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CONGENITAL HEART DISEASE

CLINICAL CASE

Percutaneous Atrioventricular Valve Repair With MitraClip in Failed Adult Fontan Circulation for Tricuspid Atresia

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ABSTRACT

A 60-year-old woman with tricuspid atresia had previously undergone palliation with a Glenn procedure and a Blalock-Taussig shunt, and finally correction with a Fontan procedure. Twenty-five years later, severe atrioventricular valve regurgitation with concomitant valvular prolapse and cleft leading to heart failure was successfully treated using 1 single MitraClip XTW, confirming its effectiveness as a lower-risk alternative to surgery. The patient showed marked improvement of heart failure-related symptoms, with residual mild to moderate regurgitation and improved liver function. (JACC Case Rep. 2025;30:103272) © 2025 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/).

A 60-year-old woman was admitted to the cardiology department because of worsening exertional dyspnea, swollen legs, and liver dysfunction during the past 3 months. Physical examination revealed bilateral peripheral edema, ascites with enlarged liver, and irregular heartbeat with holosystolic murmur at the apex (3/6 Levine).

PAST MEDICAL HISTORY

At 3 years of age, the patient underwent a Glenn procedure to palliate tricuspid atresia (TA), followed at 29 years of age by a left Blalock-Tussig shunt. A Fontan procedure (bicaval-right atrial appendage-pulmonary artery anastomosis) was performed when she was 35 years old, with the contextual closure of the BT shunt. In the immediate postoperative period,

TAKE-HOME MESSAGES

- In older congenital heart failure patients with prohibitive surgical risk, the MitraClip procedure may be considered to effectively reduce the degree of valvular regurgitation and mitigate Fontan failure, alleviating symptoms and improving quality of life.
- In experienced hands, a single clip can potentially be used even in complex valvular anatomies, such as those with concurrent clefts and leaflet prolapse, leaving a 2-clip strategy as a valuable additional choice to mitigate future problems.
- However, although an improvement in heart failure symptoms has been observed, the long-term survival benefits have yet to be established.

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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

AF = atrial fibrillation

ASD = atrial septal defect

AVV = atrioventricular valve

LVEF = left ventricular ejection fraction

PDA = patent ductus arteriosus

PFO = patent foramen ovale

PMVR = percutaneous mitral valve repair

PVR = pulmonary vascular resistance

TA = tricuspid atresia

TEER = transcatheter edge-to-edge repair

VSD = ventricular septal defect

a postsurgical stenosis caused by a suture point at the proximal anastomosis of the left pulmonary artery was treated with stenting. During the previous 7 years, the patient had been admitted twice to the hospital on account of new-onset atrial fibrillation, with permanent atrial fibrillation being managed by β-blockers. In the 6 months before admission, she experienced right-sided heart failure and portal hypertension. She had been taking 2.5 mg apixaban twice daily because of low body weight (45 kg) and renal dysfunction, 100 mg atenolol once daily, and 75 mg furosemide twice daily.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for heart failure in late Fontan includes worsening of pulmonary hypertension, atrioventricular valve (AVV) regurgitation, and ventricular dysfunction.

