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CASE REPORT

ADVANCED

CLINICAL CASE

Transcatheter Edge-to-Edge Repair of Systemic Tricuspid Valve in Extracardiac Fontan Circulation

First in Human

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ABSTRACT

Transcatheter edge-to-edge valve repair can improve clinical outcomes in otherwise high-risk surgical patients. This is a first-in-human procedure outlining transcatheter edge-to-edge valve repair of a systemic tricuspid valve in an extracardiac Fontan patient born with hypoplastic left heart syndrome with prohibitive surgical risk. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:221–225) © 2022 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/).

HISTORY OF PRESENTATION

A 13-year-old girl was admitted with weeks of dyspnea following posterior spinal fusion for severe idiopathic scoliosis. On admission, she was hypoxic, with evidence of pansystolic murmur and bilateral lung crepitations.

LEARNING OBJECTIVES

- To consider TEER in the Fontan population, even in HLHS with tricuspid valve incompetence with high surgical risk.
- To use multimodal imaging, including contrast cardiac computed tomography and transesophageal echocardiography, so that procedural planning for atrial access and valve repair is optimized.

MEDICAL HISTORY

The patient was born with hypoplastic left heart syndrome (HLHS) and underwent a fenestrated extracardiac Fontan procedure at 3 years of age. Other history included device fenestration closure at 4 years of age, left pulmonary artery (PA) stenting and coiling of a venovenous collateral vessel.

DIFFERENTIAL DIAGNOSIS

Decompensated heart failure due to tricuspid regurgitation (TR) was suspected. Right-to-left shunting and respiratory causes of hypoxemic respiratory failure were evaluated.

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ABBREVIATIONS AND ACRONYMS

CHD = congenital heart disease

HLHS = hypoplastic left heart syndrome

PA = pulmonary artery

TEER = transcatheter edge-to-edge repair

TR = tricuspid regurgitation

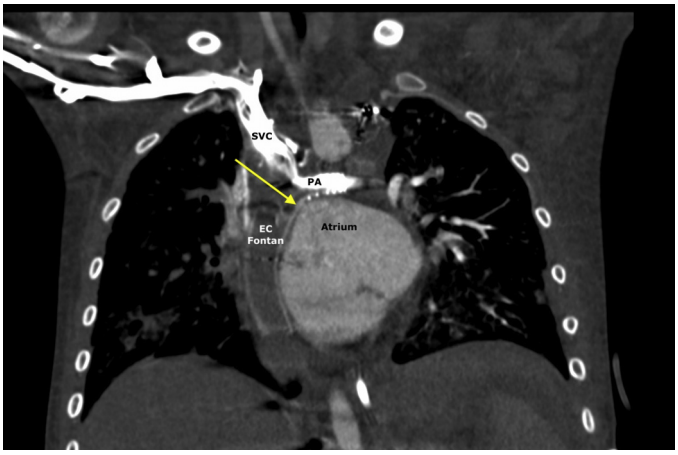
INVESTIGATIONS

Chest radiography revealed pulmonary edema with pleural effusions. Transthoracic echocardiography demonstrated mildly reduced single right ventricular function with severe systemic atrioventricular valve TR. She underwent invasive cardiac catheterization, demonstrating good Fontan hemodynamic status. Fontan pressure was 14 mm Hg, and pulmonary vascular resistance was 0.8 WU. The patient had unobstructed bilateral PA flow and no significant arteriovenous malformations. She continued to have increasing oxygen requirements and developed acute renal failure. This culminated in intubation, inotropic support, and chest tube insertion, and TR intervention was contemplated. Transcatheter edge-to-edge repair (TEER) of the tricuspid valve with off-label MitraClip (Abbott Vascular) use was considered and recommended. For preprocedural planning, contrast cardiac computed tomographic imaging was performed, demonstrating proximity of the proximal left PA to the Fontan systemic atrium (**Figure 1**).

