

Giant coronary artery fistula connecting the left coronary sinus with the superior atriocaval junction

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A left Valsalva sinus-right atriocaval junction giant congenital fistula was diagnosed in a young man with recent bacterial endocarditis. Surgical closure of the fistula was performed to correct the left-to-right shunt, to reduce the risk of recurrence of endocarditis and to prevent aortic root distortion. Surgery was preferred to percutaneous treatment because of the perceived risk of coronary complications. The dimensions of the residual chamber connected with the aortic root appeared unchanged at the 1-year follow-up echocardiography. Simple closure, with no procedure on the proximal coronary tract of the fistula may offer an effective and definitive correction of giant fistulas.

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Introduction

Coronary artery fistula (CAF) is defined as an abnormal communication through which coronary arterial flow is shunted into a cardiac chamber, great vessel or other vascular structures. This congenital anomaly is rare, the reported incidence being < 0.5%. The hemodynamic importance of this lesion and its clinical manifestation are quite variable. Usually CAF is asymptomatic, but may present during infancy with heart failure requiring surgical closure. Untreated CAF may be complicated by cardiac failure, bacterial endocarditis or coronary steal. Although closure by transcatheter embolization represents a primary therapeutic option, surgical treatment may offer excellent short- and long-term results when the unfavorable anatomy of CAF renders the percutaneous procedure inapplicable or too dangerous because of the risk of occlusion of important coronary branches. In this report we describe the surgical treatment of a rare giant CAF performed with the aim of correcting the left-to-right shunt by the interruption of the anomalous vascular connection avoiding any surgical procedure on the tract originating from the left Valsalva sinus.

Case report

A 30-year-old black male was admitted with fever and chills. Echocardiography

showed an anomalous vascular structure 1 cm in diameter associated with a significant left-to-right shunt from the left Valsalva sinus to the right atrium. Two small vegetations were detected on the left coronary aortic cusp and the inferior atriocaval junction. Mild aortic and tricuspid regurgitation were present. Streptococcal endocarditis was diagnosed and penicillin treatment was initiated leading to clinical recovery with complete regression of the vegetations.

Selective angiography confirmed the presence of an anomalous winding vascular structure, originating from the left aortic sinus and draining into the upper part of the right atrium. After its origin the fistula curved rightwards to reach the right atrium following a straight course. The left circumflex and intermediate coronary arteries originated from the left side of the twisted tract of the fistula (Fig. 1). The left anterior descending coronary artery had a separated origin from the left sinus. The right coronary artery originated normally from the right sinus. The cardiac chambers and great vessels were morphologically normal. A left-to-right shunt was detected by oximetry with a QP/QS ratio of 2:1.

Two months following the resolution of the streptococcal endocarditis the patient was scheduled for surgical treatment of the giant fistula. The aims of surgery were 1) to correct the significant shunt, 2) to decrease the probability of recurrence of endocardi-

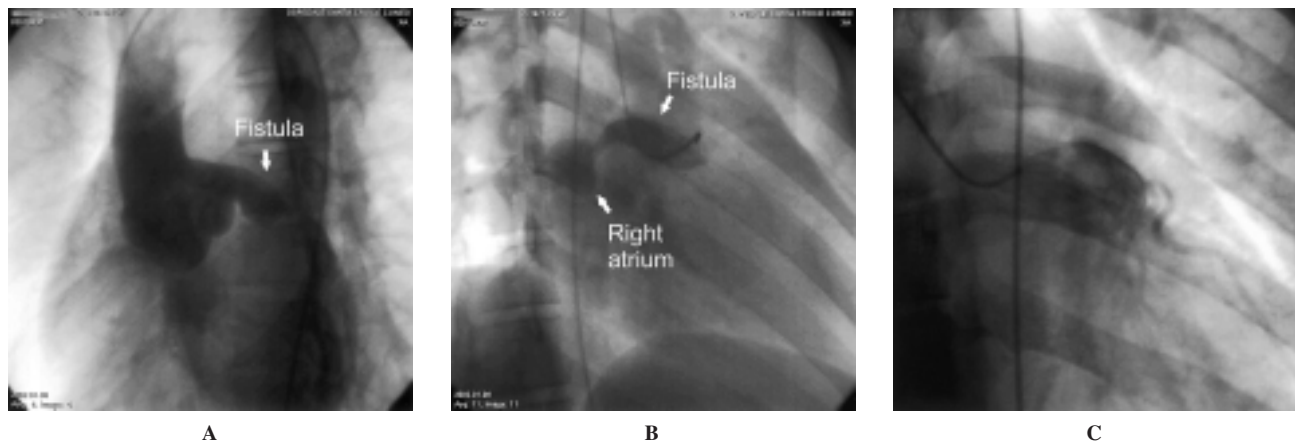


Figure 1. Selective angiography of the fistula showing its origin from the left aortic sinus (A), drainage of the fistula into the atriocaval junction (B), the origin of intermediate and marginal branches from the twisted tract of the fistula (C).

tis, and 3) to prevent aortic distortion and/or coronary artery aneurysm.

