

Case Report

Successful Surgical Correction of a Patient with Congenital Coronary Arteriovenous Fistula Between Left Main Coronary Artery and Right Superior Cavo-Atrial Junction

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The case of a 7-year-old female with a congenital left main coronary arteriovenous fistula to the right superior cavo-atrial junction, presenting with congestive cardiac failure, is reported for its rarity. The surgical importance of this anomaly is highlighted.

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Congenital coronary arteriovenous fistulas are rare, with a reported incidence of 0.1 to 0.2%.¹ The right coronary artery is the most common site of origin and the right ventricle (approximately 40%) and right atrium are the most common draining chambers.²⁻⁵

We present the surgical treatment of a giant left main coronary arteriovenous fistula draining into the right superior cavo-atrial junction. We were able to find only six other reports of cases with an anomalous left main coronary vessel draining into the superior cavo-atrial junction.⁶⁻¹¹ The extreme rarity of the disease entity, the use of contrast echocardiography and angiography for diagnosis, the successful surgical correction and a brief literature review of the outcome after surgical repair form the basis of this report.

Case presentation

A 7-year-old girl weighing 18 kg was referred to our cardiac clinic for progressively increasing dyspnoea and palpitations. She had been dyspnoeic for one year and was in

New York Heart Association (NYHA) class III at the time of surgery.

On examination, she was well nourished, but had tachypnoea and tachycardia. Blood pressure was 118/80 mmHg. Clinically, she had cardiomegaly, a systolic-diastolic precordial thrill, and a continuous machinery murmur, loudest at the left 4th intercostal space. Chest radiography revealed cardiomegaly, with a cardiothoracic ratio of 0.70. The electrocardiogram showed a QRS axis of +90°, right axis deviation and right ventricular hypertrophy.

Two-dimensional echocardiography and colour flow imaging were performed using a Hewlett-Packard Sonos 5500 with a 3.5 MHz transducer. The examination revealed a dilated left main coronary artery (1-1.5 cm in diameter) arising from the aorta. A normal sized right coronary artery was seen in the atrioventricular groove. A large and tortuous extracardiac vessel posterior to the aorta and running in the atrioventricular groove was also noted. The vessel terminated in the medial portion of the superior cavo-atrial junction. Color Doppler showed a continuous turbulent flow pat-

tern of high intensity throughout the cardiac cycle, with equiphasic systolic and diastolic peaks maximally in the region of the superior cavo-atrial junction.

Oximetry showed increased oxygen saturation of 88% at the lower end of the superior *vena cava*, with high right ventricular filling pressures, and the QP/QS ratio was calculated at 2.6:1. Selective coronary arteriography demonstrated a large vessel originating from the left sinus of Valsalva in the region of the left coronary orifice. It had a tortuous course and passed posteriorly, medially, and superiorly, ultimately joining the superior *vena cava* at its junction with the right atrium (Figure 1). Computerised tomographic angiography revealed a fistulous vessel arising from the left coronary sinus, coursing to the right posteriorly, and draining into the superior *vena cava* (Figure 2).

Intraoperatively, after opening the pericardium, the fistula was identified running along the upper margin of the right atrium adjacent to the sino-atrial nodal artery and terminating with an aneurysmal bulge at the medial aspect of the superior cavo-atrial junction (Figure 3A). Because of the intra-myocardial course and the proximity of the sino-atrial nodal artery, it was decided to ligate the fistula under cardiopulmonary bypass using aorto-bicaval cannulation. The right atrium was opened with the heart perfused and beating and the fistula opening was easily identified at the superior cavo-atrial junction. After clamping the aorta, myocardial protection was achieved using antegrade cardioplegia with digital pressure on the fistula and topical cooling. It was considered too dangerous to attempt to ligate the fistula proximally to the left main coronary artery. The fistulous opening was closed

using two pledgeted mattress sutures (Figures 3B-3E). Post bypass there was no step up in oxygen saturation in the right atrium or the superior and inferior *venae cavae*. The patient had an immediate marked reduction of cardiomegaly and an uneventful postoperative course. After 16 months of follow up, she is in NYHA functional class I and is receiving no medications.

