Anomalous origin of a coronary artery from the opposite sinus with an interarterial course between the great arteries (AOCA) is associated with myocardial ischemia and sudden cardiac death, particularly in adolescents and young adults (1–18). There have been several recent reports of prospective identification of this anomaly by transthoracic echocardiography (TTE) (19–27). This information is obviously critical in a patient who presents with symptoms of myocardial ischemia, but it is less clear what the course of action should be in patients who have this anomaly identified without symptoms. In this report, we discuss our experience with AOCA, focusing on TTE findings and the surgical approach to repair.

**METHODS**

The cardiology database at Children’s Hospital of Wisconsin was reviewed for the diagnosis of coronary artery anomalies. From this patient population, medical records, echocardiographic and angiographic imaging, and surgical reports were reviewed to identify patients with AOCA.

**Findings.** **Patient Population.** From September 1997 through August 2002, 10 patients were identified with AOCA. All patients were children/adolescents with a median age at the time of diagnosis of 12.2 years and a range of three months to 20 years (Table 1). Only 3 of the 10 patients were <10 years at the time of presentation. The median weight was 45.3 kg with a range from 4 to 72 kg. There were eight males and two females.

**Patient Presentation.** Symptoms of cardiac ischemia were present in 4 of the 10 patients and resulted in...
cardiology evaluation that led to the diagnosis of an anomalous coronary artery (Table 1). A 13.5-year-old female presented with repeated episodes of exercise-induced syncope during basketball practice. A 14.9-year-old male presented with exercise-induced syncope during track and field practice, and a 15.9-year-old male presented with sudden cardiac death while playing “pickup” basketball. Finally, a 20.1-year-old male had exercise-induced syncope and chest pain while doing calisthenics as a Navy recruit during training; TTE identified the coronary artery anomaly prospectively in three of the four patients. The 20-year-old was found to have electrocardiogram and enzyme evidence of myocardial infarction in a local emergency room and underwent emergent cardiac catheterization at an outside institution with the diagnosis made by that technique. The other six patients had TTE for suspected congenital heart disease or musculoskeletal chest pain and were found serendipitously to have AOCA.

**ECHOCARDIOGRAPHIC/ANATOMIC FINDINGS.** Four of the 10 patients had anomalous origin of the right coronary artery from the left sinus of Valsalva with the anomalous coronary coursing between the great arteries (Table 1). All four of these patients had an intramural course of the anomalous right coronary artery within the anterior aortic wall. All were asymptomatic at the time of presentation, and all were prospectively diagnosed by TTE using Sequoia ultrasound equipment (Siemens AG, Mountain View, California). The anomalous coronary was best imaged from a short-axis view in the plane of the aortic root at the level of the coronary artery origins. The anomalous right coronary artery was initially suspected by two-dimensional imaging as a vessel running within the anterior sinus of Valsalva (Fig. 1A). Color Doppler flow mapping confirmed the presence of an anomalous right coronary artery as a linear diastolic flow signal within the wall coursing toward the right sinus of Valsalva (Fig. 1B); flow in the anomalous vessel was best visualized when the color Doppler Nyquist limit was lowered to 20 to 40 cm/s.

Six of the 10 patients had anomalous origin of the left coronary artery from the right sinus of Valsalva with the anomalous coronary coursing between the great arteries (Table 1). Five of those six patients had an intramural course of the left coronary artery within the anterior aortic wall (Fig. 2), and one patient had an intramyocardial course within the muscular sulcus between the great arteries (Fig. 3). Five of the six patients were prospectively diagnosed by TTE using two-dimensional imaging and color Doppler techniques; the other patient was diagnosed by cardiac catheterization as discussed in the prior text. Four of the six patients had symptoms of myocardial ischemia at the time of diagnosis.

