# 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, and Yves d'Udekem, MD, PhD, Washington, DC

From the Division of Cardiac Surgery, Children's National Hospital, The George Washington University School of Medicine and Health Sciences, Washington, DC.

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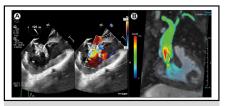
Address for reprints: Yves d'Udekem, MD, PhD, Cardiac Surgery, Children's National Hospital, The George Washington University School of Medicine and Health Sciences, 111 Michigan Ave NW, Washington, DC 20010 (E-mail: yves.dudekem@childrensnational.org).

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

#### 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.

► Video clip is available online.

Double-outlet right ventricle (DORV) is a cardiac anomaly in which 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. Alternative approaches can be used 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 patient with DORV with previous Fontan palliation. Institutional review board approval was waived.

# **CASE REPORT**

A 7-year-old boy with the diagnosis of a DORV, noncommitted 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 5 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 16-mm 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 subaortic 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 50 mm Hg) 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

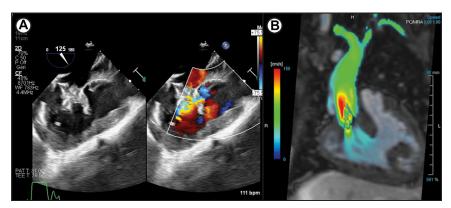
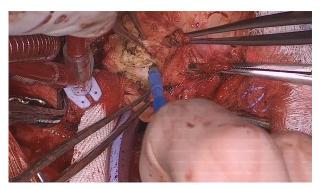


FIGURE 1. A, Preoperative echocardiography with outflow tract obstruction and B, 4-dimensional magnetic resonance flow imaging.

transected. After the inspection of the outflow tract, a sub-aortic 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. To preserve the AVV function and prevent conduction system-related complications, no further manipulations were attempted. The oversewn pulmonary artery root was reopened. 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 18-mm 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. The aortic root and the homograft were then anastomosed to the proximal part of the ascending aorta, creating a



**VIDEO 1.** Creation of a double-barrel outflow tract for complex subaortic obstruction in single ventricle. Video available at: https://www.jtcvs.org/article/S2666-2507(22)00525-9/fulltext.

double-barrel DKS connection. The rings on the Fontan conduit were also removed (Video 1). Weaning from cardio-pulmonary bypass, chest closure, and the immediate postoperative course were free of complications. The patient was extubated on the first postoperative day. Postoperative transe-sophageal echocardiography revealed normal neopulmonic 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.<sup>3</sup> 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 singleventricle patients deteriorates ventricular function. In cases in which 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.<sup>4,5</sup> We propose a unique surgical alternative for the patients who underwent singleventricle 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

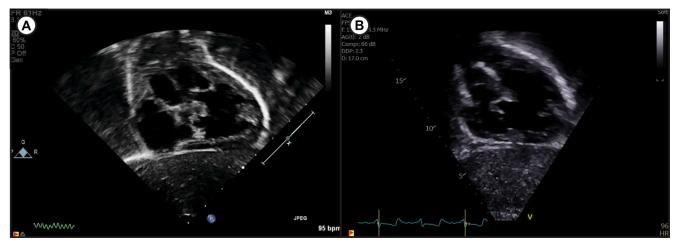


FIGURE 2. A, Preoperative echocardiography versus B, post-Damus-Kaye-Stansel constellation.

pressure and allow a catheter intervention in case of failure due to calcification or valvar dysfunction.

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