

## Congenital Left Main Coronary Artery to Coronary Sinus Fistula

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### ABSTRACT

Congenital coronary artery fistula is an extremely rare anomaly that may involve any of the coronary arteries and any of the cardiac chambers. We report the case of a 14-year-old female patient with a symptomatic congenital coronary fistula starting from the left main coronary artery and draining to the coronary sinus. The patient underwent surgical ligation of the fistula and had an excellent outcome.

### INTRODUCTION

A coronary artery fistula is defined as an abnormal communication between a coronary artery or its branches and another cardiovascular structure, including any of the coronary vessels and any of the cardiac chambers. Several combinations of the involving structures have been reported. The most common origin is the right coronary artery (60%), followed by the left coronary artery (35%). It usually drains into the right heart chambers, most commonly the right ventricle (40%), followed by the right atrium (25%), pulmonary artery (15% to 20%), and the coronary sinus (7%) [Kirklin 2003]. We describe the case of a 14-year-old girl with coronary artery fistula originating from the left main artery draining into the coronary sinus. The left anterior descending artery (LAD) and left circumflex artery (LCR) originated from the right and the left side of the fistula, respectively, after its origin. The right coronary artery originated normally from the anterior left main coronary artery. The fistula was ligated surgically with the beating heart technique on cardiopulmonary bypass (CPB). She had an uneventful postoperative course, and at 24-month follow-up, she remains free of cardiovascular symptoms.

### CASE REPORT

A previously healthy 14-year-old girl presented with a recent history of 2 episodes of syncope. Clinical evaluation

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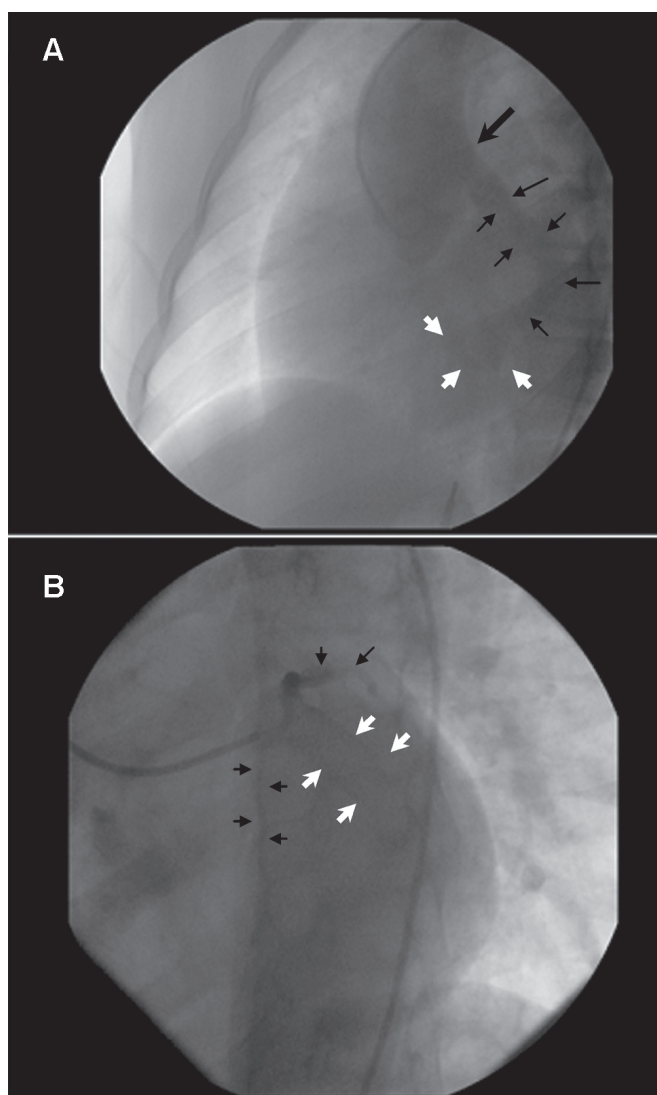


Figure 1. A, Cardiac catheterization showing the presence of the fistula (thin black arrows), which originates from the left main coronary artery (thick black arrow) and drains to the coronary sinus (white arrows). B, The same examination depicting the fistula (white arrows) and its relationship with the left anterior descending artery (lower black arrows) and the circumflex artery (upper black arrows).

