

Original Article

Anomalous Coronary Artery from the Opposite Coronary Sinus in Young Children

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ABSTRACT

Objective: To report the management of anomalous coronary artery (ACA) from the wrong aortic sinus and review the literature

Background: ACA is a rare congenital lesion. It is associated with sudden death, particularly in young adolescents.

Design: Retrospective study (1990 - 2004)

Methods: All patients with interarterial single coronary artery arising from the aortic sinus were studied. Seven patients were identified. Transthoracic and transesophageal echocardiography were performed using high frequency transducers. Other tests used were: A 12-lead echocardiogram, 24-hour Holter, coronary angiography, exercise and Dobutamine stress testing and single-photon emission computed tomographic myocardial scintigraphy (SPECT). Extracorporeal membrane oxygenation (ECMO) was used for severe left ventricular dysfunction.

Results: There were five boys and two girls. All had an

intramural course of the coronary arteries. Their age range was between 2 weeks to 15 years. In four out of seven cases, it was an anomalous left coronary artery from the right anterior aortic sinus (ALCA-RAS). The other three cases were of an anomalous right coronary artery from the left aortic sinus (ARCA-LAS). In one patient, Doppler evidence of the intramural coronary artery stenosis was documented at cardiac arrest and ECMO support was needed.

Conclusion: As part of first echocardiography study, all patients, regardless of their age, should have their coronary arteries screened in the echocardiography laboratories. Once coronary artery abnormalities are detected (i.e., arising from opposite sinus), efforts should be made to look for any evidence of ischemia either through noninvasive or invasive techniques. Anomalous left coronary origin from the right sinus of Valsalva can result in significant myocardial ischemia and infarction.

KEYWORDS: anomalous coronary artery, echocardiography, intramural coronary artery**INTRODUCTION**

The origin of one of the coronary arteries from the wrong aortic sinus of Valsalva with a proximal course between the aorta and pulmonary artery is a rare congenital lesion^[1-4] (Fig. 1). The incidence of ectopic coronary origin from the aorta in the population is unknown. Its estimates in cardiac catheterization laboratory populations approximate 0.6%^[5]. It is associated with life-threatening conditions or sudden death, particularly in young adolescents^[6-8] where the majority of cases are diagnosed at autopsy^[4,6] [9-11]. Some authors^[12] report that this anomaly is the second most common cause of sudden death among young athletes. The diagnosis is usually made either after an acute event, or more recently as an incidental finding on routine echocardiography screening for other forms of congenital heart disease. We report seven cases where routine screening by echocardiography detected this anomaly and discuss potential

management implications in this group. Also, we describe the findings in a case who presented with a sudden cardiac event.

METHODS

From our database (n Kuwait and Canada) we identified all cases of ALCA from the wrong aortic sinus between 1990-2004. All patients represented incidental cases that were being evaluated for reasons not associated with the abnormal coronary artery. The following tests were performed:

- **Echocardiography:** A complete transthoracic two-dimensional echocardiogram was performed in each case using an Advanced Technology Laboratory (HDI 5000) or a Hewlett Packard (HP 5500) imaging system utilizing high frequency transducers. In three cases, transesophageal echocardiography was utilized to obtain further images of the abnormal coronary artery as well as Doppler velocity profiles.

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Table 1: Summary of the cases of ACA

Case	Age	Sex	Diagnosis	Symptoms	12-Lead ECG	Echo-Cardio-graphy	Angio-graphy	DSE	Myocardial Perfusion scan	Outcome
1	2-weeks	M	ALCA-RAS	Murmur	Normal	TTE	No	No	No	Alive
2	4 years	F	ALCA-RAS	Cardiac arrest	Severe ST-depression & Left anterior hemi block	TTE&TEE	No	No	No	Death post surgery
3	10 years	M	ALCA-RAS	Murmur	Normal	TTE&TEE	Yes	Yes	Yes	Alive
4	14 years	M	ALCA-RAS	Pre-renal transplant assessment	Normal	TTE	Yes	No	Yes	Alive
5	15 years	M	ARCA-LAS	Near syncope	Normal	TTE&TEE	Yes	Yes	No	Alive
6	8 years	M	ARCA-LAS	Murmur	Normal	TTE	No	No	No	Alive
7	2-weeks	F	ARCA-LAS	Murmur	Normal	TTE	No	No	No	Alive

ALCA-RAS = anomalous left coronary artery from right aortic sinus, ARCA-LAS = anomalous right coronary artery from the left aortic sinus

DSE = dobutamine stress echocardiography, F = female, M = male, TEE = transesophageal echocardiography, TTE = transthoracic echocardiography

- 12-lead ECG: All patients had a resting electrocardiogram (ECG).

