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CASE REPORT
Adult congenital heart disease

First described mitral clip in an adult extracardiac Fontan patient: a case report

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Background The use of transcatheter edge-to-edge repair (TEER) in patients with advanced heart failure has been shown to reduce hospitalizations and increase survival. As patients with Fontan circulations grow older, a significant proportion of them will develop severe atrioventricular (AV) valve regurgitation in the systemic ventricle. Conventional surgical repair and transplant carry high mortality risk for the adult Fontan patient with progressive heart failure.

Case summary A 51-year-old female extracardiac Fontan patient developed severe AV valve regurgitation and progressive functional decline. Based on her operative risk for conventional surgical intervention or transplant, TEER using the Abbott MitraClip device was performed. The degree of mitral regurgitation was decreased from severe to moderate regurgitation.

Discussion This is the first known case describing the use of a successful TEER in an adult patient with an extracardiac Fontan. Given the increasing numbers of patients surviving into adulthood with a Fontan circulation, transcatheter interventions may provide an alternative treatment option to conventional surgeries and medical therapies.

Keywords Fontan • Transcatheter edge-to-edge repair • Adult congenital heart disease • Mitral clip • Heart failure • Case report

ESC Curriculum 9.7 Adult congenital heart disease • 4.3 Mitral regurgitation • 2.2 Echocardiography • 7.4 Percutaneous cardiovascular post-procedure

Learning points

- Evaluation of novel transcatheter procedures in a complex adult congenital patient should be done with a multidisciplinary team of congenital cardiologists, surgeons, imaging, and interventional experts
- Advanced planning for case approach, including 3D modelling, may be advantageous for successful outcomes
- Transcatheter edge-to-edge repair for significant atrioventricular valve regurgitation in the Fontan patient may provide an additional treatment option for the 'failing' Fontan patient

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Introduction

Progressive atrioventricular valve regurgitation (AVVR) is found in up to 20% of single ventricular patients after Fontan palliation, and significant AVVR can lead to a two-fold increase in Fontan failure.[1,2] The use of transcatheter edge-to-edge repair (TEER) of the mitral valve (MV) in adult patients with heart failure and moderate to severe or severe mitral regurgitation (MR) despite optimal medical therapy has been shown to improve mortality and resulted in lower rates of hospitalization.[3] The Fontan operation for single ventricle patients has undergone several iterations since its inception in 1971. The extracardiac Fontan repair is currently favoured at many centres due to potentially superior fluid dynamics and ease of surgical intervention (see Supplementary material online, *Figure S1*).[4,5] However, since the extracardiac conduit connects systemic venous return directly to the pulmonary arteries, intracardiac transcatheter intervention becomes more challenging. In this report, we describe our approach to access the systemic atrioventricular (AV) valve of a Fontan patient via extracardiac conduit puncture, making TEER a possible therapeutic option in Fontan patients with significant systemic AV valve regurgitation.

Timeline

Time	Event
1970	Born with tricuspid atresia with malposed great arteries and VSD
1974	Pulmonary artery band
1990	Bi-directional Glenn
1996	Extra-cardiac Fontan
2016	Moderate (2+) mitral regurgitation by ECHO, able to walk 3 km without difficulty
2017	Patient began to have more fluid retention, required escalating doses of diuretics
2019	Moderately severe (3+) mitral regurgitation by ECHO
2020	Severe (4+) mitral regurgitation by ECHO
2020–2021	Multiple hospital admissions for heart failure with volume overload
Spring 2021	Patient evaluated in clinic, NYHA Class III symptoms, had abdominal and leg swelling, evidence of liver cirrhosis by imaging
Summer 2021	Obtained a chest CT and TEE to plan for transcatheter edge-to-edge repair. Discussed management options with multi-disciplinary team, including imaging, adult congenital cardiology, paediatric and adult interventional, heart failure experts, and patient
Fall 2021	Patient underwent mitral clip procedure with a total of three clips placed. Mitral regurgitation reduced from 4+ to 2+ in severity
Spring 2022	Follow-up ECHO shows mild to moderate (1-2+) mitral regurgitation with improved systemic ventricular function and no further heart failure admissions to date

Case presentation

A 51-year-old woman with a history of tricuspid atresia, D-transposition of the great arteries, and a ventricular septal defect status post-Fontan palliation presented with severe MR. Her past medical history includes a pulmonary artery band at age one, a bidirectional Glenn at the age of 20, and an extracardiac Fontan at age 26.

