Unusual method of creation of a transcatheter fenestration in an extracardiac conduit Fontan circulation

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ABSTRACT

Failing Fontan physiology in univentricular hearts manifest with protein-losing enteropathy, plastic bronchitis, low cardiac output, and recurrent effusions. Transcatheter creation of fenestration in a failing Fontan may be useful in alleviating the symptoms by improving the cardiac output. It is traditionally achieved by puncturing through the conduit from femoral or jugular venous access. In the absence of good venous path, transhepatic access provides a direct route for needle puncture of the conduit. If marked intimal ingrowth into the conduit results in increasing rigidity and makes the conduit nonyielding, alternative approaches may be needed. A successful creation of pulmonary artery to atrial roof communication through the potential space of transverse sinus using a covered stent is presented.

Keywords: Failing Fontan, protein-losing enteropathy, stent fenestration

INTRODUCTION

Although Fontan completion offers effective palliation in most patients with univentricular hearts by improving oxygenation and growth, the resultant high venous pressures may lead to peculiar problems including protein-losing enteropathy, plastic bronchitis, and persistent effusions in some patients. The acutely failing Fontan circulation requires urgent management to decompress the systemic venous pressures and augment cardiac output. Surgical takedown may involve the use of extracorporeal membrane oxygenation support to preserve life. Catheter-based effective Fontan takedown is an alternative to surgery in these patients, who are acutely unstable, and offers the potential for an adjustable fenestration.[1] Creation of fenestration through an extracardiac prosthetic conduit such as Dacron or polytetrafluoroethylene Dacron (VASCUTEK, a TERUMO Company. Scotland, UK) may be technically challenging because of the angulation from femoral and jugular venous access.[2] We report a novel method of stenting from pulmonary artery directly into the atrium through transverse sinus of the heart, a potential space in the setting of a failing Fontan circulation.

CASE REPORT

A 14-year-old girl weighing 55 kg was diagnosed to have d-transposition of great arteries, hypoplastic left ventricle, large ventricular septal defect, and severe pulmonary stenosis; she was initially palliated with bilateral bidirectional nonpulsatile Glenn shunt and atrial septectomy at 4 years of age. Three years later, her cardiac catheterization showed pulmonary artery pressure of 11 mmHg and ventricular end-diastolic pressure of 8 mmHg. She underwent extracardiac conduit Fontan completion with 18-mm Dacron tube graft. Five years after her Fontan surgery, she had exertional dyspnea,

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pedal edema, and facial puffiness. Her oxygen saturation was 96%. Her serum protein and albumin levels were 3.8 and 1.8 g/dl, respectively, but the liver enzymes were not elevated. Although a fecal α-1 antitrypsin assay was not done, the clinical and other laboratory parameters suggested a diagnosis of protein-losing enteropathy.

During cardiac catheterization, the pulmonary artery pressures were 16 mmHg and inferior vena caval (IVC) pressure was 18 mmHg. The conduit was uniformly narrowed and the gradient was 2 mmHg [Figure 1, Movie 1]. A computed tomogram with contrast showed marked thickness of the conduit indicative of diffuse tissue intimal ingrowth with a thickness of 6-8 mm. After discussions with surgical team about morbidity involved in surgical revision of conduit in a patient with systemic right ventricle, transcatheter approach was adopted after informed consent from the patient. The conduit was stented with Palmaz 4014–18-mm MaxiLD balloon combination (Cordis Corporation, Miami, Florida, USA) and postdilated at 16 atmospheres with 18-mm Atlas balloon (Bard PV, Tempe, Arizona, USA) [Figure 2, Movie 2]. As the stenosis was confined to the conduit because of tissue ingrowth, the stent was deployed avoiding any protrusion into the pulmonary arteries superiorly. This reduced the IVC pressure to 15 mmHg. An attempt to fenestrate the conduit through the side struts of the stent into the atrium to reduce the Fontan circuit pressure was not successful in view of very tough nature of the conduit. She transiently improved with diuretic and warfarin with improvement in her serum albumin levels, but minimal edema persisted.

Six months after the procedure, she reported back with increasing symptoms and declining serum proteins. In view of reappearance of symptoms, it was planned to fenestrate the Fontan circuit. All attempts from femoral venous access to perforate the conduit vein, by increasing the curvature of the Brockenbrough needle (Medtronic INC. Danvers MA) to improve the contact of the needle tip on the stented conduit, adding electrocautery to the transseptal needle to improve the efficiency of the needle, and approach from right jugular vein to alter the angle between the needle tip and the conduit, failed because of enormous thickness of the conduit, previously assessed by the tomographic scan. It was decided to perforate the undersurface of the right pulmonary artery to enter the potential space of transverse sinus of the heart and then puncture through the roof of the atrium to gain an access from the pulmonary artery to the atrium.

