JACC: CASE REPORTS VOL. 29, 2024
© 2024 THE AUTHORS. PUBLISHED BY ELSEVIER ON BEHALF OF THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION. THIS IS AN OPEN ACCESS ARTICLE UNDER THE CC BY-NC-ND LICENSE (http://creativecommons.org/licenses/by-nc-nd/4.0/).

CONGENITAL HEART DISEASE

CASE REPORT: CLINICAL CASE SERIES

Catheter-Based Fontan Completion Without Prior Surgical Preparation

Is This the Way Forward?

Muthukumaran Chinnasamy Sivaprakasam, CCST,[a] Rajaguru Ganesan, MD, DRNB,[a] Ramkishore S, DNB, DRNB,[a] Saranya A, MD,[a] Haritha Girija, MD,[a] Arun V, MD,[a] Mahitha V, DNB,[a] Neville Solomon, MS, MCH[b]

ABSTRACT

We report 3 cases of transcatheter Fontan completion in conventional Glenn anastomosis patients without surgical preparation by creating a tunnel between the pulmonary artery and right atrium. Cavocaval anastomosis is established by placing a covered stent graft within the scaffold created with the uncovered stents at the pulmonary artery–right atrium and inferior vena cava–right atrium junction. (JACC Case Rep. 2024;29:102657) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Prior surgical preparation followed by hybrid transcatheter Fontan completion has been described in the literature.[1-3] We describe the technique of transcatheter Fontan completion in patients without surgical preconditioning and their outcomes along with patient selection criteria.

PATIENT SELECTION AND PREPROCEDURE IMAGING

We undertook transcatheter Fontan in single-ventricle patients with conventional Glenn anastomosis. Echocardiogram was performed to assess the ventricular function, valvular regurgitation, and flow in the Glenn anastomosis and branch pulmonary artery (PA). Preprocedure computed tomographic pulmonary angiogram (CTPA) was done to assess the PA anatomy, proximity of the right PA (RPA) to the right atrium (RA) roof, alignment of the PA and RA roof, hepatic venous drainage along with suprahepatic inferior vena cava (IVC) diameter, and length from the PA end to the IVC-RA junction. We excluded patients with

- nonalignment of the RPA with the RA roof with a distance of >5 mm between them,
- one and a half ventricle repair with Glenn anastomosis,
- weight of <15 kg,
- extensive pulmonary-arteriovenous malformations,
- heterotaxy syndrome with an interrupted IVC, and
- hepatic veins draining directly into the atrium.

Cardiac catheterization was performed before the procedure to assess Glenn anastomosis pressure and

TAKE-HOME MESSAGE

- Transcatheter Fontan in unprepared patients is a feasible option in selected cases with proper preprocedure evaluation.

From the [a]Department of Pediatric Cardiology, Apollo Children's Hospital, Chennai, Tamilnadu, India; and the [b]Department of Cardiothoracic Surgery, Apollo Children's Hospital, Chennai, Tamilnadu, India.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received May 23, 2024; revised manuscript received July 19, 2024, accepted August 15, 2024.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2024.102657

ABBREVIATIONS AND ACRONYMS

CTPA = computed tomographic pulmonary angiogram

IVC = inferior vena cava

PA = pulmonary artery

RA = right atrium

RIJV = right internal jugular vein

RPA = right pulmonary artery

left ventricular end-diastolic pressure. Angiograms were done in the Glenn anastomosis, IVC, and systemic ventricle to assess hepatic venous drainage and branch PA anatomy, to rule out pulmonary-arteriovenous malformations, and to check forward flow across the PA.

PROCEDURE

All patients had undergone their first palliative surgery (Glenn surgery) with forward flow occlusion before the procedure. After institutional Ethics Committee approval and consent from the parents, the procedure was performed as a palliation after explaining the risk and benefits. The procedure was carried out under general anesthesia after intravenous antibiotic administration (cefuroxime 25 mg/kg) using bilateral femoral venous and right internal jugular venous (RIJV) access with 6-F sheaths. The details regarding the patient characteristics, preprocedure anatomy, and hemodynamics are described in Table 1.

Catheter-based Fontan creation involves 3 steps.

