HISTORY OF PRESENTATION AND PAST MEDICAL HISTORY

A 31-year-old woman was admitted for chest pressure, episodes of presyncope, lower extremity edema, and dyspnea on exertion. She had a history of severe bicuspid aortic stenosis requiring the Ross procedure at age 16, including a 24-mm right-ventricle-to-pulmonary-artery (RV-PA) homograft (Figure 1 A); however, this was complicated by left coronary artery (LCA) endarteritis and pseudoaneurysm, requiring a second surgical repair. The present work-up revealed severe RV-PA homograft stenosis (Figure 1 B, Video 1).

She was taken to the catheterization lab for transcatheter pulmonary valve (TPV) implantation. Serial balloon dilation of the RV-PA homograft was performed (18 mm x 2 cm Vida™, 20 mm, and 22 mm x 4 cm Atlas® Gold balloons, Bard Peripheral Vascular, Inc.) with excellent hemodynamic recovery after balloon deflation. However, on final balloon inflation (24 mm x 4 cm Atlas® Gold), she had no return of arterial pressure, and rhythm degenerated into ventricular fibrillation (VF).

DIFFERENTIAL DIAGNOSIS

(1) RV-PA homograft rupture with free bleeding into mediastinum
(2) Acute coronary occlusion due to external compression from fractured RV-PA homograft fragment
(3) Acute RV failure due to serial occlusion of RV outflow
(4) Distal pulmonary arterial perforation with uncontrolled hemorrhage

INVESTIGATIONS AND MANAGEMENT

Following defibrillation, the patient degenerated back into VF, requiring multiple rounds of defibrillation and cardio-pulmonary resuscitation (CPR). During spontaneous circulation, aortic root and pulmonary angiography (Figure 2, Video 2) revealed no free bleeding from the RV-PA homograft, but it did show contrast extravasation from the neoorta via the LCA into the RV-PA homograft. The patient was cannulated for extracorporeal membrane oxygenation (ECMO) and transferred to the operating room (OR). There she was placed on cardiopulmonary bypass and her heart arrested, at which time pulmonary arteriotomy revealed a longitudinal split posteriorly in the wall of the homograft. Behind this, several pledgets were visible from the prior LCA pseudoaneurysm repair. The LCA itself had been completely disrupted as a result of the homograft split.
The LCA was ligated and a bypass performed via saphenous vein graft to the left anterior descending artery. The posterior wall of the RV-PA homograft was reconstructed using a bovine pericardial patch, and a 23-mm bovine pericardial valve was implanted (Perimount Magna Ease, Edwards Lifesciences Corp.). A pericardial patch was sewn onto the anterior aspect of the homograft conduit.

Initially, the patient could not be weaned from bypass and returned to the intensive care unit on ECMO, and she continued to require ECMO support on postoperative day (POD) 1. She was taken to the hybrid OR on POD 2 for decannulation, but echocardiography revealed severe prosthetic pulmonary valve stenosis with a velocity of 5 m/s (Figure 3). Through a purse-string suture, intraoperative angiogram via direct RV puncture revealed a restricted orifice in the valve’s anterior (Figure 4 A) with a filling defect or fixed leaflet.

A Palmaz P4010XL stent (Cordis) was deployed on a 22-mm x 4-cm Balloon in Balloon catheter (NuMED) within the bioprosthetic valve, completely expanding the orifice. A 22-mm Melody valve (Medtronic) was implanted with no evidence of regurgitation on final evaluation (Figure 4 B-C, Video 3).

Figure 2.
Balloon dilation of right-ventricle-to-pulmonary-artery (RV-PA) homograft. (A) Lateral view of left ventricular outflow tract (LVOT) angiogram during 24-mm balloon inflation. The left coronary artery (LCA) was not well seen (yellow arrow). (B) Neoaortic (Ao) root angiogram post-balloon deflation demonstrates contrast extravasation from the left coronary ostium to the pulmonary artery (PA). (C) Frontal view of LVOT angiogram in retrospect demonstrates compression of the neoaorta by the inflated balloon rather than poor contrast filling due to obstructed cardiac output. RCA: right coronary artery; LV: left ventricle

Figure 3.
Prosthetic pulmonary valve stenosis following surgical replacement. On extracorporeal membrane oxygenation, the newly-placed surgical bioprosthetic pulmonary valve exhibited high velocity (5 m/s) on transesophageal echocardiogram: (A) two-dimensional, (B) color, and (C) continuous-wave Doppler.
The patient was decannulated from ECMO on POD 3 and was extubated and ambulating on POD 4. Postoperative imaging showed normal RV size and function with normal velocity and pressure gradient across the pulmonary valve (Figure 5) and significant improvement of regurgitation (Video 4). She was discharged in excellent condition and was clinically well at 6-month follow-up.

**DISCUSSION**

RV-PA homograft degeneration can be seen in up to 50% to 55% of patients within 10 years of initial surgical repair and 5 to 6 years after the second. TPV can be an alternative to reduce the open surgeries these patients require but is in the highest risk category of congenital heart interventions. Although a key component of TPV, balloon dilation of the RV-PA homograft can cause complications such as conduit tear/rupture and coronary arterial compression. Conduit rupture has been reported during balloon dilation in approximately 9% of cases; although balloon expandable covered stents can be used to mitigate these events, in some cases such as ours, surgical rescue may still be required. Risk factors include unusual angles in
the right ventricular outflow tract (RVOT), use of bicuspidized homografts, heavy calcification, conduit type and size at implant, stenotic diameter, and balloon size.3,4,6

Coronary arterial compression can complicate up to 5% of RV-PA balloon dilations.6 Simultaneous aortic root or coronary angiogram with balloon inflation in the RV-PA conduit before stent/TPV deployment allows intraprocedural assessment for coronary compression.6 Generally, coronary compression during balloon dilation of the RV-PA conduit is relieved after balloon deflation if the stent/TPV is not implanted.

Importantly, VF is unusual in RV-PA homograft rupture, and to our knowledge this is the first report of RV-PA homograft rupture into a reimplanted LCA. This catastrophic event raises several questions: (1) Should an aortogram have been performed at smaller-diameter balloon dilation? (2) Could the patient have been managed using transcatheter rescue with covered stents in both the LCA and RV-PA homograft? (3) Could rescue procedures have been better prepared or managed more efficiently?

Close multidisciplinary collaboration through rapid defibrillation, CPR, ECMO, and surgical repair was critical to this patient’s survival. Immediate transfer to the OR enabled cardiac protection (cardioplegia) and venting of the left atrium, which was important to the outcome because the patient continued to have myocardial ischemia with the LCA-to-homograft shunt and cardiac demand due to left ventricular load on the ECMO circuit. Likewise, hybrid intervention on the hyperacutely failed bioprosthetic pulmonary valve was another remarkable result of the adult congenital heart disease (ACHD) heart team approach.7

Also remarkable was the rapid failure of the freshly implanted bioprosthetic valve. Our surgical team hypothesized that reduced RVOT flow while on ECMO likely played a role in its hyperacute failure. Failure of bioprosthetic valves while on ECMO has been reported, with evidence of fusion of the bioprosthetic cusps in valves implanted in the mitral position; this is thought to be related to thrombus formation in the low-flow state.6,9 Alternatively, cranial malrotation of the pulmonary prosthetic valve has been reported as causing functional valve failure.10

CONCLUSIONS

Interventional procedures such as TPV are important advancements in ACHD and can be an alternative to open surgery, but they are not without risk. Close collaboration with a heart team at a specialized ACHD center is required to prepare for and manage complications in these complex cases.
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