A right pulmonary arteriogram in anterior–posterior and lateral views through jugular access delineated the anatomy of right pulmonary artery and atrium in the levophase, and indicated the thickness of the potential space. A 6-French Mullin sheath tip (Cook Medical, Bloomington, Indiana, USA) with Brockenbrough needle was positioned in the right pulmonary artery and a sustained pressure was applied on the needle to perforate into the atria. Once the needle tip entered the atria as confirmed by give way of the needle and contrast injection from the needle tip, an extra support 0.014″ coronary guidewire was advanced into the atria to avoid inadvertent damage to the atrioventricular valve and other intracardiac structures from the needle tip. A further sustained pressure over the needle enabled the dilator and subsequently the sheath to be tracked from the right pulmonary artery into the atrium, thereby stitching the wall of the pulmonary artery to the atria by the snugly fitting Cook sheath. This ensured that there is no bleeding into the potential space around the puncture. A premounted 7 mm × 21 mm covered Advanta V12 stent (Atrium Medical, Hudson, New Hampshire, USA) was taken across the Mullin sheath and positioned across the new fenestration, guided by angiogram and dilated to 14 atmospheres [Figure 3, Movie 3].

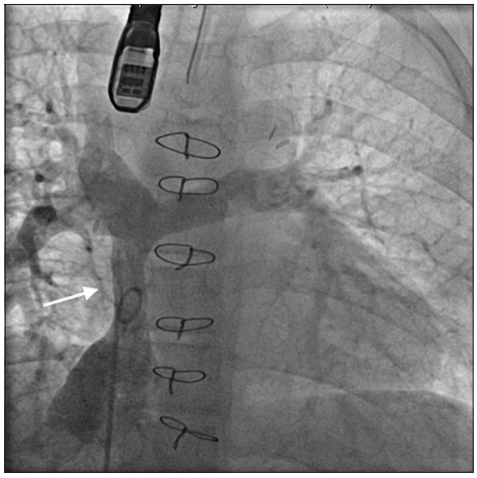

Figure 1: Angiogram of Fontan circuit in posteroanterior view showing the uniformly narrowed conduit (arrow). (See also Movie 1.)

Figure 2: Angiogram in posteroanterior view showing good flow through the well open stented conduit. (See also Movie 2.)

Postprocedure angiogram [Figure 4, Movie 4] showed good flow through this fenestration from the pulmonary artery to the atrium that was confirmed in the echocardiogram [Figures 5 and 6, Movies 5 and 6]. The pressure in the Fontan circuit reduced from 15 to 12 mmHg. The transpulmonary gradient reduced from 7 to 4 mmHg following the fenestration creation. The oxygen saturation dropped from 96% to 80% postprocedure. The diuretic dose reduced and the patient was discharged on warfarin and aspirin. Sildenafil was added to favorably influence her pulmonary vascular resistance. The reduction in edema reduced her weight to 51.5 kg at discharge. At 12 months of followup, she is marginally less symptomatic of edema, but still has low albumin levels.

DISCUSSION

Failing Fontan circulation clinically present with protein-losing enteropathy, plastic bronchitis, or persistent effusions. Fenestration in the circuit decompresses the elevated venous pressure and improves the cardiac output, at the cost of systemic hypoxia.[3,4] As surgical creation of fenestration in Fontan circuit is more morbid, it is often done in the catheterization laboratory using Brokenbrough needle puncture of the conduit followed by stent placement.[5-7] If the angulation of the conduit from femoral venous route is unfavorable, or if there is lack of femoral venous access as in venous occlusion or left isomerism, transhepatic route gives a more perpendicular access to the conduit for a needle puncture.[8,9] In our patient, puncture was difficult

Figure 3: Angiogram in posteroanterior view showing position of the partially inflated premounted 7 mm × 21 mm covered Advanta V12 stent across right pulmonary artery to atrium fenestration (arrow). (See also Movie 3.)

Figure 4: Angiogram in posteroanterior view showing good flow across right pulmonary artery to atrium stent fenestration (arrow). (See also Movie 4.)

