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CASE REPORT

ADVANCED

CLINICAL CASE

Transcatheter Mitral Valve Repair in a Tricuspid Atresia Patient With Potts and Glenn Shunts

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ABSTRACT

Our patient was a 50-year-old woman with tricuspid atresia who had undergone palliation with a Potts shunt to the left pulmonary artery as an infant and a classic Glenn shunt to the right pulmonary artery as a young child. Under general anesthesia, she underwent transcatheter edge-to-edge repair of the mitral valve for severe symptomatic mitral regurgitation. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:1379–1383) © 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

We present a 50-year-old woman with progressive heart failure symptoms and cardiac cachexia in the setting of congenital heart disease. After multidisciplinary discussion and planning, she was admitted to our hospital (Emory University Hospital, Atlanta, Georgia, USA) for transcatheter edge-to-edge repair (TEER).

On admission, examination was notable for a continuous murmur in the left upper sternal border and a holosystolic apical murmur with a displaced apex. Her pulse was irregularly irregular. Her lungs were clear to auscultation. Cyanosis and digital clubbing were present. Her oxygen saturation was 75% on room air.

LEARNING OBJECTIVES

- To understand the physiology and chronic hemodynamic consequences of aortopulmonary shunts, including chronic LV volume overload and LV dilation in adult patients with palliated congenital heart disease.
- To recognize the importance of a multidisciplinary team-based approach in the care of adult congenital heart disease patients and the application of established therapies, including transcatheter procedures, in novel ways.

PAST MEDICAL HISTORY

The patient was born with tricuspid atresia. She underwent a Potts shunt to the left pulmonary artery (LPA) at 13 days of age and a classic Glenn shunt to the right pulmonary artery (RPA) at 2.5 years of age (Supplemental Figure 1). Other history included permanent atrial fibrillation and pulmonary hypertension. She had chronic cyanosis and secondary erythrocytosis.

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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.

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ABBREVIATIONS AND ACRONYMS

ASD = atrial septal defect

EROA = effective regurgitant orifice area

LPA = left pulmonary artery

LV = left ventricular

MC = MitraClip

MR = mitral regurgitation

PEEP = positive end-expiratory pressure

RPA = right pulmonary artery

TEE = transesophageal echocardiography

TEER = transcatheter edge-to-edge repair

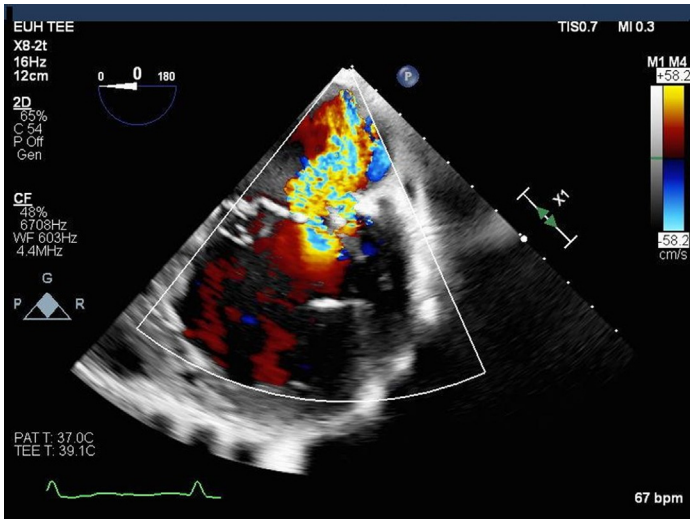
DIFFERENTIAL DIAGNOSIS

Her clinical decline was attributed to worsening mitral regurgitation (MR) and left ventricular (LV) dilation, which resulted from long-standing LV volume overload from the Potts shunt and left lung overcirculation.

INVESTIGATIONS

Her admitting chest radiograph was unchanged compared with previous examinations. Transesophageal echocardiography (TEE) showed an LV ejection fraction of 55%. There was severe MR with a restricted posterior leaflet and chordal attachments of P1 and P3 (heart rate, 88 beats/min) (**Figure 1**). Pulmonary vein systolic flow was severely blunted (**Figure 2**). Her left atrial volume index was 336 mL/m^2. The MR effective regurgitant orifice area (EROA) was 75.81 cm^2. Tricuspid atresia with a hypoplastic right ventricle and a large secundum atrial septal defect (ASD) were present. Cardiac computed tomography angiography showed a widely patent Glenn shunt and Potts shunt. There was severe biatrial enlargement (**Figure 3,** Video 1).