MANAGEMENT

Tricuspid TEER was performed via the right internal jugular vein because of concerns of the lack of "height" using an inferior vena cava approach, under transesophageal echocardiographic guidance. Transesophageal echocardiography demonstrated a moderately dilated and hypertrophied right ventricle with new moderately decreased systolic function. Tricuspid leaflets were thickened with severe central TR (Video 1), which was believed to be functional from annular dilation (**Figure 2**). Vena contracta width was 0.7 cm, proximal isovelocity surface area radius was 1 cm, and effective regurgitant orifice area was 0.53 cm^2, with a regurgitant volume of 60 mL, all consistent with severe regurgitation. The main coaptation gap was between the septal and posterior leaflets and was 0.4 cm. An SL1 sheath (Abbott Laboratories) was positioned in the PA via the internal jugular vein. Through this, a BRK needle (Abbott Laboratories) was used for "transseptal" access through the PA into the systemic atrium (**Figure 3**), and the sheath was advanced. Through this, a stiff wire was placed in the ventricle. A 7-mm balloon was used for "septostomy" at the PA-atrial access site. Fontan pressure was 22 mm Hg, with oxygen saturation of 30% despite 100% fraction of inspired oxygen. Arterial pressure was 99/61 mm Hg, and atrial pressure was 11 mm Hg (v-wave 14 mm Hg). Arterial saturation was 96%, and starting Qs was 1.2 L/min/m^2. Access was upsized to a 24-F steerable guide catheter, which was positioned in the atrium. Because of jugular access, each clip was miskeyed 90° counterclockwise, and each clip was directed to the valve with the M/L knob moving the device anteromedial (M) or posteromedial (L) and the A/P knob controlling anterolateral (A) and posterolateral (P) positioning. Clockwise rotation moved anteriorly, and counterclockwise moved posteriorly. Full straddle was achieved, as the Fontan atrium was massively enlarged. A MitraClip XTW clip (Abbott Vascular) was deployed at the anterior and septal leaflets. Given difficulty grasping posterior and septal leaflets simultaneously, a MitraClip XTW on the anterior and posterior leaflets was placed (Video 2) to then allow a third MitraClip XTW on the posterior and septal leaflets (**Figure 4**). TR was reduced from 5+ (torrential) to 2+ (moderate) (Videos 3 and 4). Mean gradient on transesophageal echocardiography across the tricuspid valve was 3 mm Hg at end of case. Fontan and atrial pressure were unchanged, and Qs had improved (1.44 L/min/m^2). A 7-F delivery sheath was inserted through the guide catheter, and a 6-mm septal occluder device was

FIGURE 1 Computed Tomography



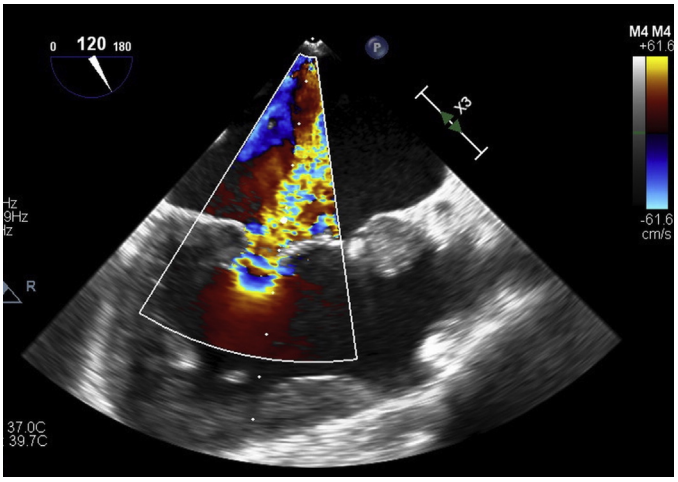
Contrast computed tomography demonstrating pulmonary artery (PA) adjacent to the systemic atrium with a good site for access to the atrium **(arrow)**. Superior vena cava (SVC) and extracardiac (EC) Fontan conduit are also labeled.

delivered for closure of iatrogenic PA-atrial communication (Figure 5). Hemostasis of the right internal jugular site was achieved by manual compression. There was no procedural complication.

