Revascularization of coronary circulation in pulmonary atresia with intact ventricular septum and right ventricular-dependent coronary circulation

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Disclosures: S.A. is the site Principal Investigator for a multicenter study led by the University of Michigan but reports no salary support. All other authors reported no conflicts of interest.

The Journal policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

Read at the 103rd Annual Meeting of The American Association for Thoracic Surgery, Los Angeles, California, May 6-9, 2023.

Received for publication Feb 28, 2023; revisions received April 6, 2023; accepted for publication April 8, 2023.

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J Thorac Cardiovasc Surg 2023;■:e1-5
0022-5223/$36.00
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https://doi.org/10.1016/j.jtcvs.2023.04.007

CENTRAL MESSAGE

Coronary revascularization in pulmonary atresia-intact ventricular septum and right ventricular-dependent coronaries can stabilize coronary flow while awaiting transplant or the next surgical stage.

See Commentary on page XXX.

CASE PRESENTATION AND METHODS

This full-term, 4.4 kg, 3-week-old neonate with PA-IVS and RVDCC was maintained on prostaglandin infusion following birth. With decrease in her pulmonary vascular resistance in her first couple of weeks of life, she had significant myocardial ischemia with ST segment changes and elevated troponin levels requiring intubation for stabilization. Heart transplantation was elected as a definitive pathway. To stabilize the precarious coronary circulation, a graft from the aorta to TV was planned in addition to a mBTT shunt. After median sternotomy, a 3.5-mm polytetrafluoroethylene right mBTT shunt was placed between the innominate artery and right pulmonary artery off pump. A 5-mm saphenous vein homograft progression to the next stage of single-ventricle palliation and precluding the need for cardiac transplantation. The institutional review board or equivalent ethics committee of the Cleveland Clinic did not approve this study because it is a Video Case Report. The subject’s parent provided written informed consent for the publication of the study data.

Pulmonary atresia with intact ventricular septum (PA-IVS) and right ventricle-dependent coronary circulation (RVDCC) is a challenging condition that classically has mandated single-ventricle palliation without RV decompression. Some patients with evidence of coronary ischemia, ventricular dysfunction, or coronary ostial atresia require primary listing for transplantation to allow for survival out of early infancy. However, even with a transplant listing, the coronary circulation in this anatomy is very precarious and puts these patients at high risk of coronary compromise while awaiting transplantation. This is evidenced by a mortality rate of 18% in patients with PA-IVS and RVDCC within 3 months of systemic-to-pulmonary artery shunting.1 Over time, surgeons have attempted to supply the RV-dependent coronaries in ways that do not rely on blood flow via the tricuspid valve (TV) and RV cavity itself. Apart from 1 reported case performed in a desperate situation, these strategies have all been employed to stabilize the coronary circulation until the time of cardiac transplantation. We demonstrate a strategy to revascularize the RVDCC in a patient with known coronary ischemia via an aortic-to-TV graft at the time of modified Blalock-Taussig-Thomas (mBTT) shunt creation that avoids a ventriculotomy and aims to allow for successful

Video clip is available online.
was sewn to the side of the ascending aorta. The distal arch and inferior vena cava were cannulated, cardiopulmonary bypass was initiated, the ductus ligated, and the heart immediately arrested by antegrade cardioplegia to avoid myocardial ischemia given the coronary fistulas. Atrial septectomy was performed upon opening the right atrium (RA), and the saphenous vein homograft was routed into the RA through the RA wall and anastomosed directly to the 5-mm TV annulus. The patient’s TV did not have any degree of tricuspid regurgitation preoperatively, so the suture line to the annulus was sewn in a manner to intentionally make the TV incompetent and decompress the RV (Figure 1). After successful weaning from cardiopulmonary bypass, the inferior vena cava cannula was exchanged for a Carmeda-coated cannula (Carmeda AB), and left ventricular assist device support was initiated with PediMag (Abbott Laboratories) at low flow of 70 mL/kg (300 mL) per minute. The assist device was inserted to ensure stability for this innovative procedure until the next stage. The patient was successfully weaned off ventricular assist device support on postoperative day 3. She remained actively listed for cardiac transplantation as status 1Ae during her recovery.

