Multiple replacements for recurrent stuck mechanical valve in a patient with Fontan circulationtech

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Abstract

Significant atrioventricular valve insufficiency is associated with increased mortality and morbidity in patients with a single-ventricle because of the congestion associated with single-ventricle circulation. Treatment of atrioventricular valve insufficiency is mandatory for completing Fontan circulation. Although valve repair is a favorable procedure for the lesion because it preserves the valve leaflet and ventricular motion, replacement of the atrioventricular valve is the last option of treatment. The choice of a mechanical valve is often made for atrioventricular valve replacement in children. However, because children's small body size, mechanical valve implantation can cause early and late complications, including thromboembolic or hemorrhagic events, the artificial valve leaflet becoming stuck, and ventricular dysfunction. Because of these complications, valve replacement can be performed several times, and repeat surgical intervention can induce technical difficulties of artificial valve implantation as well. We present a patient who received multiple mechanical valve replacements due to repeat valve leaflet stuck after first palliation of pulmonary artery banding followed by Glenn anastomosis and Fontan completion. To overcome surgical difficulty, some technical modification was required during the repeat valve replacement.

Title page

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This case report has been approved by the institutional review board of our university.

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a favorable procedure for the lesion because it preserves the valve leaflet and ventricular motion, replacement of the atrioventricular valve is the last option of treatment. The choice of a mechanical valve is often made for atrioventricular valve replacement in children. However, because children's small body size, mechanical valve implantation can cause early and late complications, including thromboembolic or hemorrhagic events, the artificial valve leaflet becoming stuck, and ventricular dysfunction. Because of these complications, valve replacement can be performed several times, and repeat surgical intervention can induce technical difficulties of artificial valve implantation as well. We present a patient who received multiple mechanical valve replacements due to repeat valve leaflet stuck after first palliation of pulmonary artery banding followed by Glenn anastomosis and Fontan completion. To overcome surgical difficulty, some technical modification was required during the repeat valve replacement.

Introduction

Significant atrioventricular valve (AVV) insufficiency, which has been reported in up to 37% of patients with single-ventricle [1,2], is one of the major drawbacks of the completion of Fontan circulation. Congestion in the atrium resulting from atrioventricular regurgitation should be reduced before and after completion of single-ventricle circulation. Valve repair is preferable for surgical intervention to the lesion, because valve and ventricle function can be preserved, and the risk of complications related to anticoagulant therapy after mechanical valve implantation might be reduced. Furthermore, an artificial valve frame fixes the AVV annulus without expected annulus growth after implantation, which results in the need for repeat artificial valve replacement depending on the body growth of the patient. Valve replacement is frequently associated with a moderately high mortality and various complications, including the need for permanent pacemaker implantation and repeat valve replacement [3]. However, valve replacement is often used as treatment for irreparable valve lesions.

Case description

The patient was diagnosed with a single right ventricle and mitral atresia and received pulmonary artery banding (PAB) during the infantile period for reduction of pulmonary blood flow and ventricle preload. Moderate AVV regurgitation was already observed at that time. Almost one month after PAB, replacement of the AVV with a mechanical valve (diameter 18 mm) was required for uncontrollable AVV regurgitation. After Glenn anastomosis as second-stage palliation followed by pulmonary artery augmentation with the e-PTFE patch, Fontan circulation was completed (age 2 years 5 months). Two years and 5 months after Fontan operation (age 4 years 10 months), repeat valve replacement was performed using a mechanical valve with a 21-mm diameter because of a stuck valve leaflet. One month after this repeat valve replacement, a stuck valve was again observed, another valve replacement was needed. During the same hospitalization, the stuck valve was observed yet again, and yet another valve replacement was performed using a mechanical valve with a 21-mm diameter. Because the patient's ventricular structure possibly caused the disturbance of the valve leaflet, the bottom of the mechanical valve was raised with an artificial graft. Although repeated valve leaflet stuck was observed, laboratory examination did not reveal any coagulopathy. Repeated valve replacement resulted in severe ventricular dysfunction, which might result in congestion of blood flow through the valve leaflet. The patient was placed on much medication, including diuretics and β-blockers for severe heart failure. When the patient was 6 years 5 months old, a stuck valve was again observed, and rather than valve replacement, fibrous tissue around the stuck valve leaflet was removed, which resulted in recovery of motion of the valve leaflet. Because of the patient's severe ventricular dysfunction, it was believed that it would be hard for the patient to be removed from cardiopulmonary bypass after AVV replacement (AVVR) and AVVR was not indicated then. However, one month after the operation, re-re-re-re valve replacement was needed because of a stuck valve during the same hospitalization. During the operation, an e-PTFE graft of the Fontan route was transected to obtain the surgical field for valve implantation, and then a bottom-raised mechanical valve with a 19-mm diameter implanted. Because of the excessive fibrous tissue observed on the AVV annulus, which could have hindered the valve leaflet motion, the same-size valve could hardly be implanted. The left atrial (LA) space was too small for the bottom-raised valve, and thus the LA wall was augmented with an e-PTFE patch. To increase cardiac output, right atrium to LA communication was

