

Symptomatic newborn coronary fistula repair without cardiopulmonary bypass



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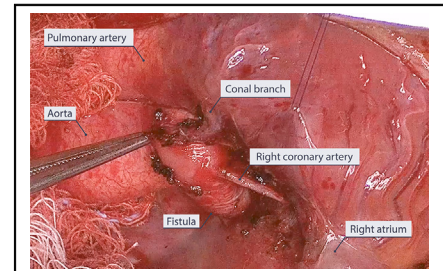
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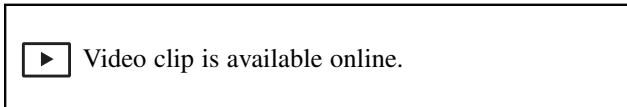
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A coronary fistula is shown, as well as the surrounding cardiac structures.

CENTRAL MESSAGE

In selected cases of coronary artery fistula, repair can be safely performed without the deployment of cardiopulmonary bypass, provided that myocardial ischemia is precluded.



Congenital coronary artery fistula (CAF) is a rare connection between a coronary artery and a cardiac chamber. Single or multiple fistulae can be present at once and may originate from any coronary artery. The most common CAF is between the right coronary artery (RCA) and right heart chambers.¹ The blood flow from a coronary artery into a low-pressure cardiac chamber through a fistula produces a coronary steal and left ventricular volume overload, both of which can lead to myocardial insult. In addition, the amplified flow due to pressure gradient can induce aneurysm formation and coronary intimal injury. Interventional catheterization with the aim of closure is used frequently; however, certain cases necessitate surgical intervention with or preferably without cardiopulmonary bypass (CPB).

METHODS

Institutional review board approval was waived. Our patient was diagnosed antenatally with an aortopulmonary fistula. Considering the high intrauterine stress, an urgent cesarean delivery was performed, with cardiac surgery team on standby in the delivery room for possible surgical intervention. The baby was born in good condition and remained hemodynamically stable for the first week of life in the critical care unit. An elective diagnostic catheterization confirmed the presence of a large tubular segment originating from the right coronary sinus and giving rise to both the RCA and a conal/marginal branch that continued to the right atrium. On the eighth day, the patient underwent surgical repair. Following midline

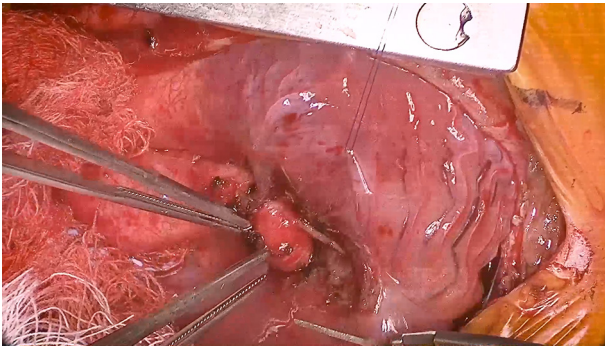
sternotomy and partial removal of the thymus, the fistula and the 2 branches were carefully dissected free on the beating heart. A small bulldog clamp was placed in the immediate vicinity of the RCA takeoff and the other distally on the fistula. The fistula was divided, and both ends of the fistula were closed using continuous monofilament sutures. Preceding the removal of the clamps, a longitudinal suture line was placed on the long axis of the main tubular segment to facilitate a size reduction (Video 1).

RESULTS

The postoperative course was unremarkable, with 2 days of cardiac intensive care and 4 days of hospital stay. The postoperative echocardiography confirmed the absence of residual fistular flow and normal systolic and diastolic ventricular function. The downsizing of the ectatic RCA segment caused no significant electrocardiogram or cardiac enzyme alterations. The patient has been discharged home and doing well at the last 4-month follow-up.

DISCUSSION

Fetal or newborn coronary fistulae are exceedingly rare and usually don’t require early intervention. The most common CAF is between a coronary artery and a right heart chamber, as in this case. Both in the fetus and early neonatal period right ventricle (RV) pressure is high; therefore, left-to-right flow through the fistula is hindered and the



VIDEO 1. Symptomatic newborn coronary fistula repair without cardiopulmonary bypass. Video available at: [https://www.jtcvs.org/article/S2666-2507\(22\)00582-X/fulltext](https://www.jtcvs.org/article/S2666-2507(22)00582-X/fulltext).

normal coronary blood flow is sustained. After birth, the pulmonary vascular resistance, as well as the RV pressure, decreases. Together with the increase in coronary steal phenomenon and the volume overload of the left ventricle, those patients with CAF may become symptomatic. Therefore, close monitoring of these patients, especially in the first few days of life, is warranted. Coronary angiography is a helpful tool in hemodynamically stable patients (1) for the confirmation of the diagnosis; (2) for the planning of potential intervention; and (3) for the visualization of the collaterals. The presence of a sizeable RCA distal to the fistula suggested adequate collateral perfusion from the left coronary system, and this was also confirmed angiographically. Repair of the coronary fistula and downsiz-

ing of the proximal ectatic RCA to prevent potential thrombus formation was possible without the use of CPB and myocardial ischemia. If the anatomy of the fistula was not deemed to be operated without CPB and the presence of the collaterals was ambiguous, the use of CPB is a viable alternative. In this case, before the administration of cardioplegia, both the vena cavae and the pulmonary arteries should be controlled with snares to prevent myocardial ischemia.

CONCLUSIONS

Asymptomatic coronary-to-right heart CAF can become symptomatic subsequent to the drop in pulmonary vascular resistance and RV pressure in the neonatal period. Therefore, these patients should be monitored closely, especially in the first week of life. The repair can be safely performed and should be preferred^{2,3} without the deployment of CPB, provided that the myocardial ischemia is precluded.

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