Diagnosis of Intramural Coronary Artery in Transposition of the Great Arteries Using Two-dimensional Echocardiography

Luciano Pasquini, MD; Ira A. Parness, MD; Steven D. Colan, MD; Gil Wernovsky, MD;
John E. Mayer, MD; Stephen P. Sanders, MD

Background. An intramural coronary is an uncommon but potentially significant risk factor for transfer of the coronary arteries as part of the arterial switch operation for transposition of the great arteries. Preoperative diagnosis is advantageous because it helps prevent accidental injury to the intramural coronary artery during transection of the aortic root and excision of the coronary artery ostium from the aorta. Therefore, we investigated the reliability of two-dimensional echocardiography for detecting an intramural coronary artery in infants with d-transposition of the great arteries.

Methods and Results. All infants with d-transposition of the great arteries who underwent echocardiography and primary surgical repair at this institution between January 1987 and June 1992 were identified by search of the cardiology data base. From this group, patients were diagnosed with an intramural coronary artery by review of the echocardiographic, surgical, and autopsy reports. Among 435 infants with transposition, 29 infants were diagnosed as having an intramural coronary artery. In 27 cases, the diagnosis was confirmed at surgery or autopsy, and there were two false-positive echocardiographic diagnoses (specificity, 99.5%). Twenty of the 27 patients with an intramural coronary artery were correctly diagnosed prospectively by echocardiography (sensitivity, 75%), including 17 of 23 patients with an intramural left coronary artery or left anterior descending coronary artery and 3 of 4 patients with an intramural right coronary artery. Two primary diagnostic criteria were identified: a major coronary artery arising from the contralateral septal sinus, near the usually intercoronary commissure, and a course for this vessel within the posterior aortic wall between the great arteries, creating a “double-border” appearance. Retrospective review using these criteria identified 26 of the 27 intramural arteries with no false-positive diagnoses.

Conclusions. We conclude that coronary echocardiography is a very promising technique for detecting an intramural coronary artery in transposition of the great arteries. Careful prospective application of the identified diagnostic criteria should greatly improve the diagnostic accuracy. (Circulation. 1993;88:1136-1141.)

Key Words • echocardiography • arteries

Coronary artery anatomy in common transposition of the great arteries (D-TGA) is highly variable.1-6 Transplantation of the coronary arteries is an integral and critical part of the arterial switch operation,7-9 which is currently considered to be the treatment of choice for most patients with D-TGA.10-12 Most coronary arterial patterns seen in D-TGA pose no problem for performance of the arterial switch operation.13-15 However, the presence of an intramural segment of a coronary artery can present a significant challenge for the surgeon.11,16-19 Preoperative diagnosis of an intramural coronary artery is important in patients otherwise suitable for an arterial switch operation. Selective coronary angiography can be difficult to perform in the neonate with D-TGA, even using a transvenous approach.20 Aortic root angiography does not always adequately delineate an intramural segment, even using the recently described modifications that enhance diagnostic accuracy.21,22 Therefore, we investigated the reliability of two-dimensional echocardiography for detecting an intramural coronary artery in infants with D-TGA. Although we have reported the echocardiographic diagnosis of this important coronary arterial pattern in a few patients,23 to our knowledge this is the first detailed description of the echocardiographic appearance.

Methods

Patients

All patients with D-TGA and a ventricular d-loop24 who underwent echocardiographic examination and corrective surgery or autopsy at this institution between January 1987 and June 1992 were identified through a search of the computerized cardiology data base. Patients with the following characteristics were selected: situs solitus of viscera and atria, two patent atroventricular valves, two ventricles of approximately normal size,
and two patent semilunar valves. Patients who had undergone a corrective heart operation previously were excluded. From this large group, all patients diagnosed with an intramural course of either the left or right coronary artery by echocardiography or at surgery or autopsy were selected as the subjects of this study.

Echocardiography

A complete two-dimensional echocardiogram was performed in each patient using either a Hewlett-Packard 77020 or Sonos 1000 cardiac imager or an Acuson 128 Cardiac Imager equipped with a 7.5- or 5-MHz transducer focused appropriately for body size. The examinations were recorded on ½-in. videocassette tape for review in real-time, slow motion, or stop-frame modes. Sedation with chloral hydrate was used when necessary.