STEP 1: ESTABLISHING A CONNECTION BETWEEN THE RA AND PA. A schematic diagram of catheter-based Fontan and its modification in our patients is shown in Figure 1. The first step was to create communication between the PA and RA. In patient 1, we used an 8-F Swartz transseptal guiding introducer (St. Jude Medical) along with the dilator. Hand angiograms were done to check the tenting of the PA over the roof of the RA. After confirmation in the frontal and lateral planes, the PA was perforated using a transseptal puncture needle, and the RA was entered, as both were adherent to each other. A tunnel was created connecting the PA and RA by using a 0.0250-inch guidewire and dilator through the Swartz introducer. The complete step-by-step process of creating a tunnel between the PA and RA roof is described in detail in our first case report.[4] In patients 2 and 3, we changed our strategy for creating communication between the PA and RA after analyzing the CTPA and cardiac catheterization images, as there was a gap between the RPA and RA roof, as shown in Figures 2 and 3. Through the right femoral vein, a tent was made in the roof of the RA close to the RPA with the 8-F Swartz transseptal guiding introducer along with the dilator. The hard end of a 0.014-inch Asahi Gaia third wire (Asahi Intecc Ltd) was tented against the RA roof, and using cautery, we perforated the RA roof and entered the PA using the wire, which was exteriorized through the RIJV route using a guiding catheter and snare. Using the

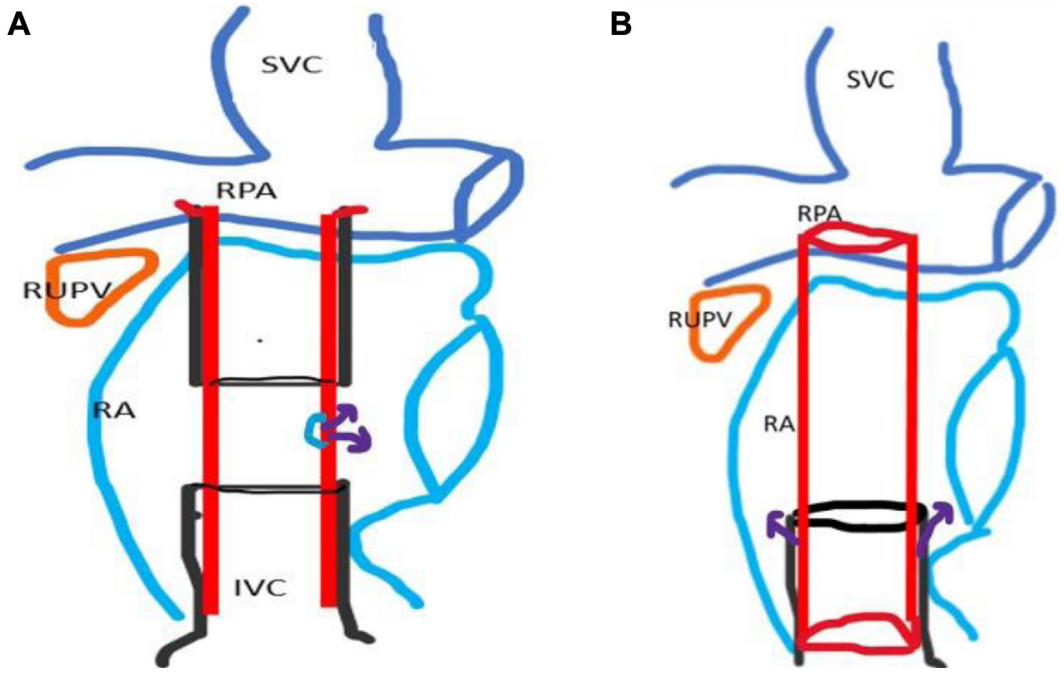
TABLE 1 Patient Characteristics and Preprocedure Details

	Patient 1	Patient 2	Patient 3
Age, y	7	7	12
Sex	Female	Female	Female
Weight, kg	19	18.5	21
Height, cm	120 cm	127 cm	135 cm
Primary diagnosis	DORV-VSD/TGA type with hypoplastic RV with PS	DORV–inlet VSD with hypoplastic RV with severe PS	DORV–inlet VSD and borderline RV with D-malposed great arteries with severe PS
Preprocedure			
Saturation, %	70	75	72
Glenn pressure, mm Hg	18	15	13
LVEDP	13	12	10
Distance between RPA and RA roof, mm	PA adherent to RA	1.5	2.5
Suprarenal IVC diameter, mm	15.3	15	14.8
IVC diameter near the RA junction, mm	18	18	21
Intercaval axis longitudinal length,[a] mm	48	49	60
Prior transcatheter intervention	LPA stenting	None	Forward flow occlusion of the MPA

[a]The intercaval axis is the distance between the RPA and the IVC-to-RA junction.