Over the next several weeks, the patient had evidence of re-emergence of myocardial ischemia. On angiography, the aorta-to-TV graft exhibited stenosis and needed stenting at the transit site into the RA through the RA wall (Figure 2). The infant subsequently needed stenting of the aorta-to-TV graft 1 additional time along with stenting of the mBTT shunt once. During the ensuing months, she grew well and had nice clinical stability, but slowly her oxygen saturation began to decline into the low 70s, and she required increasing amounts of supplemental oxygen. As such, at age 6 months, it was determined that the patient should undergo the second stage of single-ventricle palliation with a bidirectional cavopulmonary shunt, and, at the same operative setting, have upsizing of her aorta-to-TV graft. Aortobicaval cannulation was performed, and CPB was established. The mBTT shunt was ligated. The heart was arrested in antegrade fashion. The former aorta-to-TV graft was removed, and the TV leaflets were excised completely. Inspection the coronary ostia within the aorta revealed the absence of a right coronary ostium. A 6-mm ringed polytetrafluoroethylene graft was chosen and sewn to the TV annulus that was now free of any residual TV leaflet tissue, thus creating a large, unobstructed end-to-end anastomosis. The graft was brought through the RA wall and then sewn to the aorta in end-to-side fashion. The entrance of the graft into the RA wall was secured with a purse-string suture. The mBTT shunt was excised, and a bidirectional cavopulmonary shunt was performed (Figure 3). After reperfusion, the heart regained normal sinus rhythm spontaneously and was successfully weaned off of cardiopulmonary bypass support without the need for any mechanical circulatory support. The patient was extubated on postoperative day 1. She was subsequently weaned to room air with stable saturations and transitioned to the floor where she progressed, fed, and grew well. After multidisciplinary team discussions, she was subsequently removed from the heart transplant waiting list at age 9 months (Video 1).
DISCUSSION

Most patients with PA-IVS and RVDCC can successfully undergo staged single-ventricle palliation without RV decompression with 15-year survival rates exceeding 80%, rates that exceed the survival of heart transplantation in this patient population. The type and location of coronary artery stenoses or atresia present in PA-IVS and RVDCC are important to appreciate because the more proximal the stenoses, the worse the risk of mortality, with bilateral coronary ostial atresia at the aorta (0.3% of all PA-IVS patients) being the highest-risk variant. As such, a subset of these patients will exhibit evidence of coronary ischemia or develop ventricular dysfunction over time. Along with those patients with coronary ostial atresia, these patient subsets often cannot survive long enough to undergo any palliation beyond systemic-to-pulmonary artery shunting or patent ductus arteriosus stenting. These specific patients have long been directed through the heart transplantation pathway for definitive management. Despite entering this pathway, mortality while awaiting transplantation is not low as the risk of ongoing coronary ischemia is not mitigated in any way while waiting. This has led to interest in establishing alternate sources of blood flow to the RV-dependent coronary arteries in this anatomy.

Beginning with Freeman and colleagues in 1993 and Laks and colleagues in 1995, multiple groups have created aorta-to-RV conduits at various points in the single-ventricle palliation pathway to provide oxygenated, antegrade blood flow to the myocardium via the RVDCC. In 2018, Sakurai and colleagues advanced this concept and applied it to a patient with bilateral coronary ostial atresia with the goal of completely avoiding the need for cardiac transplantation. This patient had placement of a 3-mm polytetrafluoroethylene aorta-to-RV shunt at the time of mBTT shunt creation and then underwent successful bidirectional cavopulmonary shunt at age 4 months. The aorta-to-RV shunt was subsequently upsized to 5 mm in size, and the patient was discharged home and did not require any additional interventions by age 9 months. Most recently, Said and colleagues further modified this concept in the setting of a patient with PA-IVS and RVDCC with bilateral coronary ostial atresia by taking the risk of tricuspid regurgitation out of the equation by sewing the aorta-to-TV shunt directly to the orifice of the TV via the RA. That particular patient required a period of temporary extracorporeal mechanical support 1 month postoperatively and remained on the transplant list and was successfully transplanted 2 months after her initial double-shunt operation.
From a technical perspective, the graft used for the initial revascularization in our case was a saphenous vein homograft. The transition of this graft into the RA cavity proved to be vulnerable and required stenting twice. We believe a ring-reinforced polytetrafluoroethylene graft should be used from the outset of the first operation to ensure enough stiffness to transit the atrial wall and maintain graft patency.

