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Damus-Kaye-Stansel Procedure with Extracardiac Fontan: Successful Recruitment of Previously Ligated Pulmonary Valve

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ABSTRACT

We present a case of a 2.5-year-old-girl with complex congenital heart disease: tricuspid atresia (TA), bulboventricular septal defect (VSD), hypoplastic right ventricle, d-transposition of the great arteries (d-TGA) with aortic outflow from redundant RV. Due to II/III degree atrioventricular block induced after diagnostic cardiac catheterization, an epicardial pacemaker was implanted during the Glenn procedure. Because of severe left ventricle outflow tract obstruction, she was finally referred for extracardiac TCPC (extracardiac Fontan type) with recruitment of PV and Damus-Kaye-Stansel anastomosis. Intraoperatively, the pulmonary trunk stump was opened and a competent pulmonary valve with flaccid leaflets was found. Simple ligation of the pulmonary trunk with a preserved pulmonary valve enabled an effective aorto-pulmonary bridging of systemic outflow tract with the use of natural fully competent ventricle-arterial valves. The relief of single ventricle outflow tract obstruction led to final stabilization of spontaneous sinus rhythm recovery after 2 years of pacemaker stimulation.

INTRODUCTION

Patients with univentricular heart (SV), hypoplastic right ventricle, and transposition of the great arteries (TGA) are at risk of slowly progressing subaortic stenosis (SAS), especially when intracardiac bulboventricular septal defects coexist with stenosis of the aortic arch [Matitiau 1992]. The preferred solution is elective Damus-Kaye-Stansel (DKS) procedure, but optimal timing of surgery remains questionable [McElhinney 1997].

The problem of systemic outflow obstruction is of great importance in newborn patients with diagnosed restrictive bulboventricular septal defect and aortic outflow from a redundant ventricle. In this anatomic variant of single ventricle (SV), the restriction over bulboventricular defect results in interventricular pressure gradients, which promote heart failure (HF). Thus intracardiac blood flow should be meticulously controlled in every patient with SV, especially while an initially diagnosed anomaly is presented with unrestricted, or borderline septal defects in early infancy. Moreover, complicated single ventricle morphology could be presented with a variety of atrioventricular (AV) conduction disturbances, mostly related to staged surgical and interventional treatment.

We report an univentricular heart patient with AV-block and restrictive bulboventricular defect who was ultimately

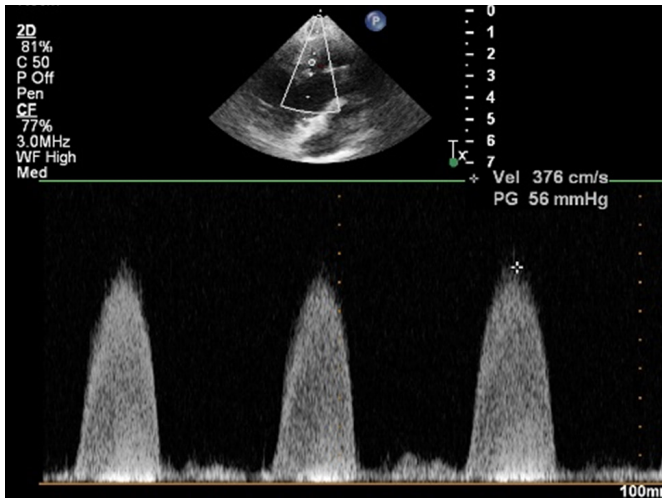

Figure 1. Peak gradient over bulboventricular septal defect (VSD) was around 50 mmHg, measured by echocardiography.

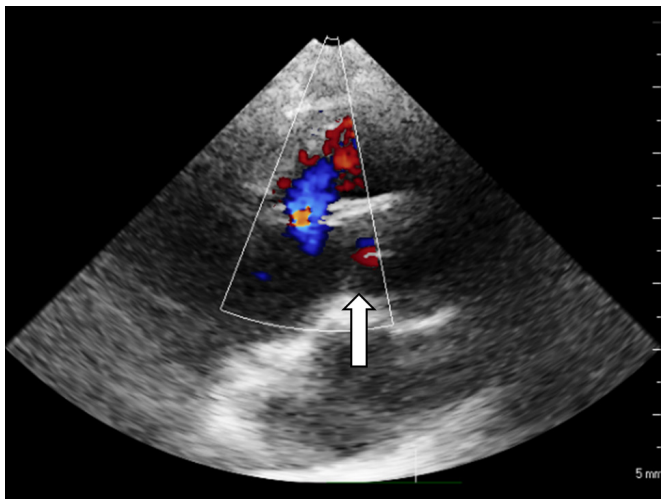

Figure 2. Competent pulmonary valve (arrow) with flaccid leaflets.