Approximately 3 years prior to presentation, the patient had moderate MR and was able to walk 3 km without difficulty. She began to have more fluid retention in her abdomen and became dependent on escalating doses of diuretics. One year prior to the presentation, the MR progressed to severe and she had multiple hospital admissions for Fontan failure.

At her evaluation in 2021, she was in New York Heart Association Class III heart failure and complained of abdominal and leg swelling. An echocardiogram showed severe MR, systolic flow reversal into the pulmonary veins, and mildly to moderately reduced systolic function at 45% by method-of-discs of her systemic ventricle. Medications included bumetanide, spironolactone, lisinopril, metoprolol, warfarin, and potassium. Baseline vital signs were notable for an oxygen saturation of 87% on room air. Baseline haemoglobin and haematocrit were 16.9 g/dL and 50.4%, respectively. She had chronic kidney disease Stage III with a baseline creatinine of 1.04 mg/dL. Her other medical history included obstructive sleep apnea requiring continuous positive airway pressure and bipolar disorder. Liver magnetic resonance imaging showed Stage III hepatic fibrosis and numerous lesions consistent with focal nodular hyperplasia.

Given the patient's progressive heart failure symptoms in the setting of severe MR and liver cirrhosis that would make any conventional surgical repair or multi organ transplant high risk, the proposal was made to proceed with a TEER of the MV. The patient's data were reviewed by a multidisciplinary group of adult congenital heart disease, radiology, cardiac surgery, and structural cardiology interventional experts. The options were discussed with the patient, and she elected to undergo TEER procedure.

A chest computed tomography (CT) scan with contrast was obtained of the patient's cardiac anatomy (*Figure 1*). From this scan, a 3D model was created to help guide planning (*Figure 1*). To allow for optimal function of the mitral clip delivery system, a Fontan conduit puncture was planned that would place the delivery system approximately 4–5 cm above the plane of the MV (*Figure 1*). Using the inferior vena cava (IVC)/Fontan conduit anastomosis as an anatomic landmark, a puncture site 3–4 cm above the anastomosis site was designated.

Cardiac catheterization demonstrated a cardiac index of 3.0 L/min/m^2 by Fick with a Fontan pressure of 25 mmHg. The systemic atrial pressure was 15 mmHg with a v-wave to 22 mmHg. There were no significant gradients throughout the Fontan pathway or aorta. Her indexed pulmonary vascular resistance was 1.5 WU/m^2. Her mixed venous saturation was 62% with a systemic saturation of 87%.

A transesophageal echo (TEE) probe was inserted to help guide the procedure. Initial TEE evaluation showed severe regurgitation and systolic flow reversal into the left upper and right upper pulmonary veins. There was no significant MV inflow gradient. The mechanism of the MR appeared to be due ventricular enlargement with myxomatous MV leaflets, resulting in poor central coaptation. The calculated MV area was 8 cm^2 by TEE.

Standard right femoral venous access was obtained and pre-closure was performed with two Perclose Proglide devices. A 6F pigtail was advanced to the inferior margin of the Fontan and angiography was performed to evaluate fluoroscopic markers for puncture. A BRK-1 Brockenbrough needle was advanced through an SL1 catheter into the Fontan conduit, approximately 3 cm above the IVC/Fontan conduit anastomosis. The puncture of the Fontan conduit into the systemic