Discussion

Since the first description of congenital coronary artery fistula by Krause in 1865, these abnormal communications have been reported to be present in 1 in 50,000 live births (1% to 2% of the general population) and are reported to occur in 0.2% to 0.25% of patients undergoing coronary arteriography.¹⁻¹⁷

Coronary artery fistulas may be isolated (55% to 80% of cases), or associated with other congenital cardiac anomalies (20% to 45% of cases).¹⁻¹⁷ Associated anomalies include tetralogy of Fallot, atrial septal defect, patent *ductus arteriosus*, ventricular septal defect and superimposed coronary artery disease (35%).¹⁻⁵ Coronary artery fistulas may be single (74% to 90%), multiple (10.7% to 16%), or may originate from both coronary arteries (4% to 18%).¹⁻¹⁷

No definite pathogenetic explanation has been described for this anomaly. It has been postulated that it may be due to failure of obliteration of the intra-myocardial trabecular sinusoids with anomalous development of the intra-trabecular spaces, through which blood is supplied to the myocardium during intrauterine life.¹⁻⁹

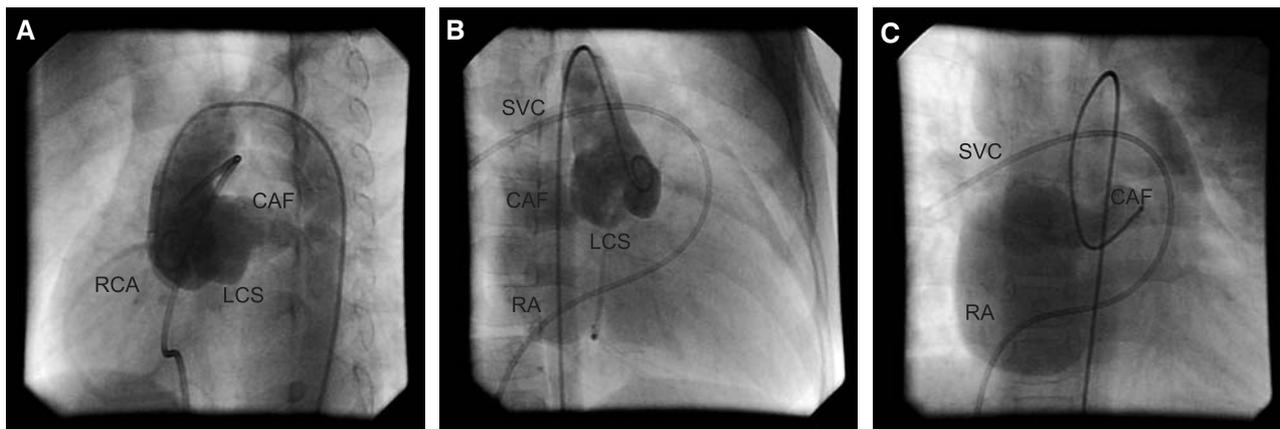


Figure 1. A, B. Aortogram in left anterior oblique projection shows dilated and tortuous coronary artery fistula (CAF) arising from the left coronary sinus (LCS) and draining into the lower end of the superior vena cava (SVC). In comparison, the right coronary artery (RCA) is normal in calibre. C. Selective injection into the left coronary artery, showing the dilated and tortuous fistulous (CAF) tract arising from the left coronary sinus (LCS) and emptying into the superior cavo-atrial junction. Note dense opacification of the right atrium (RA).

