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ORIGINAL ARTICLE

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# Congenital coronary artery fistula: Presentation in the neonatal period and transcatheter closure

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## Abstract

**Background**: Congenital coronary artery fistula is a rare coronary anomaly. Most commonly, such fistulae drain into the right side of the heart or the pulmonary artery. Children with coronary artery fistulae are generally asymptomatic, although they may have left ventricular enlargement in the setting of a moderate sized left to right shunt. Symptoms of congestive heart failure or ischemia are very rare in neonatal period, and suggest the presence of a very large shunt and/or coronary steal.

**Methods**: Single center retrospective review of transcatheter intervention on coronary artery fistulae presenting with symptoms in the neonatal period from January 2000 to March 2018. Patients with associated congenital heart diseases (except atrial septal defect) were excluded. Patient records, catheterization data, angiograms and noninvasive imaging were reviewed.

**Results**: Two patients underwent transcatheter intervention for symptomatic coronary artery fistula in the first few weeks of life. The first patient had multiple right and left coronary artery to right ventricle fistulae and presented with severe biventricular systolic dysfunction. Transcatheter closure of the fistulae was performed using multiple Gianturco coils. The second patient had a large left main coronary artery to left ventricle fistula (with left anterior descending and circumflex coronary artery atresia) presenting with symptoms of ischemia. This large fistula was closed using one Amplatzer Vascular Plug type-II and two Micro-Vascular Plugs. Both patients had improvement in symptoms post intervention and are doing well at the last follow up at 12 years and 7 months respectively.

**Conclusions**: We hereby describe the rare presentation of symptomatic coronary artery fistulae in the neonatal period and their successful transcatheter management. This is also the first description of left anterior descending and circumflex coronary artery atresia in the setting of a large left main coronary artery to left ventricle fistula.

KEYWORDS
congenital heart disease, coronary anomaly, coronary fistula

  

## 1 | INTRODUCTION

Coronary artery fistulae are congenital or acquired coronary artery anomalies in which blood is shunted bypassing the myocardial capillary network into a cardiac chamber (coronary-cameral fistula). These are relatively rare with an incidence estimated to be around 0.1%-0.2%[1] of all patients who undergo selective coronary angiography. These fistulae are commonly asymptomatic in neonatal period, although congestive heart failure may present in children with relatively large shunts. Treatment is often advocated to reduce the risk of myocardial ischemia, congestive cardiac failure, infective endocarditis, and other rare complications such as rupture of the aneurysmal fistula at a later age. Surgical ligation with or without coronary artery bypass grafting or percutaneous closure using a variety of devices have been reported in children previously.[2,3] Transcatheter closure of coronary fistulae in the first few weeks of life is only rarely reported due to the rarity of this lesion presenting with symptoms in the neonatal period.

