Hybrid approach to neonatal repair of large symptomatic congenital coronary artery fistula

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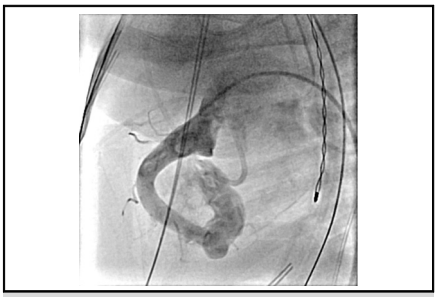


Postnatal catheterization showing RCA-RV fistula.

CENTRAL MESSAGE

Neonatal repair of large symptomatic coronary artery fistula is aided by guidewire placement to define the fistula and intraoperative angiography to confirm repair.

See Commentaries on pages 298 and 299.

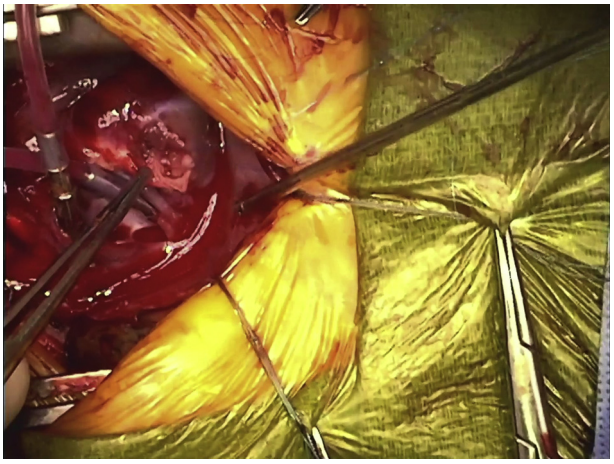
Video clip is available online.

Coronary artery fistula are rare lesions that account for 0.2% to 0.4% of all congenital heart disease.[1] The most common coronary fistula presentation is a right coronary artery (RCA) to right atrium/ventricle (40%) in an asymptomatic adult.[2] Rarely, neonates present with large fistula in extremis.[3] In these situations, a multidisciplinary approach between cardiology, interventional cardiology, and cardiothoracic surgery is optimal to facilitate early successful surgical repair.[4] We present a neonate with a significant RCA to right ventricle (RV) fistula who had an excellent outcome using this hybrid approach (Video 1).

CASE PRESENTATION

A 37-week, 2.65-kg female patient was born with a prenatal diagnosis of large RCA-RV fistula. After emergent cesarean delivery, the baby was intubated at 1 minute of life for respiratory distress and bradycardia. She was taken to the cardiac catheterization lab on escalating inotropes and catheterization showed a right dominant heart with a small posterior descending artery and collaterals from the normal left anterior descending. A large RCA connected to the posterior atrioventricular AV groove and entered underneath the tricuspid valve into the posterior/basal aspect of the RV. Hemodynamic measurements calculated a cardiac index of 8.0, pulmonary vascular resistance of 3.4 iWU, and a pulmonary blood flow to systemic blood flow of 1.2. Right ventricular pressure was near systemic at 53 mm Hg compared with the systemic pressure of 57 mm Hg. There was holodiastolic flow reversal in the aortic arch and ST changes on electrocardiogram. The patient was fully heparinized, and a 0.014" whisper guidewire was left in the right femoral artery through the fistula to help guide operative repair.

Informed consent was obtained from the family and the patient was transferred to the operating room. Transthoracic echocardiography showed a proximal RCA of 0.55 cm, z score = 15.67, and distal RCA 0.35 cm. In the operating room, the fistula was temporarily controlled with a surgical clip on cardiopulmonary bypass and antegrade cardioplegia was delivered from the aortic root. An arteriotomy was made from the epicardial surface of the RCA where the fistula entered the RV (Figure 1). The guidewire was identified and a right ventriculotomy was then performed to ensure the tricuspid valve chordal apparatus would not be disturbed while closing the fistula. The guidewire was palpated and visualized within the heart. The fistula was closed with a running 8-0 PROLENE suture (Ethicon, Somerville, NJ) from within the RCA and



VIDEO 1. Postnatal echocardiogram, aortogram, operative video, and on-table completion angiogram of novel hybrid management of neonatal congenital coronary artery fistula. Video available at: https://www.jtcvs.org/article/S2666-2507(20)30347-3/fulltext.

several additional figure-of-eight stitches were placed from the ventricular side. A right atriotomy was performed to inspect the tricuspid valve and it was floated to evaluate its competence. The right ventriculotomy and right atriotomy were closed in a double running fashion. On-table angiogram revealed a residual fistula and a 6-0 running ligation stitch was placed in the epicardium between the RCA and RV. Re-angiogram confirmed no residual fistulous tract and demonstrated good flow in the coronaries. Epicardial echocardiogram demonstrated mildly depressed function with no tricuspid regurgitation. Crossclamp time was 81 minutes, and total cardiopulmonary bypass time was 103 minutes. The patient did well postoperatively, was extubated on postoperative day (POD) #3, transferred to the floor on POD #6, and discharged home on POD #17. Cardiac computed tomography performed at 5 months of age showed normal caliber coronaries (proximal RCA 0.17 cm, z = 0.99, and the distal RCA 0.12 cm) (Figure 2), and echocardiogram showed good biventricular function. The patient continues to be asymptomatic and progress normally at 18 months of life (Video 1).