- 24-hour Holter: A Holter monitor was performed in four cases.

- Exercise testing: An exercise stress test was done in two cases.

- Coronary angiography: In three cases, selective coronary artery angiograms were done.

- Dobutamine stress test: This was done in two cases.

- Single-photon emission computed tomographic myocardial scintigraphy (SPECT): Performed with intravenous administration of ^{99m}Tc-Sestamibi during exercise and at rest 24 hours later in two cases.

Extracorporeal membrane oxygenation (ECMO) was used in one patient for severe left ventricular dysfunction presenting with evidence of myocardial infarction and life threatening arrhythmia leading to cardiac arrest.

RESULTS

Presentation and clinical features

Seven patients were identified (Table 1). Their ages ranged between two weeks to 15 years (median = 10 years) and there were five males and two females. Anomalous left main coronary artery from the right aortic sinus was identified in four cases and anomalous right coronary from the left sinus in three. There was only one death (case 2) during the course of the study.

In four cases, the reason for referral was a cardiac murmur, in one it was for evaluation of a patent ductus arteriosus in a neonate with subsequent surgical ligation (case 1). An episode of near syncope was the reason for referral (case 6), which was initially felt to be vasovagal. This patient had no complaints of chest pain or palpitations. In case 4 the patient had the abnormality detected as part of a pre-renal transplant assessment in a case with nephrosclerosis. The last patient (case 2) had a

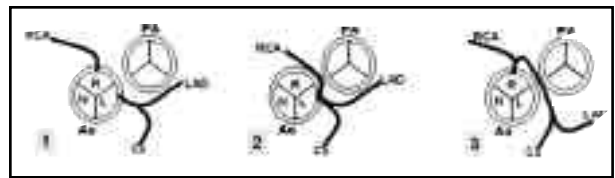


Fig. 1: Coronary artery sketch: Fig.1-1: Normal coronary artery anatomy where the left coronary artery arises from the left aortic sinus and the right coronary artery arises from the right coronary sinus (appropriate sinuses). Fig. 1-2: Origin of the right coronary artery from the left aortic sinus. Fig. 1-3: Anomalous left coronary artery from the right aortic sinus. Notice the abnormal course and compression of the segment of the coronary artery between the aorta and the pulmonary artery (inappropriate sinuses). Ao = Aorta, PA= Pulmonary artery, L=Left, R=Right, N=non-coronary cusp, LAD=Left anterior descending, Cx= Circumflex artery, RCA= Right coronary artery.

sudden cardiac event outside our institution where the diagnosis was made by an outside pediatric cardiologist in the emergency room. The patient was subsequently transferred to our intensive care unit for further management and required ECMO support for low cardiac output.

All patients were not related to one ethnic group. Incidentally, all patients who were referred for evaluation of a cardiac murmur turned out to have an innocent murmur. Parents reported no similar problem in their close families. All patients had no birth defects. Only one patient (case 1) had PDA ligation and no other associated cardiac defects were detected in others. To date, none of the patients on follow up required surgery, except for the deceased one (case 2).

Electrocardiograms (ECG's) and exercise tests: All were normal at presentation except in case 2, which demonstrated severe ST-segment depression in all the precordial and inferior leads with evidence of left anterior hemi-block. All recorded 24-hour Holter monitors and exercise tests were normal.

Echocardiography: All patients were diagnosed initially by transthoracic echocardiography (Table 1 and 2). In three cases transesophageal echocardiography

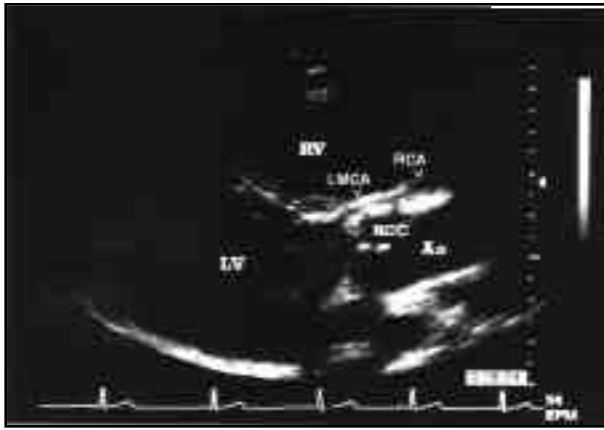


Fig. 2: (A) Parasternal long axis view through the aortic root in a boy with anomalous left main coronary artery (LMCA) with an intramural segment of the LMCA giving the double border appearance of the aortic wall (arrowhead). Seen is a single ostium with an origin from the right coronary cusp. Right coronary artery=RCA, aorta=Ao, left ventricle=LV, right ventricle=RV.