DORV = double-outlet right ventricle; IVC = inferior vena cava; LPA = left pulmonary artery; LVEDP = left ventricular end-diastolic pressure; MPA = main pulmonary artery; PA = pulmonary artery; PS = pulmonary stenosis; RA = right atrium; RPA = right pulmonary artery; RV = right ventricle; TGA = transposition of the great arteries; VSD = ventricular septal defect.

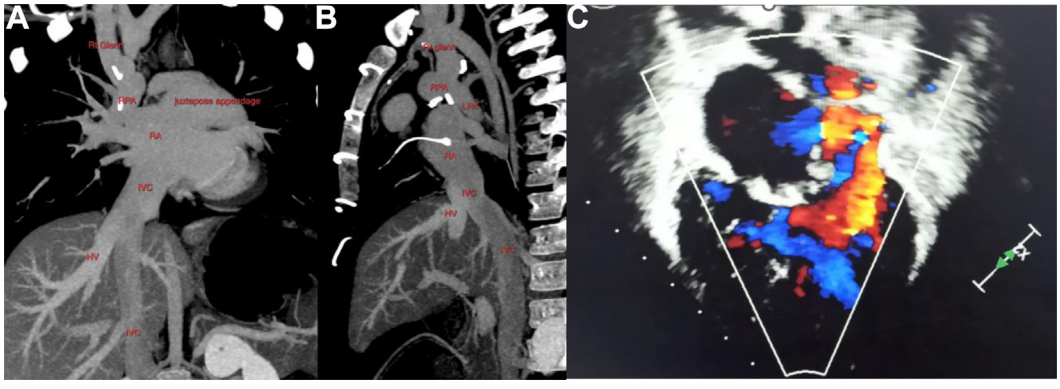
FIGURE 1 Schematic Representation of Transcatheter Fontan Creation



(A) Stent graft placed within the anchoring stents (red). (B) Modification of the technique by using a single covered stent placed from the RPA-RA junction to the uncovered anchoring stent in the IVC, with fenestration equivalent to that in A, shown with arrow marks.
IVC = inferior vena cava; RA = right atrium; RPA = right pulmonary artery; RUPV = right upper pulmonary vein.

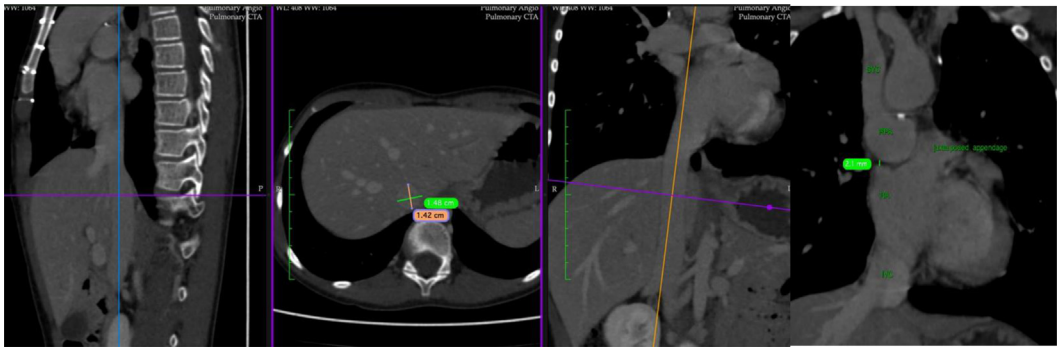
coronary wire as a railroad, a Rubicon catheter 14 (Boston Scientific) was used to enter the PA from the right femoral vein and exteriorized through the RIJV. Because the communication created was very small, we never had desaturation after the initial perforation. After the initial perforation, angiograms were done in the Glenn anastomosis and RA to assess for any extravasation of contrast, and an echocardiogram

