

MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

ADVANCED

CASE REPORT: DA VINCI CORNER

Complex Catheter-Based Structural Heart Reconstruction in a Patient With Tricuspid Atresia and Björk Palliative Conduit



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ABSTRACT

Björk conduit failure is a common reason for reintervention after a Björk modification of the Fontan procedure. We describe a first performed in human percutaneous procedure for the treatment of a failing Björk circuit in an adult with congenital heart disease and complex anatomic features. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:212–6) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

A 48-year-old woman, with a modified-Björk Fontan circulation, was admitted repeatedly to Rabin Medical Center, Israel, for worsening right-sided heart failure. Her physical examination showed signs of volume overload, including bilateral pedal edema and ascites.

PAST MEDICAL HISTORY

Our patient was born with tricuspid atresia in situs solitus and with concordant ventriculoarterial

connections. She had undergone multiple palliative surgical procedures, including a Waterston central shunt connecting the pulmonary artery to the aorta. This was later converted to a classical Glenn procedure, connecting the superior vena cava to the right pulmonary artery, which also remained partially connected to the right atrium (RA). Later the patient underwent a Björk procedure, connecting the RA to the right ventricular outflow tract (RVOT) with a nonvalved conduit (Björk conduit).

LEARNING OBJECTIVES

- To understand the complexity of Björk conduit failure management.
- To describe a first performed in human percutaneous treatment of a failing Björk circuit with complex anatomic considerations.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of worsening volume overload in our patient with a Björk modification of the Fontan circulation included stenosis or regurgitation of the Björk conduit, arrhythmias, Fontan-associated liver disease, and protein-losing enteropathy.

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INVESTIGATIONS

Over time, the hypoplastic right ventricle grew to become a pumping chamber of normal size, with a significant stroke volume (Figure 1). This caused an escalating problem of systolic backward flow in the Björk conduit resulting from the lack of a valve in the canal (Figure 2).

Her functional status subsequently deteriorated to New York Heart Association functional class III to IV, resulting in frequent hospitalizations for overt right-sided heart failure. During these hospitalizations, she underwent repeated abdominal paracentesis for removal of refractory ascites and received intravenous diuretic agents. There was consensus that the patient was a poor candidate for surgical reintervention or organ transplantation.

The pathophysiology was caused by the absence of a tricuspid valve and backward congestion, similar to that of “free” tricuspid regurgitation. This was confirmed on hemodynamic cardiac catheterization. A procedure to reconstruct a tricuspid valve was required. There were 3 main anatomic dilemmas in this patient.

The main obstacles to percutaneous valve implantation were the elliptical shape of the Björk conduit and the proximity of the right coronary artery to the Björk conduit (Figure 3). This anatomy required pre-stenting, which was to be done using multiple stents to build a scaffolding from the inferior vena cava (IVC) proximally to the Björk conduit, thus allowing transcatheter heart valve (THV) implantation proximal to the elliptical conduit.

Another major concern was ensuring adequate sealing of the gap between the outer stent graft and a THV. We obtained permission to use a self-expanding pre-stent device (Alterra Adaptive Prestent, Edwards Lifesciences, Irvine, California). This device has a narrow waist and acts as a docking adaptor for the Edwards SAPIEN 3 THV (Edwards Lifesciences). This device was to be placed inside the planned scaffold, thus eliminating the diameter gap between the stent graft and the THV and preventing possible right coronary artery compression.

Third, it was important to preserve the patency of the hepatic veins and coronary sinus. A custom-made self-expandable bare-metal stent (Innoventric, Nes Ziona, Israel) was commissioned to be used for the section of the scaffolding from the IVC to the proximal RA part to ensure hepatic vein drainage. This stent and the pre-stent device were provided for use in this patient on the basis of compassionate use after

obtaining ethical permission from the Institutional Review Board.

MANAGEMENT

This procedure was performed by a multidisciplinary team and was broadcast live to the Edwards Lifesciences proctor, who was unable to be present because of restrictions related to the coronavirus disease 2019 pandemic.