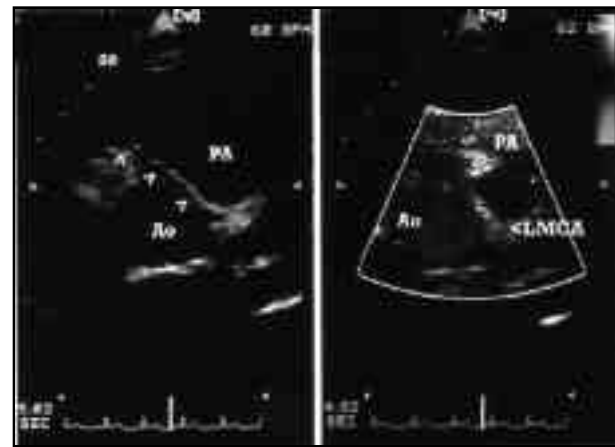


Fig. 2:(B) Parasternal short axis view showing the intramural segment of part of the long course of the left main coronary artery (arrowheads). Aorta=Ao, pulmonary artery=PA.

Table 2: Summary of echocardiographic features

Parameter	ALCA-RAS	ARCA-LAS
Origin	RAS	LAS
Ostium	Mostly large single	Usually two separate
Orifice (3D-image)	Rounded or slit-like	Mostly slit-like
Intramural	Segment course long interarterial	Short interarterial
Proximity to commissure	close	Very close
2D image:		
Parasternal long axis	Double border appearance of aortic wall	No
Parasternal short axis	Take off at 10 o'clock at 110 degrees followed by acute angle LMCA take off (hockey-stick appearance)	Take off at 12 or 1 o'clock at 160 degrees (spear-like appearance)
Doppler color flow	Aliasing usually seen before LMCA bifurcation	No
Spectral Doppler flow	Usually low pandiastolic & slightly increased systolic flow	Normal flow pattern

ALCA-RAS= Anomalous left coronary artery from right aortic sinus
 ARCA-LAS= Anomalous right coronary artery from left aortic sinus
 LMCA= left main coronary artery

graphy was performed to further delineate the anatomy including a 3-dimensional reconstruction in case 1.

In those patients with anomalous left coronary artery from the right aortic sinus the parasternal long axis view demonstrated the large single ostium, which then bifurcated into right and left main coronary arteries. The left main coronary artery then ran between the aorta and pulmonary artery with an intramural segment within the aortic wall resulting in a double border appearance of the anterior aortic wall (Fig. 2A). Similarly, the parasternal short axis view demonstrated a single large ostium that arose from the right anterior aortic sinus, which then bifurcated, into right coronary artery and a long interarterial / intramural segment (Fig. 2B). In all cases the left main coronary artery formed an acute angle with its origin. The

intramural segment was clearly seen in all cases.

In those patients with anomalous right coronary artery from the left aortic sinus, the parasternal long axis view did not show the double border appearance of the aortic wall. The parasternal short axis view revealed the slit like orifice of the anomalous right coronary artery, which was even more clearly demonstrated by transesophageal and three-dimensional echocardiography (Fig. 3 A & B). In both cases the orifice was very close to the commissure between the right and left sinuses which was in contrast to cases with anomalous origin of the left coronary artery from the right aortic sinus.