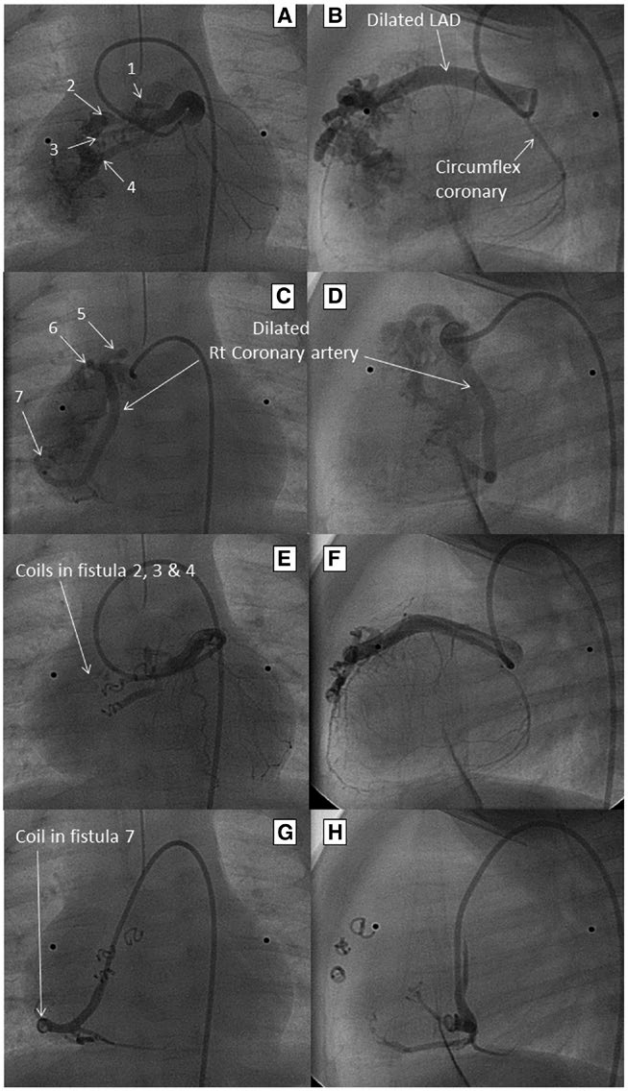


**FIGURE 1** Coronary angiograms of Patient 1. (A, B) AP and lateral projections of left coronary angiogram depicting the dilated left anterior descending (LAD) coronary artery and normal circumflex coronary artery. There are four fistulae (1-4) from the left coronary artery to the right ventricle; (C,D) AP and lateral projections of right coronary angiogram showing the diffusely dilated right coronary artery with proximal (5, 6) and distal (7) fistula into the right ventricle. The right coronary artery is also diffusely dialted; (E, F) AP and lateral projections of left coronary angiograms after coil occlusion of the fistulae 2, 3 and 4 from the left anterior descending coronary artery to the right ventricle using 0.038″ × 2 cm × 3 mm Gianturco coils (Cook Medical, Bloomington, IN); (G, H) AP and lateral projections of right coronary angiogram after coil occlusion of the distal fistula (7) to the right ventricle

## 2 | METHODS

We performed a retrospective review of neonates who were symptomatic from coronary artery fistulae and underwent transcatheter intervention at Texas Children's hospital, Houston, Texas from January 2000 to March 2018.

Coronary artery fistulae were initially diagnosed on the basis of echocardiogram or advanced imaging modalities in the form of cardiac computed tomography (CT) or magnetic resonance imaging. Confirmation of the diagnosis was performed using conventional angiography. Patients were included if they had symptoms ascribable to coronary artery fistulae in the form of congestive heart failure, myocardial ischemia, infective endocarditis or rupture. Those with associated congenital heart disease, except atrial septal defect or patent foramen ovale, were excluded. Those with iatrogenic coronary artery fistulae were also excluded.

Demographic and clinical information was accessed using electronic medical records. Medical records were reviewed for reported symptoms ascribable to the coronary artery fistulae in the neonatal period. Noninvasive imaging and cardiac catheterization data including the angiograms were reviewed. Both patients underwent transcatheter intervention for closure of the fistulae.

## 3 | RESULTS

Case 1: A term female born to a 35-year-old multi-gravida mother was diagnosed to have marked cardiomegaly on fetal ultrasound and further evaluation revealed torrential retrograde flow in the ascending aorta due to a large right and left coronary artery fistulae draining into her right ventricle. There was no known maternal medical disease or family history of any cardiac condition. There was significant concern about the patient developing heart failure after birth when separation from the placental circulation would result in an immediate increase systemic vascular resistance, which could potentiate the shunt through the coronary fistulae.