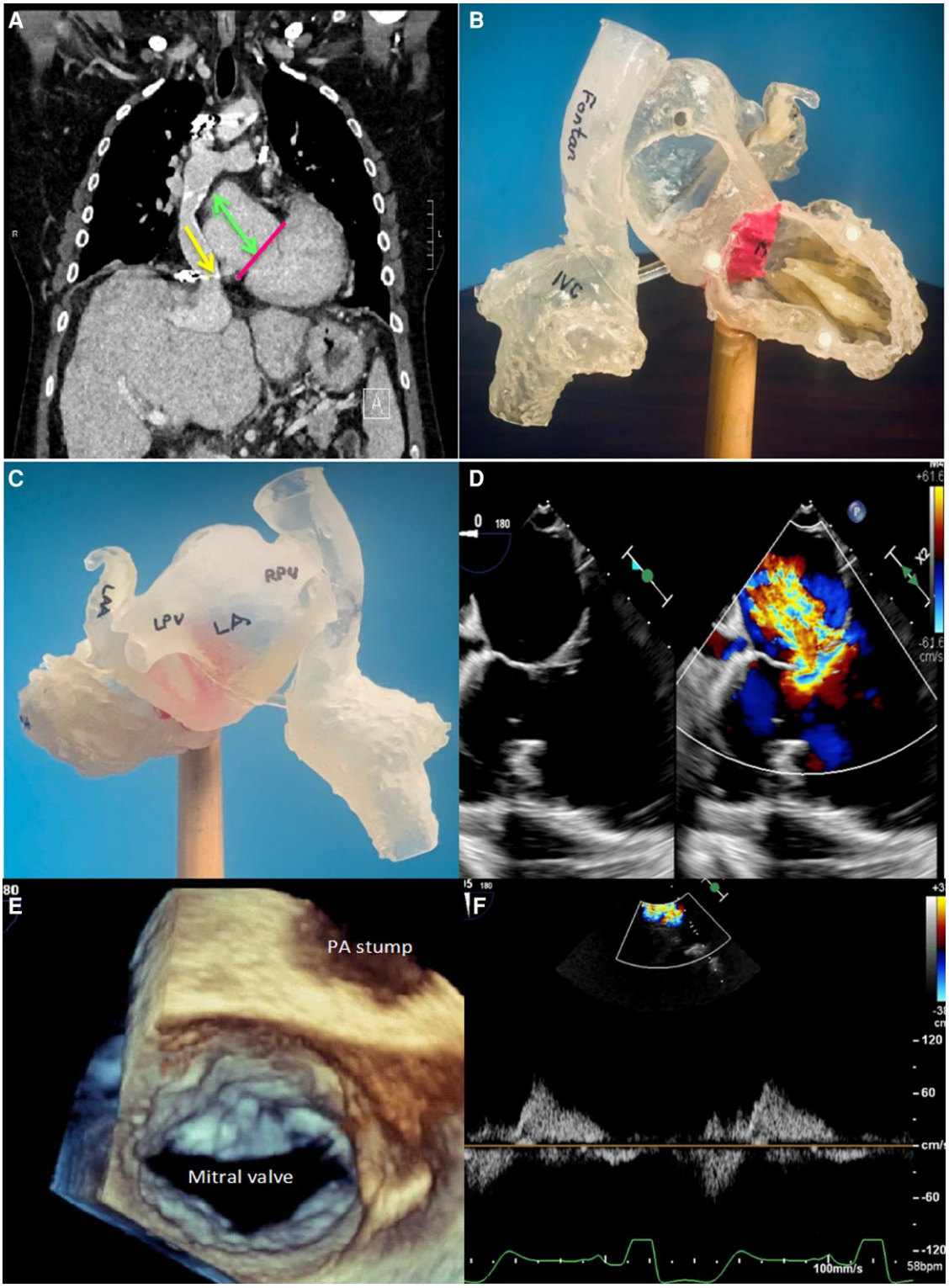

Figure 1 (*A*) CT angiography: Puncture site identified (single headed, yellow arrow) 3-4cm from IVC/extracardiac Fontan anastomosis to allow Mitraclip device to enter systemic atrium (double headed, green arrow) approximately 4-5cm above mitral valve (MV- single line, pink). (*B*) 3-D printed model: Demonstration of anatomic relationship of extracardiac Fontan conduit relative to systemic atrium, IVC, and MV annulus (single line, pink). (*C*) 3-D printed model: Demonstration of extracardiac Fontan baffle relative to right pulmonary vein (RPV), left atrium (LA), left upper pulmonary vein (LPV) and left atrial appendage (LAA). (*D*) TEE: Commissural view of MV with color Doppler demonstrating severe MR (*E*) TEE: 3-D surgeon view (left atrial view) of MV with pulmonary artery (PA) stump used for anterior orientation. (*F*) TEE: Pulse wave Doppler of left upper pulmonary vein (LUPV) showing systolic flow reversal.