FIGURE 2 Pre-Procedural Evaluation of Patient 2 by ECHO and CTPA



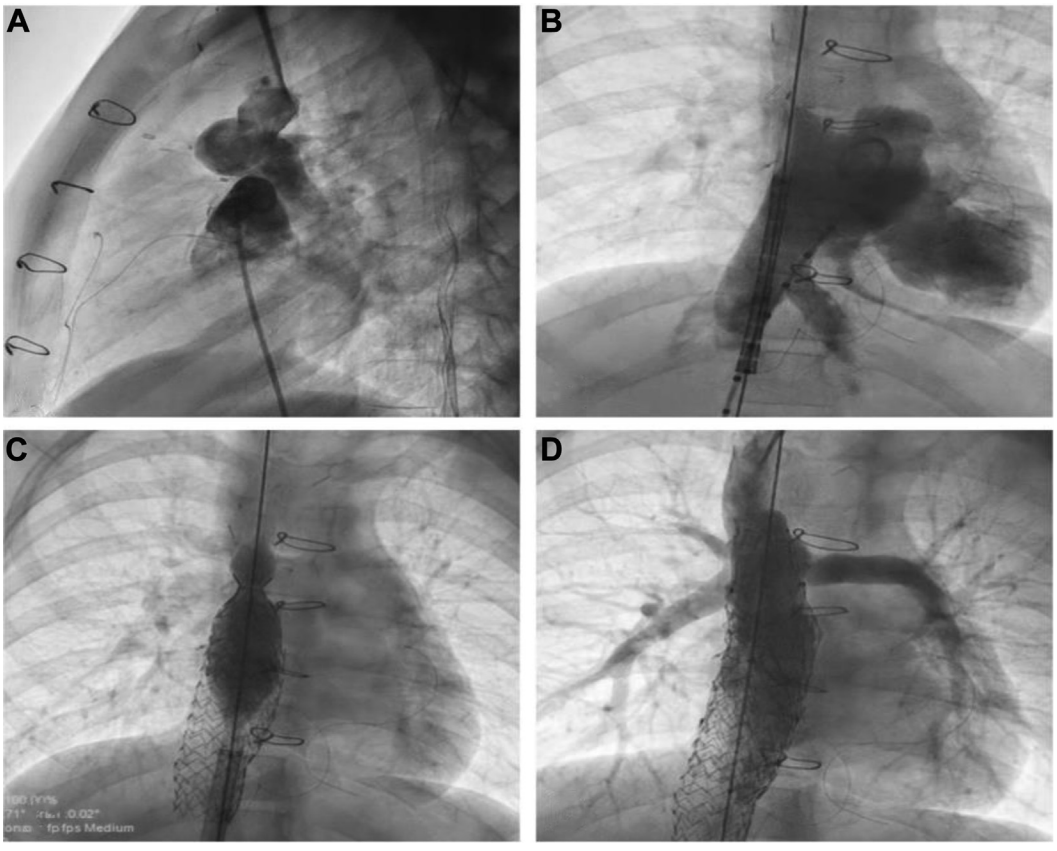
Computed tomography images of patient 2 showing (A) the anatomy of Glenn anastomosis and (B) the inferior vena cava. (B) Echocardiogram showing the intracardiac anatomy with inlet ventricular septal defect and hypoplastic right ventricle.

