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Large Left Coronary Artery to Right Ventricular Fistula in A Paediatric Patient: A Watchful Approach and Successful Closure Utilising Modern Imaging Modalities

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Abstract

Background: Coronary artery fistulae are rare anomalies with an incidence of 0.2-1.3%. Management for symptomatic patients is clearly defined, however; in asymptomatic patients, indications and timing for intervention as well as suitable type of intervention remains debated.

Case presentation: We describe a 3 months old male infant referred with an asymptomatic systolic murmur. The infant diagnosed with a large left coronary artery fistula communicating with the right ventricle. On serial review; the infant showed evidence of progressive left heart volume loading on the transthoracic echocardiogram, by 15 months of age.

Computed tomography was integral to pre-operative planning. The fistula was uneventfully closed by percutaneous trans-catheter embolization using an 8x7mm second generation Amplatzer® vascular plug.

Follow up angiography and echocardiogram showed no residual flow.

Conclusion: We describe a toddler presenting in infancy who underwent successful transcatheter closure of a large left coronary artery to right ventricular fistula, following an initial conservative management approach. Left-sided volume loading was alleviated following percutaneous closure. Pre-operative imaging facilitated successful planning and closure.

Keywords: Coronary artery fistula, catheterisation, trans-catheter embolization, percutaneous, congenital heart disease.

Background

Coronary artery fistulae are rare cardiac anomalies with an incidence of 0.2-1.3% [1,7]. We describe an infant diagnosed with a large left coronary artery to right ventricular fistula who was initially managed conservatively before definitive trans-catheter device closure was performed.

Case Presentation

A 3-month old boy was referred to our Paediatric Cardiology clinic with a murmur. He was asymptomatic and had been feeding well with good weight gain since birth. Cardiac examination revealed an active precordial impulse with a continuous 3/6 murmur throughout the left sternal border. He had no hepatomegaly and had an otherwise unremarkable examination.

A transthoracic echocardiogram showed a large left coronary artery to right ventricular fistula (Figure 1), following along the course of the left circumflex artery territory (Figure 2) and entering into the base of the right ventricle below the septal leaflet of the tricuspid valve (Figure 3). The left anterior descending artery originated from the proximal part of the coronary fistula (Figure 1). There was minimal left ventricular volume loading. There was normal biventricular systolic function with no regional wall abnormalities (Figure 1-3).

Computed Tomography (CT) angiography confirmed a large left main coronary artery to basal right ventricle coronary artery fistula (Figure 4,5). The coronary arrangement was conventional with distal bifurcation of the left main coronary artery into the left anterior descending and circumflex arteries, and a normal right coronary artery origin and course. The left main coronary artery was significantly dilated (Figure 4), measuring 5.3mm (z-score +10.5), with the LAD tapering acutely to a normal calibre within the anterior interventricular groove. The fistula coursed in the circumflex artery territory, measuring 7.7mm proximally (z-score +19.3) and tapering distally to 3.7mm at its minimal diameter (Figure 6,7). There was mild dilatation of the left ventricle.

Given the stable clinical condition and minimal left sided volume loading, the infant was followed clinically with a view to performing a trans-catheter device embolization electively.

Although he remained asymptomatic, there was progressive left-sided volume loading on serial echocardiograms. At 15 months, his echocardiogram demonstrated a moderately dilated left atrium and mildly dilated left ventricle. There was holodiastolic flow reversal in the descending and abdominal aorta Appearances of the fistula appeared unchanged with restrictive continuous left to right flow, with no evidence of thrombus, ischaemia or regional wall motion abnormalities. The ECG showed left ventricular hypertrophy with no evidence of ischaemia.

The toddler was brought forward for percutaneous device embolization of the coronary fistula. Access was obtained through the right femoral artery and right femoral vein using 4 French and 5 French sheaths. 50 IU of heparin was given once arterial access obtained. Coronary angiogram identified a large fistula from the proximal left coronary artery coursing inferiorly in the inter-ventricular groove and entering the right ventricle below the septal leaflet of the tricuspid valve (Figure 8,9). Left to right shunt was calculated at 1.6 L/min/m^2 with normal pulmonary vascular resistance of 2 Wood Units.

