Creation of a Double-barrel Outflow Tract for Complex Subaortic Obstruction in Single Ventricle

Mahmut Ozturk, MD; Aybala Tongut, MD; Sofia Hanabergh, MSc; Can Yerebakan, MD, PhD; Yves d’Udekem, MD, PhD

Division of Cardiac Surgery, Children’s National Hospital, The George Washington University School of Medicine and Health Sciences, Washington, DC, USA

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Corresponding Author:

Yves d’Udekem, Md, PhD

Cardiac Surgery, Children’s National Hospital

The George Washington University School of Medicine and Health Sciences

111 Michigan Ave NW,

Washington, DC 20010

Phone: +1 (202)758-5305
Central Message

We propose a unique surgical alternative for the patients who underwent single ventricle palliation and developed an outflow tract obstruction with the involvement of an atrioventricular valve.

Central Picture Legend

Outflow tract obstruction in echocardiography and in 4D magnetic resonance flow imaging

Glossary of Abbreviations

Atrioventricular valve (AVV)
Double-outlet right ventricle (DORV)
Ventricular septal defect (VSD)
Damus-Kaye-Stansel (DKS)
Main pulmonary artery (MPA)
Abstract

Single ventricle outflow tract obstruction in Fontan circulation can jeopardize survival rates. Involvement of one of the atrioventricular valves in the obstruction may necessitate replacement of the valve if it is not amenable to repair. In patients with Fontan circulation, valve replacement is associated with heart block and high short and long-term mortality. Here, we present a unique case of right ventricular outflow tract reconstruction with a double barrel Damus-Kaye-Stansel technique interposing an aortic homograft in pulmonary position in a DORV patient with Fontan palliation to avoid the risks associated with the valve replacement or pacemaker implantation.
Introduction

Double-outlet right ventricle (DORV) is a cardiac anomaly where both great vessels mainly originate from the right ventricular cavity. In the DORV constellation, the anatomy, and the alignment of intracardiac structures also determine the clinical presentation of the affected patient. When palliated into Fontan, the entire systemic circulation relies on an unobstructed outflow tract and its restriction is a feared condition. If the atrioventricular valve (AVV) is causing the obstruction, conventional strategy suggests surgical relief with resection and replacement of the valve with prosthesis. However, AVV replacement in these patients carries a significant heart block, in-hospital and long-term mortality risk\(^1\). Alternative approaches can be utilized to preserve the valve function. Herein we present right ventricular outflow tract reconstruction with double-barrel Damus-Kaye-Stansel (DKS) procedure using a homograft in pulmonary position in a DORV patient with previous Fontan palliation. IRB approval was waived.

Case Report

A seven-year-old boy with the diagnosis of a double-outlet right ventricle, non-committed ventricular septal defect at birth was palliated into Fontan circulation in domo. On the sixth day of life, he underwent banding of the main pulmonary artery (MPA) and ligation of ductus arteriosus. Additionally, a balloon atrial septostomy was conducted. At five months of age, the MPA band was taken down. Bidirectional Glenn anastomosis was performed with pulmonary valve excision, division, and oversewing of the MPA. The patient had at the time no systemic outflow tract obstruction. The Fontan completion with a 16mm ring supported, fenestrated intra/extracardiac conduit was conducted at two years and two months of age. The patient had a middle cerebral artery stroke following the Glenn surgery.
During his routine follow up, he presented with a decrease in systemic oxygen saturation. A sub-aortic stenosis, caused by mobile accessory tissue on the anterior and septal leaflets of the right AVV with moderate flow acceleration across the systemic outflow tract (peak velocity < 4 m/s under sedation, peak gradient 50mmHg) was diagnosed using transesophageal echocardiography. (Figure 1) The decision for the reoperation was made by a multidisciplinary heart team.

The operation was performed through median sternotomy. Aortic, femoral and innominate venous cannulation and cardioplegic arrest were established. The aorta was transected. After the inspection of the outflow tract, a subaortic stenosis caused primary chordal tissue intimately related to the right AVV was identified. The right AVV was also inspected through a right atriotomy. The presence of a subvalvular apparatus with valvular and chordal involvement was confirmed. A limited myectomy was performed, however it was not sufficient to alleviate the outflow tract obstruction on intraoperative inspection.

Resection of the subvalvular and chordal tissue without impairing the right AVV function was not obtainable. In order to preserve the AVV function and prevent conduction system-related complications, no further manipulations were attempted. The oversewn pulmonary artery root was re-opened. The left main coronary artery and a sizable branch of the right coronary was in immediate proximity to the pulmonary root. Therefore, we refrained from excising the root completely. An 18mm decellularized aortic homograft was placed into the pulmonary root with the inclusion technique in a similar fashion with what we previously described for the Ross procedure\textsuperscript{2}. The aortic root and the homograft were then anastomosed to the proximal part of the ascending aorta, creating a double barrel Damus-Kaye-Stansel connection. The rings on the Fontan conduit were also removed. (Video 1) Weaning from cardiopulmonary bypass,
chest closure, and the immediate postoperative course were free of complications. The patient was extubated on the first postoperative day. Postoperative transesophageal echocardiography revealed normal neo-pulmonic and aortic valve velocities. The patient was discharged on the seventh operative day. He is at home and doing well. (Figure 2)

**Discussion**

An unobstructed systemic outflow tract is the arch stone in Fontan circulation. Systemic outflow tract obstruction may cause ventricular hypertrophy, which is a risk factor for mortality in the Fontan patient group. The intention to alleviate the subaortic stenosis by resecting the subaortic membrane or myectomy can result in injury to the conduction tissue and the pacemaker implantation in single-ventricle patients deteriorates ventricular function. In cases, where an AVV leaflet is involved in the pathophysiology of the outflow tract obstruction, a meticulous risk assessment for the method of treatment is needed to preserve the integrity of the AVV, as AVV regurgitation in single ventricle increases mortality and the need for transplantation. In addition to the risk of heart block, AVV replacement in these patients is also associated with notable in-hospital and late mortality. Therefore, in this particular case, we proceeded with the DKS procedure. Novel techniques like the use of valved conduits in adults and tissue-engineered homograft for semilunar valve replacement after the DKS surgery in Fontan palliation or after Ross and Truncus arteriosus repairs have been reported. We propose a unique surgical alternative for the patients who underwent single ventricle palliation and developed an outflow tract obstruction with the involvement of one of the atrioventricular valves. An interposed autograft in the pulmonary root would potentially show less dilation under the systemic pressure and allow a catheter intervention in case of failure due to calcification or valvar dysfunction.
Disclosures

No funding.
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References


Figure Legends

Figure 1: a) Preoperative echocardiography with outflow tract obstruction and b) 4D magnetic resonance flow imaging

Figure 2: a) Preoperative echocardiography versus b) post DKS constellation

Video Legends

Video 1: Creation of a Double-barrel Outflow Tract for Complex Subaortic Obstruction in Single Ventricle