After vaginal delivery, she did not need any significant resuscitation. She had a grade III/VI continuous murmur in the right upper and right lower sternal borders. Her echocardiogram confirmed the prenatal findings and demonstrated severe dilation of the right

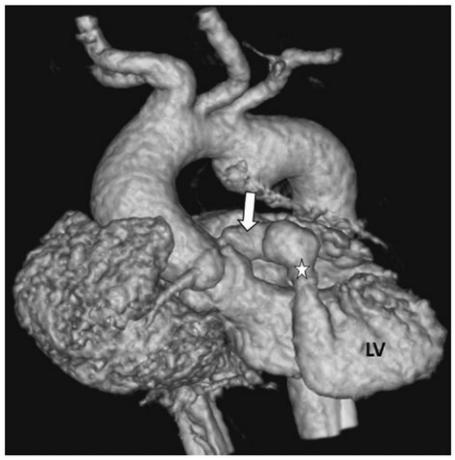


**FIGURE 2** The 3D reconstruction image from the computed tomography scan of Patient 2 demonstrating the aneurysmal proximal left main coronary artery (arrow) draining into the left ventricular cavity (LV). The fistula (*) measured 4 mm in minimal diameter, 8 mm in maximal diameter and 16 mm in length

ventricle and bowing of the interventricular septum into the left ventricular cavity, with biventricular systolic dysfunction. She developed increasing tachypnea and tachycardia over the next few days, chest x-ray showing massive cardiomegaly (Figure S1). The complexity of her numerous coronary fistulae and her presentation with heart failure and systolic dysfunction in the neonatal period was estimated to carry a very poor prognosis, and she was initially believed to be inoperable. She was eventually referred to the cardiac catheterization laboratory for hemodynamic assessment and intervention at the age of 5 weeks, weighing 3.8 kg. Aortic root angiogram demonstrated multiple large coronary to right ventricle fistulae (Videos S1 and S2). Using a 4 Fr catheter in the femoral artery, selective coronary angiography revealed four fistulae from the dilated left anterior descending coronary artery, all draining into the anterior right ventricular outflow tract Figure 1. Three of these fistulae were closed using 0.038″ × 2 cm × 3 mm Gianturco coils (Cook Medical, Bloomington, Indiana) advanced through a 4F JB1 catheter (Terumo, Somerset, New Jersey) using a 0.035 Teflon-coated wire (Cook). The fourth fistula was smaller and less significant and therefore not occluded. In addition, the right coronary artery had three fistulae draining into the right ventricle,the largest of these was occluded using a similar coil. The Qp:Qs ratio decreased from 2.4:1 preprocedure to 1.6:1 post-procedure. Her left ventricular systolic function and cardiomegaly improved after the procedure. She was maintained on digoxin and furosemide at home and was growing well.

A follow up echocardiogram at the age of two years showed a mildly dilated left ventricle with normal systolic function. She was brought to the cardiac catheterization laboratory for further evaluation in view of left ventricular dilation. There was trivial residual flow across the coils in the three fistulae from the left anterior descending coronary artery into the right ventricle. The fourth fistulous communication from the LAD to the right ventricle (which had not been previously occluded due to its small size) was still small with trivial amount of flow. In the right coronary artery, the previously coiled fistula had no residual shunting, which the two other fistulae that had not been previously occluded were very tortuous with minimal shunting. The Qp:Qs was calculated to be 1.1:1. Therefore, no intervention was performed. She continued to grow well and remained asymptomatic. However, she continued to show evidence of progressive left ventricular dilation on the follow up echocardiograms. She was therefore brought to the catheterization laboratory again at the age of 5 ½ years. Her Qp:Qs was measured to be 1.2:1. The residual fistula from the left anterior descending coronary artery to the right ventricle remained small with minimal residual flow. However, at this time she was noted to have a modest amount of shunting through the two remaining fistulae from the right coronary artery to the right ventricle, both of which were occluded (using one 0.035″ × 5 cm × 5 mm and two 0.035″ × 4 cm × 4 mm MReye coils (Cook) in one fistula, one 0.035″ × 3 cm × 3 mm MReye coil in the other, delivered using a 4F JR catheter. At her most recent follow-up at the age of 12 years, she remains asymptomatic, her left ventricular dimension has normalized and she has normal left ventricular systolic function.