Doppler flow patterns:

Two spectral Doppler flow characteristics were identified. Firstly, in those with no evidence of obstruction the appearance was that of low velocity diastolic flow with a short but slightly increased systolic flow velocity. This appearance correlated with mild color aliasing seen in the intramural segment of the left main coronary just proximal to its bifurcation (Fig. 4 A & B). In the case with a cardiac event there was clear evidence of an obstructive flow pattern in the intramural segment, with laminar flow in the proximal interarterial segment. Of interest, this was only seen on the transesophageal study, as the intramural segment of the left coronary artery was stenotic and difficult to image from the transthoracic location. As the left main coronary artery exited from its intramural course, the vessel and its proximal branches became dilated with evidence of re-laminization of the Doppler flow signal. Spectral Doppler flow showed a high velocity systolic and diastolic flow pattern in the intramural segment with the former being 40 cm/sec and the latter 180 cm/sec. Of note, the spectral pattern at the site of origin of the left coronary artery and in the dilated segments

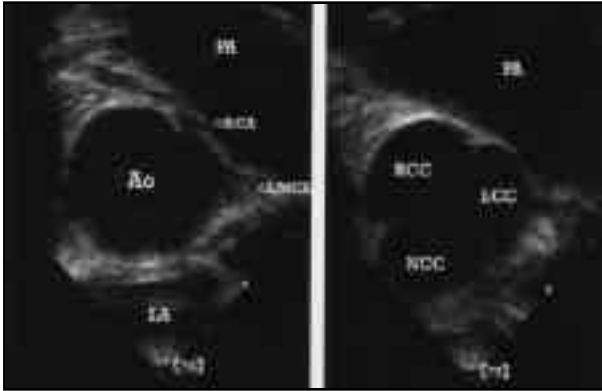


Fig. 3: (A) Transesophageal echocardiographic image at mid-esophagus in short axis view at the level of aortic sinuses showing the three sinuses and the origin of the right coronary artery (RCA) from the left aortic sinus. Note the proximity to the anterior commissure.

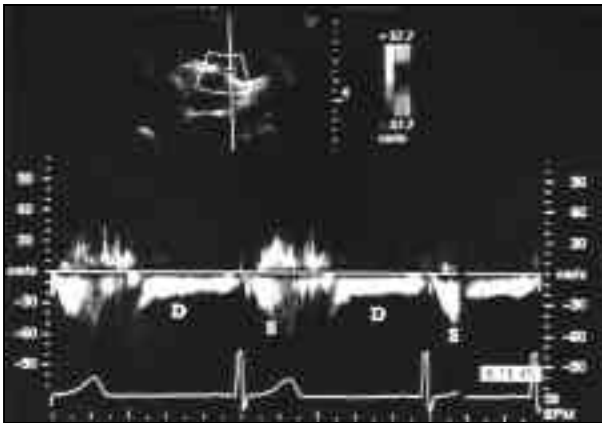


Fig. 4: (A) A normal spectral Doppler flow pattern seen in the anomalous left main coronary artery. A pandiastolic (D) and a short systolic (S) flow are seen. This shows a non-obstructed flow pattern.

demonstrated a lower velocity pattern. (Fig. 5 A& B).

M-Mode: All patients (except case 2) had normal cardiac systolic and diastolic functions at first study and on follow up. All cardiac chamber dimensions were also normal. In case 2, the patient presented with severe left ventricular dysfunction and wall motion abnormalities of the anterolateral wall. Her left ventricular ejection fraction was 5-10% requiring ECMO support.

Angiography: Three cases (3, 4, and 5) underwent selective coronary angiography. The angiographic pictures correlated well with echocardiographic images and did not add any additional information (Fig. 6).

Dobutamine stress echocardiography (DSE): Two cases (3 & 5) underwent this type of study to look for any evidence of wall motion abnormalities or ischemic changes. In case 3, the DSE was interpreted as demonstrating mild hypokinesia involving the apical anterior wall and the mid anterior septum. With an increasing dose of



Fig. 3: (B) Three-dimensional image of the coronary artery ostia in a case of ARCA-LAS. This shows the slit-like orifice of the anomalous right coronary artery and rounded orifice of the left main coronary artery, both originating from the left coronary sinus.

