Converting Fontan-Björk to 1.5- or 2-ventricle circulation

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Abstract

Patients with tricuspid atresia and ventricular septal defect have in the past occasionally undergone a Fontan with “Björk” modification to create a connection between the right atrium and the right ventricular outflow tract. While rarely performed now, patients with this physiology often face severe complications requiring re-intervention. We hypothesize that surgical conversion to a 2-ventricle or 1.5-ventricle circulation can improve hemodynamics, clinical status, and thus increase time to transplant. We present two successful cases to illustrate this idea.

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Surgical interventions for patients with single ventricle congenital heart disease have evolved over time. Patients with tricuspid atresia (TA) and ventricular septal defect (VSD) occasionally are treated with the “Björk” modification, which includes creation of a valveless right atrial (RA) to right ventricular outflow tract (RVOT) communication [1]. Long-term complications occur due to the free regurgitant flow, calcification and/or stenosis of the homograft, RA dilatation, atrial arrhythmias, liver congestion, thromboembolism, and ventricular dysfunction [2,3]. In the majority of patients after Björk modification, no additional surgery is considered, other than orthotopic heart transplant. We present two cases of successful Fontan take-down to either a 1.5-ventricle or 2-ventricle anatomy late after Björk operations.

Case Reports

Case 1

A man born with TA and VSD presented with signs and symptoms of a failing Fontan at age 32 years. In infancy he had undergone bilateral Blalock-Taussig shunt placement followed by Fontan with Björk modification at four years of age. Figure 1A demonstrates forward flow through the RVOT and Figure 1B shows reverse flow. By age 17 years, the patient had developed significant RA dilatation, recurrent atrial flutter, and sinus node abnormalities requiring pacemaker. He was New York Heart Association (NYHA) class II. Echocardiogram revealed moderately dilated RV with right ventricular end diastole (RVDD) measuring 3.42 cm with mildly diminished function. He underwent placement of a 29-mm porcine valve in the “tricuspid” position in the RA-RVOT conduit and concurrent radiofrequency atrial maze procedure, converting him to 2-ventricle physiology. Echocardiographic image in Figure 2 demonstrates unobstructed flow through the implanted valve in the RA-RVOT conduit. RV size and function remained stable with RVDD measuring 3.49 cm. Post-operatively, NYHA class improved to class I within 6 weeks. He had some recurrence of atrial flutter that was managed medically. He clinically did well for 15 years until degradation of tissue valve led to biventricular

dysfunction, recurrent atrial flutter, and right pleural effusion. At 32-years-old he underwent tricuspid valve replacement with further radiofrequency atrial ablation and has been doing well without hospital readmission since re-operation fifteen months ago.

Case 2

A man born with TA and VSD presented with signs and symptoms of a failing Fontan at age 42 years. He had undergone left BT shunt in infancy, Waterston shunt at six-years-old, and Fontan with Björk modification at nine-years-old. By age 42 years, he developed RA enlargement complicated by permanent atrial fibrillation and liver congestion. He underwent cardiac MRI, which revealed RV end-diastolic volume of 73 mL/m^2 and ejection fraction of 31% with an RV:LV end-diastolic volume ratio of 1. He was NYHA class II-III. He underwent Fontan conversion with creation of a 1.5-ventricle circulation by creating a bidirectional Glenn anastomosis (superior vena cava to pulmonary artery) and implanting a 29-mm porcine valve between the RA appendage and the RVOT as can be seen in Figure 3. He had a radiofrequency maze procedure performed during the same operation but unfortunately remained in atrial fibrillation post-operatively albeit with improved rate control. He has done well post-operatively without hospital readmission in the ten months since Fontan conversion and is now NYHA class I.

Comment

Surgical interventions for single ventricle patients, specifically TA with VSD patients, has evolved over the past few decades. While Björk-Fontans are rarely performed now, there are patients who are now adults facing the long-term consequences of Björk circulation. There are currently no guidelines as to when to intervene or what intervention to conduct. A transplant would be the most definitive therapy. However, given the lack of donor organs for patients with

complex congenital heart disease and the comorbidities associated with transplantation, strategies to delay transplant or alternatives to transplant are clearly needed.

Ono et. al. reviewed 41 patients with Fontan-Björk physiology and identified positive clinical and hemodynamic results from having pulsatile flow from the RV to the PA, including fewer interventions for tachyarrhythmias, higher oxygen uptake on exercise stress testing, higher systolic PA pressure, and decreased cardiac decompensation compared to those with non-pulsatile flow [4]. Thus, for patients with adequate RV size and function, placement of a valve in the “tricuspid” position resulting in conversion to a 1.5- or 2-ventricle system could reduce complications of Fontan physiology or prolong time for need for transplant.

For some patients with Fontan-Björk physiology who face complications due to the regurgitant flow in the RA-RV connection, a percutaneous valve in the “tricuspid” position is a possible intervention [2,5-7]. Shah et al. described 16 patients who underwent percutaneous valve implantation for RA-RV regurgitation and RA dilation. While these patients showed hemodynamic improvement, the patients who remained symptomatic were those with uncontrolled atrial flutter [2]. Studies have shown that patients who undergo Fontan conversion with surgical treatment of tachyarrhythmias at the time of Fontan conversion are superior to conversion without arrhythmia surgery [3,8]. Therefore, for these two patients who had developed RA dilatation from the regurgitant flow inherent to Björk physiology that was complicated by recurrent or permanent atrial arrhythmias, the team of surgeons and cardiologists offered surgical conversion with combined atrial arrhythmia surgery.

The first patient underwent a 2-ventricle repair because he had reasonable RV size and function and the second underwent a 1.5-ventricle repair because of concern that it would be difficult to come off bypass postoperatively without a Glenn due to small RV size and decreased RV function. Both patients underwent maze procedures for treatment of medically-resistant arrhythmias. Because they needed arrhythmia intervention, percutaneous valve placement was felt not to be a good option.

These two cases illustrate a surgical option for 1.5- or 2-ventricle Fontan conversion in selected patients with the Björk modifications who either would benefit from atrial arrhythmia surgery or for those who are otherwise not candidates for percutaneous placement of a valve in the tricuspid position. While data is limited in this subset of patients, we hypothesize that implanting a durable valve in this location would reduce complications from Fontan-Björk circulation, namely liver congestion and right atrial dilatation with resultant atrial arrhythmias.

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Figure legends

Figure 1: Echocardiogram images of the Björk modification with A showing forward flow through the RVOT and B showing reverse flow.

Figure 2: Echocardiogram image showing unobstructed flow through the “tricuspid valve” in the RA-RV conduit.

Figure 3: Cardiac MRI data with A showing the pre-operative SVC to RA junction and B shows the post-operative SVC to PA anastomosis, smaller RA, and new porcine bioprosthetic “tricuspid valve.”