Coronary echocardiography was performed using the views and methods described previously. In general, a modified parasternal short-axis view through the aortic root was used to display the origins and proximal segments of the coronary arteries. Slight clockwise rotation (10 to 30°) of the transducer improved visualization of the left coronary artery, whereas a similar amount of counterclockwise rotation was useful to image the right coronary artery. The left anterior descending coronary artery usually was best seen by angling toward the left shoulder in a parasternal long-axis view until the sector plane intersected the anterior interventricular groove tangentially.

The echocardiographic diagnosis recorded prospectively on the initial echocardiographic report was used for analysis. Although the echocardiogram of each patient was reviewed as part of this study, the original prospective interpretation was used for analysis of diagnostic accuracy.

Surgical Observation

The surgical summary for each patient was reviewed to determine the coronary artery pattern observed at surgery. If the description was not completely clear, the summary was reviewed with the surgeon.

Autopsy Observation

The autopsy report for each patient who underwent post mortem examination was reviewed to determine the coronary arterial pattern. In addition, each specimen was examined by two of the authors.

Terminology

Intramural coronary artery. The intramural coronary artery is a coronary artery, the proximal portion of which courses within the wall of the aorta so that the medial layers of the coronary artery and aorta are not separated by an adventitial layer. The ostium of the coronary artery is separated from the site at which the coronary artery exits through the aortic adventitial layer by the length of the intramural segment.

Aortic sinuses of Valsalva. The two aortic sinuses of Valsalva adjacent to the aorticopulmonary septum usually contain the coronary ostia. The more leftward (and usually anterior) of these is the left septal (facing) sinus; the more rightward (and usually posterior) of these is the right septal (facing) sinus.

Approval for this study was granted by the Human Investigation Committee on July 13, 1992.

Results

Four hundred sixty-one patients were identified with D-TGA who underwent echocardiography and surgery during the study period. An adequate description of the coronary artery anatomy was available from surgery and/or autopsy in 435 of these patients. Twenty-seven of these patients had an intramural coronary artery by direct surgical inspection or at autopsy and constitute the study group. The ages of the 27 patients ranged from 1 day to 8 months (mean, 20 days), and the weights ranged from 1.8 to 8.5 kg (mean, 3.9 kg). Nineteen patients were male, and 8 were female.

Echocardiography

The coronary arteries could be imaged in all 27 patients by echocardiography. The intramural coronary
artery was correctly diagnosed prospectively in 20 of the 27 patients with seven false-negative diagnoses (sensitivity, 74%) (Fig 1). There were two false-positive diagnoses of intramural coronary artery by echocardiography in the 435 patients with surgical or autopsy confirmation (specificity, 99.5%). (An intramural coronary artery was observed by echocardiography in four additional patients, but the anatomy was not examined at surgery or autopsy in these patients, and they were not included in the analysis.) The incidence of an intramural coronary artery in our patients with D-TGA was 27 of 435 (6.2%). (If the 4 additional patients are included, then the incidence is 31 of 439 patients [7.1%].)

An intramural left coronary artery arising from the right septal sinus with normal branching pattern (Figs 1A and 2) was correctly diagnosed in 12 of the 16 patients with this anatomy. An intramural left anterior descending coronary artery from an ostium in the right septal sinus with the left circumflex artery arising from the right coronary artery and pursuing a retropulmonary course (Fig 1B) was correctly diagnosed in 4 of 6 patients. An intramural left anterior descending coronary artery arising from an ostium in the right septal sinus with the left circumflex artery arising from the left circumflex artery (Fig 1C) were correctly diagnosed in the only patient with this anatomy. An intramural right coronary artery arising from an ostium in the left septal sinus (Figs 1D and 3) was correctly diagnosed in both patients. An intramural right coronary artery from an ostium in the left septal sinus, with the left circumflex artery arising from the right coronary artery and pursing a retropulmonary course (Fig 1E), was correctly diagnosed in the single patient with this anatomy. Finally, an intramural right coronary artery arising from the right septal sinus near the intercoronary commissure (Fig 1F) was missed in the only patient with that anatomy.

The two false-positive diagnoses were intramural right coronary artery from an ostium in the left septal sinus in a patient with origin of the right coronary artery high in the right septal sinus, immediately adjacent to the intercoronary commissure, with no intramural segment seen at surgery, and intramural right coronary artery from an ostium in the right septal sinus in a patient with usual coronary artery anatomy for D-TGA and no intramural segment seen at surgery.

