


CASE REPORT

An unexpected circular shunt: Novel method to treat semilunar valve insufficiency in a single ventricle patient on mechanical circulatory support

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Abstract

Severe semilunar valve insufficiency in single ventricle patients supported with mechanical circulatory support while awaiting transplant remains a complex clinical scenario with few favorable options for management. We present the first case, to our knowledge, of transcatheter closure of the pulmonic valve in a patient palliated with a hybrid stage 1 procedure for hypoplastic left heart syndrome.

KEYWORDS

interventional devices/innovation, mechanical circulatory support, percutaneous intervention, pulmonary valve disease, ventricular assist device

1 | INTRODUCTION

Palliated single ventricle heart disease represents a high proportion of the patients treated by interventional pediatric cardiologists. Patients with single ventricle palliation and significant hemodynamic compromise are often supported by some form of mechanical circulatory support (MCS) system such as a ventricular assist device (VAD) or extracorporeal membranous oxygenation as a bridge to transplant or an interval palliative operative procedure.^{1,2} In the setting of a VAD, severe semilunar valve insufficiency (SVI) can create the substrate for a circular shunt (Figure 1). A small case series reported that surgical closure of the systemic semilunar valve is a viable option in children awaiting transplant supported by a VAD; notably, one of those patients underwent unsuccessful transcatheter valve occlusion before surgical valve closure.³ To the best of our knowledge, this is the first report of successful transcatheter closure of the pulmonic valve in a patient with hypoplastic left heart syndrome (HLHS) palliated with a hybrid stage 1 procedure and supported with MCS.

2 | CASE REPORT

The patient was a 3-month-old male born prematurely at 34 weeks and 2 days gestation weighing 2.3 kg with a prenatal diagnosis of HLHS with mitral stenosis and aortic atresia (MS/AA). After delivery, he was found to have severe tricuspid valve regurgitation and a bicuspid pulmonic valve with mild stenosis and regurgitation, consistent with findings on fetal echocardiography. There was also a concern for coronary cameral fistulae with the bidirectional flow in the left coronary system and the patient had developed progressive hydrops with bilateral pleural effusions, a circumferential pericardial effusion and abdominal ascites. Given the guarded prognosis, the patient was discussed at our multidisciplinary surgical conference and the decision was made to proceed with a hybrid stage 1 and VAD implantation as a bridge to transplantation. His initial intervention included an operative atrial septectomy, Frater stitch pulmonary valve repair, bilateral pulmonary artery bands, and PediMag[®] VAD placement with a 16 Fr venous cannula in the right atrium and 10 Fr

Abbreviations: CICU, cardiac intensive care unit; HLHS, hypoplastic left heart syndrome; MCS, mechanical circulatory support; MS/AA, mitral stenosis/aortic atresia; SVI, semilunar valve insufficiency; VAD, ventricular assist device.

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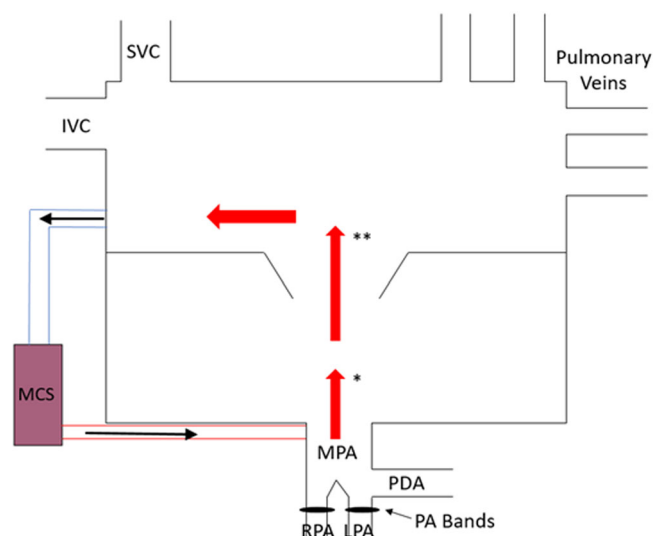