INVESTIGATIONS

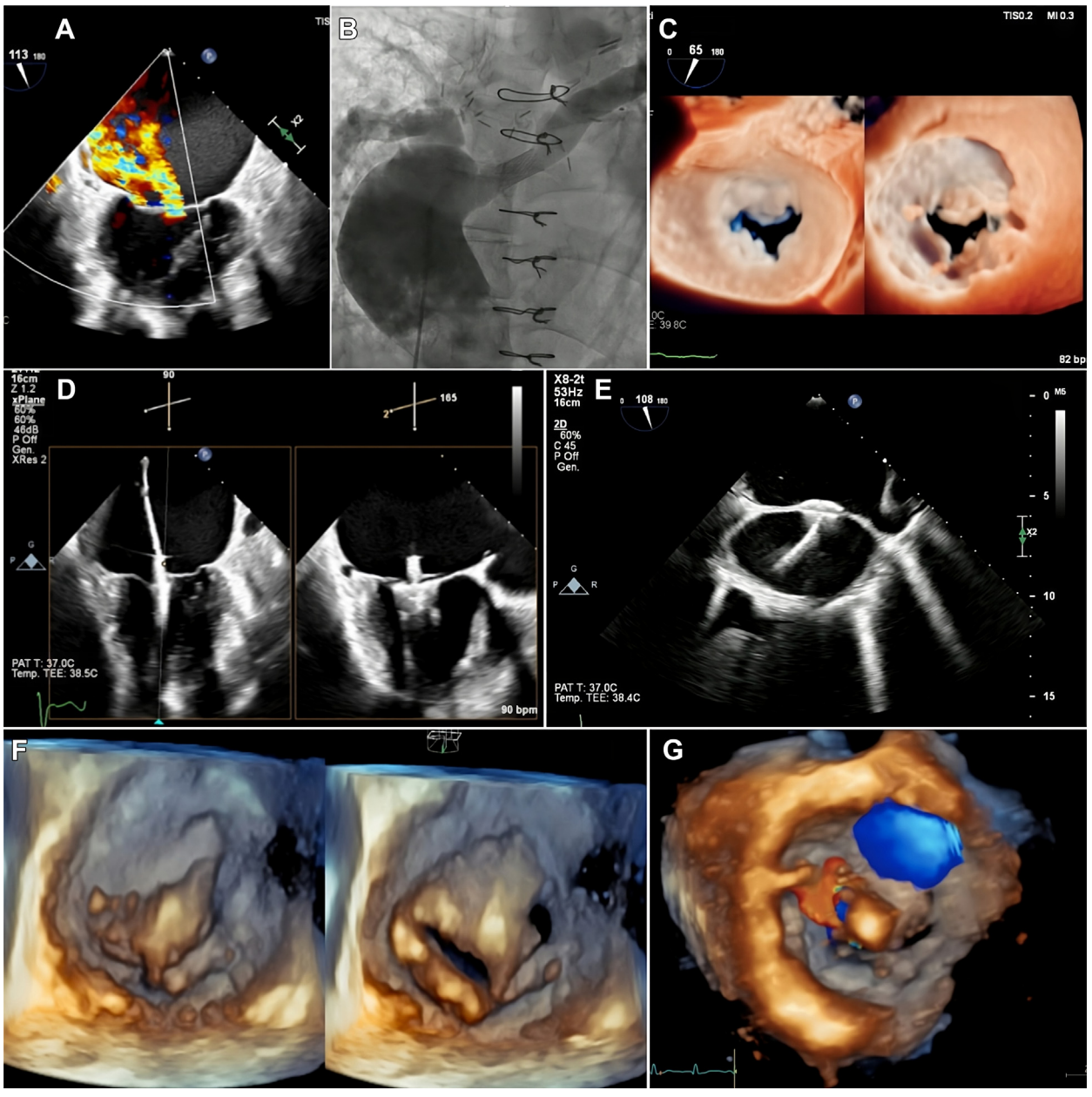
Electrocardiogram (ECG) revealed atrial fibrillation, with a mean heart rate of 75 beats/min. At transthoracic and transesophageal echocardiography, AVV was functionally tricuspid (**Figure 1**, Video 1), with a deep cleft (width, 6.5 mm) between P2and P3 and with anterior mitral leaflet (A2-A3 segments) prolapse causing severe valve insufficiency (**Figure 2**, Video 2). Annular dilatation and severe enlargement of both atria with sluggish blood flow were also present. The ejection fraction of the functionally single ventricle was preserved (LVEF, 50%-55%), with the rudimentary collateral right ventricle connected with a large VSD to the main chamber (**Figure 3**). The MVA was 5 cm^2, with a mean gradient of 2 mm Hg. Cardiac CT confirmed patency of the right atrium-pulmonary artery anastomosis and of the stent at the proximal anastomosis of the left pulmonary artery (**Figure 4**).

Right and left heart catheterization revealed increased pulmonary wedge pressures (V wave, 25 mm Hg; mean pulmonary capillary wedge pressure, 18 mm Hg), mean pulmonary artery pressure, 25 mm Hg; pulmonary vascular resistance, 3 WU; right atrial pressure, 25 mm Hg; left ventricular end-diastolic pressure, 15 mm Hg (**Figure 5**); and CO (Fick), 2.7 L/min.

MANAGEMENT

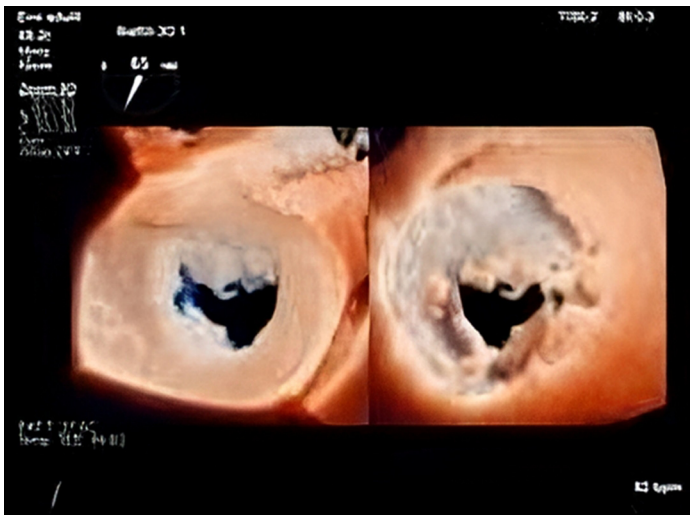
Fontan failure due to high pulmonary pressure on account of severe worsening of AVV insufficiency was diagnosed. After discussion by the heart team, the patient was considered for transcatheter edge-to-edge repair (TEER) because of the well-preserved, anatomically favorable leaflets and the high-risk profile for surgical AVV replacement. Because of relative contraindications (ascites, renal insufficiency, low weight, age, 3 previous operations, and, most importantly, moderate pulmonary hypertension with uncertain reversibility after correction), she was not added to the transplantation list. TEER was performed with the patient under general anesthesia, through transfemoral venous access. The guidewire was advanced to the interatrial septum, allowing for a central-posterior trans-septal puncture. Subsequently, a single MitraClip XTW was deployed centrally under TEE guidance (**Figure 6**). The tricuspid-type morphology of the AVV, caused by a deep cleft in the posterior leaflet, and its prolapsed anterior leaflet were additional challenges in performing the procedure. To address these issues, a single central clip was positioned slightly more clockwise than the line of coaptation, allowing the anterior leaflet to be grasped and the prolapse to be significantly reduced. Posteriorly, an attempt to grasp both leaflets separated by the cleft was made. This was partially successful, inasmuch as the lateral posterior leaflet was secured while part of the medial leaflet remained free, resulting in a mild to moderate residual regurgitation and a mean transvalvular gradient of 3 mm Hg (**Figure 7**, Video 3). The V-wave decreased from 27 mm Hg to 20 mm Hg, whereas the medium left atrial pressure went down from 18 mm Hg to 11 mm Hg. At the end of the procedure, the remaining right-to-left shunt, leading to significant desaturation, was closed with a 10-mm Amplatzer septal occluder device (**Figure 8**). The patient's postoperative course was uneventful.