FIGURE 1 Severe Mitral Regurgitation



Baseline transesophageal echocardiography with color flow Doppler demonstrating severe mitral regurgitation through a 4-chamber view.

A multidisciplinary team consisting of structural cardiology, imaging, and adult congenital cardiology completed an evaluation. She was not considered to be a surgical candidate given high mortality risk with an open approach. The team recommended she undergo TEER of the mitral valve. The discussion centered on the feasibility of controlling the guidewire around the patient's ASD and minimizing hemodynamic shunt alterations during general anesthesia. The intact septum had only a borderline height above the mitral annulus that could have been inadequate for device maneuverability; therefore, we decided to approach through the congenital ASD.

MANAGEMENT

The patient was brought to the cardiac catheterization laboratory and underwent endotracheal intubation using general anesthesia. We took care to minimize positive end-expiratory pressure (PEEP), which was mostly 0 cm/H_2O during the procedure because increasing intrathoracic pressure would impair right lung blood flow through the Glenn shunt. We also took care to keep her oxygen saturation at baseline to avoid excessive oxygenation and subsequent pulmonary edema from left lung overcirculation through the Potts shunt.

A TEE probe was inserted for imaging guidance. The left common femoral vein was accessed transcutaneously. The wire was advanced, and the large ASD was crossed by using a multipurpose catheter and a J-tipped wire (**Figure 4**). A stiff wire was then exchanged and was advanced into the pulmonary vein. The MitraClip (MC) (Abbott) steerable guide catheter was then advanced. The initial attempt to place a wide clip (NTW MC) at A2/P2 was unsuccessful. Despite the superior position of the ASD compared with the septum in respect to the mitral annulus, the guidewire was too unstable. Therefore, a more posterior transseptal puncture was performed using a Nagare (Terumo) steerable catheter and an electrified Astato wire (Asahi Intecc). This stabilized the guidewire. A single NT MC was then positioned in the A2/P2 position and was successfully deployed with capture of both leaflets (**Figure 5**). TEE confirmed the positioning, and immediate reduction of MR was noted to the mild range. There was no evidence of mitral stenosis. The left atrial preprocedural pressure was 27/31 mm Hg, and the postprocedural pressure was 20/23 mm Hg. The delivery system was removed, and her access sites were closed.

DISCUSSION

Tricuspid atresia has several morphologic variants, but the mainstay of this congenital heart disease is no direct communication between the right atrium and the right ventricle.[1] To sustain life, there are common associated septal defects to allow blood to flow from left-side circulation to right for oxygenation by the lungs. The right ventricle, which is usually the dominant ventricle in utero, remains hypoplastic because of its lack of communication with the right atrium and systemic venous blood flow.

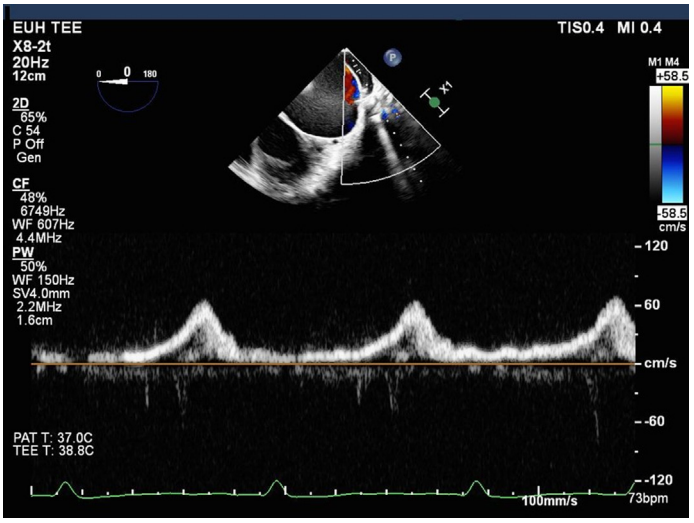
Our patient with tricuspid atresia had undergone palliation with a classic Glenn shunt to the RPA and a Potts shunt to the LPA. In the modern era, this patient would have undergone palliation with a Fontan procedure, but she was born before this was commonly done.[2]

In a classic Glenn shunt, the pulmonary arteries are divided, and the superior vena cava is connected to the RPA. Right lung pulmonary blood-flow is dependent on a central venous pressure that is higher than left atrial pressure, systemic ventricle systolic and diastolic function, low pulmonary vascular resistance, and the ability to generate negative intrathoracic pressure during inspiration. Our minimization of PEEP during the procedure reflected this because increased positive pressure from ventilation could result in reduced right lung perfusion.