Case 2: A 21-week gestation male fetus underwent a fetal ultrasound that raised concern for a congenital cardiac malformation. There was no known maternal medical disease during or prior to this pregnancy, and the parents were consanguineous of Middle East descent. A fetal echocardiogram demonstrated a 3-4 mm communication between the left ventricle and the aorta (in the left sinus of Valsalva) suggestive of a coronary fistula or a possible aorta-left ventricular tunnel. The pregnancy was closely followed and the baby was delivered by caesarian section at 38-weeks gestation. Postnatal echocardiogram confirmed the antenatal suspicion of a communication between the left ventricle and the aorta, with septal thinning and dyskinesia noted at the level of the communication into the left ventricle. A CT scan demonstrated the right dominant coronary system. Left main coronary artery was aneurysmal and seen draining into the left ventricular cavity Figure 2 at the mid basal level, across the interventricular septum. The branches of the left coronary artery (left anterior descending and circumflex coronary arteries) were not well visualized. There was normal biventricular systolic function with mild left ventricular dilation and diffuse subendocardial hypoattenuation. A patent ductus arteriosus was present and rest of the intracardiac anatomy was normal.

He developed progressive ST-T segment abnormalities in the anterior and lateral leads on serial electrocardiograms (ECG). At first the patent ductus arteriosus was thought to be implicated in potentiating the coronary steal, but the ECG changes progressed even after spontaneous closure of the ductus arteriosus at three days of life. The cardiac enzyme (Troponin-I) was within the normal limits. Cardiac catheterization was performed at 7 days of age (weight of 3.9 kg) and angiography confirmed the findings of the large left coronary to left ventricle fistula measuring 4 mm in minimal diameter, 8 mm in maximal diameter and 16 mm in length

Figure 3, Videos S3 and S4). In addition, the left anterior descending and circumflex coronary arteries were noted to be atretic with no filling from the left main coronary artery. Instead, there was retrograde filling demonstrated in the left anterior descending and circumflex coronary arteries via collaterals from the right coronary artery (Videos S4 and S5). The surgical team was consulted about the feasibility of coronary artery bypass grafting. However, given the small patient size and the rich collateral network arising from the right coronary artery to the left coronary territory, it was decided that elimination of the large shunt (physiologically similar to severe aortic insufficiency) might be sufficient to correct the ischemic changes and prevent the development of LV dilation and dysfunction. Given high-flow lesion in a large caliber fistula with minimal stenosis to anchor a device, an occlusion device with a high degree of retention was deemed necessary. This was accomplished using an 8 mm Amplatzer Vascular Plug Type II (AVPII) (Abbott, Abbott Park, Illinois) delivered within a 4 Fr 45 cm long sheath (Cook) via the femoral artery. The plug was positioned in the distal fistula just before its entry into the LV, with the distal retention skirt positioned in such a way as to lay against the LV surface so as to avoid leaving any blind-ending pouch that could serve as a nidus for clot formation in the LV and subsequent thromboembolism. Repeat angiography after elimination of the rapid run-off showed no anterograde flow in the left anterior descending and circumflex coronary arteries confirming atresia of these vessels Figure 3. The aortic end of the fistula (anatomically, this represented the dilated left main coronary) was deemed to require additional closure with softer and more conformable devices to fill in the remaining space to avoid blood stagnation that could lead to systemic thromboembolism via the aorta. This was performed using two 6.5 × 12 mm Micro-Vascular Plugs (MVP) (Medtronic, Minneapolis, Minnesota) deployed in the proximal aneurysmal portion of the left main coronary artery using a 4 Fr JL2 catheter. After placement of all the devices, there was no residual flow in the fistula, no blind-ending pouch on either end of the fistula, no protrusion of the devices into the aorta, and no aortic regurgitation.