### RESULTS

**Surgical repair.** Surgical repair was performed in eight of the 10 patients at a mean age of 13 ± 4.7 years (Table 1); the four patients who presented with symptoms of myocardial ischemia as well as four of the asymptomatic children. Two of the asymptomatic children had an associated ventricular septal defect (VSD) and had surgical repair of the anomalous coronary at the time of the VSD closure at ages four and 11 years. The other two asymptomatic patients had elective repair of the AOCA at ages 10 and 14 years because of concerns about the potential for sudden death associated with this coronary anomaly. The two youngest patients in the series (now 4.5 and 6 years of age) have remained asymptomatic and have not yet had surgical intervention. The anomalous coronary origin was confirmed by surgical inspection in all eight cases, and ostial stenosis at the origin of the anomalous coronary was identified by surgical inspection in four of the eight. In addition to TTE, five of the

**Table 1. Patient Characteristics, Anatomic Diagnoses, and Surgical Technique**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yrs)</th>
<th>Weight (kg)</th>
<th>Gender</th>
<th>AOCA</th>
<th>Symptoms</th>
<th>Dx</th>
<th>Coronary Course</th>
<th>Associated CHD</th>
<th>Surgery</th>
<th>Surgical Technique</th>
<th>Ostial Stenosis</th>
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<tr>
<td>1</td>
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<td>3.4</td>
<td>M</td>
<td>RCA</td>
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<td>TTE</td>
<td>Intramural</td>
<td>VSD</td>
<td>No</td>
<td>Unroofing</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>13.5</td>
<td>52.5</td>
<td>F</td>
<td>LCA</td>
<td>Syncope</td>
<td>TTE</td>
<td>Intramural</td>
<td>None</td>
<td>Yes</td>
<td>Patch</td>
<td>Yes</td>
</tr>
<tr>
<td>3</td>
<td>14.9</td>
<td>53</td>
<td>M</td>
<td>LCA</td>
<td>Syncope</td>
<td>TTE</td>
<td>Intramyocardial</td>
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<td>Yes</td>
<td>Patch</td>
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<td>64.8</td>
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<td>Bicuspid aortic valve</td>
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</tr>
<tr>
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<td>19.6</td>
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<td>TTE</td>
<td>Intramural</td>
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<td>No</td>
<td>Unroofing</td>
<td>—</td>
</tr>
<tr>
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<td>36</td>
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<td>TTE</td>
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<td>VSD, MVP</td>
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<td>7</td>
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<td>Unroofing</td>
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<td>8</td>
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<td>72</td>
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<td>LCA</td>
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<tr>
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<td>F</td>
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<td>Intramural</td>
<td>VSD</td>
<td>Yes</td>
<td>Unroofing</td>
<td>No</td>
</tr>
</tbody>
</table>

AOCA = anomalous origin of coronary artery; Cath = cardiac catheterization; CHD = congenital heart disease; Dx = technique used for diagnosis; LCA = left coronary artery; MVP = mitral valve prolapse; RCA = right coronary artery; TTE = transthoracic echocardiography; VSD = ventricular septal defect.
surgical repair had an intramural course of the AOCA and underwent successful unroofing of the intramural portion to relocate the ostia in the appropriate sinus (Fig. 4). In each case, the intramural portion of the anomalous coronary was visualized within the anterior aortic wall, exiting the aortic wall near its normally anticipated origin from the appropriate sinus. The intramural segment was unroofed by excising the common wall between the anomalous coronary and the aorta until the coronary exited the aorta. This resulted in a large neo-orifice from the appropriate aortic sinus with a perpendicular course of the coronary as it exited the aortic root. The intima of the coronary at the neo-orifice was attached to the intima of the aorta wall using interrupted suture to prevent dissection. The affected coronary commissure in the area of the unroofing required resuspension in four of the seven patients because of disruption of the commissure by the unroofing incision (Fig. 4). All patients had good biventricular function by transthoracic echocardiography at completion of surgery after separation from cardiopulmonary bypass, and, in each case, an unobstructed orifice in the appropriate sinus could be visualized by imaging and color Doppler flow (Fig. 2C).

