operation is preferred for patients who are hemody-
namically stable. Conventional surgical repair consists of
direct closure of the defect with mattress sutures and
Teflon felt or, alternatively, infarctectomy and closure
either primarily or with a patch. Recently, a sutureless
patch technique has been developed with good short-
term and long-term results

In the majority of cases, combined surgical repair with CABG is frequently
advocated, given that 80% of patients who experience
cardiac rupture have multivessel coronary disease.

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Right Coronary Artery From Right Sinus of Valsalva and Ventricular
Tachycardia
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Congenital coronary anomalies in which the coronary
artery arises from the contralateral aortic sinus of Val-
salva are associated with fatal arrhythmias and sudden
death. The mechanism causing sudden death has been
debated. We describe a case of an adolescent patient with
right coronary artery arising leftward, but from the right
sinus of Valsalva, with an acute-angle takeoff from the
aorta and an intramural course, who presents with
exercise-induced ventricular tachycardia.

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Congenital anomalies of the coronary arteries in which a coronary artery arises from the wrong sinus of Valsalva are relatively rare and are associated with an increased risk of sudden death, especially in the young and usually during or just after exertion [1–3]. The exact cause of myocardial ischemia and likely resultant ventricular tachyarrhythmias is not known, but hypotheses include compression of the anomalous vessel during its course between the great vessels and an acute-angle takeoff with a slitlike orifice that can close or compress as the vessel runs intramurally in the wall of the aorta. It is unclear, however, if this pathologic substrate requires that the coronary artery origin be located in the wrong sinus of Valsalva. There have been no case reports of a child with a right coronary artery arising leftward but in the correct sinus of Valsalva with an acute-angle takeoff and intramural course with ventricular tachycardia. We present this unique and instructive case whose clinical presentation was rare and unexpected.

A 16-year-old male athlete of Asian descent presented to the outpatient pediatric cardiology clinic for evaluation of an irregular heartbeat. A transthoracic echocardiogram was performed, which showed normal intracardiac structure and function, but there was suspicion for an anomalous right coronary artery from the left sinus of Valsalva. A cardiac magnetic resonance imaging study was performed, which demonstrated the right coronary artery arising leftward within the right sinus of Valsalva associated with an acute angulation of the coronary artery origin (Fig 1). Virtual angioscopy post-processing of the magnetic resonance imaging three-dimensional dataset demonstrated a crescentic orifice consistent with stenosis (Figs 2, 3). An exercise test was performed with nuclear myocardial perfusion. Near peak exercise, the patient had five discrete episodes of monomorphic ventricular tachycardia with right bundle-branch block morphology at a rate of 150 to 230 beats/min (Fig 4). He was asymptomatic during the tachycardia. The perfusion scan was negative.

Because of the arrhythmia along with the acute angulation and coronary stenosis, the patient was referred for surgery. In the operating room, the surgeon confirmed that the coronary artery arose in the correct (right) sinus of Valsalva, but more leftward than normal with an acutely angulated right coronary ostium, giving it a “fishmouth” appearance, and a 3-mm intramural course. The surgeon performed the unroofing procedure, whereby the intramural segment of the right coronary artery was resected, relieving the ostial stenosis; a 3.5-mm coronary probe was easily passed into the neo-ostium. In the 16 months since his surgery, he has had a normal

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Fig 1. Magnetic resonance imaging bright blood coronary angiography without contrast enhancement demonstrates that the left coronary artery (LCA) originates normally from the left sinus of Valsalva. The right coronary artery (RCA) origin is located more leftward than normal. On this image it is difficult to discern whether the origin of the RCA is within the right or left sinus as the intercoronary commissure is not clearly observed. (Ao = aorta; MPA = main pulmonary artery.)

Fig 2. (A) Virtual angioscopy technique is applied to Figure 1 with the virtual angioscopy camera focused on the right and left coronary origins from a position within the center of the aorta from a surgeon’s perspective. (B) The image demonstrates that the left coronary artery (LCA) origin is normal appearing: round and patent. In contrast, the right coronary artery (RCA) origin has an abnormal elliptical shape consistent with ostial stenosis. The abnormal RCA ostium cannot fully be observed from the center of the aorta. Furthermore, note that the RCA ostium is located to the right of the intercoronary commissure (ICC) within the right sinus of Valsalva. (I = inferior; L = left; R = right; S = superior.)
exercise test and Holter monitoring results and has resumed all physical activity.

Comment

The potential for sudden death from a congenital coronary anomaly has been well documented [1, 4–6]. However, ischemia does not occur in everyone with this anomaly; the reason behind this remains unclear. Currently, there is no accepted standard to stratify patients with a potentially higher risk from those with a lower risk anomaly, largely because the exact mechanism underlying the ischemia has not been fully elucidated. One postulated cause of ischemia and malignant arrhythmias is the way the coronary artery arises from the aorta, forming a slitlike orifice because of an acute-angle takeoff (ie, proximal coronary artery angle with the aorta <45 degrees), which is often found in anomalous coronary arteries [1, 5, 6]. With aortic root distention, this can lead to closure of the ostium. This is in contradistinction to the round orifice, commonly found in normally arising coronary arteries. Multiple autopsy series have suggested that the acute-angle takeoff with subsequent compression of the coronary artery at the origin is universally found in patients with coronary anomalies who die suddenly [1, 5, 6]. Of interest is an autopsy study by Virmani and colleagues [7] in which the authors analyzed the hearts of people 20 to 79 years of age who experienced sudden cardiac death and compared these hearts with those from control subjects who died of noncardiac causes. They found that those who experienced sudden cardiac death were statistically more likely to have an acute-angle takeoff of the coronary artery and also ostial valvelike ridges. These were in hearts in which both coronary arteries arose from their normal aortic sinus; as well, the acute-angle takeoff was more likely to be found with the right coronary artery [7].