FIGURE 3 Preprocedure Computed Tomography Analysis of Patient 3



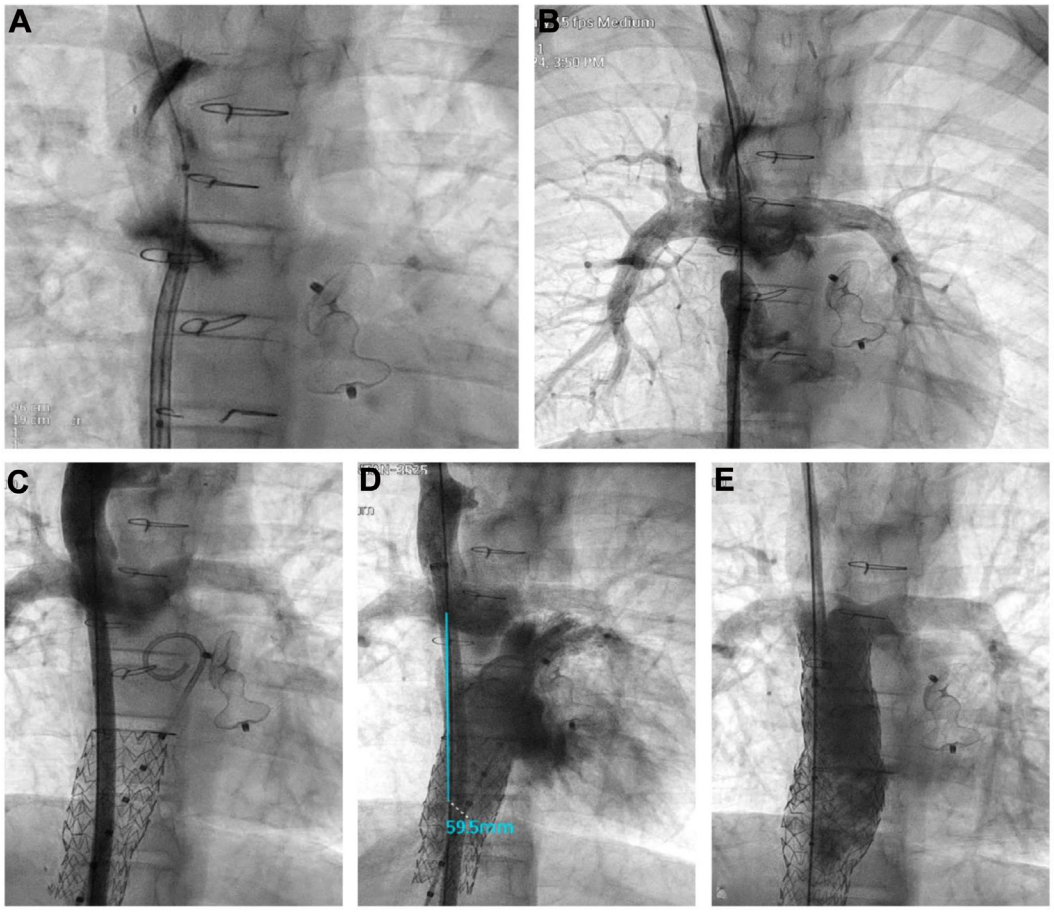
Computed tomography showing systemic venous drainage with measurement of the inferior vena cava diameter at the infrahepatic region and the relation of the Glenn anastomosis to the right atrium and the intervening distance between them.

FIGURE 4 Stepwise Catheter-Based Fontan Completion in Patient 2



Angiographic still images of patient 2 showing (A) simultaneous right atrium Glenn injection with (B) implantation of anchoring stents in the inferior vena cava and pulmonary artery. (C) The RA junction. (D) The final angiogram of the Fontan circuit after stent graft deployment.

FIGURE 5 Stepwise Modification of Catheter Based Fontan Completion in Case 3 Using a Covered Stent



(A, B) Angiographic images of patient 3 showing the creation of the tunnel using a Swartz catheter. (C, D) Posttunnel creation and inferior vena cava anchoring stent deployment angiogram of Glenn anastomosis through the sheath and pigtail angiogram of the inferior vena cava simultaneously to measure the length of required covered stent. (E) Final Fontan connection angiogram showing the low through the branch pulmonary arteries and Fontan circuit.

was done to rule out pericardial or pleural collection before proceeding to further steps. A Lunderquist wire (Cook Medical) was taken through the Rubicon catheter and exteriorized through the RIJV, forming a venous loop (Figures 4 and 5).

STEP 2: ANCHORING STENTS IN THE IVC. Anchoring stents in the IVC were deployed to act as a frame for the stent graft placement. The suprarenal diameter of the IVC was measured, and a balloon 6 mm larger than the diameter was chosen to deploy the uncovered stent. The length of the stent was determined from the suprarenal part of the IVC to the mid-RA. We used a Zephyr XL (SLTL) uncovered stent as the anchoring stent in the IVC-RA junction.

STEP 3: PA-RA JUNCTION STENTING/STENT GRAFT DEPLOYMENT. The PA-RA junction stenting was done with an uncovered Zephyr XL stent in patients 1 and 2. The length was determined from the base of the RPA to the middle of the RA so that it overlaps the IVC stent, thus forming a scaffold for the stent graft deployment. An Endurant II (Medtronic) stent graft was used in patients 1 and 2. The diameter of the stent graft was chosen to be 2-4 mm larger than the suprahepatic IVC diameter. Stent grafts were used in the initial 2 patients because it was easy to create a fenestration in the graft before deployment. However, in patient 3, we used a covered stent from the RPA-RA junction (Figure 1) to the suprahepatic IVC, which eliminated the need for anchoring stent in the