DISCUSSION

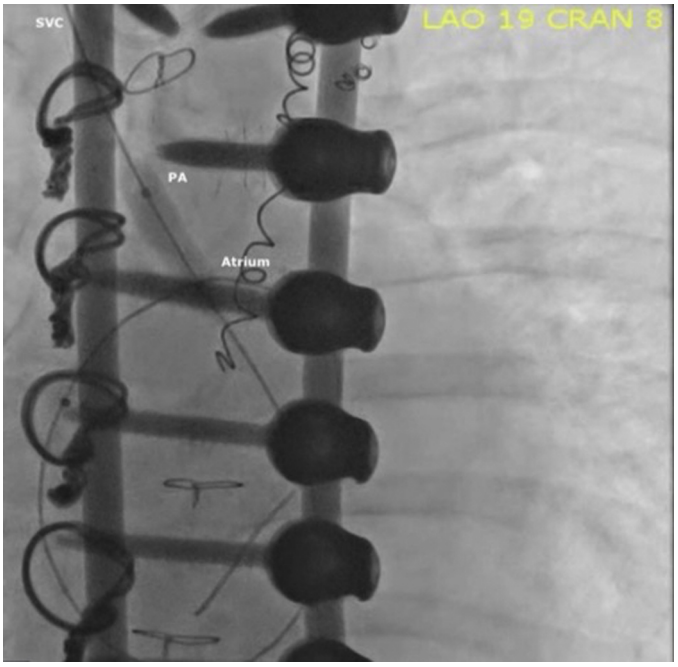
Transcatheter intervention has revolutionized the management of patients with congenital heart disease (CHD), who have often undergone multiple surgical procedures and are at high operative risk. To the best of our knowledge, this is the first report describing TEER in a patient with extracardiac Fontan physiology for HLHS to repair severe systemic tricuspid valve insufficiency. This innovative approach is a proof of concept for management of critically ill patients with CHD with significant valvular heart disease. The Fontan procedure for redirection of blood flow in patients with CHD is indicated when biventricular repair is

FIGURE 2 Tricuspid Regurgitation



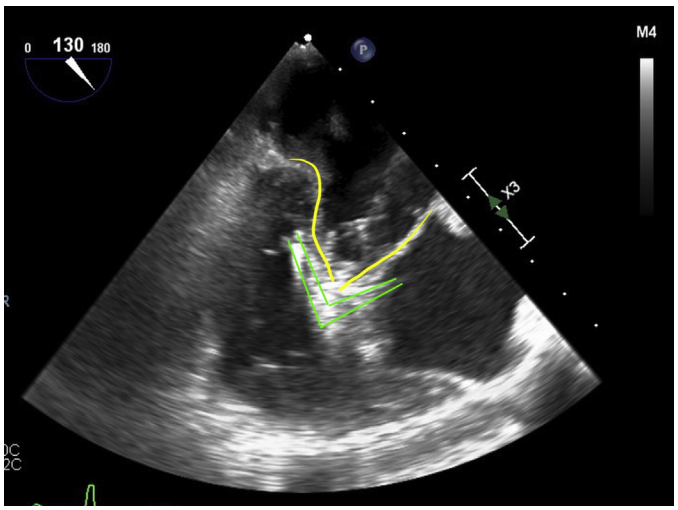
Transesophageal echocardiography with color flow demonstrating broad vena contracta with severe tricuspid regurgitation.

FIGURE 3 Septostomy



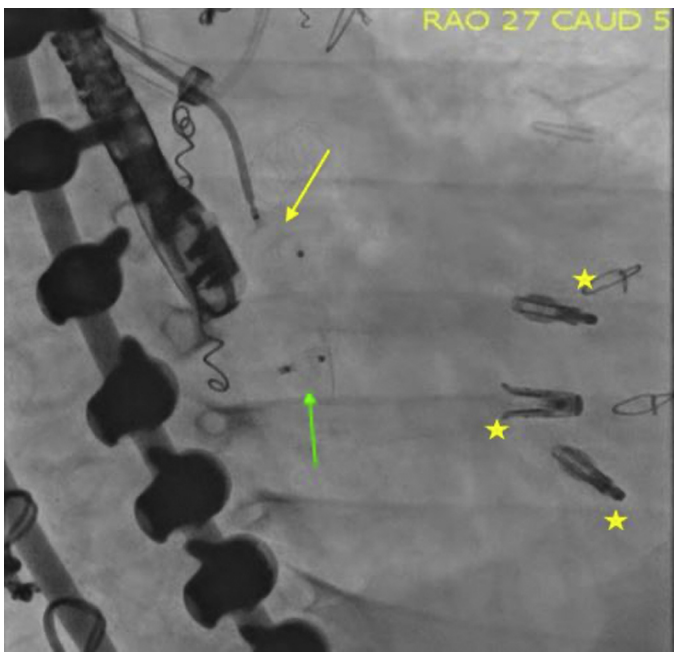
Fluoroscopy of balloon septostomy with access from the right internal jugular vein into the superior vena cava (SVC), then the pulmonary artery (PA) and systemic atrium. CRAN = cranial; LAO = left anterior oblique.