SURGICAL OUTCOME. All seven patients who underwent unroofing were asymptomatic at a median follow-up interval of 1.5 years with laminar flow into the proximal coronary by serial postoperative TTE. All but the 4-year-old patient have undergone postoperative ETT without evidence of myocardial ischemia, and three have had cardiac catheterization after surgery documenting a widely patent coronary neo-ostia. No patient who underwent the unroofing procedure is currently restricted from activities. Of the four patients who underwent commissure resuspension after unroofing, aortic insufficiency is trivial in three and absent in one by TTE at follow-up.

DISCUSSION

Coronary artery anomalies have been estimated to occur in 0.6% to 1.2% of the population based on reviews of large series of angiographic studies (28,29); these series likely underestimate the true incidence because many people never undergo angiography. Anomalous origin of the left coronary artery from the right sinus of Valsalva is quite rare (with an estimated incidence of 0.03% to 0.05%), but it is frequently associated with early cardiac death, especially during vigorous exercise (1–9). In patients with this anomaly who had exercised-induced sudden cardiac death, all were adolescents, and 50% were asymptomatic without previous cardiac complaints (3,4). Anomalous origin of the right coronary artery from the left sinus of Valsalva is more common (incidence estimated at 0.1%) and is also associated with sudden cardiac death (1–6,10–18). Sudden cardiac death has been described in 30% of patients identified with this anomaly, and the vast majority were adolescents who were asymptomatic before the episode (4). These reviews suggest that AOCA of either coronary artery carries a significant

Figure 1. Transthoracic echocardiography from a short-axis plane in a patient with anomalous origin of the right coronary artery from the left sinus of Valsalva and an intramural course of the anomalous coronary. The anomalous right coronary artery can be seen arising from the left sinus of Valsalva (A) near the origin of the left main coronary artery (LMCA) and coursing intramurally within the anterior aortic wall (small arrows) between the aorta (Ao) and the pulmonary artery (PA) towards the right sinus of Valsalva. Color Doppler imaging (B) shows the linear diastolic flow of the anomalous coronary within the anterior aortic wall (arrow), the red color signal confirms anomalous coronary flow towards the transducer anteriorly, consistent with the coronary originating from the left sinus and coursing towards the more anteriorly positioned right sinus. For the accompanying videos corresponding to Figure 1 (Videos 1 and 2), please see the July 2 issue of JACC at www.cardiosource.com/jacc.html.

eight surgical patients had undergone cardiac catheterization preoperatively, and none was suspected of having ostial stenosis by any imaging technique. Two of the four patients with ostial stenosis were asymptomatic and had normal exercise treadmill testing (ETT) before surgery; both of those patients had anomalous origin of the right coronary artery from the left sinus of Valsalva.

The one patient with an intramycardial course of AOCA had patch augmentation at the origin of the anomalous coronary at an outside institution and has been lost to follow-up. The other seven patients that underwent
risk of sudden cardiac death, particularly for the young athlete, and that symptoms are frequently absent before the sudden death episode.

Improvements in ultrasound technology now allow delineation of coronary artery anatomy in many patients using TTE. Prospective identification of anomalous origin of the left main coronary artery from the right sinus of Valsalva (20–25) and anomalous origin of the right coronary artery from the left sinus of Valsalva (19,26,27) have been described using TTE. Two-dimensional TTE frequently must be combined with color Doppler flow mapping to successfully identify anomalous origin of a coronary artery (19,20,30). In many cases, two-dimensional imaging may be misleading in patients with AOCA that have an intramural course because the anomalous coronary can appear to arise normally from the appropriate sinus as it exits the aortic wall (20). Color Doppler is particularly useful in diagnosing AOCA with an intramural course because the technique can give the additional information of direction of flow in the intramural segment. This helps in differentiating whether the anomalous coronary arises from the right or left sinus.

