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Partition of Common Atrioventricular Valve in a Patient With Dextrocardia and Univentricular Circulation

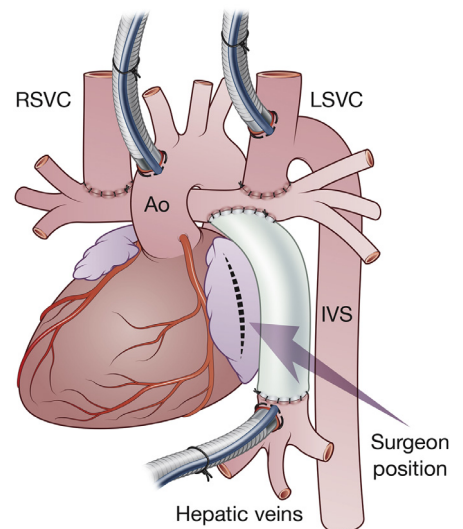
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Long-term outcomes in children with atrioventricular septal defect (AVSD) and univentricular palliation are of concern, with <60% survival at 25 years.¹ Common atrioventricular valves (AVV) often become insufficient in patients with univentricular physiology, leading to heart failure.^{1,2} We have recently observed that outcomes of children with AVSD who reach Fontan circulation are not as bad as previously thought, provided that the AVV remains competent.¹ Common AVV surgery is associated with substantial mortality and reoperation rates.³ Although successful AVV repair is associated with better survival and freedom from reoperation, good quality repair is difficult to achieve in univentricular circulation,³ especially in patients with dextrocardia.⁴ Herein, we report a patient with unbalanced AVSD and dextrocardia who underwent AVV repair using the “polytetrafluoroethylene (Gore-Tex, W.L. Gore & Associates, Flagstaff, AZ) bridge” technique⁵ with excellent early outcome.

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CLINICAL SUMMARY

An 8-year-old boy (weight 37 kg, body surface area 1.17 m²) with Fontan circulation presented for AVV repair. He was diagnosed at birth with hypoplastic left ventricle, unbalanced AVSD, transposition of great arteries, left atrial isomerism, dextrocardia, pulmonary stenosis, bilateral superior vena cavae, and hemiazygos continuation of the inferior vena cava to the left superior vena cavae. He underwent univentricular palliation with central shunt at 1 month of age, Kawashima operation with bilateral cavopulmonary anastomosis and pulmonary artery banding at 7 months of age, and nonfenestrated extracardiac Fontan operation (Fig. 1A) at 4 years of age. Post-Fontan echocardiography revealed only mild-to-moderate AVV



Repair of atrioventricular valve in dextrocardia: access from the left side of the patient.

Central Message

Competence of atrioventricular valve in univentricular physiology allows better long-term outcomes and can be achieved even in the difficult setting of dextrocardia with appropriate surgical strategy.

regurgitation and normal biventricular function. However, his common AVV became progressively more regurgitant. Echocardiography and cardiac magnetic resonance imaging at 3 years after Fontan operation demonstrated moderate-to-severe AVV regurgitation with a regurgitant fraction of 31%, predominantly due to superior-inferior leaflet malcoaptation with adequate mural leaflet (Fig. 2A). Although the patient remained asymptomatic and had normal ventricular function, we elected to perform AVV repair to prevent ventricular dilatation and failure. Intraoperatively, the common atrium was opened and the AVV was approached from the left side of the patient due to dextrocardia (Fig. 1A). There was significant dilatation of the annulus and central malcoaptation of the leaflets (Fig. 1B). A 36 mm Cosgrove-Edwards annuloplasty ring (Edwards

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CONGENITAL – PARTITION OF COMMON ATRIOVENTRICULAR VALVE IN A PATIENT

