

## Anomalous Origin of the Left Coronary Artery From the Noncoronary Cusp: Not a Benign Lesion

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**Abstract** This case report describes two patients with a very rare condition who presented with pathologic symptoms. Anomalous origin of the left coronary artery from the noncoronary cusp has been described as a “benign” lesion by some authors in the past, although rare cases of morbidity/mortality are described in the literature. Both reported patients underwent surgical repair for the lesion and at this writing are asymptomatic at follow-up evaluation. These two patients presenting with pathologic symptoms and undergoing surgery afford novel descriptions. The authors believe these descriptions add to our knowledge of this rare disorder.

**Keywords** Anomalous coronary artery · Arrhythmia · Ischemia

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Anomalous origin of the left coronary artery (LCA) from right coronary cusp with an interarterial and intramural course is associated with sudden death. An opposite sinus origin of the right coronary artery from the left sinus of Valsalva with an interarterial and intramural course also is associated with sudden death, but individuals with this anomaly generally are considered to be at less risk than the former group.

Origin of the LCA from the noncoronary sinus of Valsalva is an extremely rare disorder, with only case reported in the literature [1–11] and an incidence estimated variously at 0.0008 [10], 0.004 [1], and 0.012% [2]. Some authors [10, 11] have suggested that a coronary artery arising from the noncoronary sinus is a benign disorder, although three pediatric case reports in the literature have described sudden death [4, 5] or life-threatening arrhythmia [7] associated with this condition. The patient with the life-threatening arrhythmia had a cardiac defibrillator placed.

Origin of the LCA from the noncoronary cusp has not been described as intramural (shared intima and media between the aortic wall and the coronary artery). An intramural coronary artery is postulated to be one of the factors associated with sudden death for patients with coronary anomalies and lends itself to surgical repair by an unroofing procedure in which the common wall is removed.

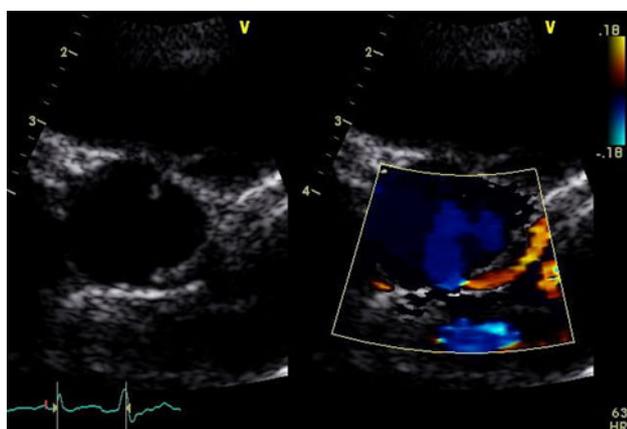
We report two cases of anomalous origin of the LCA from the noncoronary sinus of Valsalva with an intramural course in which the patients presented with pathologic symptoms. The first patient was referred for premature ventricular contractions (PVCs). The anomaly was diagnosed via transthoracic echocardiography and computerized tomography angiography (CTA). A positron emission tomography (PET) scan detected myocardial ischemia in the left anterior descending (LAD) artery and left circumflex (Cx) distribution. The second patient presented with

multiple episodes of syncope and wide QRS tachycardia and subsequently experienced hemodynamic instability requiring emergency surgery.

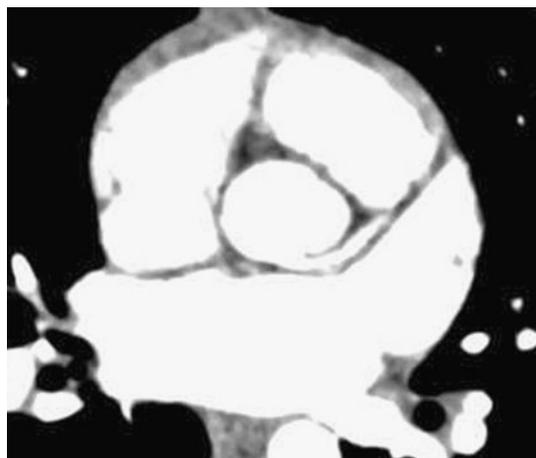
## Case Reports

### Case 1

A 6-year-old girl was referred for evaluation of an irregular heartbeat. The patient reported a history of occasional tachycardia spells and chest pain. Her physical exam was notable for irregular heartbeat with occasional extrasystoles but was otherwise normal. A 12 lead electrocardiogram (ECG) showed PVCs in bigeminy, and follow-up 24 h Holter monitoring showed 45,984 PVCs. A transthoracic echocardiogram (Fig. 1) showed the LCA originating from



**Fig. 1** Case 1. Preoperative echocardiogram showing the origin of the left coronary artery from the noncoronary sinus of Valsalva in two dimensions and with color Doppler



**Fig. 2** Case 1. Preoperative computed tomography (CT) angiogram showing the origin of the left coronary artery from the noncoronary sinus of Valsalva

the noncoronary cusp with an acute angle take-off and a probable intramural segment proximal and distal to the commissure between the noncoronary and left coronary cusps. The girl's biventricular function was normal with no regional wall motion abnormalities. A cardiac computed tomographic (CT) angiogram (Fig. 2) showed anomalous origin of the LCA from the noncoronary sinus of Valsalva with inability to confirm an intramural course. An ammonia stress PET perfusion scan showed signs of ischemia involving the LAD artery and Cx distribution.