In a Potts shunt, the descending aorta is connected to the LPA to provide left lung blood flow. The Potts shunt is one of several aortopulmonary shunts to provide pulmonary blood flow when this is inadequate. These shunts can cause pulmonary overcirculation and can lead to pulmonary hypertension and/or LV volume overload and its associated complications, as seen in our patient with left lung pulmonary hypertension and severe LV dilation, dysfunction, and resultant MR.[3]

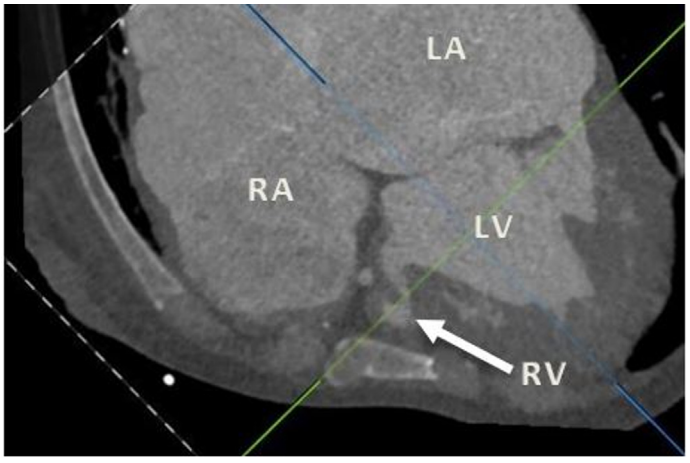
The advanced cardiopulmonary disease of adult congenital heart disease patients who have undergone many previous palliative surgical procedures puts them at high risk for complications of typical open-heart surgery. The emergence of transcatheter approaches for valvular diseases has been a major innovation to mitigate some of these risks.[4] We present the first case to our knowledge of MC placement in a patient with tricuspid atresia.

FIGURE 2 Pulmonary Vein Waveform



Baseline transesophageal echocardiography demonstrating severely blunted systolic pulmonary vein flow.

FIGURE 3 Cardiac Computed Tomography Angiography



Imaging demonstrating the hypoplastic right ventricle communicating with the left ventricle and the severe biatrial dilation. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

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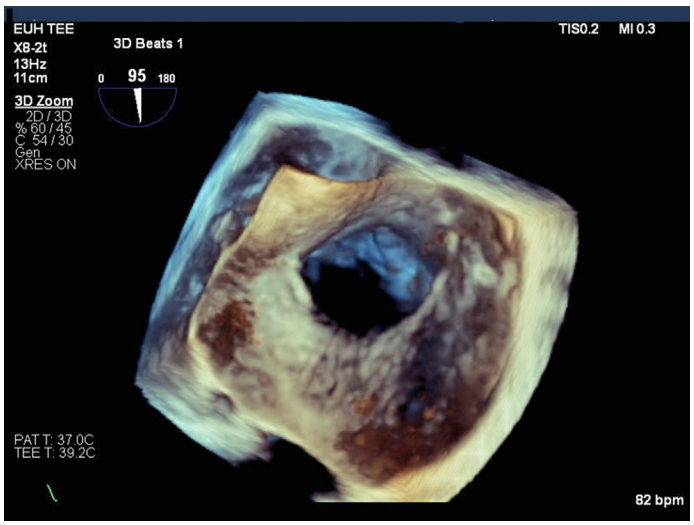
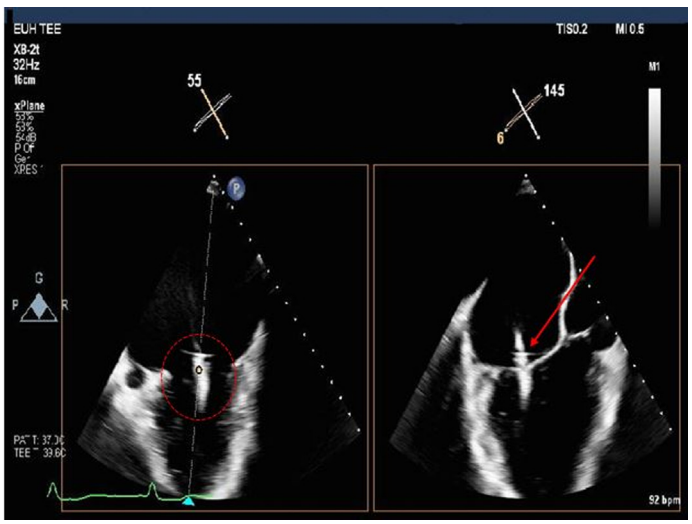
FIGURE 4 Congenital Atrial Septal Defect