A 0,035" Terumo wire was passed through the fistula from the left main coronary artery, into the right ventricle and left pulmonary artery, where it was snared and exteriorised via the right ventricle to right atrium and right femoral vein to create an atrioventricular loop. A long 4F Flexor sheath was placed over the wire into the mid-fistula from the venous side. An 8mm Amplatzer Vascular Plug (AVPII) was delivered into the fistula and positioned at the distal end. The ECG was monitored prior to release with no evidence of ectopy or ischaemia. Post deployment aortic root angiography confirmed a stable device in good position with no residual flow (Figure 10,11). Post procedure transthoracic echo confirmed normal tricuspid valve function and good biventricular systolic function (Figure 12). There were no arrhythmias or ischaemic changes during or post-procedure.

He had an uncomplicated post–operative recovery and continues to do well 6 months following his procedure. He remains on aspirin empirically. There was no residual flow through the fistula

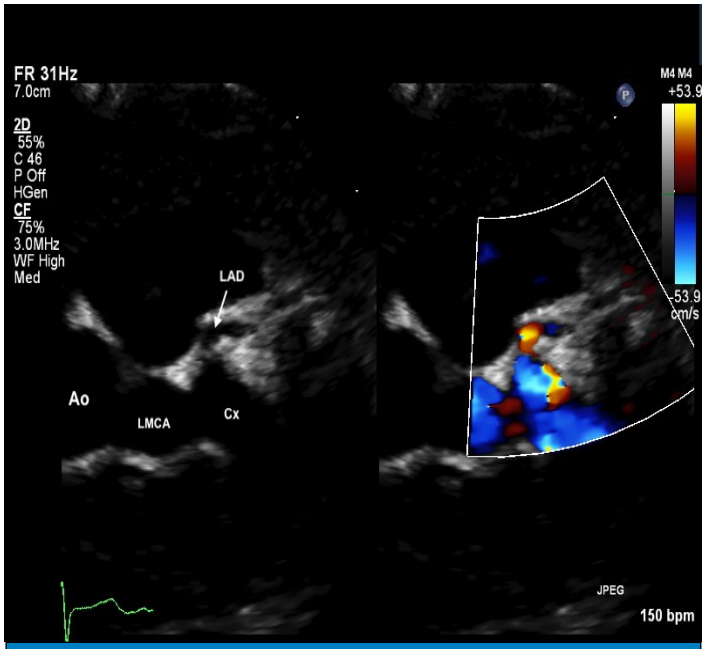


Figure 1: Parasternal short axis view demonstrating the origin of the coronary fistula following along the course of the left circumflex artery territory and the LAD originating from the proximal part of the fistula. (LAD – left anterior descending, Ao – aorta, LMCA – left main coronary artery, Cx – circumflex).

