

Simultaneous Atrioventricular Valve Repair Compelling Creation of Unconventional Fenestration During Extracardiac Total Cavopulmonary Connection

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Abstract

Severe regurgitation of the atrio-ventricular valve and borderline pulmonary vascular resistance are risk factors for failure of univentricular repair. We believe that fenestration in the Fontan pathway plays an important role in avoiding postoperative catastrophes-immediate and late, in such high-risk substrates. Rarely, the intra-operative circumstances compel the surgeon to opt for an unconventional method of fenestration.

We report an interesting case of high-risk univentricular repair, had previously undergone pulmonary artery banding and bidirectional Glenn shunt, who now required a simultaneous repair of the regurgitant atrio-ventricular valve compelling the creation of an unconventional fenestration during Stage III palliation.

Keywords: Atrio-ventricular valve regurgitation; Completion fontan procedure; Unconventional fenestration.

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INTRODUCTION

Atrioventricular valve (AVV) regurgitation increases the risk of failure of Fontan circulation in patients with single ventricle congenital cardiac disease.¹⁻⁴ Almost one third of the patients are likely to experience AVV regurgitation beyond the 3rd decade of life.⁵ Adoption of fenestration in Fontan pathway remains controversial with some centres opting for it while others opting out.⁶

We report a patient who underwent stage III palliation with simultaneous repair of AVV, forcing to opt for an unconventional technique of fenestration in Fontan circuit.



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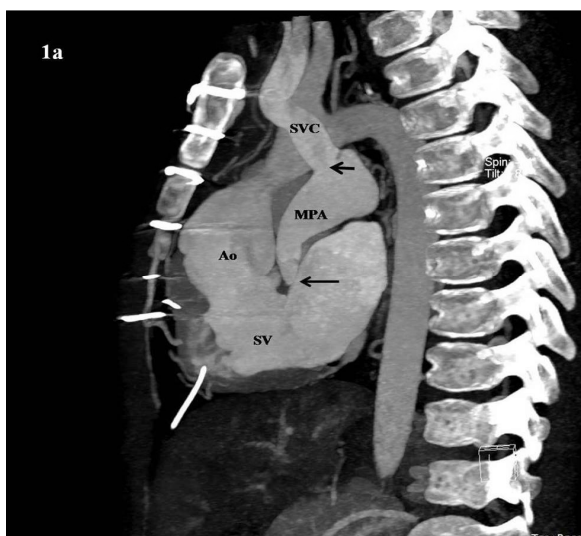
CASE REPORT

A 2-month-old female baby came to us with a well-established diagnosis of single large ventricular septal defect, extending from inlet to the outlet septum, with a rudimentary inter-ventricular septum at the apex, amounting to single ventricle. She had associated severe pulmonary hypertension for which she had undergone pulmonary artery banding as an initial palliation.

At the age of 4 months, she required further tightening of the band for persistent increased

pulmonary blood flow and pulmonary hypertension. At the age of 18 months, she underwent a delayed bidirectional Glenn shunt with atrial septectomy (Stage II palliation) with further tightening of the pulmonary artery band. Trivial right AVV regurgitation was noted during this hospital admission.

The AVV regurgitation gradually increased to a severe grade in the next couple of years mandating intervention. A catheterization study revealed borderline pulmonary vascular resistance. Contrast-enhanced computerized tomographic scan was also done (Fig. 1a).



1a) This is a sagittal view of contrast-enhanced computerized tomography scan showing widely patent superior cavo-pulmonary connection (short arrow) and banded main pulmonary artery (long arrow). One can appreciate the absence of interventricular septum, and the tiny collateral vessels around the heart. Ao Aorta; MPA Main pulmonary artery; SV Single ventricle; SVC Superior vena cava.



1b) This is an intraoperative photograph taken from the head-end of the patient, showing the heart in diastolic cardioplegic arrest and ascending aorta cross-clamped. The right atrium is opened and retracted with stay sutures. The right atrioventricular (AV) valve is being repaired (arrow). Pericardial adhesions can be noted. **1c)** This is another intraoperative photograph taken from the right side of the patient, showing the extracardiac expanded polytetrafluoroethylene conduit anastomosed to the distal open end of the inferior vena cava. Kreutzer's technique of fenestration (arrow) has been performed, wherein the proximal open end of the inferior vena cava is sutured to an opening made in the adjacent part of the extracardiac Fontan conduit.

The baby was taken up for 3rd redo-sternotomy, repair of right AVV and completion Fontan surgery with a fenestration. Standard cardiopulmonary