Transesophageal echocardiography 3-dimensional reconstruction of congenital atrial septal defect.

FIGURE 5 Leaflet Grasp at Mitral Valve



Transesophageal echocardiography demonstrating device **(circled)** positioned at the mitral valve and grasping of leaflets for clip deployment **(red arrow).**

This unconventional approach required a multidisciplinary team and extensive planning both preoperatively and postoperatively for a beneficial outcome. We hope this serves as a proof of concept for future endeavors of transcatheter mitral valve therapies in patients with complex congenital heart diseases.

FOLLOW-UP

The patient was extubated and transported to the cardiac intensive care unit for further monitoring and postprocedural care. Her remaining course was uncomplicated, and she was discharged home.

At her 3-month follow-up, she had notable symptom improvement and weight gain. Her New York Heart Association functional classification improved from IV to III. Her diuretic therapy requirements decreased. A transthoracic echocardiogram of the valve demonstrated good seating of the clip with mild to moderate residual MR and a mean gradient of 2 mm Hg (**Figures 6A and 6B**). The left atrial volume index was 180 mL/m^2. The MR EROA was 32.60 cm^2.

CONCLUSIONS

We believe this is the first reported case of successful MC deployment in a patient with tricuspid atresia and severe MR as the result of previous palliations. This case demonstrates, with a multidisciplinary team-based approach, that a patient who previously would have been considered too high a risk can undergo successful general anesthesia and transcatheter cardiac surgery.

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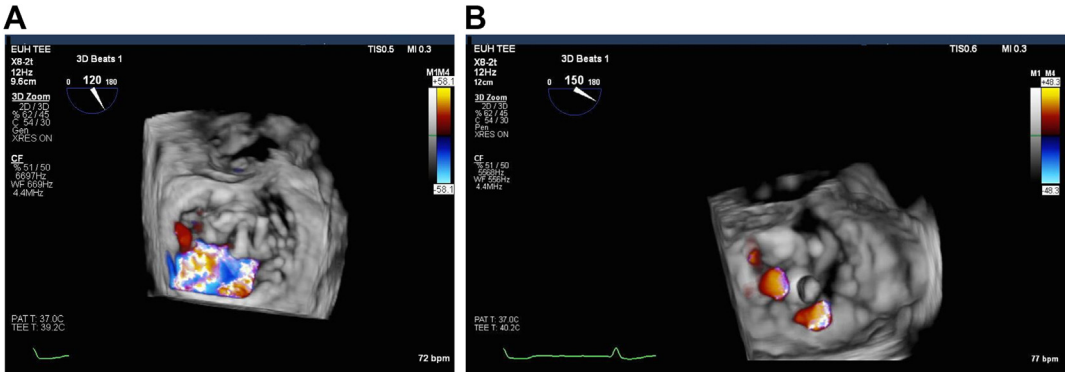
FIGURE 6 **Before and After 3-Dimensional Transesophageal Echocardiography Reconstruction With Color Doppler**



Transesophageal echocardiography (TEE) imaging demonstrating **(A)** severe mitral regurgitation with broad jet with significant improvement and **(B)** mild mitral regurgitation post-transcatheter edge-to-edge repair. Abbreviations as in Figures 1 and 4.

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KEY WORDS congenital heart disease, cyanotic heart disease, echocardiography, mitral valve, pulmonary circulation, systolic heart failure

APPENDIX For a supplemental figure and a video, please see the online version of this article.