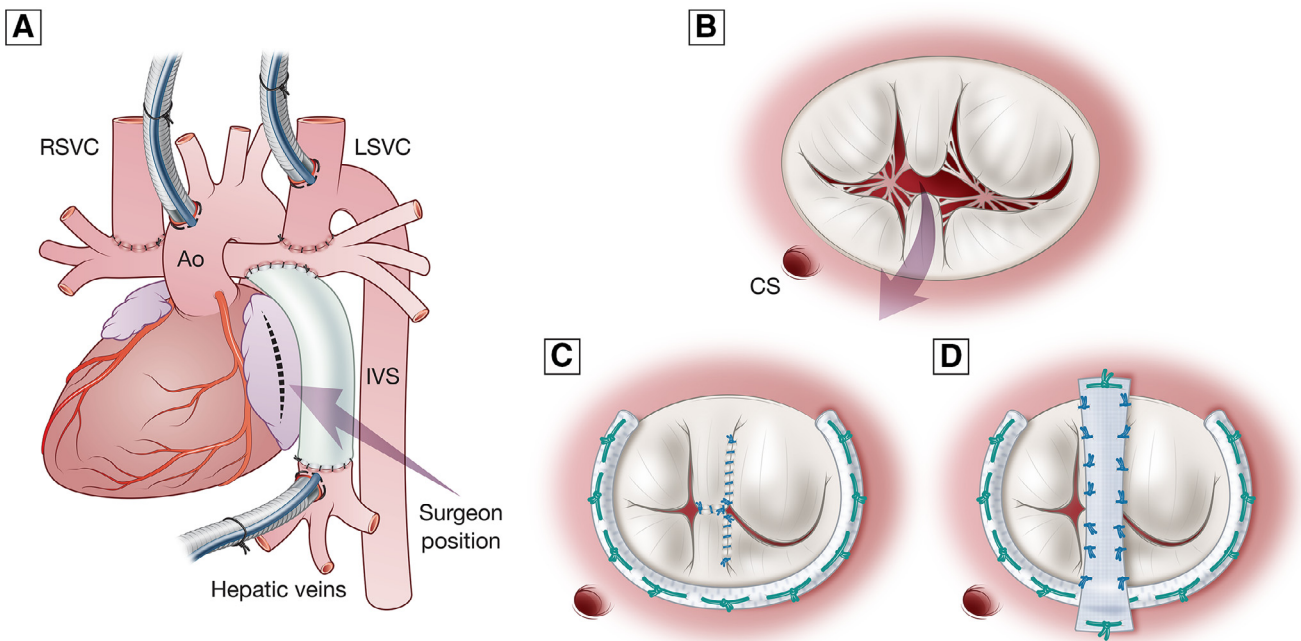


Figure 1. Due to dextrocardia, access to the atrioventricular valve was gained through the atriotomy performed from the left site of the patient (A). Malcoaptation of the bridging leaflets and severe annular dilatation was observed (B). The annulus was reduced with annuloplasty ring. The valve was partitioned by approximation of the bridging leaflets and reinforced with Goretex bridge (C, D). CS, coronary sinus.

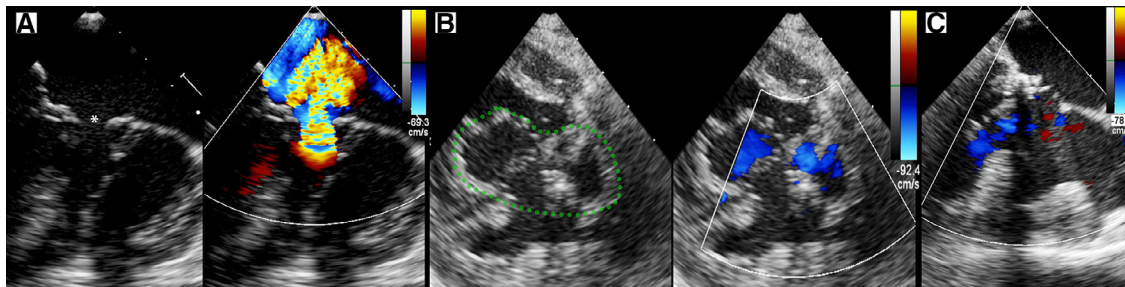


Figure 2. Preoperative echocardiography (A): coaptation defect (*) and broad regurgitation jet on the common atrioventricular valve. Postoperative short axis (B) showing the partitioned atrioventricular valve (green dotted line), with no regurgitation in long axis (C). (Color version of figure is available online.)

Lifesciences, Irvine, CA) was implanted. As a usual strategy to avoid the conduction system, which location is highly unpredictable in this anatomical setting, sutures were placed superficially in the area of the coronary sinus. The clefts were closed and the bridging leaflets were approximated, effectively separating the common valve into mitral and tricuspid components (Fig. 1C). To ensure durability of the repair, a Gore-Tex strip was placed across the annulus and secured with 5-0 Prolene sutures (Ethicon Inc., Somerville, NJ) to the bridging leaflets and with 3-0 Ethibond sutures (Ethicon Inc., Somerville, NJ) to the atrioventricular valve annulus (Fig. 1D). Cardiopulmonary bypass time was 159 minutes. Aortic crossclamp time was 103 minutes. Postoperative course was uneventful. The patient remained in sinus rhythm. Postoperative echocardiography demonstrated partitioned AVV orifices (Fig. 2B) with no AVV regurgitation (Fig. 2C) or stenosis. At 5-month follow-up, the

patient had mild AVV regurgitation and normal ventricular function.

DISCUSSION

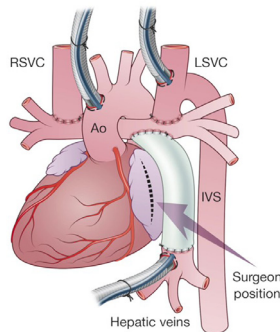
Moderate-to-severe AVV regurgitation is an independent predictor of mortality and reoperation after single-ventricle palliation.³ Patients with single-ventricle physiology and common AVV are at risk of progressive AVV insufficiency after Fontan operation,² as occurred in the patient described herein. We have previously demonstrated that AVV repair is especially difficult in unbalanced AVSD³ and dextrocardia⁴ using conventional techniques. It has now become evident that successful achievement of valve competency via repair or replacement is important for improved survival and freedom from reoperation.³ In light of these findings, we elected to perform a preventive operation on our patient. We applied a “Gore-Tex

bridge” technique to ensure durability, which we have previously described in detail.⁵ This technique was particularly useful in our patient to stabilize the annulus and support the bridging leaflets, given the presence of a central regurgitation related to right ventricular enlargement and restriction of the tricuspid valve component. Although there are no guidelines on the sizing of annuloplasty ring for the repair of common AV valve, we generally apply the same principle as used in mitral valve repair. That is: the reconstructed common anterior leaflet should cover the area of the common AV valve and provide reliable coaptation with the components of the reconstructed posterior leaflets. The technique of AVV repair described herein may be a valuable alternative to valve replacement in challenging patients with univentricular circulation, particularly those with dextrocardia.

SUPPLEMENTARY MATERIAL

The following is the supplementary data to this article:

8-year-old boy
 Unbalanced AVSD with hypoplastic LV
 Transposition of great arteries
 Left atrial isomerism
 Dextrocardia
 Pulmonary stenosis
 Bilateral SVC
 Hemi-azygos continuation of IVC to left SVC
 Central shunt at 1 month of age.
 Kawashima operation with bilateral cavo-pulmonary shunts and PA band at 7 months of age.
 Non-fenestrated extra cardiac Fontan operation at 4 years of age.



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Video S1. Repair of common atrio-ventricular valve after Fontan operation in a child with dextrocardia and unbalanced atrio-ventricular septal defect.