Management of a Large Coronary Artery Fistula in a Neonate

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Central Message

Coronary artery fistulas are rare cardiac anomalies that can be treated by transcatheter or surgical methods. Here we present a case of surgical closure following an attempted transcatheter approach.

Glossary of Abbreviations

AV – atrioventricular
CAF – coronary artery fistula
CPB – cardiopulmonary bypass
CTA – computed tomography angiography
POD – post-operative day
RCA – right coronary artery
RV – right ventricle
TR – tricuspid regurgitation

Coronary artery fistulas (CAF) are exceedingly rare. They are defined as an abnormal communication between a coronary artery and another cardiac structure or major thoracic vessel. While largely presumed to be asymptomatic, a recent study showed that a majority of neonates/infants presented with heart failure-type symptoms, albeit many of them had a larger fistula size. Indications for intervention on CAFs have been controversial because many patients are asymptomatic, however delaying intervention is associated with a significantly higher risk of death pre-operatively and an increased risk of morbidity and mortality post-
Therefore the majority of CAFs are addressed when they are identified. Transcatheter and surgical approaches have been both been used with good success rates, however the number of neonates undergoing repair by either method remains low. Here we present a case of surgical repair of a large CAF in a neonate after an attempted transcatheter closure resulted in iatrogenic tricuspid regurgitation (TR). The Institutional Review Board (IRB) of Children’s Healthcare of Atlanta approved the study protocol and publication of data (approval number: STUDY00001462, date approved: 07/18/2022). Patient written consent for the publication of the study data was waived by the IRB as it was determined to be a research not involving human subjects.

Clinical Summary

A 2.6 kg female born at 39 weeks was found to have a murmur. Her hemodynamics at that time were acceptable without any cardiovascular or respiratory support. She was then transferred to our facility for further management.

Echocardiography revealed diffuse dilation of the right coronary artery (RCA), a large fistulous connection between the RCA and the right ventricle (RV) outflow tract, three small left to right atrial shunts, normal left ventricular size and function, a mildly dilated RV with normal function, and holodiastolic flow reversal in the aortic arch (video 1). Computed tomography angiography (CTA) detailed the fistulous connection between the RCA and RV infundibulum, measuring 0.7 x 0.58 cm (figure 1A,B). Cardiac catheterization further illustrated the fistula, measuring 0.7 x
1.09 cm, as well as a normal size and course for the RCA distal to the fistula (figure 1C, video 2).

Due to persistently low diastolic blood pressure, on day of life 4, we attempted transcatheter closure, first with a microvascular plug-7Q device (MVP-7Q, Medtronic) which left significant residual flow through the fistula. Coil embolization was attempted, but high flow through the fistula precluded accurate coil position. We then tried a 12 mm Amplatzer vascular plug II (AVP-11, Abbott) which unfortunately embolized into the RV (figure 1D, video 3). The device was moved to the right atrium for retrieval, resulting in severe tricuspid regurgitation (TR) noted on post-procedure echocardiography.

On day of life 5, she was taken to the operating room for surgical repair (video 4). The fistula was too large to suture ligate externally. The entrance to the RVOT was visible through the tricuspid valve, but not amenable to closure. Therefore, we divided the epicardium overlying the fistulous tract which revealed the coronary and ventricular openings to the fistula. These were closed with polytetrafluoroethylene patches.

The tricuspid valve was then repaired by resuspending the anterior leaflet, reimplanting a major chord to the septal wall, and performing a partial commissuroplasty. The atrial septal defect was closed primarily. Post-repair echocardiography confirmed no residual fistulous connection, mild to moderate TR, and a tiny residual atrial level shunt.
Her post-operative course was uneventful. She was extubated and weaned off inotropes and vasoactive support by post-operative day (POD) 4. Follow-up echocardiography showed two tiny additional RCA fistulas to the RV and moderate TR. She tolerated gastric feeds well and was transitioned to oral afterload reduction. Intravenous therapeutic heparin was initiated on POD 0. She was transitioned to therapeutic lovenox on POD 6 and then discharged home on POD 11 on aspirin and lovenox.

At six months post-op, she continues to develop normally with adequate weight gain. Follow-up echocardiogram shows no residual coronary artery fistulas, a small PFO, and trace to mild TR. Repeat CTA shows no residual fistulas and mild lobular irregularity of the proximal-mid RCA (figure 2). She continues on aspirin and lovenox with eventual conversion to aspirin only in the coming months.

**Discussion**

Coronary artery fistulas are rare, and their repair in neonates is even less common. Our patient appeared to be asymptomatic initially, but given the degree of diastolic runoff through the fistula we presumed she would develop signs/symptoms of heart failure, and therefore opted to intervene in the neonatal period. Transcatheter techniques can be used to address CAFs, however they are not without risk. In hindsight, perhaps the large fistula size should have pushed us towards surgical intervention as first line treatment. In the future we will likely consider fistula diameter when deciding between approaches. The repair, however, was durable and she has recovered well with improvement in her TR. She will likely remain on lifelong antiplatelet therapy to mitigate the long-term risk of coronary thrombosis.
Figures/Videos

Central Picture. Amplatzer device deployment within an RCA fistula with residual flow through the fistula.

Figure 1. Coronal projection showing the fistula entering the RV outflow tract (blue arrow) (A). Sagittal projection showing the origin of the RCA as well as the takeoff of the fistula (blue arrow) (B). Anterior-posterior projection of coronary angiography showing a normal appearing right coronary artery distal (yellow arrow) as well as the RCA fistula (red arrow). Anterior-posterior projection showing a 12 mm Amplatzer device fully deployed with unobstructed flow to the distal RCA and residual flow around the device (D).

Figure 2. Sagittal projection six months post-repair showing no residual fistulous connection (blue arrow) (A). Three-dimensional reconstruction of coronary arteries showing lobulated proximal-mid RCA (yellow arrow), normal distal RCA, and no residual fistulous connection (B).

Video 1. Long-axis left ventricle outflow tract view showing a dilated proximal RCA and diastolic flow reversal in the aorta.

Video 2. Anterior-posterior projection demonstrating a large RCA fistula draining into the RV outflow tract, with a normal caliber RCA distal to the fistula.
Video 3. Anterior-posterior projection after deployment of the 12 mm Amplatzer device demonstrating unobstructed flow to the distal RCA and residual flow around the device.


References