After midline sternotomy and pericardiotomy, the fistula was clearly visible on the surface of the left atrial roof, but its complete isolation obliged us to open the left atrial chamber. This condition did not permit us to perform a simple ligation with a beating heart. Normothermic cardiopulmonary bypass was instituted. The ascending aorta was clamped and warm blood cardioplegia was infused into the aortic root. The right atrium was opened and the fistula outlet, a single hole (8 mm in diameter) in the posterior wall of the atriocaval junction, was detected and sutured. The fistula was isolated in the left atrial roof (Fig. 2) and incised about 1 cm from the junction with the superior vena cava. The origins of the intermediate and circumflex branches were not visible because they were more left-sided, as suggested at angiography performed preoperatively. A short tract (1 cm), which did not contain anomalous coronary branches, was excised and the two

stumps (wall thickness approximately 2 mm) were sutured.

The patient was discharged on postoperative day 7 without complications. The postoperative creatine kinase-MB peak was 14 U/l. Oral anticoagulation was instituted and continued for 6 months to prevent coronary thrombosis.

At 1 year of follow-up, the patient was asymptomatic and echocardiography confirmed the interruption of the fistula. No complications on the vessel proximal to the closure were detected. The dimensions of the proximal residual chamber of the fistula were unchanged (Fig. 3). The left ventricular wall motion was normal.

Discussion

The present case report refers to an unusual fistulous connection¹ complicated by bacterial endocarditis

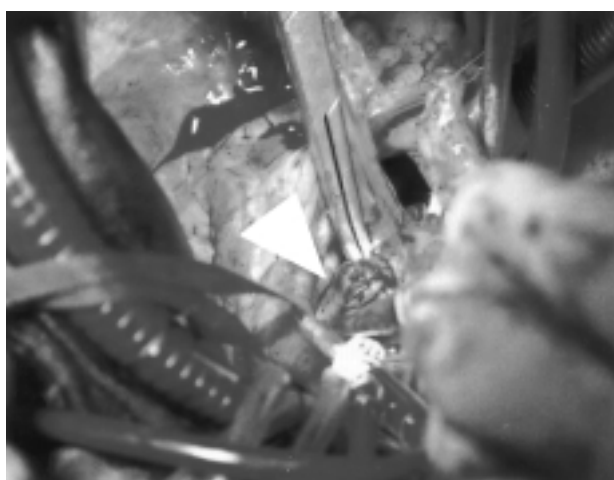


Figure 2. Intraoperative view: the white triangle indicates the fistula isolated into the left atrial roof.

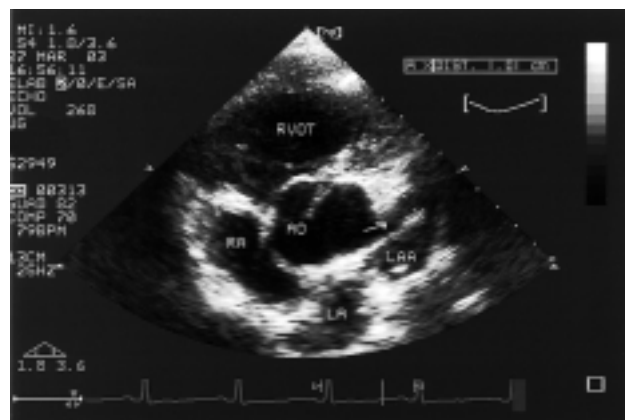


Figure 3. One-year echocardiography showing unchanged dimensions of the residual chamber. The white arrow indicates the residual tract of the fistula originating from the left Valsalva sinus. AO = aortic root; LA = left atrium; LAA = left atrial appendage; RA = right atrium; RVOT = right ventricular outflow tract.

which resolved without the need for surgery. Asymptomatic coronary fistulas complicated by bacterial endocarditis constitute a rare event.

Recent reports recommend surgery for all patients with CAF and demonstrable shunting², even if asymptomatic, because of the risk of future complications³. Successful transcatheter treatment of a fistula from the left main artery and right atrium was already reported in 1984⁴ and today percutaneous procedures have become a primary therapeutic option. However, fistulas with multiple communications or giving origin to vessels of functional importance that could be compromised during percutaneous maneuvers should better be addressed by surgery⁵. In our case, a transcatheter procedure was not considered because of the perceived risk of complications on large coronary branches. Ligation with a beating heart represents the easiest choice in the surgical treatment of this anomaly⁶. In this case cardiopulmonary bypass was instituted because of the necessity to open the left atrial roof to achieve complete isolation of the fistula. The surgical procedure was simple but left a residual chamber in communication with the aortic root. Some reports showed that proximal coronary artery dilation persists after the operation and sometimes thromboses^{2,7}. Araya et al.⁸ described the fatal rupture of the dilated proximal artery after closure of a coronary fistula. To prevent this complication a reduction aneurysmectomy has been suggested when the proximal coronary artery is very dilated and thin-walled². Because of the risk of complications on the coronary ostia, the moderate dilation and the wall

thickness of the fistula, we avoided any procedure on the proximal tract.

One-year echo follow-up confirmed the absence of an unfavorable evolution on the residual tract of the fistula and its coronary branches, showing that the simple surgical approach described may offer an effective and definitive treatment.

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