created. On the 45th postoperative day, the patient was discharged from the hospital on warfarin potassium to maintain a prothrombin time–international normalized ratio between 2.5 and 3.0.

Discussion

Artificial valve implantation in small children with a single-ventricle is associated with an increased risk of mortality [4]. The rigid frame of the artificial valve disturbs the motion of the ventricle and prevents the growth of the AVV annulus. In the small ventricle space, the leaflet motion can be hindered by the ventricular structure, such as chordae, and in the small atrial space, the bloodstream through the valve leaflet is also obstructed. Considering the growth of children, it might be preferable to select a larger size valve for implantation to secure an adequate valve orifice area. However, this strategy could result in valve dysfunction caused by bloodstream obstruction and a stuck leaflet. In addition to these drawbacks, implanting a large artificial valve could result in restriction of ventricle motion due to the rigid frame of the valve followed by ventricle dysfunction. Ventricle dysfunction may cause congestion of blood stream, which result in artificial valve leaflet stuck as well. Implantation of a larger size artificial valve in smaller children is a major risk factor for mortality [4] for these reasons.

We also encountered much technical difficulty in removing the old valve and implanting the new one because the direction of the cardiac apex could not be changed to enable a good surgical field due to severe adhesion caused by repeat heart surgery and after the multiple insertion of an artificial valve, a thickening of the AVV annulus can often be observed, which results in difficult insertion of a new valve on the annulus.

In our case, a severe thickening of the AVV annulus and small LA space were noted; thus, a smaller size valve with a diameter of 19 mm was selected and implanted in the supra AVV annulus position. We were concerned that the structure in the ventricle could hinder the opening of the artificial valve leaflet. To reduce the chance of valve leaflet dysfunction, the bottom of the mechanical valve was raised with an e-PTFE graft. The LA wall was augmented with an e-PTFE patch because the artificial valve was projected into the LA space. Various technical modifications in mechanical valve implantation are necessary for the case who had received repeated valve procedure.

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Figure legend

Figure 1

(A) Artificial graft of the Fontan route (inferior vena cava to pulmonary artery) was transected to obtain an adequate surgical field for the valve replacement.

(B, C) The atrial wall was augmented with an e-PTFE patch, and the transected artificial graft of the Fontan route was interposed with another artificial graft.

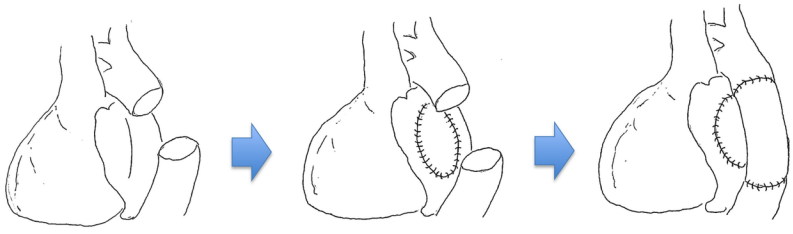

Figure 1