TABLE 2 Preprocedure and Postprocedure Details of the Patients

	Patient 1	Patient 2	Patient 3
Preprocedure			
Route of tunnel creation	PA-to-RA approach	RA-to-PA approach	RA-to-PA approach
IVC-RA stent	24 × 59-mm Zephyr XL	59-mm Zephyr XL premounted stent over 24 × 60-mm balloon	49-mm Zephyr XL over 22 × 50-mm BiB balloon
PA-RA stent	20 × 29-mm Zephyr XL	39-mm Zephyr XL over 20 × 40-mm Z med balloon	59-mm Zephyr covered stent over 20 × 60-mm balloon
Covered stent graft	25 × 49-mm Endurant II	25 × 49-mm Endurant II	–
Fenestration size	5 mm in the stent graft	5 mm in the stent graft	Nonfenestrated
Procedure complication	None	Foreshortening of the stent graft at the IVC-RA junction Residual leak covered with 39-mm Zephyr CS stent over 22 × 40-mm Z med balloon	None
Postprocedure			
Saturation on room air, %	92	91	95
Complications	None	None	Hematoma
Duration of ICU stay, h	12	12	16
Duration of hospital stay, d	5	4	4
Fontan pressure, mm Hg	17	15	13

Manufacturer information: Zephyr XL (SLTL), BiB balloon (NuMed), Endurant II (Medtronic), Z med (NuMED Inc).
ICU = intensive care unit; other abbreviations as in Table 1.

RPA-RA junction, and there was no profound desaturation either. The covered stent was anchored over a 12-mm/8-cm Mustang Balloon (Boston Scientific corp) initially. Then, we dilated the IVC end of the stent serially and stopped when saturation reached 90%, thus accepting the gap between the covered stent and IVC stump as fenestration (Figure 1B). Procedure and postprocedure details of the patients are described in Table 2.

FOLLOW-UP

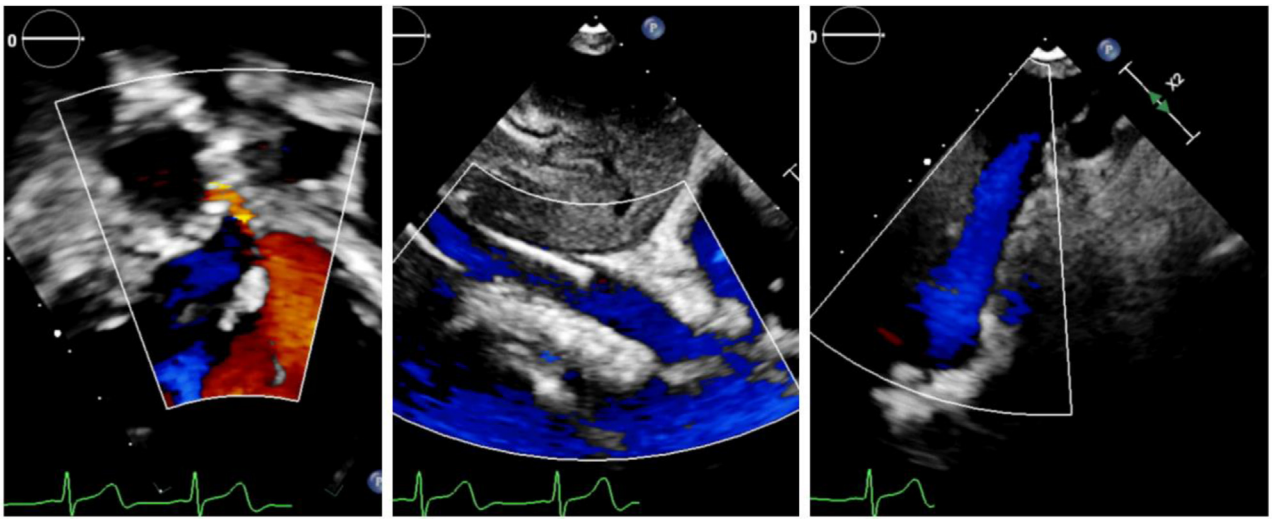
All patients were extubated on the table and monitored in the intensive care unit postprocedure. They were managed with intravenous fluids and intravenous heparin (10 IU/kg/h) overnight and were mobilized on the following day. Echocardiogram showed unobstructed flow in the Fontan circuit and no flow reversal in the hepatic veins, with good flow in the

FIGURE 6 Post Fontan Follow Up Evaluation With CTPA of Our Index Case



Computed tomographic pulmonary angiogram images of patient 1 at the 1-year follow-up showing a patent Fontan pathway with unobstructed hepatic venous drainage (bold yellow arrow).