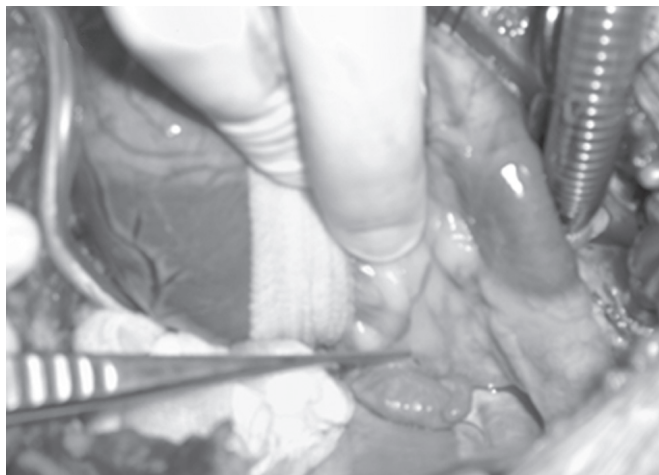


Figure 2. Intraoperative photo showing the fistula (tip of the forceps).

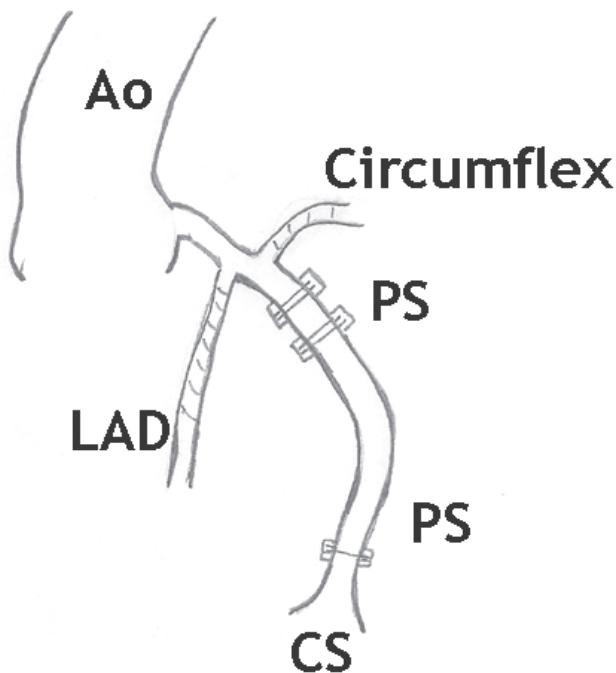


Figure 3. Schematic representation depicting the sites of the fistula, where the pledgeted sutures were placed. LAD indicates left anterior descending; PS, pledgeted sutures; CS, coronary sinus; Ao, Aorta.

revealed a continuous murmur at the left sternal border. The rest of the clinical examination and routine laboratory tests were unremarkable. Transthoracic echocardiography showed an anomalous vascular structure associated with a significant left to right shunt originating from the left main coronary artery, draining to the coronary sinus. The ejection fraction was 60%, and the cardiac chambers were normal without dilatation. Further investigation with magnetic resonance imaging (MRI) and selective angiography were performed (Figure 1), which confirmed the presence of the fistula originating from

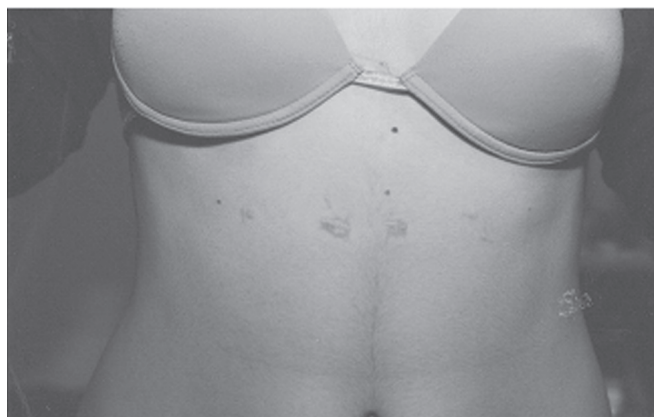
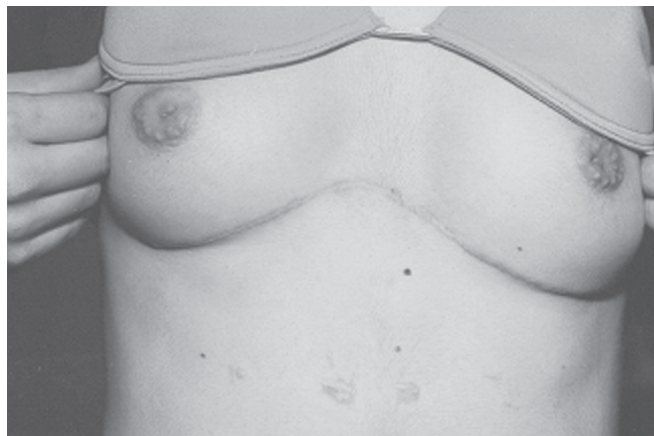