Diagnostic Criteria

Based on our experience over the study period and especially the retrospective review of the echocardiograms of the study patients, we identified three echocardiographic criteria that are important in the diagnosis of an intramural coronary artery.

Identification of a major coronary artery coursing between the two arterial roots. This criterion was present in all 20 cases correctly diagnosed prospectively by echocardiography and, in retrospect, in 6 of the 7 patients in whom the diagnosis was missed by echocardiography (Figs 2 and 3). This finding was also present in one case of a false-positive diagnosis of intramural right coronary artery from the left septal (facing) sinus. With this criterion alone, 26 of the 27 cases could have been diagnosed correctly with one false-negative and one false-positive diagnosis.

FIG 2.  Echocardiographic parasternal short-axis views through the aortic root in an infant with an intramural left coronary artery (LCA). Left, Ostium of the right coronary artery (RCA) is seen centrally located in the right facing or septal sinus of Valsalva (RFS). Note that there is no significant displacement of the endothelial orifice of the coronary artery and the adventitial exit of the coronary artery. Middle, Origin of the LCA from the RFS is seen immediately adjacent to the usually intercoronary commissure. Note the “double-border” appearance of the posterior aspect of the aortic root where the proximal LCA runs in the wall of the aortic root. The endothelial orifice of the coronary artery is displaced several millimeters to the right of the adventitial exit of the coronary artery, seen best on the right. Right, Exit point of the intramural coronary from the posterolateral aspect of the aortic root and the bifurcation of the LCA into the left anterior descending coronary (LAD) and left circumflex (LCCA) arteries. Ao indicates aorta; LFS, left facing or septal sinus of Valsalva; and PA, pulmonary artery.
Origin of a coronary artery from the contralateral septal (facing) sinus adjacent to the posterior (usually intercoronary) commissure at an acute angle to the aortic wall and with the proximal segment of the coronary artery running perpendicular to the posterior commissure and parallel to the aortic wall. This causes a “double-border” appearance of the posterior aspect of the aortic root (Figs 2 and 3). This finding was present along with the above criterion (it appears that this criterion cannot be present without the above criterion) in all 20 cases diagnosed correctly and in 6 of the 7 cases missed prospectively. This finding was not present in either of the false-positive cases. Therefore, use of both criteria could have resulted in correct diagnosis of all except one case with no false-positive diagnoses.

Origin of a major coronary artery high in the sinus of Valsalva, near the supravalvar ridge, as seen in a long-axis view through the aortic root. This finding was present in 16 of the 20 patients diagnosed correctly and in 6 of the 7 patients in whom the diagnosis was missed prospectively (Fig 4). The origin of the left coronary artery was high in one of the false-positive cases as well.

**Time Period**

Four of the 7 cases in which the diagnosis of intramural coronary artery was missed were among the first 10 cases seen (40% error rate) during the first 2 years of the study period. During the most recent 3 years, only 3 of 16 cases were missed (17% error rate), including the patient with an intramural segment of the right coro-

**Associated Anatomic Features**

The ventricular septum was intact in 19 of the 27 patients with an intramural coronary artery, and a ventricular septal defect was present in the remaining 8 patients. The aorta was anterior and rightward of the pulmonary artery in 17 patients and directly anterior to the pulmonary artery in 9 patients, and the great arteries were side-by-side in 1 patient. Three patients had other major associated anomalies: interruption of the aortic arch in 2 patients and coarctation of the aorta in the other patient.

**Surgical Results**

Eleven of the 27 patients with an intramural coronary artery underwent a Senning operation with four hospital deaths, 12 patients underwent an arterial switch operation with no hospital deaths, 3 patients underwent the Aubert modification of the arterial switch operation\(^\text{27}\) with two hospital deaths, and 1 patient survived a Rastelli operation. Both patients in whom a false-positive diagnosis of intramural coronary artery was made by echocardiography successfully underwent an arterial switch operation after careful inspection of the coronary artery anatomy at surgery.