FIGURE 4 Leaflet Grasping



Transesophageal echocardiography demonstrating an attempt at grasping of 2 leaflets **(yellow)** within the clip **(green)**.

FIGURE 5 Fluoroscopy



Fluoroscopy of pulmonary artery–atrial access site device closure **(yellow arrow)** with a septal occluder device. Three clips seen on tricuspid valve **(stars)** and previous Fontan fenestration closure device seen **(green arrow)**. CAUD = caudal; RAO = right anterior oblique.

not possible, as is the case in HLHS. First described in 1971 by Fontan and Baudet,[1] it has become the backbone of treatment for many complex congenital heart defects. In HLHS, in which there is a single morphologic right ventricle, ventricular dysfunction and associated tricuspid valve insufficiency are common before adulthood and are independent risk factors for mortality in the Fontan population.[2] Furthermore, right ventricular dominance and mitral atresia have been shown to be predictive of atrioventricular valve failure in Fontan patients,[3] as seen in this case.

In the non-CHD population, TEER in high-risk patients with mitral regurgitation is beneficial,[4,5] and more recent data show improved outcomes in patients undergoing tricuspid TEER.[6] This case demonstrates feasibility, periprocedural safety, and success using the current TEER system. With appropriate preprocedural imaging and planning, we overcame anatomical difficulties such as alternative venous access, atrial access through the Fontan conduit, and tricuspid valve TEER to achieve reduction in valve regurgitation. The best timing for these procedures remains to be determined. We believe that it may be when there is severe systemic atrioventricular valve regurgitation with symptoms of compensated heart failure on medical therapy. It is possible that this patient's cardiac dysfunction was too advanced to achieve improvement. This patient had decompensated and required hospitalization, had deteriorating ventricular function, but did not have very high "v" waves, a finding likely affected by simultaneous renal ultrafiltration. Further endeavors to improve transcatheter interventions in the Fontan population could revolutionize clinical outcomes in this cohort.

FOLLOW-UP

The patient returned to the intensive care unit. Day 1 postprocedural transthoracic echocardiography showed moderate to severe TR, and chest radiography showed 3 clips on the tricuspid valve (**Figure 6**). Despite a good procedural result and no acute complications, the patient's overall status continued to deteriorate, with multiorgan failure, likely a reflection that the intervention was performed belatedly. The patient passed away after 5 days.

CONCLUSIONS

This appears to be the first published case of the use of TEER through a jugular approach in a patient with an extracardiac Fontan circulation and a systemic tricuspid valve. This is a proof-of-concept example that confirms technical ability and procedural safety of a minimally invasive transcatheter valve repair in a complex patient with Fontan physiology.

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

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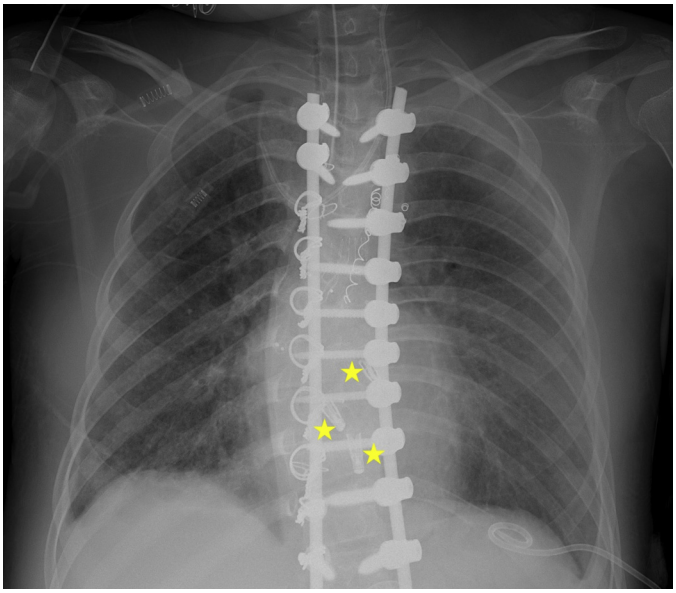
FIGURE 6 **Chest Radiography**



Postprocedural chest radiography demonstrating 3 clips **(stars)**.

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KEY WORDS congenital heart defect, pulmonary edema, tricuspid valve, valve repair

APPENDIX For supplemental videos, please see the online version of this paper.