The procedure was performed in 4 stages. The first step was to insert a large, self-expandable, bare-metal stent (Innoventrics stent 76 × 40 mm) inside the IVC to the RA as an anchoring apparatus. Transesophageal echocardiography confirmed that neither the hepatic veins nor the coronary sinus were obstructed. To ensure stability of this stent further, another bare-metal stent (Andra XXL 43 mm, Macromed, Accrington, United Kingdom) was implanted inside the proximal part of the stent in the IVC.

The second step was to insert a stent graft (Valliant Captiva 42 × 42 mm diameter and 100 mm length, Medtronic, Minneapolis, Minnesota) from the Björk conduit to the bare-metal stent with a margin of overlap. A second stent graft (Valliant Captiva 44 × 44 mm and 100 mm length) was then implanted from the line of anastomosis of the Björk conduit with the RVOT and into the first stent graft. This completed

ABBREVIATIONS AND ACRONYMS

- IVC = inferior vena cava
- RA = right atrium
- RVOT = right ventricular outflow tract
- THV = transcatheter heart valve

FIGURE 1 Cardiac Computed Tomography Images Showing a Substantial Right Ventricle

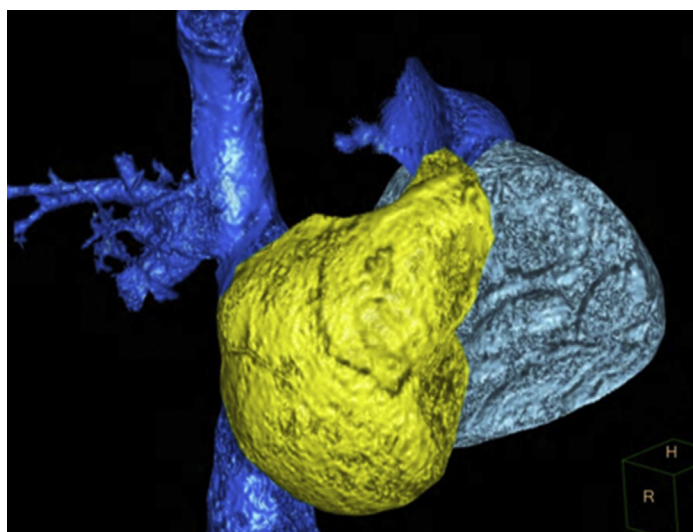
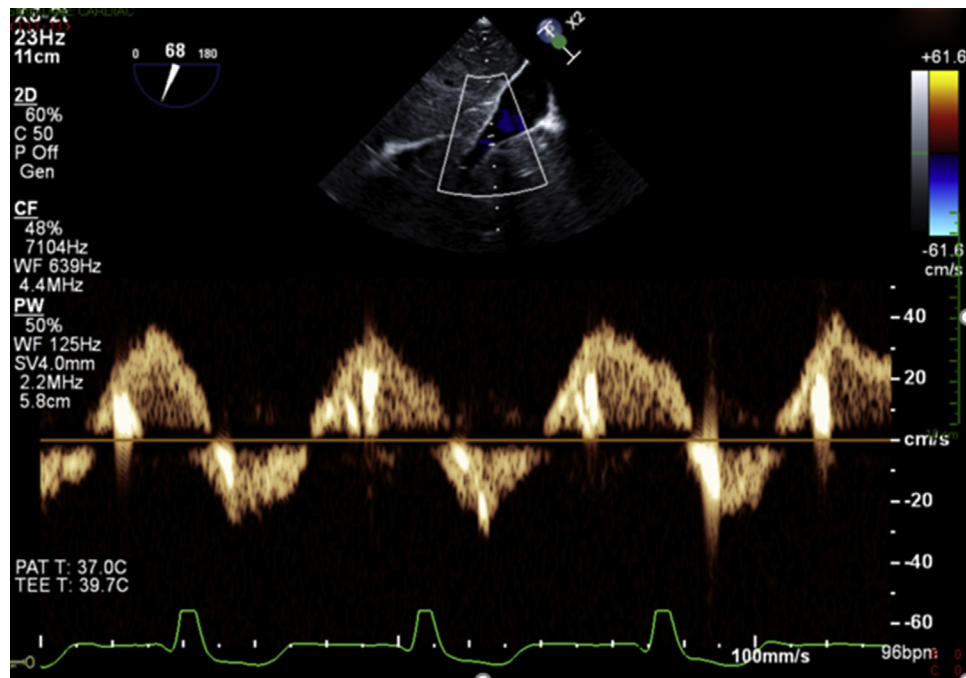


FIGURE 2 Transesophageal Echocardiography Before Intervention Showing Severe Systolic Flow Reversal in Hepatic Vein

the entire telescoping scaffold from the IVC to the RVOT, with a bare-metal proximal section and a covered-stent distal section.