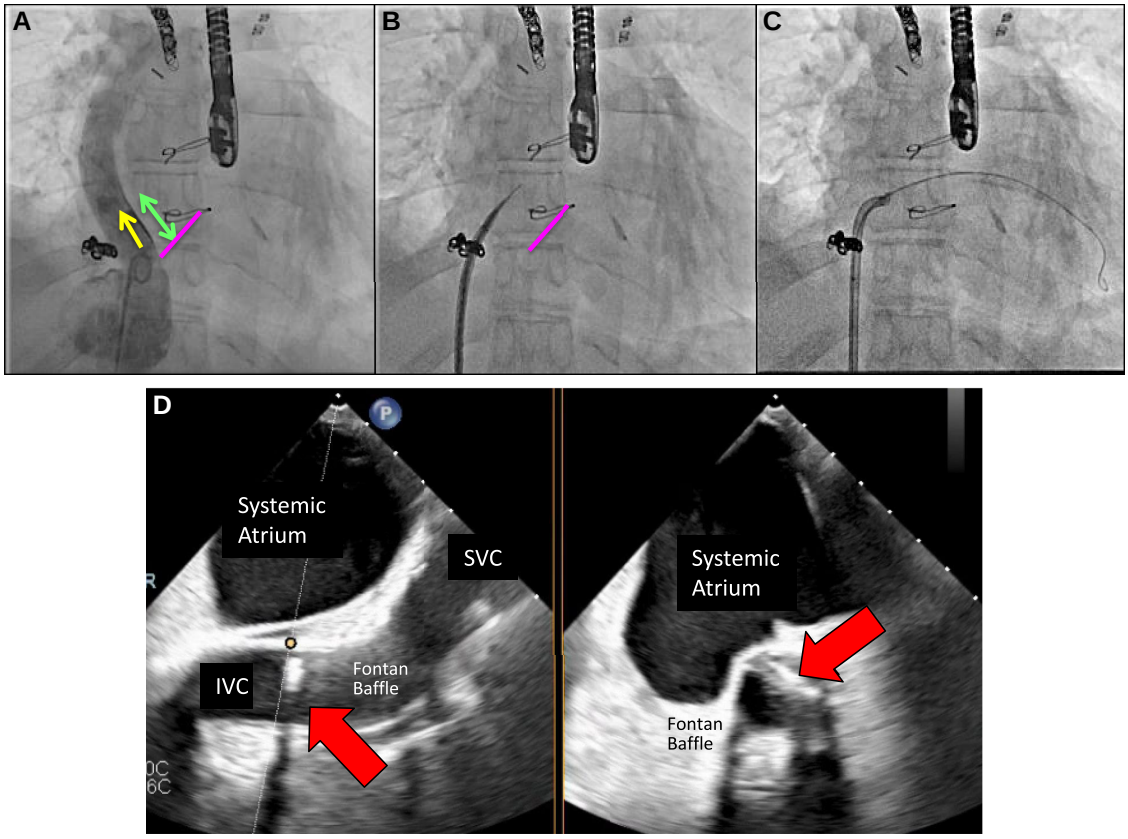

Figure 2 (*A*) Fluroscopy: contrast injection of extracardiac Fontan conduit. Puncture site identified 3–4 cm above the initial MitraClip XTW clip was manipulated into a medial position along A2/P2 and deployed after securing both the anterior and posterior leaflets within the clip. Following initial deployment, the regurgitation remained significant due to the broad span of the regurgitation jet and the myxomatous leaflets. In total, three MitraClips were placed to ensure a sufficient reduction in regurgitation. Two Abbott XTW clips were secured A2/P2, and 1 Abbott NTW clip was placed across A1/P1 (*Figure 3*). IVC/extracardiac Fontan anastomosis (single headed, yellow arrow) to allow the Mitraclip device to enter systemic atrium (double headed, green arrow) approximately 4–5 cm above the MV (single line, pink). (*B*) TEE: relationship of extracardiac Fontan conduit relative to systemic atrium. (*C*) Fluoroscopy: puncture through extracardiac Fontan conduit into the systemic atrium approximately 4–5 cm above MV with wire advanced to maintain position. (*D*) TEE: bi-plane image of BRK-1 Brockenbrough needle in extracardiac Fontan conduit (single headed, large red arrow) with puncture into systemic atrium.

atrium was difficult and required the use of Bovie cautery and manual force, but was successfully performed and a wire was placed into the systemic ventricle (*Figure 2*). The conduit puncture site was then balloon dilated to facilitate smooth passage of the Abbott MitraClip delivery system. The delivery system was advanced into the systemic atrium.