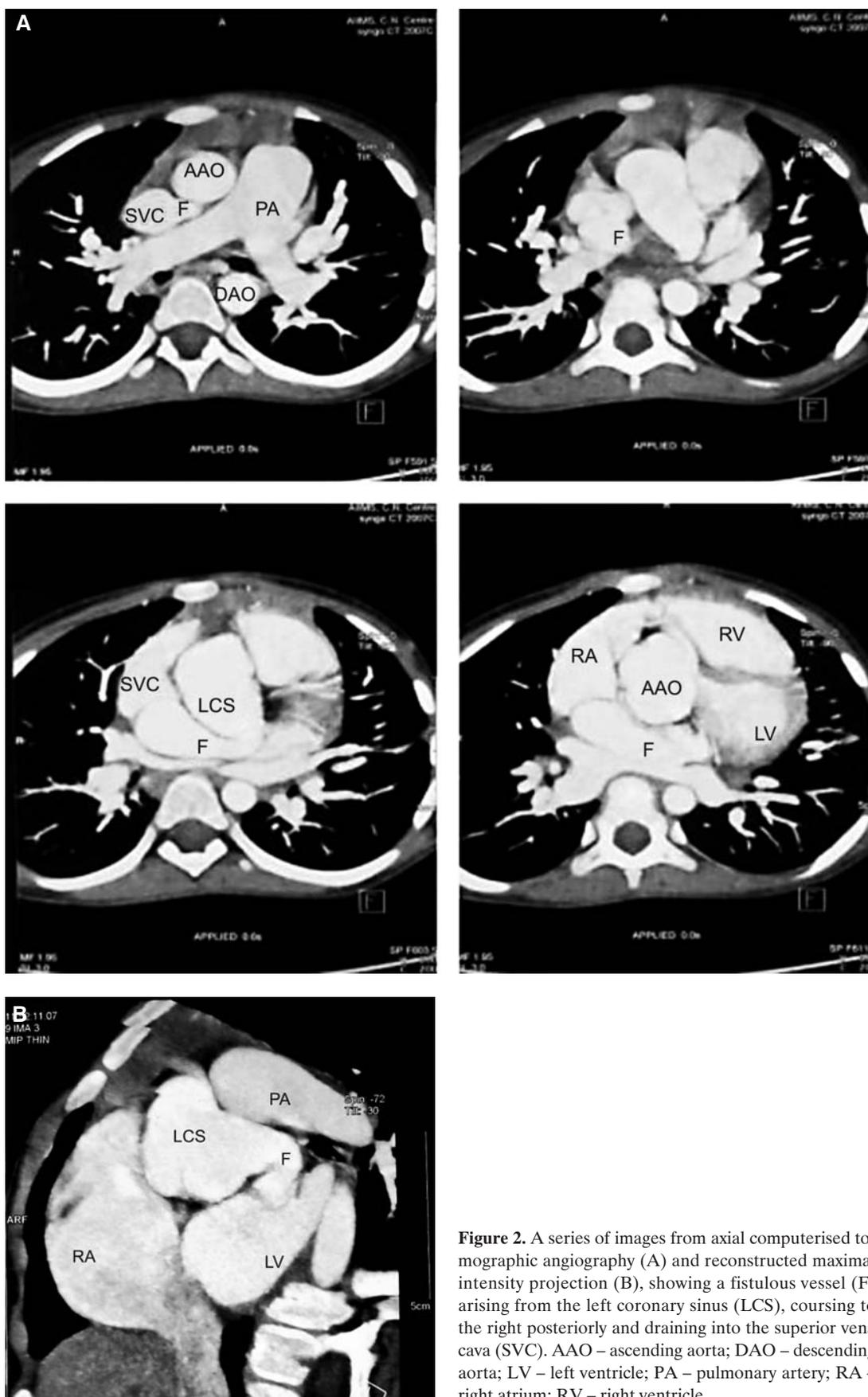


Figure 2. A series of images from axial computerised tomographic angiography (A) and reconstructed maximal intensity projection (B), showing a fistulous vessel (F) arising from the left coronary sinus (LCS), coursing to the right posteriorly and draining into the superior vena cava (SVC). AAO – ascending aorta; DAO – descending aorta; LV – left ventricle; PA – pulmonary artery; RA – right atrium; RV – right ventricle.