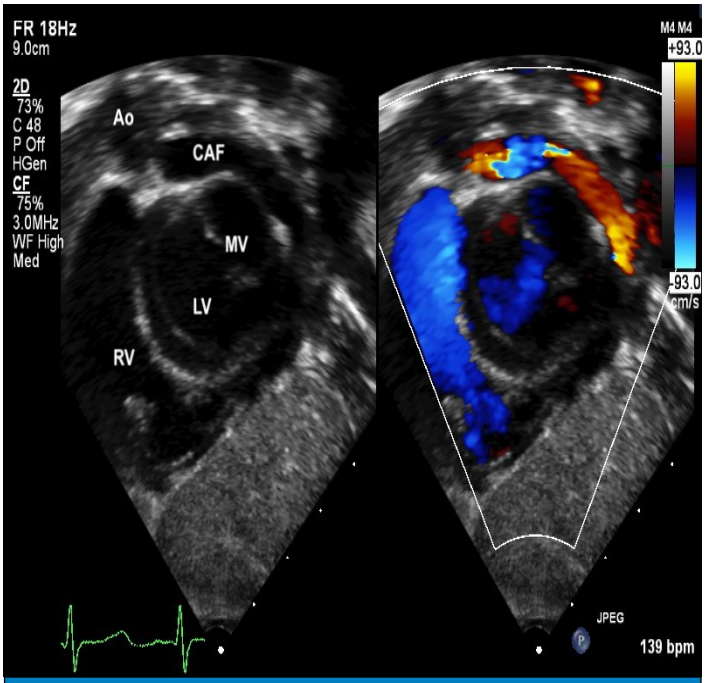


Figure 2: Subcostal view demonstrating the course of the coronary fistula along the circumflex artery territory. (Ao – aorta, CAF – coronary artery fistula, MV – mitral valve, LV – left ventricle, RV – right ventricle).

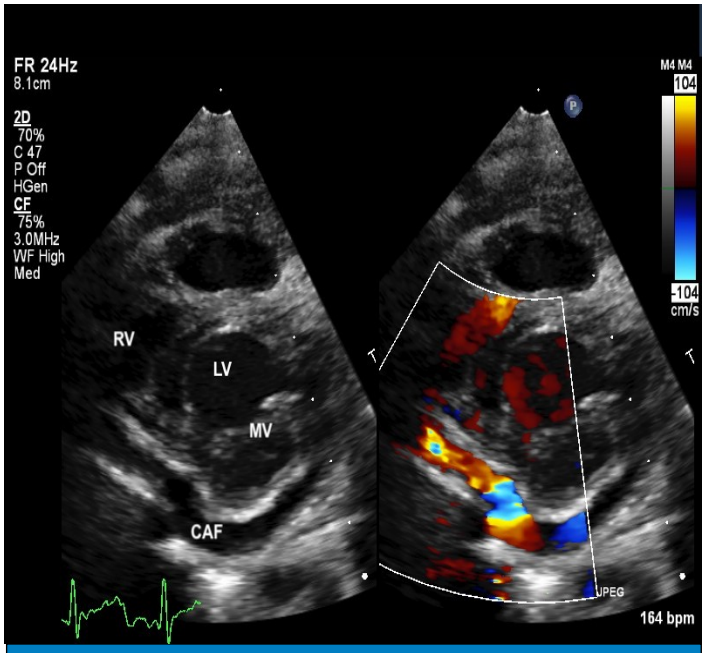


Figure 3: Short axis parasternal view demonstrating the insertion of the coronary artery fistula into the right ventricle. (RV – right ventricle, LV – left ventricle, MV – mitral valve, CAF – coronary artery fistula).

Figure 4: Coronal CT (Computed Tomography) view demonstrating the origin of the coronary artery fistula off the left main coronary artery proximally. (PA – pulmonary artery, Ao – aorta, LMCA – left main coronary artery, LV – left ventricle, RAA – right atrial appendage).

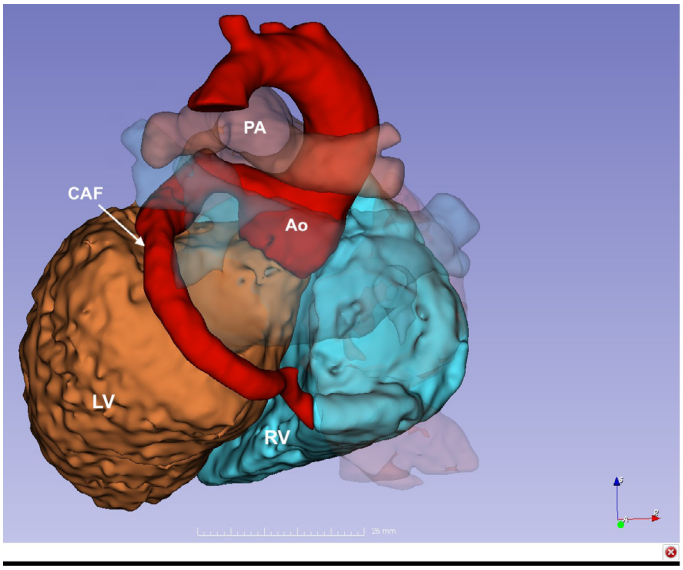
and no thrombus to proximal coronary arteries. Left ventricular systolic function remains normal with no regional wall motion abnormalities.