This particular patient manifested with significant clinical signs of myocardial ischemia shortly after birth in the setting of a nondecompressed RV. Despite the RV being adequately pressurized, this ischemia persisted. Given her ongoing ischemia in the setting of RVDCC and a fully pressurized RV, the benefit of this operation was that it delivered more highly oxygenated blood to the RV and the RVDCC directly from the aorta at aortic pressure. As such, this graft

FIGURE 3. Stage 2 palliation with bidirectional cavopulmonary shunt and upsizing of aorta-to-tricuspid valve graft.

provides a more optimal source of coronary blood flow in patients with the most significant forms of RV dependence, including those with coronary ostial atresia, and also should be considered in patients showing clinical signs of coronary ischemia despite a fully pressurized RV.

This particular patient did not have any degree of tricuspid insufficiency before her initial procedure. At the time of her first operation when the aorta-to-TV graft was sewn in place, the TV leaflets were intentionally made incompetent to decompress the RV and allow for unimpeded blood flow into the cavity of the RV. Given that the orifice of the TV was essentially very diminutive, any incompetence would not have been a large clinical concern. Overall, when the pulmonary vascular resistance drops, the diastolic pressure becomes lower, which may compromise the existing coronary perfusion. In addition, any decrease in cardiac output will result in lower mixed venous blood feeding the coronaries, which will compound the ischemia and lead to possible arrest.

The patient in our present report remained listed for transplantation in the initial postoperative period after bidirectional cavopulmonary shunt given the uncertain durability of the Fontan pathway compared with transplant with this anatomy. However, this approach has clearly demonstrated to us that it is not only durable but also highly effective in achieving remarkable clinical progress in this patient. She has normal left ventricle function, no evidence of any ongoing coronary ischemia, and is growing robustly and meeting all appropriate milestones. As such, she has the clinical stability and progress of an outpatient who has successfully recovered from a major operation and is now proceeding forward with normal daily life. In the absence of any new clinical changes in follow-up as would be evaluated for any postoperative cardiac surgery patient, we believe that this patient should not remain on the transplant list and should instead progress forward with the intention of ultimate Fontan completion as the definitive palliative management. Yet, once patients with PA-IVS with RVDCC successfully achieve bidirectional cavopulmonary shunt circulation, we must acknowledge their risk-to-benefit ratio of cardiac transplantation changes. This now provides a window where this same risk-benefit ratio may also substantially change for patients with RVDCC and myocardial ischemia that could potentially benefit from the aorta-to-TV graft approach.

It is important to recognize that the work of Sakurai and colleagues demonstrates that successful single-ventricle palliation can be pursued even in the most extreme variant of this disease with bilateral coronary ostial atresia at the aortic level and can even allow for an outpatient status in these patients. We believe that the reports of Sakurai and colleagues, Said and colleagues, and our present group provide good evidence to suggest that a different paradigm of care, what we call coronary revascularization, can potentially be applied to patients presenting with this complex anatomy. Not only can this proposed new approach stabilize the coronary circulation during transplant waitlist time, but also it has the potential to be a definitive RVDCC revascularization that may allow even the most complex and at risk of these patients with PA-IVS to achieve successful staged single-ventricle palliation and obviate the need for transplantation in early childhood altogether.

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