Figure 4. Postoperative photo showing the cosmetic result.

the left main artery and draining to the coronary sinus. Pulmonary to systemic flow ratio ( $Q_p/Q_s$ ) was 1:1.6. The patient was referred for surgical correction. Considering the cosmetic result, a bilateral submammary incision and a midline sternotomy were performed.

The fistula coursed in the left atrioventricular groove (Figure 2). We decided to proceed with the beating heart technique on CPB, because when lifting the heart, hemodynamic instability occurred. The fistula was double ligated with pledgeted 5-0 prolene sutures after the take off at the LAD and circumflex branches (Figure 3). Temporary discontinuation of CPB and examination of 12-lead electrocardiogram (ECG) showed no ischemia. Afterward, ligation of the fistula at its draining site before its entrance to the coronary sinus was performed. Weaning from CPB revealed again no ECG changes. Transesophageal intraoperative echocardiography (TEE) confirmed no residual shunt with no wall motion abnormalities.

The patient had an uncomplicated postoperative course. She was extubated on postoperative day 1 and was discharged home the fifth postoperative day. At 6-, 12-, and 24-month follow-up, she is free from cardiovascular symptoms, and echo examination revealed no residual shunt flow. In addition, an excellent cosmetic result was achieved (Figure 4).

## DISCUSSION

More than 300 cases of coronary fistulae have been reported in the literature since the first report in 1865 [Krause 1865]. This communication may be congenital or acquired, resulting from atherosclerosis, Takayasu's arteritis, trauma, or cardiac operations [Said 1999]. Angiographic series show an incidence of 0.3% to 0.8% of this congenital malformation [Gowda 2006]. Most fistulae are small, cause no symptoms, and are found incidentally during cardiac catheterizations, but larger fistulae may cause significant coronary steal phenomena, resulting in myocardial ischemia or remarkable left to right shunt with volume overload and congestive heart failure. The clinical presentation includes angina, syncope, infective endocarditis, arrhythmias, congestive heart failure, rupture of the proximal dilated aneurysmal coronary vessel or of the fistula, superior vena cava syndrome, and sudden death [Hong 2004; Gowda 2006]. Diagnostic investigation includes echocardiography, selective angiography, and/or computed tomography/MRI. Symptoms, complications, and significant shunt are the main indications for treatment. The natural history is variable. Spontaneous closure, although uncommon, has been reported [Hackett 1984]. However, in view of the fact that these fistulae may increase in size and produce symptoms, heart failure, and potentially lethal complications, the presence of a coronary artery fistula is an indication for operation unless the shunt is small [Kirklin 2003].

Surgical correction is a safe and effective treatment modality with excellent results [Balanescu 2001; Kirklin 2003]. The usual approach is through a median sternotomy. In our case, a bilateral submammary skin incision was applied for a better cosmetic result. CPB is used when the artery is dilated and when the fistula is relatively inaccessible or involves the distribution of the circumflex or distal right coronary artery. In addition, CPB can be used when the fistula is in the course of the artery rather than in its termination and an aneurysm is present. The fistula may be safely closed without CPB when it occurs in the termination of a major coronary artery branch into an accessible site. TEE is a valuable tool in the

operating room because it can reveal the residual fistulous flow [Stevenson 1994]. Transcatheter embolization has been reported as an alternative technique and is considered the therapy of choice for appropriately selected cases with small single fistulas with no proximity to vessels of functional importance and appropriate neck [Hong 2004]. Catheter closure may be performed with a variety of techniques including balloons, coils, and stents [Gowda 2006]. The safe and effective results of both interventional and surgical approaches support the option for elective closure of clinically significant fistulas promptly [Gowda 2006]. Long-term follow-up is essential due to the possibility of recurrence, dilatation of the coronary artery, and myocardial ischemia.

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