The initial MitraClip XTW clip was manipulated into a medial position along A2/P2 and deployed after securing both the anterior and posterior leaflets within the clip. Following the initial deployment, the regurgitation remained significant due to the broad span of the regurgitation jet and the myxomatous leaflets. In total, three MitraClips were placed to ensure a sufficient reduction in regurgitation. Two Abbott XTW clips were secured A2/P2, and 1 Abbott NTW clip was placed across A1/P1 (*Figure 3*).

At the completion of the case, moderate residual MR was observed by TEE. Final intra-procedural mean MV inflow gradient was 4 mmHg (heart rate 64 b.p.m.). Due to a significant right-to-left shunt across the procedural Fontan conduit, an 8 mm Amplatzer atrial septal defect occluder device was placed (*Figure 4*). Final mean Fontan pressure decreased to 16 mmHg following TEER intervention and final systemic atrial pressure was similarly reduced to 8 mmHg with a v-wave to 12 mmHg. A follow-up transthoracic echo showed a decrease in MR from severe to moderate regurgitation (*Figure 5*).

As of the time of this publication, the patient had no further congestive heart failure admissions. Her last transthoracic echocardiography, performed 4 months after the TEER procedure, showed mild to moderate residual mitral regurgitation, a mean MV gradient of 5 mmHg (HR 58 b.p.m.), and mild improvement in her systemic ventricular function.

Discussion

Nearly one-third of Fontan patients will have failure of their AV valve by 30 years of age.[1] Conventional surgical repair in the adult Fontan patient is high risk due to the presence of multiple comorbidities and prior sternotomies. For patients that undergo surgical AV valve repair, up to a third will have failure of their valve repair. Surgical AV valve replacement and repair is also associated with early mortality and morbidity.[1,6,7] Attempts to use transcatheter interventions to