DISCUSSION

This case demonstrates a rare presentation of coronary artery fistula with a good result. More importantly, it describes a successful algorithm for dealing with an infrequent lesion where expediency can minimize potential cardiovascular collapse and/or infarct. Prenatal diagnosis of large coronary artery fistula is extremely valuable in preparing for the birth of an unstable newborn.[5] Patients with suspected large fistulae should be transferred to tertiary care hospitals with readily available pediatric interventional cardiology and cardiac surgery. Coordination between the primary cardiologist, interventional cardiology, and cardiothoracic surgery where all are present for the echocardiogram and catheterization greatly reduces confusion regarding decision-making. Leaving a wire through the fistula reduces crossclamp and bypass time by removing the trial-and-error aspect from surgical ligation. On-table angiography ensures that no residual fistula remains and the coronary arteries have not been compromised. Intraoperative echocardiography assures good ventricular function. These key steps can help prepare all involved for this rare lesion and lead to early, safe, and successful neonatal repair. To the authors knowledge this is the first hybrid approach to repair of coronary artery fistula reported in a neonate.

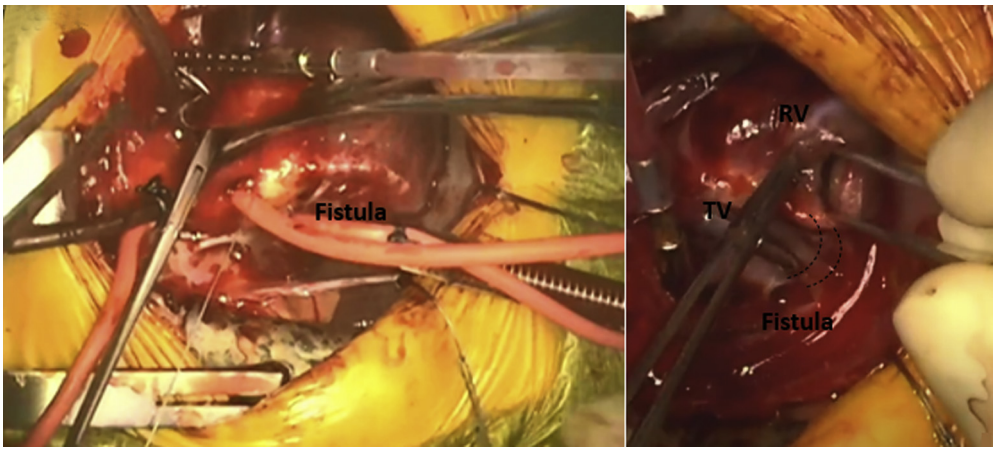


FIGURE 1. Surgeon's view of the large RCA to RV fistula. On the *right* image, the fistula has been opened and the tricuspid valve is in view. The fistula entered the RV underneath the tricuspid valve in between the chordae. It was unable to be ligated through the valve, so a right ventriculotomy (forceps) was performed to avoid causing tricuspid regurgitation. The *dotted line* marks the approximate course of the fistula. *RV*, Right ventricle; *TV*, tricuspid valve.

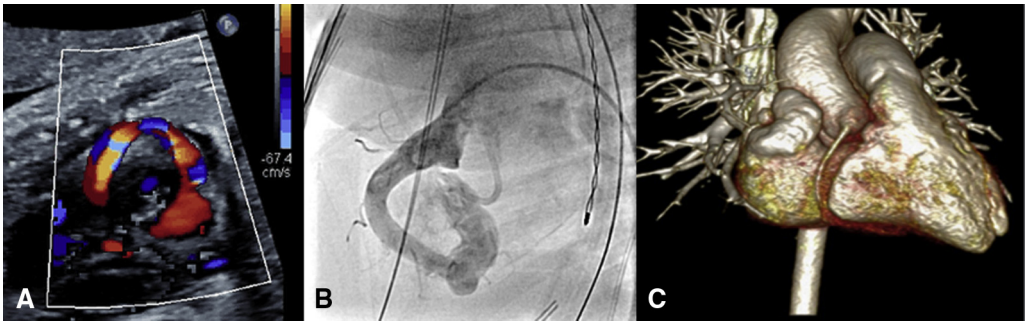


FIGURE 2. A, Fetal echocardiogram demonstrating large RCA fistula wrapping around the RV. B, Postnatal aortogram showing large hemodynamically significant RCA-RV fistula. The neonate was in extremis and the fistula was repaired via a novel hybrid approach. C, Computed tomography scan showing normal-caliber RCA at 5 months' postoperation.

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