The third step was to insert the self-expanding pre-stent device (Alterra Adaptive PreStent, 40 × 45 mm)

inside the stent graft scaffold. A stent graft balloon (Reliant, 46 mm, Medtronic) was used to ensure full expansion of the pre-stent device.

The fourth step was to implant the Edwards SA-PIEN 3 29-mm THV within the pre-stent device within

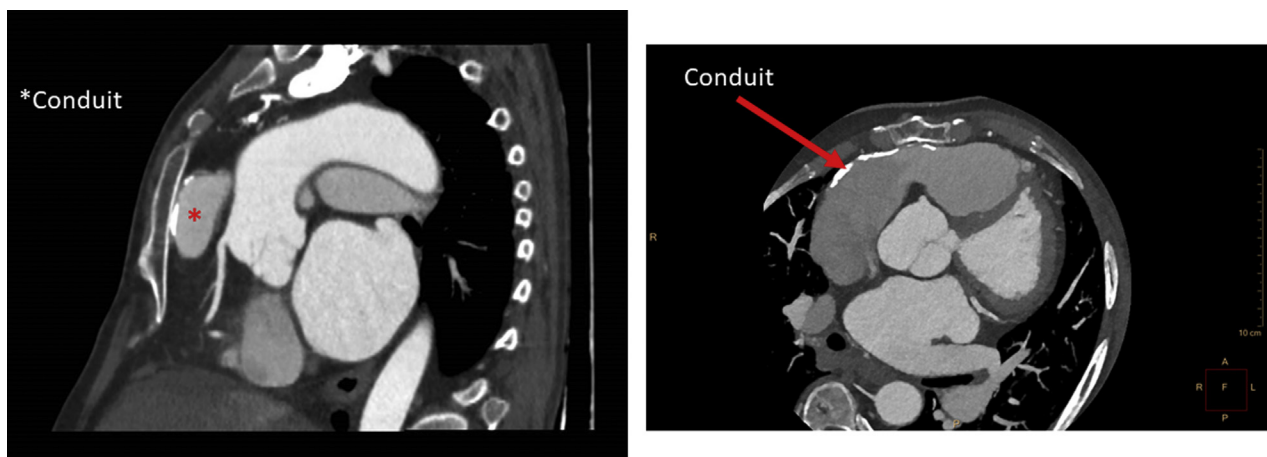
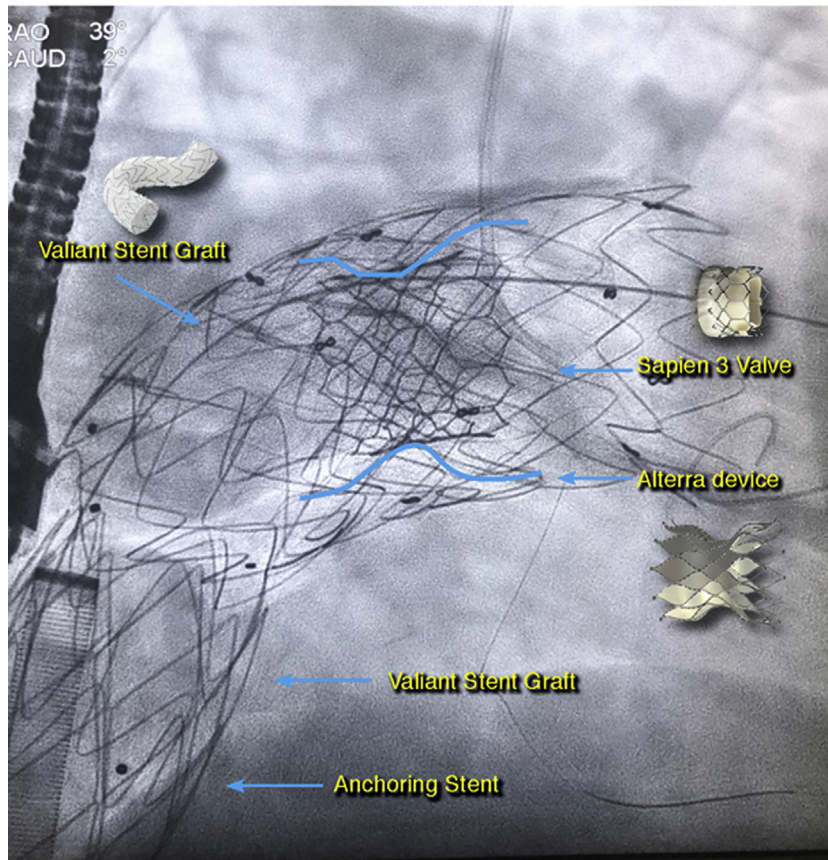
FIGURE 3 Cardiac Computed Tomography Image Showing the Elliptical Björk Conduit