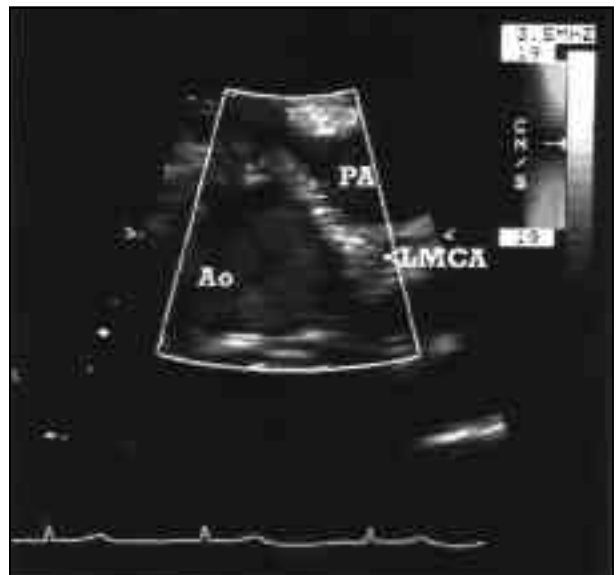


Fig. 4: (B) Doppler color flow mapping in the same patient, which shows very slight color aliasing of the intramural segment of the left main coronary artery (LMCA).

Dobutamine, there was progressive increase in contractility involving all segments, apart from those mentioned above which remained mildly hypokinetic, though improved compared to baseline. This was interpreted as consistent with prior myocardial damage. In case 5, the study was interpreted as being normal.

Myocardial perfusions scan: In two cases (3 & 4) single-photon emission computed tomographic myocardial scintigraphy (SPECT) was performed with intravenous administration of ^{99m}Tc -Sestamibi during exercise and at rest 24 hour later. Both showed no evidence of perfusion defects.

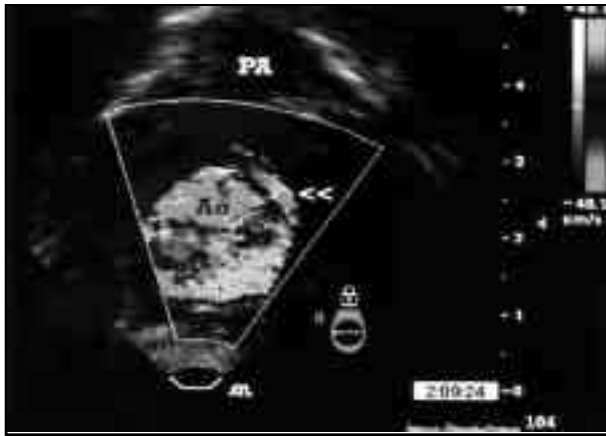


Fig. 5: (A) In comparison to the case in Fig. 3, severe stenosis of the interarterial, intramural segment of the left main coronary artery before its bifurcation in a patient who presented in cardiac arrest. The transesophageal Doppler color flow pattern showing color aliasing at the stenotic intramural segment.

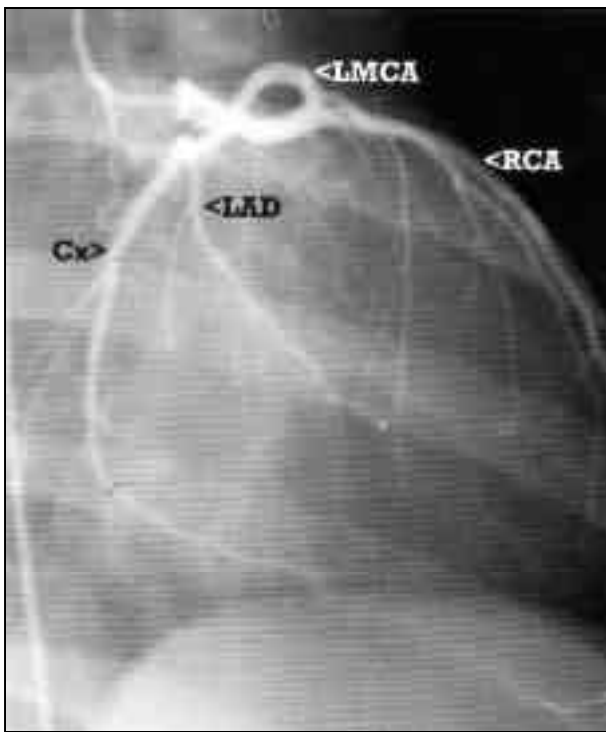


Fig. 6: Right anterior oblique projection of the single coronary angiograms. This shows an unusually long segment of the left main coronary artery initially coursing posteriorly.

