right coronary artery was diagnosed incorrectly.

Echocardiographic diagnostic criteria for usual coronary artery anatomy accompanying D-TGA included:

1. Visualization of separate origins of the right and left coronary arteries from the right and left posterior sinuses of Valsalva, respectively, in parasternal short-axis and/or long-axis views (7/7).

2. Identification of a coronary artery passing between the pulmonary root and the mitral valve in a parasternal short-axis, apical four-chamber, or left oblique subxiphoid view (9/9) (figure 2), with or without definition of its origin.

3. Visualization of the origin of the left circumflex artery from the right coronary artery a few millimeters after its origin and passing posteriorly and leftward in parasternal short-axis or subxiphoid left oblique view (5/6) (figure 3).

4. Inability to visualize the normal bifurcation of the left coronary artery in either the parasternal short-axis view or parasternal long-axis view angled to the left. This is a weak criterion because it was present in only 50% of patients with usual coronary artery anatomy for D-TGA.

Single right coronary artery was correctly diagnosed in one of three patients by two-dimensional echocardiography. In the other two patients the incorrect diagnosis of origin of the left circumflex from the right coronary artery was made.

Echocardiographic diagnostic criteria for single right coronary artery included:

1. Identification of a single large coronary ostium from the right posterior sinus of Valsalva in the parasternal short- and/or long-axis views (3/3).

2. Visualization of the left coronary artery arising from the right a few millimeters from its origin and passing leftward behind the pulmonary root in parasternal short-axis or apical four-chamber view (3/3).
(3) Visualization of the bifurcation of the left coronary artery into the left anterior descending and left circumflex coronary arteries in a parasternal short-axis view and/or a leftwardly angled parasternal long-axis view (1/3) (figure 5).

Single left coronary artery was correctly diagnosed by two-dimensional echocardiography in the only patient in our series with this anatomy. In the parasternal short-axis view, a single large coronary artery was seen arising from the left posterior sinus of Valsalva and there was no coronary arising from the right posterior sinus. The single coronary artery gave rise sequentially to the left circumflex, left anterior descending, and right coronary arteries (figure 6). The right coronary artery passed anterior to the aorta to reach the right atrioventricular groove, as seen in parasternal short-axis view (figure 6). The courses of the left anterior descending and left circumflex coronary arteries were best seen in parasternal short-axis or leftwardly angled parasternal long-axis view. No coronary artery was seen between the pulmonary root and mitral valve.

Inverted origin of the coronary arteries was correctly diagnosed by two-dimensional echocardiography in the only patient in our series with this anomaly. In the parasternal short-axis view, a coronary ostium was seen in each of the posterior sinuses of Valsalva. The coronary artery arising from the left posterior sinus (figure 7) then passed anterior to the aorta to reach the right atrioventricular groove (right coronary artery), while the coronary artery arising from the right posterior sinus passed (figure 7) to the left between the pulmonary root and mitral valve.

FIGURE 4. Parasternal and subxiphoid views illustrating origin of the left circumflex coronary artery from the right coronary artery. Top left, Parasternal short-axis view showing the left circumflex artery originating from the right coronary artery and passing leftward posterior to the pulmonary root. Middle left, Subxiphoid left oblique view illustrating the same finding. Bottom left, A more posterior subxiphoid left oblique view demonstrating a greater length of the left circumflex coronary artery. The scale is as shown in the top panel. Right, Line drawing showing parasternal short-axis scan plane for displaying the origin of the left anterior descending from the right coronary artery. LA = left atrium; LV = left ventricle; i = inferior; other abbreviations as in figures 1 and 2.
right coronary artery (0 to 6%). We found a similar distribution among our patients; only two of the rarer patterns were observed.