Discussion and Conclusion

Congenital coronary artery fistulae are rare anomalies [1,8]. They can be either congenital or acquired. Haemodynamically significant coronary artery fistulae are found in less than 1% of adults. In a study by Levin et al, 50% of coronary artery fistulae were from the right coronary artery and 42% from the left coronary artery with some rarely arising from both (5%) [1]. The vast majority drain in the right heart structures (90%), mostly the right ventricle (41%) followed by the right atrium (26%) and much less likely to the pulmonary artery (16%) [1]. In 90% of cases, they are single fistulae draining into the venous system [1,8]. Symptoms are dependent on the size, exit point and restrictive nature of the defect[5] [5]. Early intervention is required in infants and neonates with clinical evidence of heart failure [2-5].

Figure 5: Sagittal CT (Computed Tomography) view demonstrating the course of the coronary artery fistula along the circumflex coronary artery territory and inserting into the base of the right ventricle close to the tricuspid valve. (Ao – aorta, LV – left ventricle, CAF – coronary artery fistula, RV – right ventricle).

Management of coronary artery fistulae depends on the size and haemodynamic significance of the shunt [6]. Most infants with coronary artery fistulas are asymptomatic. Traditionally, small asymptomatic fistulae are managed conservatively as they are relatively benign with a potential for spontaneous closure with a reported range of 1- 23% in small case series [7]. This number is no doubt underestimated given that small coronary fistulas do not give rise to symptoms or murmurs in infants making detection challenging as echocardiogram would not be indicated. Larger



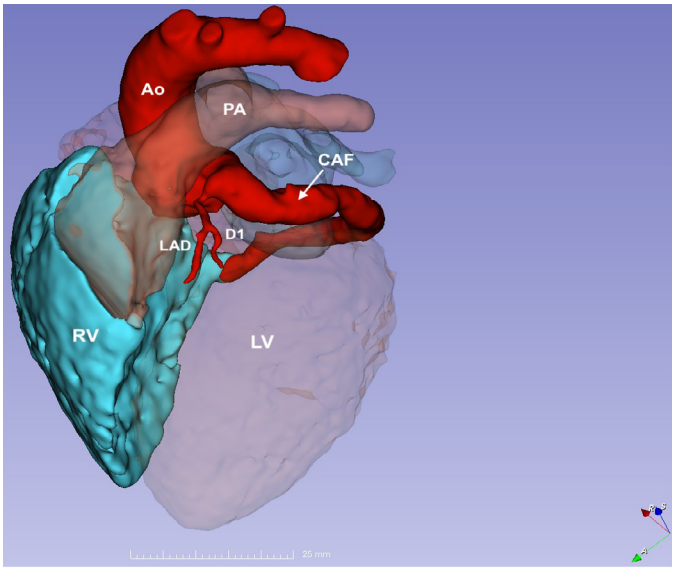


Figure 6 & 7: Reconstructed 3D (dimensional) images demonstrating the full course of the coronary artery fistula as it originates off the left coronary sinus, courses along the circumflex coronary artery territory and inserts into the base of the right ventricle. Also demonstrated in figure 7; the normal size left anterior descending coronary artery originating proximally off the fistula. (CAF – coronary artery fistula, PA pulmonary artery, Ao – aorta, LV – left ventricle, RV – right ventricle, LAD – left anterior descending, D1 – first diagonal coronary artery branch).

fistulae would require surgical ligation or trans-catheter closure [8,9]. Late complications of unrepaired fistulae including coronary steal, thromboembolism, arrhythmias, endocarditis or fistula rupture [4,5].