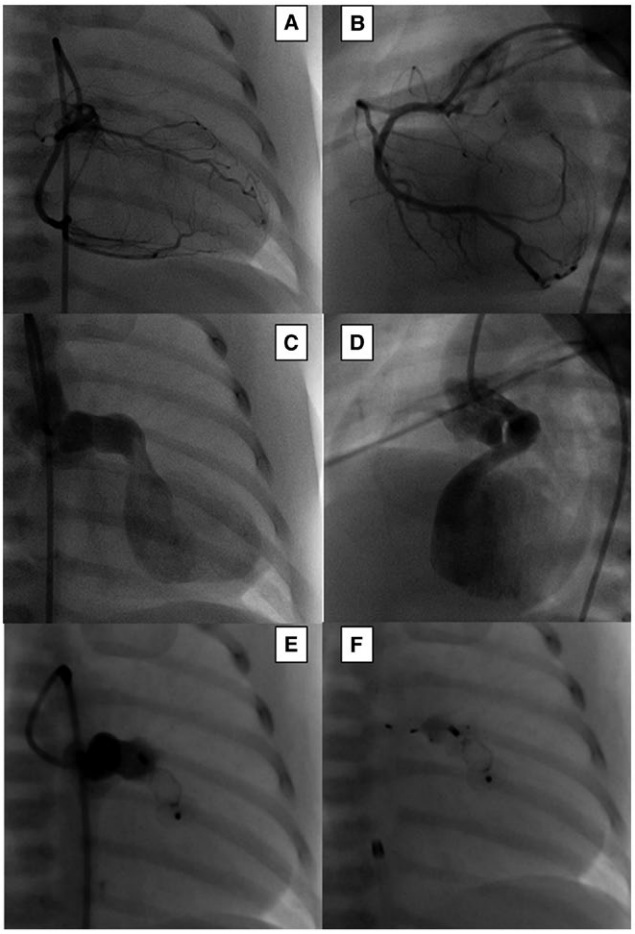


**FIGURE 3** Coronary angiography of Patient 2. (A, B) Selective right coronary angiography depicting the collaterals from the right coronary artery to the left coronary system; (C, D) Selective left main coronary artery angiogram demonstrating the left coronary artery fistula to the left ventricle; (E) Contrast injection into the proximal portion of the fistula after deployment of the 8 mm AVPII into the distal part of the fistula. There were no branches seen coming off the left main coronary artery, confirming atresia of the left anterior and circumflex coronaries; (F) After deployment of the 8 mm AVPII and two 6.5 × 12 mm MVPs into the fistula

The patient tolerated the procedure well and the ischemic changes resolved on repeat ECG on the evening after the procedure. He was started on propranolol (2 mg/kg/day) and aspirin 20.25 mg once daily. 24-hour Holter monitoring demonstrated normal sinus rhythm with frequent isolated monomorphic premature ventricular complexes which appeared to be fascicular in origin. He was stable and was discharged from the hospital one week after the procedure. He was doing very well at follow-up one month after the procedure, with no symptoms and no signs of ischemia on ECG. The echocardiogram showed no residual shunting across the fistula and normal left ventricular size and systolic function. The family returned to the Middle East, and by report was doing well with normal growth at a follow up at 7 months of age in their country. This patient was previously included in a series describing embolization using the MVP.[4]

## 4 | DISCUSSION

Coronary artery fistulae are anomalies of coronary termination as described by Angelini.[5] Most such fistulae drain into the right sided chambers or into the pulmonary artery.[6] Congenital left coronary artery-left ventricle fistulae are uncommon. Among a review of 200 patients with coronary artery fistulae, Oldham and colleagues described only two patients with left coronary artery fistulae emptying into left ventricle.[7] Xiao et al[3] have described one patient with a left anterior descending coronary artery fistula to the left ventricle in their series of eighteen patients over a span of 13 years. To our knowledge, a newborn with left coronary to left ventricle fistula and associated atresia of the branch coronary arteries (left anterior descending and circumflex coronaries) has not been described in English literature.