FIGURE 4 Angiographic Image of Devices In Situ



the conduit in the “tricuspid” position. This was done using a long 26-F DrySeal Flex introducer sheath (Gore Medical, Newark, Delaware). The fully reconstructed conduit with the adaptive device and the valve is shown in **Figure 4**. Transesophageal echocardiography showed a well-functioning prosthetic valve with a trivial leak between the pre-stent device and the stent graft (**Video 1**). The patient experienced no peri-procedural complications. The patient was restarted on warfarin anticoagulation therapy and was discharged 4 days following the procedure.

DISCUSSION

The Fontan operation, first described for the palliation of tricuspid atresia, has permitted the survival into adulthood of many patients with complex congenital heart lesions. (1).

The Björk modification of the Fontan circulation, as performed in our patient, was used in the past for palliating tricuspid atresia and normally related

arteries. It was based on the assumption that inclusion of the right ventricle in the circuit, even if hypoplastic, would enhance pulmonary blood flow by the pumping activity. It did not take into account that the hypoplastic right ventricle could grow in time because of the constant blood flow into it, and with the lack of a valve in the conduit, would result in an ever-increasing backward flow of blood in systole. Long-term results have shown that this conduit does not improve hemodynamic properties (2). Fortunately, patients palliated in this manner are uncommon nowadays because modern era palliation techniques with extracardiac or lateral tunnel Fontan circuits have become the methods of choice.

The management of Björk circuit dysfunction is complex. Many of these patients have had multiple surgical procedures and comorbidities that make them poor candidates for reoperation (3). Redo surgery is complex because of the juxtasternal position of the circuit, and it carries a high risk of mortality and adverse outcomes (4).

Transcatheter valve implantation has become an increasingly desirable therapeutic option. Successful THV implantation has been described in patients with RA-RV conduit dysfunction after modified Björk-Fontan procedure with favorable results (5). However, our patient had unfavorable anatomy (as described earlier) and required a creative and tailored personalized solution.

This report describes an innovative percutaneous procedure for the treatment of a failing Björk circuit. The procedure involved constructing a customized stent scaffolding that extended all the way from the elliptical Björk conduit to the IVC, implanting a self-expanding docking adaptor into the scaffold, and then implanting a THV within this scaffold.

This intervention is the first of its kind described or performed in humans. Adaptive pre-stent devices have been previously used to reconfigure the RVOT to allow pulmonary valve replacement in patients whose native anatomy was deemed unsuitable (6). However, the use of this device as a docking device in the Björk conduit is original.

FOLLOW-UP

At 6 weeks post-discharge, the patient had lost 7 kg of body weight, she was able to reduce the dose of diuretic agents substantially, and she described an improved exercise capacity and quality of life.

CONCLUSIONS

This report demonstrates a feasible percutaneous alternative to repeat surgery in a patient with conduit failure and complex anatomy. This procedure could be adapted to provide a percutaneous solution to other structural therapeutic dilemmas.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS Björk conduit, Fontan, transcatheter heart valve

APPENDIX For a supplemental video, please see the online version of this paper.