In summary, this is the first report of a young patient with a documented ventricular arrhythmia with exercise from a coronary artery arising from the correct sinus but with an acute-angle takeoff and intramural course. Only in the modern era, with our ability to accurately image the coronary arteries using cardiac magnetic resonance imaging with virtual angioscopy, are we able to make this diagnosis and realize the potential our patient had for sudden cardiac death. Our patient demonstrates that it is not necessarily the location per se of the anomalous coronary artery but the angle of takeoff and intramural course, which may be the underlying anatomic substrate necessary for potential ischemia. More data are needed regarding the anatomic abnormalities found in coronary arteries arising from the correct sinus but with a leftward origin to allow clinicians to adequately stratify patients according to risk and manage them correctly.

Fig 3. Virtual angioscopy applied from a position tangential to the aortic wall near the left sinus and looking directly at the right coronary artery (RCA) origin (A) confirms that the ostium is elliptical and abnormal (B). Moreover, this technique can identify the tissue to be targeted for the unroofing operation (between the arrowheads). (I = inferior; L-Post = left posterior; R-Ant = right anterior; S = superior.)

Fig 4. Electrocardiogram obtained near peak exercise during a maximal exercise stress test demonstrates short runs of ventricular tachycardia with a right bundle-branch block morphology at a rate of 150 to 230 beats/min.
Sinus node dysfunction (SND) is commonly observed after the Fontan procedure [1, 2]. Pacemaker implantation has been reported in 9.2% of Fontan patients [3]. The two options for pacemaker implantation for SND after the Fontan operation are epicardial lead placement, which allows for dual-chamber pacing, or transvenous atrial (AAI[R]) pacing [4]. Transvenous atrial lead implantation has been reported in the right atrium-to-pulmonary artery type of Fontan and in the intracardiac lateral tunnel Fontan where part of the Fontan baffle is composed of native right atrial tissue.

Epicardial lead placement, although commonly used, requires more invasive atrial exposure through a sternotomy or thoracotomy. Moreover, chronic epicardial lead performance has historically been inferior, with higher pacing and lower sensing thresholds. Higher lead failure rates are also noted [3].

The transvenous approach results in better pacing thresholds; however, the most common Fontan approach (prosthetic extracardiac conduit type) completely excludes vascular access to atrial tissues, thus precluding transvenous atrial lead implantation.

Hasaniya and colleagues [5] recently reported that construction of an extracardiac lateral tunnel (ECLT) Fontan conduit using viable pedicled pericardium is a relatively simple operation with low medium-term morbidity and death. Because this modification of the Fontan connection has the potential for normal growth, it can be applied at an earlier age and weight than a prosthetic tube. Here we report the first successful implantation of an endocardial atrial lead for SND in a patient with an ECLT Fontan.

A girl, aged 7.5 years, was monitored with SND. Holter monitor showed junctional rhythm with heart rates as low as 42 beats/min and symptoms of dizziness. Elevated filling pressures and mild diastolic ventricular dysfunction developed. Atrioventricular dyssynchrony was apparent on an echocardiogram.

She had a history of double-outlet right ventricle with malposed great vessels and pulmonary atresia with atroventricular canal. Staged palliation included: (1) 4-mm right-modified Blalock-Taussig shunt, (2) bidirectional cavopulmonary anastomoses, and (3) extracardiac autologous pericardial tunnel Fontan completion. Significant history was pertinent for bilateral phrenic nerve palsies necessitating right diaphragm plication.

Owing to symptomatic SND, the decision was made to proceed with single-chamber (atrial) pacemaker implantation. The patient’s extracardiac autologous pericardial tunnel Fontan enabled easy access to native right atrial tissue, prompting consideration of a transvenous atrial lead.

Vascular access was obtained in the left subclavian vein that facilitated entry to the Fontan baffle. An angiogram using a pigtail catheter was performed in the Fontan baffle. A quadripolar mapping catheter was used to map atrial signal/tissue in the Fontan baffle. Atrial tissue was localized in the posteromedial aspect of the Fontan baffle close to the third and fourth sternal wires on fluoroscopy.

References


Extracardiac Autologous Pericardial Tunnel Fontan Allows Implantation of an Endocardial Atrial Lead for Sinus Node Dysfunction

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Sinus node dysfunction is common after the Fontan procedure, and pacemaker implantation has been reported in 9.2% of Fontan patients. The two options for pacemaker implantation for sinus node dysfunction after the Fontan operation are epicardial lead placement, which allows for dual-chamber pacing, or transvenous atrial pacing. We report the first successful implantation of an endocardial atrial lead for sinus node dysfunction in a patient with an extracardiac lateral tunnel Fontan.


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