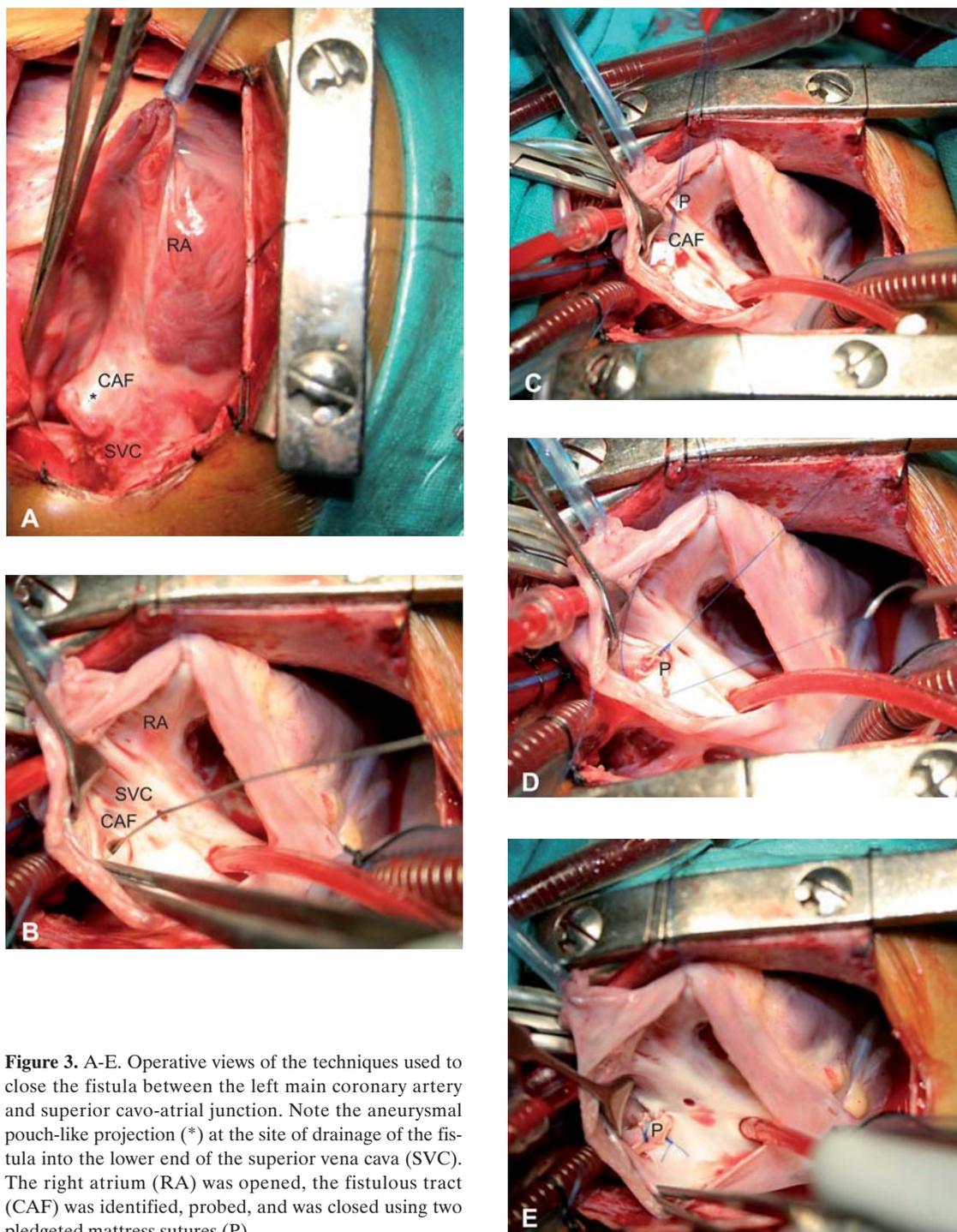


Figure 3. A-E. Operative views of the techniques used to close the fistula between the left main coronary artery and superior cavo-atrial junction. Note the aneurysmal pouch-like projection (*) at the site of drainage of the fistula into the lower end of the superior vena cava (SVC). The right atrium (RA) was opened, the fistulous tract (CAF) was identified, probed, and was closed using two pledgetted mattress sutures (P).

Approximately 92% of these coronary artery fistulas drain into the right side of the heart, whereas drainage into the left side of the heart occurs in only 8% of cases.^{2,4,14-17} Careful analysis of the published series documents the origin of the fistulous communication from various sites, including the right coronary artery (50-60%), the left anterior interventricular coronary artery (25-42%), both right coronary and

left anterior interventricular coronary artery (5%), circumflex coronary artery (18.3%), diagonal (1.9%), marginal coronary artery (0.7%), and single coronary artery (3%).¹⁻¹⁷ Its origin from the left main coronary artery is rare.¹⁻¹⁷

We analysed the published literature using a Medline search and identified only 13 instances of fistulous coronary artery drainage into the superior caval vein.