Most coronary fistulae are small and asymptomatic early in life, and are incidental findings seen on echocardiography or on angiography; however, the likelihood of symptoms increases with age due to concomitant development of atherosclerosis.[8] Fetal detection of coronary fistulae is uncommon,[9] as is neonatal presentation with heart failure or ischemia. Both our patients had a fetal diagnosis of coronary fistulae, and both were symptomatic in the first few weeks after birth. In fact, our first patient with biventricular systolic dysfunction was referred for transplant evaluation in view of the inoperable nature of the disease.

Treatment options for congenital coronary artery fistulae include surgical ligation, either isolated or in association with coronary artery bypass grafting, and interventional closure with occlusion coils, umbrellas, vascular plugs, or covered stents.[10] Catheterization and selective angiography is the optimal study for preinterventional imaging for accurate delineation of the anatomy. This was particularly useful in our second patient as the prior CT angiogram had not established that the left anterior descending and circumflex coronary arteries were actually atretic where they met the fistula itself. Nonetheless, angiographic assessment of the coronaries can be challenging when there is tremendous run-off into a fistula. Indeed, in our second case we implanted a device to occlude the fistula and repeated the angiography prior to releasing the device to prove there was no prograde flow in the left anterior descending and the circumflex coronary arteries. During coronary fistula embolization, great care is required to avoid occluding important coronary branches in the vicinity of the occlusion device, for fear of inducing a myocardial infarction in the affected territory supplied by those vessels. However, in this exceptional case of coronary fistula associated with atresia of the left anterior and circumflex coronaries, there were no at-risk vessels in the vicinity of the fistula.

The technique for embolization of coronary fistulae in children has generally involved creation of an arteriovenous loop.[3] This allows for placement of a 4 or 5 Fr catheter in the femoral artery, with delivery of the occlusion device transvenously through the bulkier delivery sheath. This approach minimizes the sheath size in the femoral artery to reduce the risk of arterial injury and avoids advancement of the occlusion device within a delivery sheath throughout the entire length of the coronary artery to the distal site of planned embolization. However, creation of an arteriovenous loop within a coronary artery in newborns may pose increased risk of coronary injury. Hence, in both cases presented herein, delivery of the occlusion devices was performed using a retrograde femoral arterial approach without use of an arteriovenous loop. In the first case, this involved placement of 0.038″ or 0.035″ coils through a 4F catheter that was advanced deep inside the branch coronary arteries arising from the left anterior descending and right coronary arteries. A flexible 4F catheter was selected, and this did not produce any coronary injury. At the time this procedure was performed (12 years ago) we did not carry detachable or microcoils, which would have been preferable for this application, as the Gianturco and MReye coils are not retrievable if malpositioned. In the second case, more modern occlusion devices were selected, including an AVPII and two MVPs, and both types of devices could be delivered retrogradely from the femoral artery using a 4 Fr sheath. Each of these devices played a different role, the AVPII acting as an initial anchor to close the distal site of the fistula, and the softer MVPs acting to fill the remaining space at the aortic end of the fistula.

## 5 | CONCLUSIONS

Coronary fistulae are rarely symptomatic in the newborn period. The presence of heart failure or ischemia in newborns with coronary fistulae should prompt a cardiac catheterization with a view to embolize the fistulae, despite the small patient size. We have shown that embolization of complex fistulae in newborns is safe and feasible using a variety of devices without requiring creation of an arteriovenous loop.

### CONFLICTS OF INTERESTS

The authors declare that they have no conflicts of interest with the contents of this article.

### AUTHOR CONTRIBUTIONS

Henri Justino devised the presented idea. Varun Aggarwal and Venkatachalam Mulukutla assisted Henri Justino in the procedure on one of the patients. Varun Aggarwal wrote the manuscript with input from all the authors. Venkatachalam Mulukutla, Athar M. Qureshi and Henri Justino contributed to the final version of the manuscript. All authors provided critical feedback and helped shape the research, analysis and manuscript.

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## SUPPORTING INFORMATION

Additional supporting information may be found online in the supporting information tab for this article.

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