Management of a large coronary artery fistula in a neonate

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Coronary artery fistulas (CAFs) 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 greater risk of death preoperatively and an increased risk of morbidity and mortality postoperatively. Therefore, the majority of CAFs are addressed when they are identified. Transcatheter and surgical approaches have been both used with good success rates; however, the number of neonates undergoing repair by either method remains low. Here, we present a case of surgical closure following an attempted transcatheter approach.

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.

CLINICAL SUMMARY
A 2.6-kg female patient 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, 3 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 detailed the fistulous connection between the RCA and RV infundibulum, measuring 0.7 × 0.58 cm (Figure 1, A and B). Cardiac catheterization further illustrated the fistula, measuring 0.7 × 1.09 cm, as well as a normal size and course for the RCA distal to the fistula (Figure 1, C, 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 1, Video 3). The device was moved to the right atrium for retrieval, resulting in severe TR noted on postprocedure echocardiography.

On day of life 5, the patient was taken to the operating room for surgical repair (Video 4). The fistula was too large to suture ligate externally. The entrance to the right ventricular outflow tract 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. Postrepair echocardiography confirmed no residual fistulous connection, mild-to-moderate TR, and a tiny residual atrial level shunt.

Her postoperative course was uneventful. She was extubated and weaned off inotropes and vasoactive support by postoperative day (POD) 4. Follow-up echocardiography showed 2 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 (enoxaparin) on POD 6 and then discharged home on POD 11 on aspirin and Lovenox.

At 6 months’ postoperatively, she continued to develop normally, with adequate weight gain. Follow-up echocardiography showed no residual CAFs, a small PFO, and trace-to-mild TR. Repeat computed tomography angiography showed no residual fistulas and mild lobular irregularity of the proximal-mid RCA (Figure 2). She was continued on aspirin and Lovenox, with eventual conversion to aspirin only in the coming months.

DISCUSSION

CAFs 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 we 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 toward 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.

References

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