**Diagnostic criteria.** The most reliable criteria for diagnosing usual coronary artery anatomy in patients with d-TGA are detection of two coronary ostia in the appropriate sinuses of Valsalva, absence of a coronary artery between the pulmonary root and mitral valve, and visualization of the bifurcation of the left coronary artery. The diagnosis was made correctly in nine of our patients fulfilling all of these criteria. It is important to avoid confusion of a diagonal branch with the origin of the left circumflex. The diagonal coronary artery branches at an acute angle and runs nearly parallel to the anterior descending coronary (figure 8), while the left circumflex coronary forms a right angle with the left anterior descending coronary artery (figure 2).

**Discussion**

Encouraging reports of the use of two-dimensional echocardiography for evaluation of congenital coronary artery anomalies, including coronary artery fistulas, anomalous origin of a coronary artery from the pulmonary trunk and origin of the left coronary artery from the right coronary artery, and acquired coronary abnormalities in Kawasaki's syndrome led us to evaluate the use of two-dimensional echocardiography for defining the coronary anatomy in d-TGA. Inability to image the coronary arteries in three of our patients resulted from overlying lung tissue that prevented acquisition of the appropriate parasternal view or from central location of the heart, with the sternum overlying the region of interest. Thus, it appears that this technique may not be applicable to all patients.

Although 28 variations of coronary artery anatomy in d-TGA have been described, three patterns account for the vast majority of cases: (1) usual coronary artery anatomy associated with d-TGA (57% to 71%), (2) origin of the left circumflex coronary artery from the right coronary artery (16% to 25%), and (3) single

**FIGURE 5.** Parasternal short-axis view through the aortic root in a patient with single right coronary artery. In this case, the left coronary passes posterior to the pulmonary artery and bifurcates into the left anterior descending and left circumflex coronary arteries. Abbreviations as in previous figures.

**FIGURE 6.** Parasternal short-axis view illustrating single left coronary artery. Top, Parasternal short-axis view showing the origin of a large diameter but very short left main coronary. This vessel bifurcates into a left circumflex coronary and a coronary trunk from which arise the left anterior descending and right coronary arteries. The right coronary passes anterior to the aorta to reach the right atrioventricular groove. Bottom, A slightly more apical cross section showing more clearly the origin of the left anterior descending coronary. The scale is the same for both panels. Abbreviations as in previous figures.
In eight other patients the diagnosis of usual coronary artery anatomy for d-TGA was made on the basis of the first two criteria only (bifurcation of the left coronary artery not visualized). The diagnosis was correct in seven of these eight patients, while in the remaining patient the left circumflex coronary artery originated from the right coronary artery.

The most reliable criteria for diagnosing origin of the left circumflex from the right coronary artery were visualization of two coronary arteries arising from the appropriate sinus of Valsalva and detection of a coronary artery between the pulmonary root and the mitral valve. When both criteria were fulfilled, the correct diagnosis was made in six of six patients. Origin of the left circumflex from the right coronary artery was diagnosed incorrectly in three of three patients in whom both criteria were not present. In one patient a large sinus node artery noted at surgery was mistaken for the left circumflex coronary artery arising from the right coronary artery. While the arterial origin and proximal course are similar in these two conditions, the left circumflex artery passes leftward behind the pulmonary root toward the left atrioventricular groove while the sinus node artery courses posteriorly and superiorly toward the atrial septum. This error was made before appreciation of the diagnostic importance of observation of a coronary artery between the pulmonary root and mitral valve.

In two patients with single right coronary artery the diagnosis of origin of the left circumflex from the right coronary artery was made, although only a single coronary ostium could be identified.

This report indicates that two-dimensional echocardiography is an extremely promising technique for definition of coronary artery anatomy in patients with d-TGA. While these results are gratifying, we anticipate that further improvement in diagnostic accuracy will come with more experience.

References
for anatomical correction of transposition of the great arteries with intact interventricular septum. Lancet 1: 1275, 1977
Diagnosis of coronary artery anatomy by two-dimensional echocardiography in patients with transposition of the great arteries.
L Pasquini, S P Sanders, I A Parness and S D Colan

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