Surgery: Only one patient underwent surgery (case 2). After a brief period of stabilization on ECMO for severe left ventricular dysfunction, the patient underwent surgical reimplantation of the anomalous left coronary artery. A vertical incision was made in the distal non-intramural segment of the left coronary artery (LCA) proper, which appeared thin-walled and measured 2-3 mm in diameter. A direct tissue connection was made between the inferior apex of the vertical aortic incision and the corresponding incision in the LCA. The anterior aspect of the connection between the LCA and the ascending aorta was augmented with

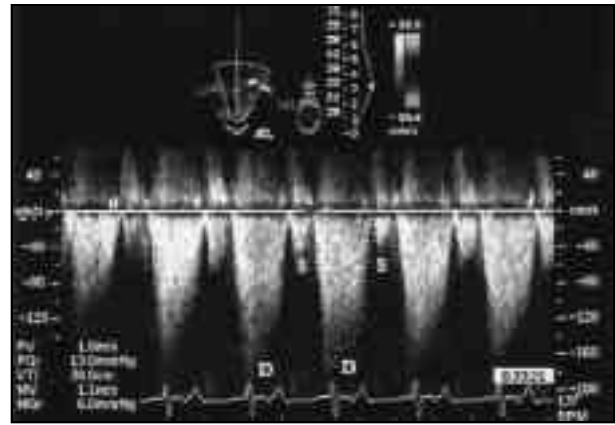


Fig. 5: (B) Spectral Doppler flow wave showed high peak diastolic and systolic velocities, which did not change with intravenous injection of nitroglycerine.

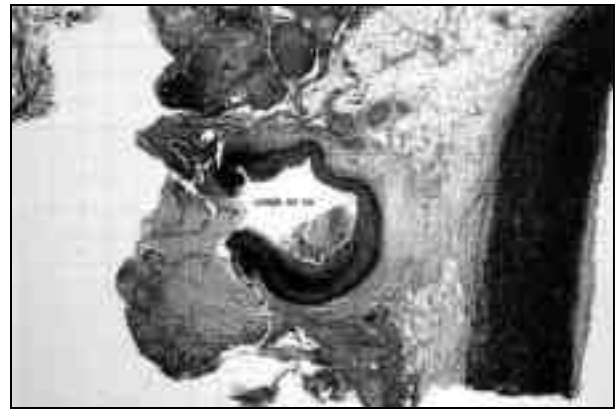


Fig. 7: Histology specimen showing the severe fibrointimal proliferation of the intramural segment resulting in severe stenosis.

a triangle-shaped patch of autologous pericardium. The segment of LCA between the origin of the LCA and the reconstructed LCA was not interrupted surgically with the LCA reimplantation. This patient died within 48 hours of surgical repair. The autopsy revealed severe and diffuse left ventricular myocardial infarction and a single coronary artery that arose from the right anterior sinus. The left main coronary artery was intramural in its course and showed stenosis at the segment just prior to LMCA bifurcation. The new LMCA ostium was narrowed at its suture line. Histological section of the narrowed intramural segment showed evidence of severe fibrointimal proliferation (Fig. 7).

DISCUSSION

When the left coronary artery arises from the right sinus of Valsalva and courses between the aorta and pulmonary artery, it invariably assumes an intramural course as it enters the aortic wall at an acute angle with typical flattening of the lumen^[6,13,14]. This anomaly has strong predominance in males with rare occurrences in females. Patients often have a slit-like orifice of the LMCA origin or valve-like ridges at the ostium^[4,6,10]. The angiographic

findings demonstrate an end-on origin of the LMCA followed by a posterior, cranial, and leftward course^[3,15,16]. Dynamic systolic compression of the intramural portion of the LMCA has been observed during angiography and may potentially contribute to periods of ischemia.

Taylor *et al*^[11,17], reported that younger patients (less than or equal to 30 years of age) with an isolated coronary artery anomaly are at risk of dying suddenly and with exercise. The association of this anomaly with exercise-related sudden death has been explained by three factors^[14,18]: Firstly, an outward expansion of the roots of both the aorta and pulmonary trunk during exertion, which can cause further compression of the ostial lumen of the anomalous artery; Secondly, the anomalous vessel can be compressed against the root of the pulmonary trunk, where it is firmly anchored to the infundibular septum, when the pulmonary trunk and the aortic root dilate during exertion; Thirdly, myocardial oxygen requirements increase with exertion and the consequence of myocardial ischemia may trigger life threatening ventricular arrhythmia and sudden death.