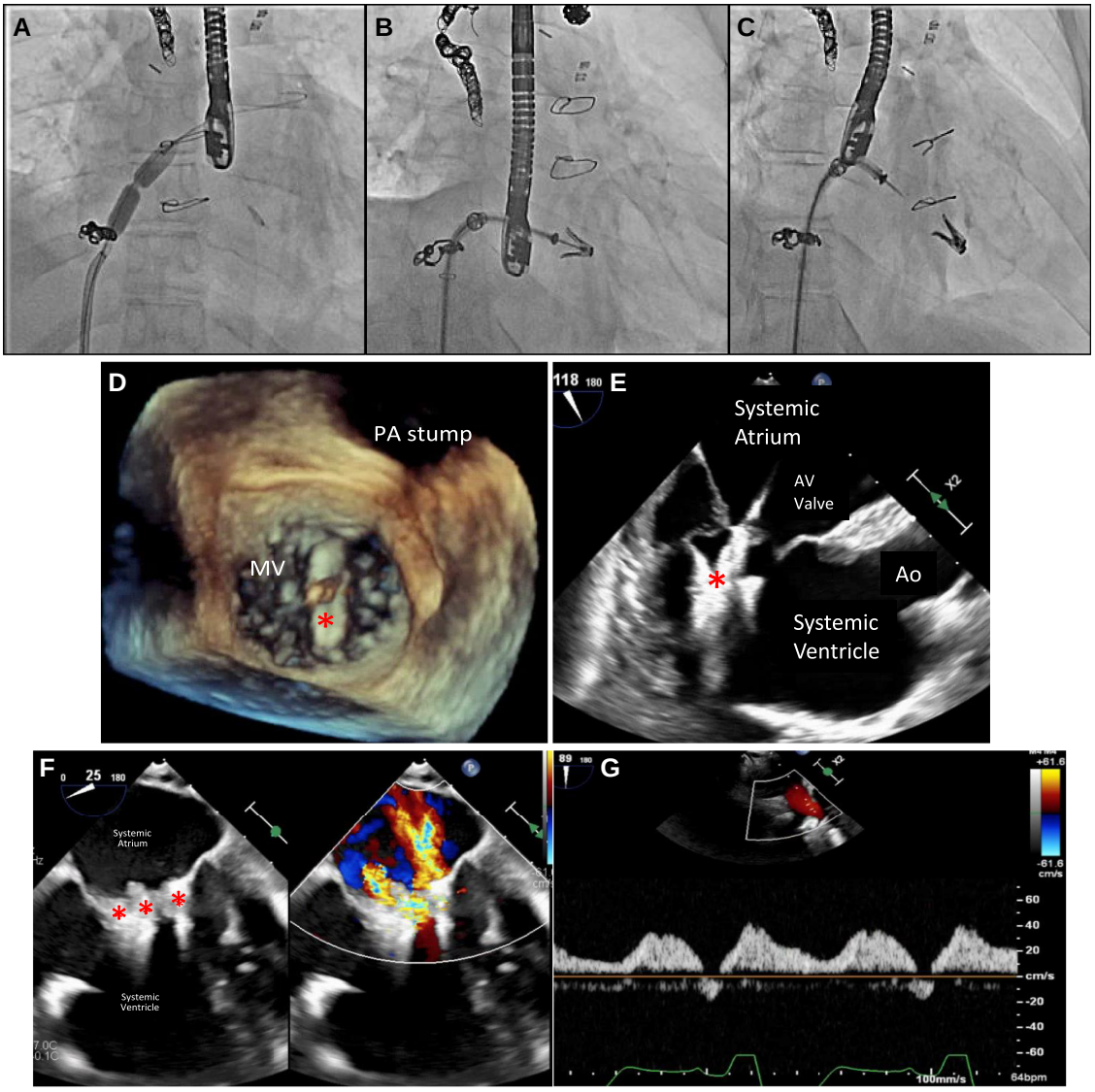

Figure 3 (*A*) Fluoroscopy: balloon dilation of Fontan puncture site into systemic atrium to facilitate smooth passage of MitraClip system. (*B*) Fluroscopy: Mitraclip positioned under mitral valve. (*C*) Fluroscopy: release of first Mitraclip. (*D*) TEE: 3D left atrial view of Mitraclip (*) positioned over anterior (A2) and posterior (P2) leaflets with the PA stump used for device orientation. (*E*) TEE: Mitraclip (*) positioned under anterior and posterior leaflets of MV to grasp leaflets. (*F*) TEE: post-procedural commissural view of MV following placement of three Mitraclips (*) with colour Doppler demonstrating moderate, residual MR. (*G*) TEE: pulse wave Doppler of LUPV demonstrating normalized systolic pulmonary vein flow.