The ACC/AHA Guidelines recommend both surgical and trans-catheter interventions for management of coronary artery fistulae in adults with no clear universal guidelines in children [10]. Trans-catheter closure is considered the treatment of choice if the anatomy of the fistula is favourable, including a single fistula with a single narrow drainage site and a clear origin proximally with no other associated structural heart disease requiring surgical intervention [11]. Full evaluation of the fistula including transthoracic echocardiography, CT angiography and trans-catheter angiography is integral to accurately determine suitability for trans-catheter embilisation [5,9].

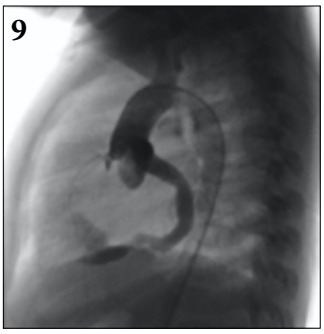
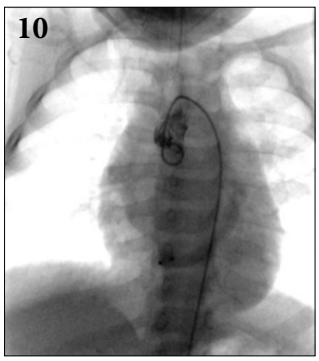


Figure 8 & 9: Aortic root angiogram on antero-posterior and lateral projections demonstrating the full course of the coronary artery fistula originating at the left main coronary artery and giving rise to a normal size left anterior descending as it courses along the circumflex coronary artery territory before entering the base of the right ventricle.



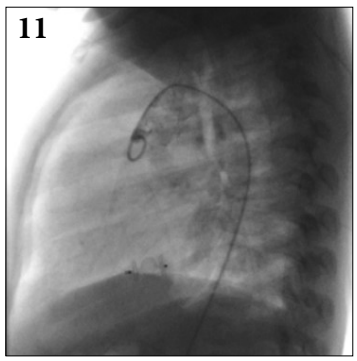


Figure 10 & 11: Aortic root angiogram on antero-posterior and lateral projections following closure of the coronary artery fistula with no residual flow into the base of the right ventricle

Cardiac catheterisation in general is well known to have a higher risk of complications in infants compared to older children ranging from major complications such as death, cerebral infarction and cardiac injury to minor transient arrhythmias during the procedure with arterial thrombosis being the most common complication of interventional cardiac catheterisation [12,13]. One reasonable size study reported arterial thrombosis rate as high as 86% in infants less than 12 months compared to 11.4% in older children [14].

Our patient was conservatively managed initially given his clinical stability. Regular monitoring with serial echocardiograms allowed us to detect early signs of progressive left-sided volume loading in the absence of clinical symptoms enabling us to intervene at an optimal time with the child reaching an age and size that would make a trans-catheter approach both more safe and effective.

In conclusion, we describe an asymptomatic infant with a large congenital coronary artery fistula who underwent successful percutaneous device embolization. Elective intervention was indicated given the progressive left-sided volume loading on serial imaging. With careful pre-procedural planning and multi-modality imaging, successful closure was achieved at the right timing allowing the child to grow and hence minimising known potential transcatheter closure complications in infancy.

Declarations

Ethical Approval

Ethic approval was not required. Consent obtained from parents at the time of intervention for use of imaging and potential publication.