Jureidini and associates^[9,19] reported color Doppler mapping of the anomalous course of the LMCA between the aorta and pulmonary artery in a child. In our series, it is clearly demonstrated that a careful echocardiographic search for coronary anomalies can aid in their detection. Indeed the routine in our laboratory is to evaluate the coronary arteries in all new cases. If the anomaly is suspected and transthoracic echocardiographic images are not clear, then transesophageal echocardiography should be performed after the patient is stabilized^[20]. Simply, identifying a coronary artery between the two arterial roots does not necessarily imply an intramural segment^[21]. Although we maintain that angiography remains the current "gold standard", magnetic resonance imaging (MRI) and ultrafast computed tomography may be useful adjunctive tests. MRI showed a complimentary role and appears a promising tool for identifying such anomalies^[18,22]. Recently, intravascular ultrasound and pressure-wire methods have been used for sub-classifying anomalous coronary arteries from opposite sinus and other coronary anomalies^[14].

The role of stress evaluation as a predictor of ischemia in these cases when detected as an incidental finding is unclear. Although we have been using dobutamine as our agent of choice in pediatric patients, adenosine, arbutamine or dipyridamole are other agents used frequently in adult patients^[13,23,24]. There is, however, enough data to support Dobutamine stress test in patients with Kawasaki disease^[25]. In case 2, it was recommended

to the family that PET (positron emission tomography) might be useful adjunct as this has the added advantage of assessing both flow and metabolic activity^[26].

While the diagnosis of this lesion has been facilitated by careful echocardiographic evaluation of the coronary arteries^[27], the indication for intervention is less clear. The use of extracorporeal membrane oxygenation (ECMO) is justified in certain cases with severely reduced ventricular systolic function as a bridge to heart transplantation^[28]. Certainly, in those with evidence of ischemia the need for surgical intervention is fairly clear. However, in pediatric patients where the finding is incidental, with no evidence of ischemia at rest or on stress, the argument for intervention is less compelling. This relates to the lack of long term data in those young patients who have undergone surgery, versus the natural history in a case where the finding is incidental. Although there are multiple case reports of the relationship between this lesion and ischemia, there are no studies that address the frequency of ischemia in asymptomatic patients. There is medium term data in complete transposition of great arteries and an intramural left coronary artery^[29]. Although an arterial switch is the current treatment of choice, postoperative ischemia is far more prevalent than cases with the more usual coronary artery patterns^[30,31]. One could argue that there are younger patients than the majority of cases identified with an intramural left coronary artery, however, a comparison of the natural history versus treated population are absent.

The operative repair is the one described by Mustafa and colleagues^[13,32] in which the intramural segment of the LMCA is unroofed after detachment of the intercoronary commissure, with creation of a neocoronary ostium in the left sinus of Valsalva. Subsequently, the intercoronary commissure is re-suspended. In adult population, the incidence of late cardiac events (late cardiac death, re-operation, myocardial infarction) was lower in patients who received internal thoracic artery graft versus saphenous vein graft and had better survival rate at eight years (91.1% versus 85.3%, respectively)^[33]. In contrast, long-term patency and re-operation rates have yet to be determined in the pediatric population. On the basis of a literature review and their own experience with this lesion, Libberthson *et al*^[5] have found that the average age of the 20 reported patients who died suddenly was 16 years (range 1 - 36 years). All of their six older patients (age 36 to 70 years) with this anomaly had angina pectoris and atherosclerotic vessels. This difference may relate to the more rigid coronary arteries in an older individual being less susceptible to torsion

and resultant ischemia in comparison to a young adolescent male. He also found that atrial pacing may reveal ischemic changes during cardiac catheterization. This has led some cardiologists to use calcium channel blocking agent or beta-blocker to limit the attainment of threateningly high heart rates. In general, management is guided by: mode of presentation, age at presentation, size of ectopic coronary artery, presence of related proximal coronary artery obstruction, and lifestyle of individual patient. Patients younger than 40 years with large ectopic arteries, functional and anatomic compromise, and ischemic or arrhythmic complications require intervention. For patients older than 50 years, specific therapy are rarely required. Lastly, for patients between 40 and 50 years care must be individualized^[34].

Our present recommendation is the following; as part of first echocardiography study, all patients, regardless of their age, should have their coronary arteries screened in the echocardiography laboratories. Once coronary abnormalities are detected (i.e., origin from the opposite sinus), efforts should be made to look for any evidence of ischemia either through invasive or noninvasive techniques. Anomalous left coronary origin from the right sinus of Valsalva can result in significant myocardial ischemia and infarction.

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