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Successful Fenestration of an Extracardiac Conduit in a Fontan Patient With the Baylis NRG RF Transeptal Needle and Creation of Fontan Fenestration with a 20-mm 535 Formula Stent

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A 16-year-old male had a history of hypoplastic left-heart syndrome and previous Fontan completion with a lateral tunnel at the age of 4 years. He developed a recurrent plastic bronchitis on optimal medical therapy. He was treated with oral prednisolone, nebulized DNase, and beclomethasone with regular bronchoscopic washes on top of lisinopril, sildenafil, and aspirin. Cardiac magnetic resonance imaging (Figure 1a and b) showed no evidence of obstruction in the pulmonary arteries, absence of Fontan thrombus, mild right atrioventricular valve regurgitation, mild neo-aortic valve regurgitation, mild single ventricular dysfunction, and no outflow obstructions. This ruled out etiologies that could be optimized with interventional treatment, and he was on full possible medical therapy. Patient's Fontan pressure was elevated from a diagnostic catheterization at 16 mmHg. After interdisciplinary team discussions with congenital surgeons and cardiologists, it was decided to attempt creation of a fenestration, to optimize systemic output and reduce venous congestion.[1,2]

In this patient with a prosthetic Fontan tunnel, puncturing would involve using one of the conventional transeptal needles made of stainless steel. With the rigid nature of the prosthetic material comprising the Fontan tunnel, puncturing would require more aggressive force than when performing a transeptal puncture. This requirement for additional force leads to higher risk of perforation of atria or pulmonary veins. By using radiofrequency (RF) needles, we can perforate the prosthesis with less force and enhanced control. Hence, we describe a case of creating a fenestration in the Fontan conduit using an NRG RF transeptal needle (NRG-E-HF-71-C0; Baylis Medical Inc, Quebec, Canada) followed by implantation of a 20-mm 535 Formula stent premounted on a 10-mm balloon (Cook Medical, Bloomington, Indiana).

Under general anesthetic, a 5F sheath was introduced in the right femoral vein. Using a 5F pigtail catheter, pressure in the inferior vena cava was measured at 16 mmHg. Angiogram showed a patent conduit with confluent branch pulmonary arteries. The conduit curved around the right atrium before inserting into the right pulmonary artery (Figure 1c and d). The femoral vein sheath was upsized to an 8.5F 63-cm FastCath (St. Jude Medical, Saint Paul, Minnesota) and advanced into the conduit. An NRG RF transeptal needle with a C-0 curve (Figure 1) was inserted through the dilator and sheath to the tip. Under transesophageal echo guidance, 3 1-second currents were applied while pushing forward gently. The needle advanced with minimum pressure to the right atrium. This was confirmed with a contrast injection through the radiofrequency needle sheath (Figure 1e and f). A 5F multipurpose A1 catheter was introduced through the sheath into the left upper pulmonary vein. Next, a 1-cm-tip 0.035-inch Amplatz Super Stiff Wire (Abbott, Abbott Park, Illinois) was introduced into the left upper pulmonary vein. Next, a 10 × 20-mm 535 Formula stent was introduced through the sheath with the first 2 strands exposed. Balloon was inflated to create a distal cone, and then the stent was pulled back to the fenestration. Stent was then uncovered, and hand injection through the sheath showed it to be across the fenestration to the lumen of the conduit. Inflation of the stent was done until the central portion reached around 4 mm

Abbreviation: RF, radiofrequency.

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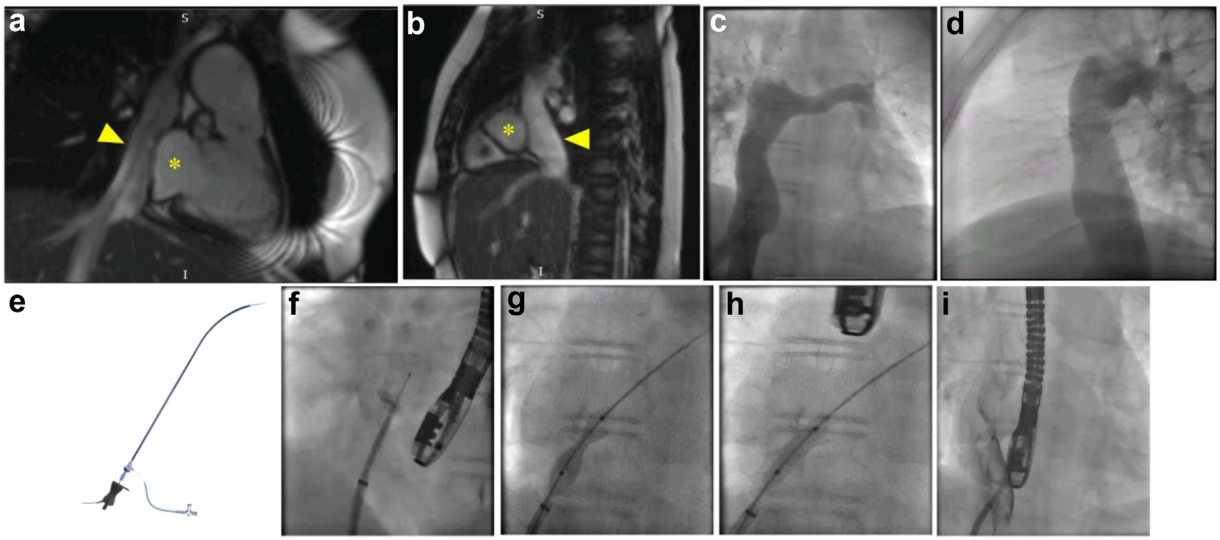

Figure 1. Fontan fenestration creation and stenting. (a) Modified coronal view of the Fontan conduit (arrowhead) connected to inferior vena cava seen adjacent to the common atrium (asterisk) on magnetic resonance imaging. (b) Sagittal view of (a). (c) Patent lateral conduit draining into right pulmonary artery (posteroanterior view). (d) Lateral view of (c). (e) An example of the NRG radiofrequency transeptal needle with a C-0 curve. The radiopaque tip allows fluoroscopic visualization. The overlying needle sheath (TorFlex) has curve availabilities from 45° to 90°, introduced through a 8.5F introducer sheath. (f) Contrast injection in the right atrium through fenestration. (g) Stent delivered through the TorFlex delivery sheath over an Amplatz Super Stiff guidewire. (h) Postdilatation of stent with 6 mm × 2-cm POWERFLEX balloon at 14 atm. (i) Contrast passing through the stent into the RA. Image 1e provided courtesy of Boston Scientific. ©2022 Boston Scientific Corporation or its affiliates. All rights reserved.

(Figure 1g). A 2.5 × 3-cm PTS-X Balloon (NuMed, Boca Raton, Florida) was then introduced and inflated to flare the distal and proximal ends. Thereafter, a 6 mm × 2-cm POWERFLEX Balloon (Cardinal Health, Dublin, Ohio) was used, inflated to 14 atm (Figure 1h). The final angiogram showed a well-seated stent with flaring of both ends and a fenestration lumen measuring around 5.5 mm with transesophageal echo and fluoroscopy (Figure 1i).

At 18-month follow-up, there was no further recurrence of the plastic bronchitis. The patient was successfully weaned off his oral and inhaled steroids over time.

For a prosthetic Fontan conduit, creating a fenestration and stenting can be facilitated by a combination of RF needle puncture followed by placement of a balloon expandable stent.

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