address AV valve regurgitation in Fontan patients, including MitraClip and valve-in-valve implantation, have been described previously.[8–11] However, to date, there are no descriptions of TEER successfully performed in an adult extracardiac Fontan patient. The available literature describing the feasibility of extracardiac Fontan transcaval puncture is best described in the electrophysiology literature, for the purpose of ablation.[12] However, the transcaval access for ablation does not require a specific entry height into the systemic atrium, and is often performed via the IVC into the atrium. In contrast, the success of the TEER procedure depends on adequate height of the device delivery system above the AV valve (approximately 4–5 cm), and in this case required puncture directly through the surgical extracardiac conduit. The use of TEER in Fontan patients may provide an alternative to surgical or transplant intervention and prevent rapid progression to Fontan failure once significant AV valve regurgitation is present. The use of 3D models to evaluate the feasibility of transcatheter device delivery in a specific patient's anatomy may help guide optimal patient selection and procedural planning. To date, there only rare case reports available describing the use of TEER intervention to improve AVVR in adult congenital heart patients.[13,14] To our knowledge, this is the first successful published example of TEER through an extracardiac Fontan baffle puncture to successfully treat AV valve regurgitation in an adult.

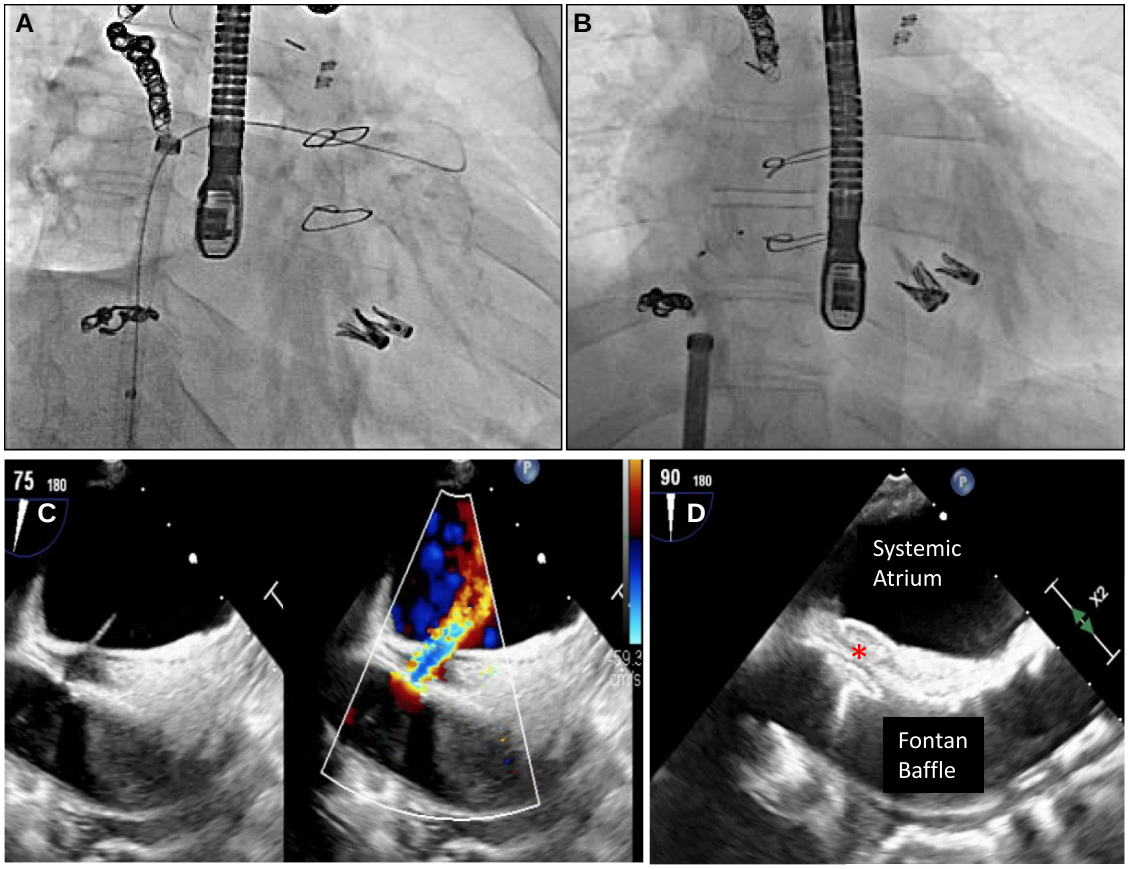

Figure 4 (*A*) Fluroscopy: three Mitraclip devices successfully deployed. Sheath positioned to deploy an Amplatzer Atrial Septal Occluder (ASO) device to close right to left shunt across Fontan conduit into systemic atrium. (*B*) Fluroscopy: release of 8 mm ASO device. (*C*) TEE: colour Doppler demonstrates right to left shunt from extracardiac Fontan baffle into systemic atrium causing significant desaturation in patient. (*D*) TEE: final position of the 8 mm ASO device (*) across Fontan conduit and systemic atrium.