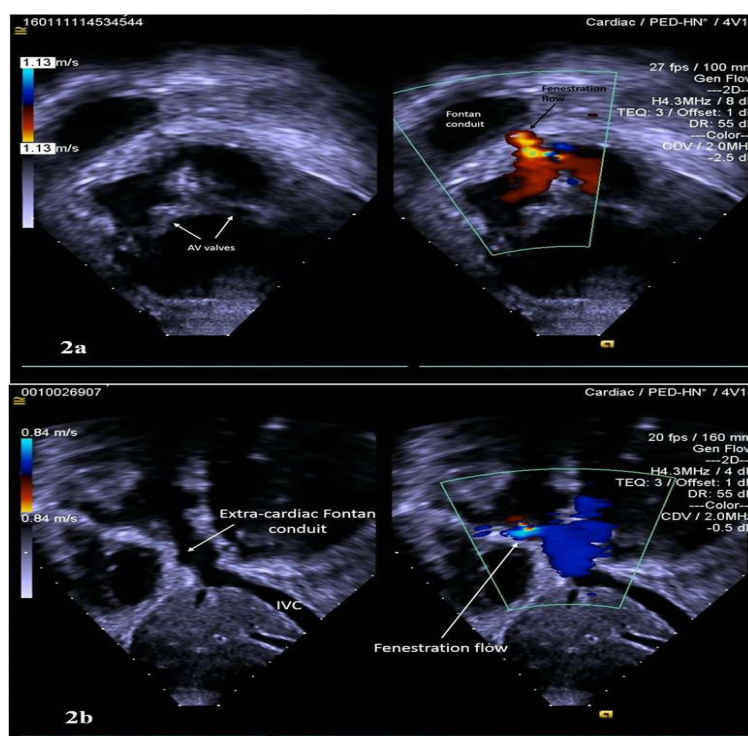
bypass was established and cardioplegic cardiac arrest was achieved. Right atriotomy was performed. A cleft in the anterior leaflet of the right AVV valve

was repaired using interrupted 5-0 polypropylene sutures. Asymmetric purse-string annuloplasty was done using 4-0 polypropylene double circumferential suture, avoiding the area of posterior and septal annulus adjacent to the conduction system (Fig. 1b). The saline injection test revealed satisfactory repair of the right AVV valve. The main pulmonary artery was interrupted after excising the valve cusps. The junction of inferior vena cava and right atrium (IVC-RA) was then divided. The distal open end of the inferior vena cava was anastomosed to an extra cardiac conduit (expanded polytetrafluoroethylene tube graft 18 mm) in standard fashion. The right atriotomy was closed in two layers of continuous polypropylene suture.

Right atrial appendage was significantly thickened (due to previous procedures, atriotomy and scarring) and retracted. In addition to that, the present atriotomy suture line made it unsuitable

for conventional fenestration.⁷ So, we employed Kreutzer's fenestration⁸ at the inferior vena caval end of the right atrium (Fig. 1c). This involved anastomosis of proximal open end of IVC-RA junction to a 4 mm hole made in the adjacent part of the extracardiac conduit. The fenestration was created using 5-0 polypropylene suture continuously. The other end of the extracardiac conduit was trimmed to an adequate length and was anastomosed to an opening made in the undersurface of right pulmonary artery, opposite to the Glenn shunt. The remainder of the procedure was completed in a standard fashion.

Post-operative 2-dimensional echocardiography revealed satisfactory AVV repair, good flow in the extracardiac Fontan conduit and well-functioning fenestration (Fig. 2a, 2b). At her most recent follow-up, the patient remains in NYHA class I, maintaining saturations between 80- 85%.



2a) Postoperative echocardiography (2-dimensional and colour doppler) in short axis view showing well-functioning and competent both atrioventricular (AV) valves and good flow in the Kreutzer's fenestration communicating to the Fontan conduit. The absence of interventricular septum amounting to a single ventricle also can be appreciated in this view. **2b)** Postoperative echocardiography (2-dimensional and colour doppler) in long axis view showing patent extra-cardiac Fontan conduit (short arrow) sutured to the inferior vena cava (IVC) and good flow in the Kreutzer's fenestration (long arrow).

DISCUSSION

Outcomes of extracardiac Fontan procedure have improved over recent times owing to improved surgical techniques and post-operative

intensive care.¹ Adopting fenestration in a Fontan pathway remains an open choice with few centres fenestrating all of them while others not employing it in any of them. Recent studies suggest that there is no survival benefit of fenestration.⁶ Despite it, we selectively perform fenestration in candidates with

higher pulmonary vascular resistance and those with regurgitant AVV.

Different types of fenestration techniques for Fontan pathway have been published in the literature.⁷⁻⁹ The conventional technique⁷ involves creating communication between the extracardiac Fontan conduit and the right atrial appendage, either directly anastomosing margin to margin, or by using an interposition tube graft. Kreutzer's technique of fenestration⁸ involves connecting the open end of proximal IVC stump to an opening made in the adjacent part of the extracardiac Fontan conduit. We have also published a novel technique of fenestration in a different surgical scenario where we created a communication between the left pulmonary artery and the left juxtaposed right atrial appendage, to act as a fenestration.⁹

We routinely employ the conventional technique of fenestration whenever indicated. We have performed Kreutzer's technique of fenestration⁸ in very selective scenarios like left juxtaposed both atrial appendages with no significant atrial mass on the right side. But this case was unique wherein the performance of simultaneous transatrial procedure for right AVV repair, and scarring of the right atrial wall due to previous procedures, deemed the right atrium unsuitable for conventional technique of fenestration.⁷ Our experience of previously performing Kreutzer's technique of fenestration⁸ turned out to be handy for an immediate on-table decision-making.

The timing of repair of regurgitant AVV remains controversial and should be individualized but AVV valve regurgitation is a proven high-risk factor for Fontan failure.¹⁻⁵ In our patient, fenestration was required for the Fontan pathway due to her severely regurgitant AVV valve and borderline pulmonary vascular resistance. We were confident of the repair procedures that we utilized, and valve replacement was not considered. We were also careful about the proximity of the conduction bundle during the valve repair to avoid conduction blocks deleterious for any Fontan circulation.

CONCLUSION

We conclude that employing the alternative technique of Fontan fenestration should be considered and works well in rare situations when conventional fenestration appears difficult to

perform. Simultaneous repair of the regurgitant AVV valve should be considered during the completion of a Fontan pathway.

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