FIGURE 1 Schematic representation of circular shunt in single ventricle palliation treated with mechanical circulatory support. IVC, inferior vena cava; LPA, left pulmonary artery; MCS, mechanical circulatory support; MPA, main pulmonary artery; PDA, patent ductus arteriosus; RPA, right pulmonary artery; SVC, superior vena cava. *Semilunar valve regurgitation. **AV valve regurgitation. [Color figure can be viewed at wileyonlinelibrary.com]

arterial cannula in the main pulmonary artery done without cardiopulmonary bypass. The patient was transferred directly to the catheterization laboratory for patent ductus arteriosus (PDA) stent placement via direct MPA access and placement of an 8 × 20 mm EV3 Protégé (Medtronic) self-expanding stent; this hybrid procedure was technically successful. Postprocedure, he had persistent mild pulmonary regurgitation and was listed status 1A for a heart transplant while well supported in the cardiac intensive care unit (CICU). The patient was extubated and more than doubled his weight to 4.4 kg. Three months later he had an acute decompensation resulting in poor cardiac output and hypotension despite high VAD output with a follow-up echocardiogram revealing severe pulmonary regurgitation prompting discussion for pulmonic (systemic semilunar) valve closure. After a multidisciplinary discussion, the group consensus was that a surgical approach was a very high risk given severe end-organ dysfunction, so the decision was made to attempt semilunar valve closure in the catheterization laboratory.

The patient was brought to the catheterization laboratory where RV angiography confirmed severe tricuspid and pulmonary regurgitation. The pulmonic valve was crossed using a 4F JR 2 Super Torque® (Cordis) catheter and an 0.018" Hi-Torque (Abbott Cardiovascular) wire initially, exchanged for an 0.035" Rosen (Cook Medical) wire to then allow for sheath exchange to a 6F 180° Amplatzer™ Torque-View™ (Abbott Cardiovascular) delivery system. The delivery system was carefully advanced across the stented PDA and positioned in the abdominal aorta. Next, a 10 mm Amplatzer™ Septal Occluder (Abbott Cardiovascular) was selected and prepared as per the manufacturer's guidelines, loaded into the delivery system, and advanced to the position of the pulmonic valve. A 10 mm

Amplatzer™ Septal Occluder (Abbott Cardiovascular) was felt to be the best device to completely occlude the pulmonic valve with a central waist of 10 mm given a pulmonic valve annulus measurement of 9 mm. Also, its short length of only 3 mm provided optimal pulmonic valve closure without significant device protrusion into the right ventricle and potential compromise of tricuspid valve function. Unfortunately, the patient's size was also self-limiting in our ability to place a transcatheter valve, which would have required a minimum 18 F delivery system. After careful manipulation, the device was deployed across the pulmonic valve, and once confirmed in appropriate position, it was released and remained in a stable position. The appropriate device position was confirmed angiographically and by echocardiography, which revealed the pulmonary root disk cupping or concave in the root with the central portion of the device spanning the pulmonic annulus and there was no residual flow through the device. The RA disk was in the body of the right ventricle and there was no evidence of impaired tricuspid valve function on echocardiography. The postdeployment assessment demonstrated minimal residual pulmonary regurgitation on the transthoracic echocardiogram (Figure 2).

Follow-up echocardiography the next day revealed no significant pulmonary regurgitation. There was a positive response to the procedure with downtrending lactate from >20 to 7.60 mmol/L, aspartate transaminase from 8875 to 5638 units/L, alanine aminotransferase from 3238 to 1503 units/L, and alkaline phosphatase from 465 to 322 units/L. Despite the successful treatment of the circular shunt, the patient had a persistent lactic acidosis, albeit improved, with low mixed venous saturation, anuria, and significant hepatitis. Over the following days, with little improvement in end-organ function, it was felt that the patient would never become a transplant candidate again and the family made the decision to withdraw life-sustaining support and the patient died peacefully.

3 | DISCUSSION

In the setting of donor shortages and prolonged wait times for patients listed for transplant, MCS has become the standard of care to support patients while awaiting transplant.² Progressive SVI in the setting of MCS for patients with palliated single ventricle heart disease awaiting transplant presents an interesting situation in which the semilunar valve can be safely sacrificed with device closure. It is important to note that eliminating forward flow with semilunar valve closure eliminates the possibility of ventricular recovery and any further single ventricle palliation, thus committing the patient to a transplant as definitive management. This also makes exchange of the pump, which is done periodically when bridging a patient to transplant with an extracorporeal continuous flow VAD, quite hazardous since there will be no cardiac output. Further, with no forward flow from the ventricle, should there be any sort of VAD failure, the patient would not survive. Given these considerations, the adult population has trended away from semilunar valve closure and now opts for valve replacements in patients with continuous-flow left