**Discussion**

Although the existence,\(^\text{28,29}\) clinical implications,\(^\text{28,30,31}\) and surgical repair\(^\text{32,33}\) of an intramural coronary artery in patients with normally related great arteries have been known for some time, this coronary artery pattern was
described in patients with D-TGA only recently,\textsuperscript{26} with an incidence of 3%. Our current study found an incidence of 6% to 7%. There are two primary reasons for the higher incidence rate in our study. First, our population is larger than those in the previous reports and is comprised of consecutive, living patients who were very young at the time of the echocardiogram. Second, referral patterns to this hospital may have introduced some selection bias toward more complex cases. Nonetheless, the incidence of an intramural coronary artery appears to be higher than previously suspected and should be considered and looked for in all patients in whom an arterial switch operation is contemplated.

Anatomic Considerations

Two anatomic patterns of intramural coronary artery accounted for the majority of our cases: intramural left coronary artery from the right septal sinus of Valsalva and intramural left anterior descending coronary artery from the right septal sinus of Valsalva. An intramural right coronary artery from the left septal sinus of Valsalva, although rare in our series, has been reported previously.\textsuperscript{26} However, an intramural right coronary artery arising from the appropriate right septal sinus is presented for the first time to our knowledge.

The most common spatial relationship between the great arteries in our patients with an intramural coronary artery was the aorta anterior and to the right or directly anterior to the pulmonary artery. Unlike a previous report,\textsuperscript{26} side-by-side great arteries occurred in only one of our patients.

Diagnostic Criteria

The most reliable criteria for diagnosing an intramural coronary artery in D-TGA appear to be a coronary artery coursing between the great arteries from the contralateral septal sinus of Valsalva with the “double-border” appearance of the posterior aortic root caused by the intramural segment of the coronary artery in the wall of the aorta. Using this set of findings for diagnosing an intramural coronary artery during a retrospective review of our cases led to a single false-negative diagnosis and no false-positive diagnoses. The false-negative case was the intramural right coronary artery arising from the appropriate right septal sinus of Valsalva (Fig 1F). We were unable to recognize the intramural segment of this coronary artery even in retrospect.

Simply identifying a coronary artery between the two arterial roots does not necessarily imply an intramural segment. One of our false-positive cases had this finding only. Similarly, origin of the coronary artery high in the sinus of Valsalva, near the supravalvar ridge, is suggestive but not diagnostic of an intramural coronary artery. In some of our cases of intramural coronary artery, the ostium was low in the sinus of Valsalva and some normal coronary arteries arose high in the sinus.

Surgical Considerations

The mechanism of sudden death in children and young adults with an intramural coronary artery and normally related great arteries is thought to be myocardial ischemia resulting from compression of the coronary artery as it passes between the great arteries or from occlusion of the ostium.\textsuperscript{34,35} Sudden death has been reported in patients with D-TGA and a single coronary artery.\textsuperscript{36} Given the relatively high incidence of an intramural coronary artery in D-TGA, it is possible that some cases of sudden unexpected death occurring late after Senning or Mustard repair of D-TGA could be due to an intramural coronary artery. If so, then coronary transplantation could be even more important in patients with an intramural coronary artery. Although surgical techniques have been reported for transplanting an intramural coronary artery in infants with D-TGA,\textsuperscript{16,18} this does not always appear to be possible, especially when the ostia of the coronary arteries are too close together (or in common) to allow separate cuffs to be taken from the aortic root.\textsuperscript{16} The techniques described by Aubert and colleagues\textsuperscript{27} and Takeuchi and Katogi\textsuperscript{37} can be applied when coronary artery transplantation is considered unsafe. Unfortunately, the theoretical mechanisms for ischemia and sudden death remain after these procedures.

Preoperative diagnosis of intramural coronary artery in D-TGA is very important for two reasons. First, foreknowledge that an intramural segment is present helps to avoid injury to the coronary artery during transection of the aortic root and excision of the coronary arterial button from the aorta.\textsuperscript{16} Second, if the anatomy does not appear favorable for an arterial switch operation, then deferral of surgery can be considered. Our own preference remains to undertake surgery in the neonatal period, and if the coronary artery pattern is not favorable, then a neonatal Senning operation is performed.

Conclusions

This report indicates that echocardiography is an extremely promising technique for defining coronary artery anatomy in patients with D-TGA and an intramural coronary artery. We anticipate further improvement in diagnostic accuracy with more experience. This anomaly appears to be more common in patients with D-TGA than previously recognized, making familiarity with the anatomy and echocardiographic appearance even more important. Because of the theoretical advantages of transplanting the intramural coronary artery, surgical techniques should be explored that would be applicable to the more difficult cases.

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