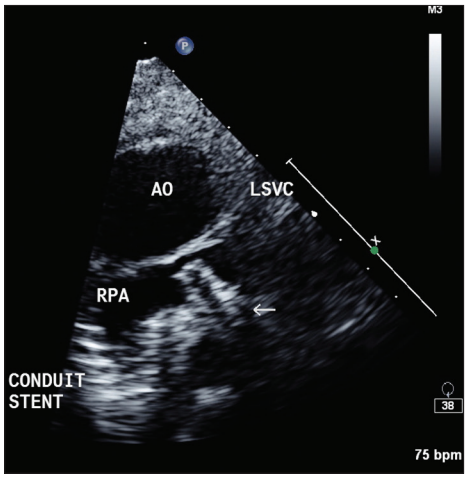

Figure 5: Two-dimensional echocardiogram in suprasternal view showing good stent position across the right pulmonary artery to atrium fenestration (arrow). (See also Movie 5.) AO: Aorta, LSVC: Left superior vena cava, RPA: Right pulmonary artery

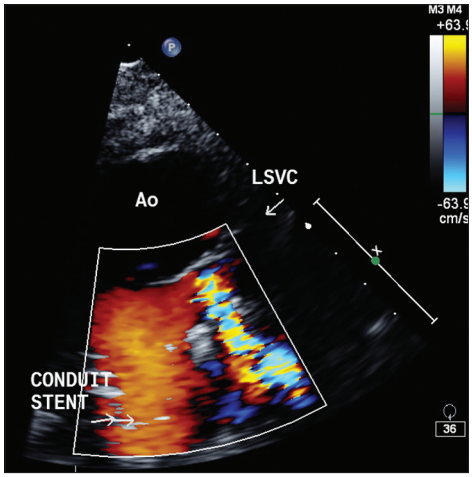

Figure 6: Two-dimensional echocardiogram with color Doppler in suprasternal view showing good flow across the right pulmonary artery to atrium fenestration (single arrow). Good flow through the conduit stent is also seen (double arrow). (See also Movie 6.) AO: Aorta, LSVC: Left superior vena cava

because of tough ingrowth within the Dacron conduit as seen on the tomographic scans.

In patients with no previous surgeries, if a transseptal needle punctures through the roof of the atria, it often leads to significant pericardial effusions because of the lack of adhesions in the potential space. Fenestrations through the extracardiac conduit also courses through this potential space between the conduit and the atrial wall, but this space is often obliterated by fibrosis because of surgical adhesions. Transverse sinus of the heart is another potential pericardial space between the pulmonary artery superiorly and atrial roof inferiorly. When patients with univentricular hearts are palliated initially with bidirectional Glenn shunts and subsequently with Fontan surgery, the right pulmonary artery is repeatedly dissected around its circumference before anastomosis with the venous system. This dissection promotes adhesions and may lead to partial obliteration of the potential pericardial space. This improves the safety of puncturing through the walls of the pulmonary artery and stitching it through the atrial wall with the Brockenbrough needle–Cook sheath assembly.[2]

The fenestration that is created improves the systemic cardiac output in all patients, but reduces the venous pressures to variable extent depending on the ventricular end diastolic pressures and pulmonary vascular resistance.[10] The success rate of this procedure to obtain durable palliation varies in different series.[11] Some patients may have partial resolution of symptoms and may require Fontan takedown or cardiac transplant. The fenestration in the latter cases acted as a bridge to the transplant.[3] New methods of fenestrations using radiofrequency energy may facilitate conventional transfemoral or transjugular fenestration of the conduit, but this technique should be handy in extreme situations of failing Fontan circulation in which such hardware is not available.[11] There is a complex interplay of factors on the Fontan circulation including pulmonary arterial anatomy and arborization, segmental lung function, pulmonary vasculature, ventricular systolic and diastolic function, and systemic vascular resistance. So fenestration of Fontan circuit should be considered as only one arm of a multimodal comprehensive management in a failing Fontan physiology.[12]

CONCLUSION

Creation of fenestration in a failing Fontan physiology transiently alleviates the symptoms and improves the cardiac output in all patients. This may be technically difficult if femoral or jugular venous access gives an unfavorable angle to fenestrate the conduit with a needle. If femoral venous access is unavailable as in venous occlusions or left isomerism, alternative access including transhepatic access can be considered. Creation of a fenestration through the potential space in the transverse sinus of the heart between the right pulmonary artery to the atrial roof is a reliable alternative in such a failing Fontan physiology. These patients, however, require close followup with multimodal management strategies for long-term outcome.

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Conflicts of interest

There are no conflicts of interest.

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