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referred for extracardiac total cavopulmonary connection (TCPC-extracardiac Fontan) combined with a delayed DKS procedure at the age of 2.5 years, which finally resolved intraventricular flow disturbances and provided complete sinus rhythm recovery after incidental AV block.

CASE REPORT

We present a case of a 2.5-year-old-girl, 11.5 kg body weight, with complex congenital heart disease: tricuspid atresia (TA), bulboventricular septal defect (VSD), hypoplastic right ventricle, d-transposition of the great arteries (d-TGA) with aortic outflow from redundant RV, and aortic coarctation with hypoplastic aortic arch (CoA-HAA), additionally suffering from Turner syndrome and hypothyroidism. Initially she underwent staged palliation: Prostaglandin E1 medication before modified extended end-to-end aortic arch repair with the pulmonary artery banding at the 10th day of life, and bidirectional cavopulmonary anastomosis (BDCPA) at the age of 7 months. Due to II/III degree atrioventricular block induced after diagnostic cardiac catheterization, an epicardial pacemaker was implanted during the Glenn procedure. The pulmonary artery (PA) was ligated in a manner to preserve the pulmonary valve (PV) orifice and effective leaflets rinsing.

Prior to the final physiologic palliation she was found to have severe left ventricle outflow tract obstruction (LVOTO) in the effect of gradual septal hypertrophy, which increased bulboventricular VSD restriction. Measured peak echocardiographic gradient over VSD was 30-50 mmHg (Figure 1).

She was finally referred for extracardiac TCPC (extracardiac Fontan type) with recruitment of PV and Damus-Kaye-Stansel anastomosis, or alternatively: surgical intraoperative VSD enlargement. Intraoperatively, the pulmonary trunk stump was opened and a competent pulmonary valve with flaccid leaflets was found (Figure 2). There were no signs of thrombus or fibrosis in the PA stump. The side-to-side anastomosis of the ascending aorta and recruited PA orifice was performed with the dome of a xenograft (Contegra, EU) patch. Her postoperative period was uneventful. The echocardiography proved unobstructed systemic single ventricle outflow and competent PV function. Pacemaker control showed the final stabilization after spontaneous sinus rhythm (SR) recovery that was partially observed in meticulous controls shortly after BDCPA. Finally, recovered natural SR firmly allowed the draw back from stimulation without a need for medical support.

DISCUSSION

Systemic ventricular outflow tract obstruction in univentricular heart with tricuspid atresia and transposition of great arteries is a frequent complication. Intraventricular gradient could appear on various stages of Fontan pathway, and is resolved in different ways. Simple enlargement of obstructed VSD, or alternatively: subaortic muscle resections are associated with a great risk of postoperative heart block, ventricle insufficiency, and predictable reoperation due to subaortic reobstruction.

In turn, more frequently reported Damus-Kaye-Stansel procedures could be performed only with competent PV; thus previous surgeries with its complete closure could exclude this option [Alsoufi 2014]. Our preferable strategy was to preserve pulmonary outflow with untouched valve orifice to postpone the decision whether to finally close, or recruit pulmonary outflow for systemic tract supplementation. Following, despite pulmonary trunk ligation, after two years PV persisted functionally in our patient, with good leaflets coaptation [Broekhuis 1999]. No evidence of thrombus was observed, which was related to meticulous antiplatelet therapy with acetylsalicylic acid (5 mg/kg b.w.) and permanent flooding of pulmonary orifice area. There is no doubt that preservation of the pulmonary valve appeared especially valuable in the presented patient with bulboventricular VSD, who was naturally considered at high risk of expected systemic outflow reobstruction with natural heart growth [Matitiau 1992].

Temporary postoperative arrhythmias after Glenn or various types of BDCPA are related to surgical maneuvers, although refractory heart blocks could appear during every percutaneous cardiac intervention in patients with univentricular heart [Reichlin 2014]. The most valuable result of the presented strategy with Fontan and delayed Damus-Kaye-Stansel simultaneous procedures, despite unobstructed systemic blood flow, was final stabilization of heart rhythm and spontaneous relief of atrioventricular block. It is worth noting that during 12-month-long regular outpatient controls, SR was regularly confirmed without any need of pacemaker resumption.

Conclusion

Simple ligation of the pulmonary trunk with a preserved pulmonary valve enables an effective aorto-pulmonary bridging of systemic outflow tract with the use of natural fully competent ventricle-arterial valves.

The relief of single ventricle outflow tract obstruction led to final stabilization of spontaneous sinus rhythm recovery after 2 years of pacemaker stimulation.

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