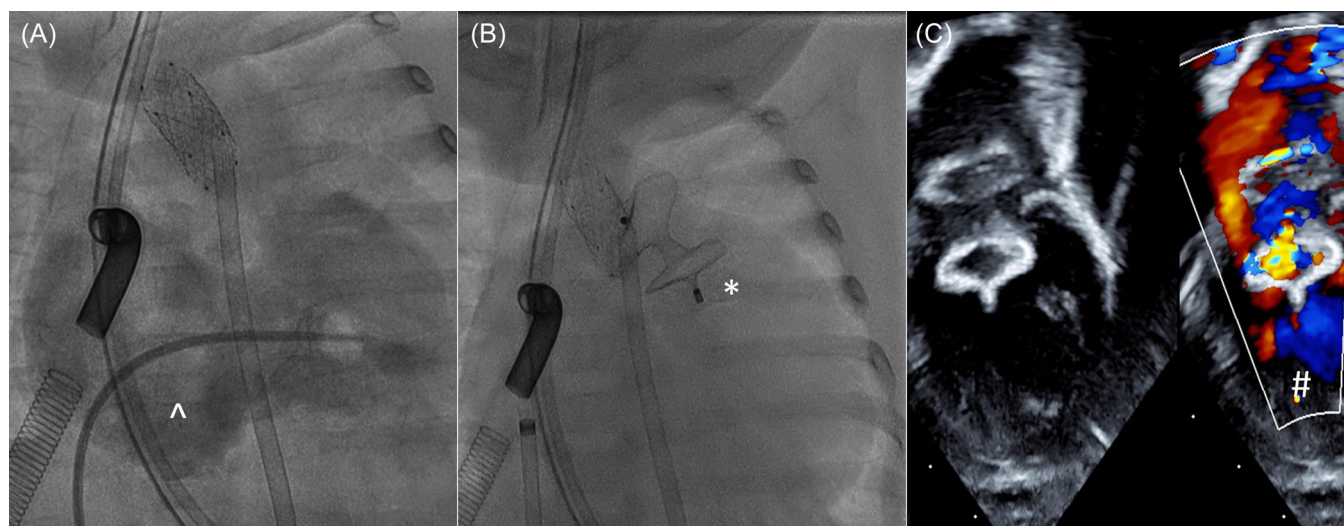


FIGURE 2 Fluoroscopic images in the anteroposterior projection showing (A) severe tricuspid valve regurgitation with a dilated right atrium (^); (B) after deployment of the 10 mm Amplatzer™ Septal Occluder (*) (Abbott Cardiovascular); (C) echocardiogram post device deployment showing stable device position with no significant residual pulmonary regurgitation (#). [Color figure can be viewed at wileyonlinelibrary.com]

ventricular assist devices (LVADs).⁴ Unfortunately, given our patient's size, a transcatheter valve replacement was not possible and the patient was felt to be too critically ill to survive an operative procedure on cardiopulmonary bypass. With these considerations in mind, the decision to permanently occlude the systemic semilunar valve should be made in concert with the family and advanced cardiac therapies team.

Previous reports have shown successful closure of the aortic valve in single ventricle disease palliated with a Norwood stage 1 and aortic insufficiency while maintaining neo-aortic valve outflow.^{5,6} Further, transcatheter closure of the aortic valve has been used as a method to ameliorate the effects of aortic insufficiency in adult patients with continuous-flow LVADs.⁷ We present the first case of transcatheter closure of the pulmonic valve in a patient with HLHS treated with a hybrid stage 1 procedure who developed an iatrogenic circular shunt with both severe SVI and atrioventricular valve insufficiency. We have shown that the technical approach to eliminating SVI in the catheterization lab can be performed safely and effectively. In our case, although technically successful, we did not achieve a favorable outcome given the severity of the patient's multiorgan failure. In the future, when caring for patients in a similarly dire situation, the decision to close the semilunar valve could be considered earlier in the clinical course so as to hopefully achieve the benefits of augmented cardiac output before irreversible damage had been done.

4 | CONCLUSIONS

Single ventricle patients treated with MCS as a bridge to transplant with progressive SVI can be effectively managed with semilunar valve closure in the catheterization laboratory. The decision to intervene

on SVI should be made early to prevent the progressive and irreversible effects of multiorgan failure.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

No analytic data were included in this case report. Angiography and hemodynamic data can be provided.

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