Within this group, we identified 3 patterns of superior vena caval fistulous communications, namely: (i) circumflex coronary artery to superior caval vein; (ii) circumflex coronary artery to persistent left superior caval vein; and (iii) right coronary artery to superior caval vein.¹⁸⁻³¹

The disease entity should be suspected when the following combination of symptoms and signs coexist: angina, dyspnoea, congestive heart failure, arrhythmias, and continuous machinery murmur at the second/third right or left parasternal border. Diagnosis of this anomaly is possible once the possibility is entertained. Two thirds of reported patients exhibited signs of ischaemia, chamber overload, infarction, premature ventricular contractions, supraventricular arrhythmias and conduction defects.¹⁻³¹ A plain chest X-ray shows cardiomegaly in two thirds of cases and left-to-right shunt with pulmonary plethora in 16.5% of cases.¹⁻³¹

The use of two-dimensional and color Doppler echocardiography has been discussed extensively in the literature.¹⁻³² Computerised tomographic angiography and magnetic resonance imaging (MRI) are new techniques available for non-invasive cardiac imaging. Despite isolated case reports of successful diagnosis, the main limitations of MRI are limited availability, relative expense and the need for specialised staff for data interpretation. Computerised tomographic angiography has been shown to be useful in demonstrating the origin, proximal fistulous tract of the left coronary artery and peripheral pulmonary vasculature. However, the entire course of the artery and the normal sized right coronary artery may not be visualised.³² The diagnostic "gold standard" remains cardiac catheterisation and coronary angiography in the study of coronary arterio-venous fistulas. These studies allow the assessment of haemodynamics, quantification of the shunt, delineation of the origin, site, and termination of coronary artery fistulas, and the definition of multiple fistulas.

The natural history of coronary artery fistulas is such that they tend to increase in size, and without surgery the patients do not have a normal life expectancy.^{1,2} In rare instances, sudden death may be the first manifestation of coronary artery fistulas.^{5,33} The reported incidence of subacute infective endocarditis in untreated patients with coronary artery fistula is 0.001% to 0.004% per patient year.⁵

In a review of published studies, Liberthson et al found 174 patients with congenital coronary artery fistulas. Overall, 39% had symptoms or complica-

tions, including dyspnoea on exertion, fatigue, angina, congestive heart failure, myocardial infarction, myocardial rupture, subacute bacterial endocarditis and death. Symptoms and complications occurred in 19% of patients aged <20 years and in 63% of patients aged >20 years. Surgical complications occurred more frequently in the latter group. Spontaneous closure of the fistula occurred in only two patients.⁵ On the basis of these data, the authors recommended closure of coronary artery fistulas during childhood even in asymptomatic patients.⁵ The complications of untreated patients with coronary artery fistula include myocardial infarction (3-11%), subacute infective endocarditis (5-20%), aneurysm formation (19-26%), rupture and death (7-14%).^{1-11,33}

Consensus exists concerning the surgical treatment of patients with symptoms, regardless of age.¹⁻³² For patients with asymptomatic fistula, however, the timing of and indications for surgery are controversial.¹⁻³³ Many and various surgical techniques have been described to deal with coronary artery fistulas. These include internal cameral closure using cardiopulmonary bypass; tangential arteriorraphy, with or without bypass; distal ligation; ligation and saphenous vein/internal mammary artery bypass.^{1,4-31} In general, the fistulas that are intracameral, short, close to a critical coronary supply, or associated with an aneurysm or congenital heart disease are closed with the use of cardiopulmonary bypass.^{1,5-31} Internal closure in the right heart chamber can be performed using cardiopulmonary bypass without cross clamping or cardioplegia on a beating or fibrillating heart.⁴ In contrast, coronary artery fistulas that are extracameral and anatomically accessible are usually controlled with ligature without using cardiopulmonary bypass.

Surgical techniques must be tailored to individual anatomy and the superiority of one technique over another has not been demonstrated so far. The successful use of percutaneous transcatheter closure devices has been described, and these are increasingly used, especially in the paediatric population.^{2,34,35} However, reports of embolisation therapy do not adequately document long-term follow up addressing the incidence of coil migration, clot propagation and recurrence.

The rarity of this case was the intra-myocardial course of the fistulous tract, voiding into the lower end of the superior *vena cava*, and the proximity of the sino-atrial nodal artery, which was readily accessible through a less invasive localised exposure under cardiopulmonary bypass.

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