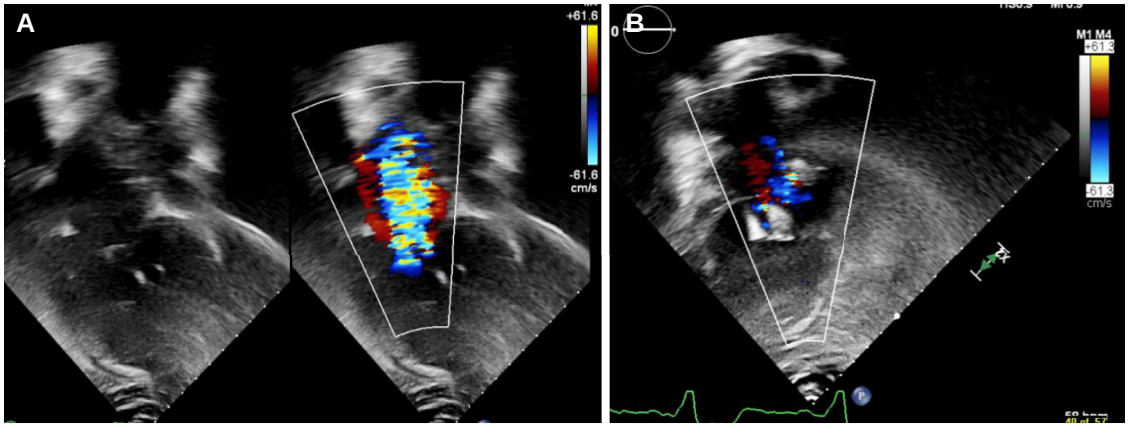

Figure 5 (*A*) TTE: pre-procedural commissural view of MV with colour Doppler demonstrating severe MR (*B*) TTE: 1-year follow-up scan of commissural view of MV with colour Doppler demonstrating mild to moderate residual MR with three Mitraclips.

Lead author biography

Dr Christiane Haeffele completed a combined medicine/pediatric residency at the Brigham and Women's Hospital and Boston Children's Hospital in Boston, MA, USA. She did her fellowship training in adult cardiology, adult congenital heart disease, and echocardiography at Stanford Hospital, Stanford, CA, USA. She is a Clinical Assistant Professor of Medicine and Pediatrics, the Director of the Structural ECHO program, and is the Program Director for the Advanced ECHO Imaging fellowship at Stanford. Her research interests include management of adults with congenital heart disease, echocardiography, structural heart disease, and catheter based interventions, and the complexities of heart transplant in the adult with congenital heart disease.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including imaging and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: Dr. Haeffele is a consultant and speaker for Edward Lifesciences. Dr. Rahul Sharma is a proctor and consultant for Abbott.

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Data availability

The data that support the findings of this study are available on request from the corresponding author, Dr. Christiane Haeffele. The data are not publicly available due to Health Insurance Portability and Accountability Act (HIPAA) privacy restrictions.

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