Data sets

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interests

Authors declare no competing interests

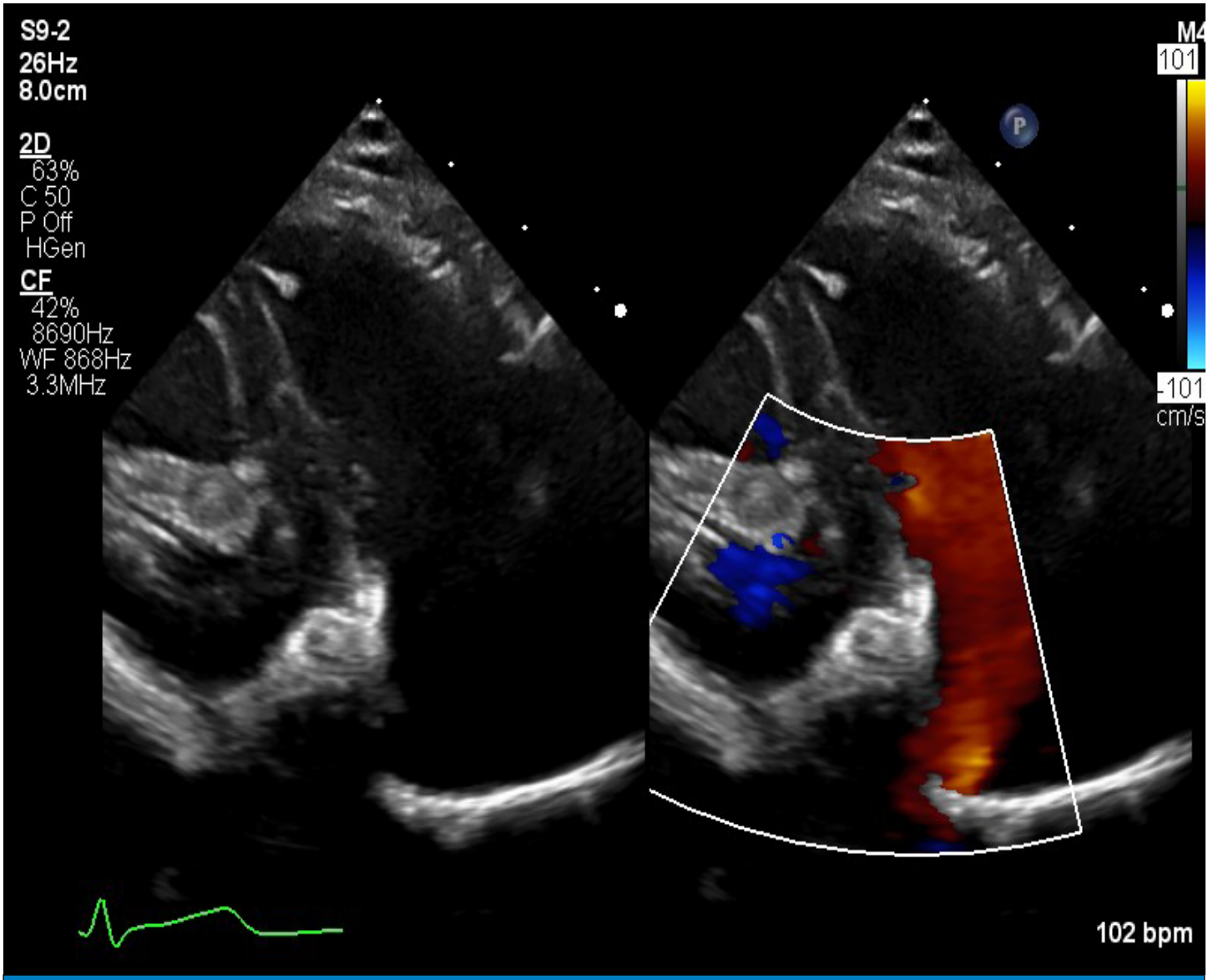


Figure 12: Parasternal long axis view of the right ventricular inflow showing the device well seated bellow the septal leaflet of the tricuspid valve (TV) with no residual flow into the right ventricle (RV).

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Authors' contributions

Ibtihalat Mohamed designed the layout and wording used in the presentation and cited references. Greg Murphy contributed to the background research and the general layout. Deane Yim reported the imaging and edited the manuscript. Steve Shipton was the intervention cardiologist who performed the trans-catheter embolization and obtained consent from parents.

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