Surgical repair of AOCA has generally been reserved for patients with known symptoms of myocardial ischemia. Multiple surgical techniques have been utilized, including coronary bypass graft placement (31–35), reimplantation of the anomalous coronary to the appropriate sinus (36,37), and the unroofing procedure (38–41) described here. The unroofing procedure was initially reported by Mustafa (38) in 1981 and has been used for both anomalous left coronary artery from the right sinus of Valsalva (38–40) and anom-

Figure 2. Transthoracic echocardiography from a short-axis plane in a patient with anomalous origin of the left coronary artery from the right sinus of Valsalva and an intramural course of the anomalous coronary. The two-dimensional image (A) shows the anomalous left main coronary artery running intramurally within the anterior aortic wall (small arrows) between the aorta (Ao) and pulmonary artery (PA) before exiting the wall in the left sinus of Valsalva and giving rise to the left anterior descending coronary branch (LAD). Color Doppler imaging (B) shows the linear diastolic flow of the anomalous coronary artery within the anterior aortic wall (arrow); the blue color signal confirms anomalous coronary flow away from the transducer, consistent with the coronary originating from the right sinus and coursing towards the more posteriorly positioned left sinus. After surgical unroofing of the intramural segment (C), a large neo-orifice has been created in the left sinus (arrow) giving rise to the left coronary system. For the accompanying videos corresponding to Figure 2 (Videos 3 and 4), please see the July 2 issue of JACC at www.cardiosource.com/jacc.html.
alous right coronary artery from the left sinus of Valsalva (41) when the anomalous coronary artery has an intramural course. This technique has several advantages over other coronary repair techniques: 1) it relieves any ostial stenosis at the origin of the anomalous coronary by unroofing the common wall between the aorta and anomalous coronary to relocate the coronary ostia in the appropriate sinus; 2) it creates a large neo-orifice of the anomalous coronary in the appropriate sinus that arises perpendicularly, rather than obliquely, from the aortic root; and 3) it removes the intramural segment of the anomalous coronary. The mechanisms by which AOCA produce myocardial ischemia are not completely understood. Ostial stenosis, the oblique angle of origin of the anomalous coronary, and compression of the coronary between the great arteries are potential risk factors that can limit coronary reserve. In addition, the intramural course of the AOCA may result in deformation of the anomalous coronary within the wall of the aorta during periods of arterial hypertension. Because wall tension is determined by the radius of the vessel, the aorta will have greater wall tension than the intramural coronary within its wall. As aortic wall tension increases with increasing aortic pressure during exercise, the intramural coronary becomes more flattened, and coronary flow may be reduced to a point where myocardial oxygen requirements are not met. Unroofing the intramural segment eliminates that risk.

The management of asymptomatic patients with AOCA remains controversial. The risk of late coronary insufficiency after coronary repair must be weighed against the risk of sudden death. We have adopted a strategy that is dictated by the course of the anomalous coronary. We feel that all patients with an intramural course of the anomalous coronary should have surgical intervention utilizing the unroofing technique. We believe that this technique creates a permanent, unobstructed neo-orifice without the risk of late ostial stenosis because no circumferential suture line is placed, and that it is ideally suited for the patient with an intramural course of the anomalous coronary. In asymptomatic children, this procedure is done electively after the age of 10 because reports of sudden death with AOCA before adolescence have only been described in young infants with severe ostial stenosis (5,7,15,16). In patients with an intramycardial coronary, neither unroofing nor reimplantation is possible because of the fixed and remote nature of the anomalous coronary as it courses within the muscular sulcus between the great arteries (Fig. 3). The other surgical options are suboptimal because there is a significant risk of late graft failure with bypass grafting,
particularly in an adolescent or young adult, and patch augmentation does not relieve the interarterial course of the anomalous coronary. For those reasons, we would reserve surgical intervention in that patient group to those with signs of myocardial ischemia by history/ETT. We anticipate that newer imaging techniques will eventually delineate ostial stenosis, allowing better risk stratification in patients with AOCA and an intramyocardial course.

We conclude that TTE can be used for prospective identification of AOCA involving either the left or right coronary artery. Identification of this anomaly is important because of the significant risk of sudden death associated with it. Surgical repair can be successfully accomplished when the anomalous coronary has an intramural course within the anterior aortic wall by unroofing the intramural segment. Because the presence of symptoms is unreliable in predicting the risk of sudden death, we feel that all patients identified with AOCA and an intramural course should undergo repair using the unroofing technique, regardless of presentation.

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REFERENCES