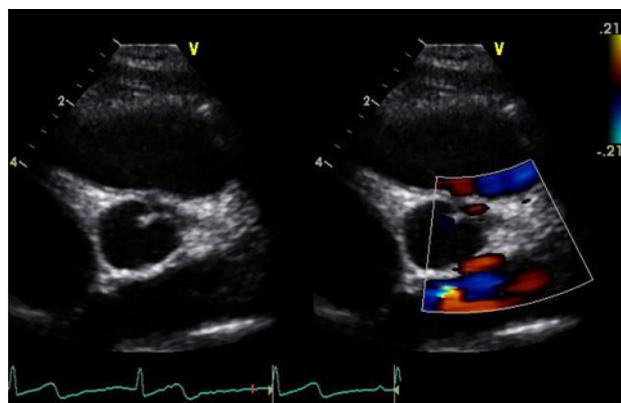
The patient was referred to cardiothoracic surgery for an unroofing of the LCA due to suspicion of an intramural course in a patient with known perfusion abnormality. At surgery, the anomalous origin of the LCA from the noncoronary sinus with an intramural course was confirmed, and an unroofing procedure was performed. Postoperative transthoracic and transesophageal echocardiograms showed laminar antegrade flow across the left main coronary artery "neo-ostium" from the left sinus of Valsalva (Fig. 3).

The follow-up ammonia stress PET scan was normal, with no ischemia noted in the LAD artery or Cx distribution. The frequency of PVCs on the follow-up Holter was markedly diminished, with a total of three PVCs compared with 45,984 before surgery.

At this writing, the girl remains asymptomatic 2 years after surgery. A follow-up stress echocardiogram was negative for myocardial ischemia.

### Case 2

An 11-year-old girl presented with multiple episodes of syncope and near syncope over a 24 h period. Rhythm strips showed a wide complex QRS tachycardia, with a differential diagnosis of ventricular tachycardia versus supraventricular tachycardia with aberrancy. She was cardioverted, intubated for pulmonary edema, and transferred to the pediatric



**Fig. 3** Case 1. Echocardiogram after the unroofing procedure of the intramural portion of the anomalous left coronary artery in two dimensions and with color Doppler

intensive care unit. A transthoracic echo was suboptimal but suggested normal coronary origins.

During anesthesia induction for an electrophysiology study, the girl had ECG evidence for ischemia, development of pulmonary edema, and worsening hemodynamics. A coronary angiography showed a filling defect in the ostium of the LCA. The LCA was engaged posteriorly, showing a normal left system.

A balloon pump was placed due to worsening hemodynamics. A transesophageal echocardiogram showed poor left ventricular systolic function and an LCA arising from the noncoronary sinus with an intramural course.

The girl was taken to the operating room, where she underwent patch angioplasty of the proximal LCA course with a glutaraldehyde-treated autologous pericardium. This was the surgical repair performed previously by the surgeon for an intramural course and interarterial course of a coronary artery. The patient was weaned from the balloon pump in 3 days. She had no further dysrhythmias, and normal cardiac function gradually developed. At this writing, she remains asymptomatic since her surgery. Cardiac magnetic resonance imaging (MRI) at her 4 year follow-up visit showed normal wall motion, with delayed enhancement negative for myocardial scarring.

## Discussion

To our knowledge, these two case reports are the first to describe an anomalous origin of the LCA from the non-coronary cusp found to be intramural and therefore amenable to an unroofing procedure. The patient in case 1 had a successful unroofing procedure with normalization of myocardial ischemia shown on ammonia stress PET compared with preoperative myocardial ischemia. She also had a marked decrease in the frequency of PVCs from almost 46,000 to 3.

The patient in case 2 had a patch angioplasty, which was the surgery of choice for an intramural anomalous coronary artery at the time of her repair at the institution that admitted her. At this writing, she remains asymptomatic and arrhythmia free after 4 years of follow-up evaluation, with normal left ventricular function, wall motion, and no scarring shown on cardiac MRI.

Contrary to suggestions in the literature, we do not believe that an anomalous LCA from the noncoronary

sinus of Valsalva is a universally benign lesion. As the cases demonstrate, this abnormality may be associated with myocardial ischemia, life-threatening arrhythmias, and sudden death for children. The association of this anomaly with an intramural course probably is underdiagnosed but important to determine because this lesion is amenable to an unroofing procedure. Children with this coronary anomaly should be followed in a coronary registry to obtain a better understanding of the natural history of this abnormality.

**Conflict of interest** None.

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