FIGURE 7 Echocardiographic Images of Patient 1 Showing a Well-Functioning Fontan With No Flow Reversal in the Hepatic Veins With Patent Fenestration at the 1-Year Follow-Up



branch PAs. The patients were discharged on oral antibiotics for 5 days along with oral warfarin and aspirin. At the 1-, 3-, and 6-month follow-ups, echocardiograms showed well-functioning Fontan circulation without any complications. Post 1-year follow-up of patient 1 with echocardiogram and CTPA showed good flow across the Fontan pathway without complications, as shown in **Figures 6 and 7**.

DISCUSSION

Transcatheter Fontan completion was described in surgically prepared patients. Many modifications were done surgically to create a foothold for percutaneous Fontan completion later.[3,5,6] In our first patient, we planned for a nonsurgical approach because it was a high-risk candidate for surgical palliation because the PA pressures were high; hence, we were searching for alternative methods of final palliation. Sizarov Aleksander et al[7] studied the cardiac morphology and suitability for transcatheter Fontan in post-Glenn patients and found that interatrial curved stent grafts were needed for successful total cavopulmonary anastomosis without any complications in children. Hence, we used a stent graft and succeeded in creating a connection between the PA-RA junction and completed percutaneous Fontan.[4] We analyzed echocardiogram and CTPA images of our first patient after 1 year, which showed that the stents were in position without any stenosis or fracture and without causing obstruction to the right upper pulmonary vein or hepatic venous drainage. With this success, we defined the selection criteria and completed transcatheter Fontan in our next cases. We initially used an Endurant stent graft for the Fontan completion because it was easy to create fenestration. However, this will require a scaffold of uncovered stents. Moreover, severe desaturation happens after stenting the PA-RA junction, and then the stent graft must be deployed quickly, which can sometimes be tricky. Therefore, in patient 3, we used a Zephyr covered stent and then intentionally did not seal the lower end of the covered stent with the IVC stent. This acted as a fenestration with minimal desaturation. We believe that in the future, if leak requires sealing, then expanding the lower end of the stent with a balloon would be enough to seal the IVC leak completely. Short-term outcomes in our patients are showing promising results; however, the long-term outcomes need to be assessed.

CONCLUSIONS

Transcatheter Fontan in surgically unprepared patients can be done in selected patients having favorable anatomy with good early outcomes.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Muthukumaran Chinnasamy Sivaprakasam, Department of Pediatric Cardiology, Apollo Children's Hospital, Thousand Lights, Chennai, Tamilnadu 600006, India. E-mail: ach.paedcardio@gmail.com.

REFERENCES

1. Galantowicz M, Cheatham JP. Fontan completion without surgery. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu*. 2004;7:48-55.

2. Sidiropoulos A, Ritter J, Schneider M, Konertz W. Fontan modification for subsequent non-surgical Fontan completion. *Eur J Cardiothorac Surg*. 1998;13:509-512.

3. Alsoufi B, Alfadley F, Al-Omrani, et al. Hybrid management strategy for percutaneous Fontan completion without surgery: early results. *Ann Thorac Surg*. 2011;9:566-572.

4. Ganesan R, Muthukumaran CS, Viswanathan S, Lakhani Z. Modified technique of percutaneous fenestrated Fontan completion without custom made stent or prior surgical preparation—a case report. *Prog Pediatr Cardiol*. 2023;69:101617.

5. Metton O, Calvaruso D, Stos B, et al. A new surgical technique for transcatheter Fontan completion. *Eur J Cardiothorac Surg*. 2011;39:81-85.

6. Boudjemline Y, Malekzadeh-Milani S, Van Steenberghe M, et al. Novel method of surgical preparation for transcatheter completion of Fontan circulation: creation of an extracardiac pathway. *Arch Cardiovasc Dis*. 2014;107:371-380.

7. Sizarov A, Raimondi F, Bonnet D, Boudjemline Y. Cardiovascular anatomy in children with bidirectional Glenn anastomosis, regarding the transcatheter Fontan completion. *Arch Cardiovasc Dis*. 2018;111:257-269.

KEY WORDS cavopulmonary anastomosis, single ventricle, transcatheter Fontan, transcatheter palliation

EBAC ABIM CME MOC ACCREDITED

Go to http://www.acc.org/jacc-journals-cme to take the CME/MOC/ECME quiz for this article.