VISUAL SUMMARY **Central Illustration of Atrioventricular Valve Repair With MitraClip in Failed Adult Fontan Circulation**



(A) Severe atrioventricular valve insufficiency. (B) Right heart catheterization. (C) Tricuspid-like morphology. (D) Deployment of single MitralClip XTW. (E) Deployment of 10-mm Amplatzer occluder device. (F) Clockwise position of MitralClip with movement of medial posterior leaflet. (G) Mild degree of residual regurgitation.

FIGURE 1 Atrioventricular Valve Morphology With Severe AVV Regurgitation



The patient's particularly challenging AVV morphology: deep cleft in the posterior leaflet at P2 level, resulting in a tricuspid-type morphology, and prolapse of the anterior leaflet, especially at A2. AVV = atrioventricular valve.

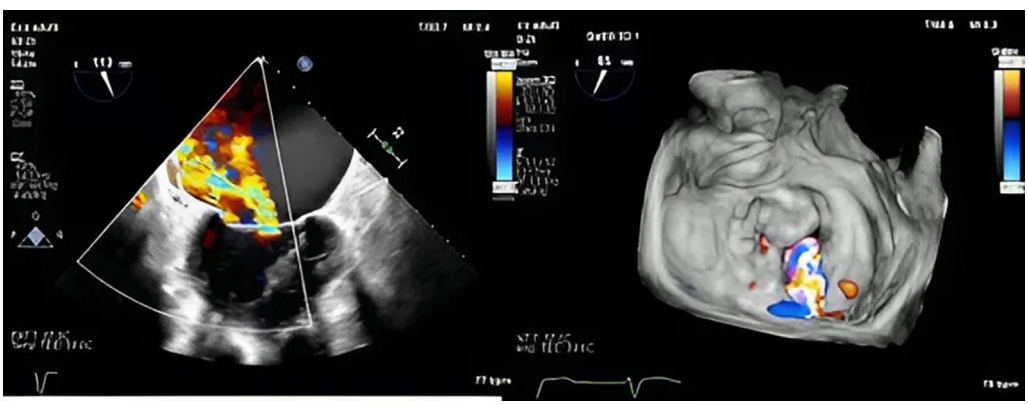
OUTCOME AND FOLLOW-UP

At her 2-month follow-up visit, the patient reported improvement of symptoms (from NYHA functional class IV to class III); no sign of right-side heart failure was present. Transthoracic echocardiography confirmed correct placement of the MitraClip XTW at the site of deployment and indicated mild, stable regurgitation with a normal anterograde gradient (**Figures 9 and 10**, Videos 4 and 5). Ventricular function decreased slightly, with a LVEF of 45% to 50%. The patient reported 1 transient ischemic attack, but CT scans were normal. No further episodes occurred after apixaban was increased to 5 mg.

DISCUSSION

Tricuspid atresia, a complex congenital heart defect characterized by absence of the tricuspid valve, requires multiple early palliation culminating in a Fontan procedure in order to guarantee survival.[1] Despite a 10-year survival rate that can approach 90%, Fontan patients can experience a rise in central venous pressure and, as a consequence, mixed pulmonary hypertension, leading over time to other cardiovascular complications, such as heart failure, cyanosis, arrhythmias, pulmonary embolism, and portal hypertension.[2] Treatment options for Fontan failure worsened by severe AVV regurgitation typically include surgical mitral valve repair or replacement and, in advanced cases, heart transplantation.[3,4] In our patient, the decision against surgery was made on account of her high morbidity

FIGURE 2 2D and 3D TEE Showing Severe AVV Regurgitation

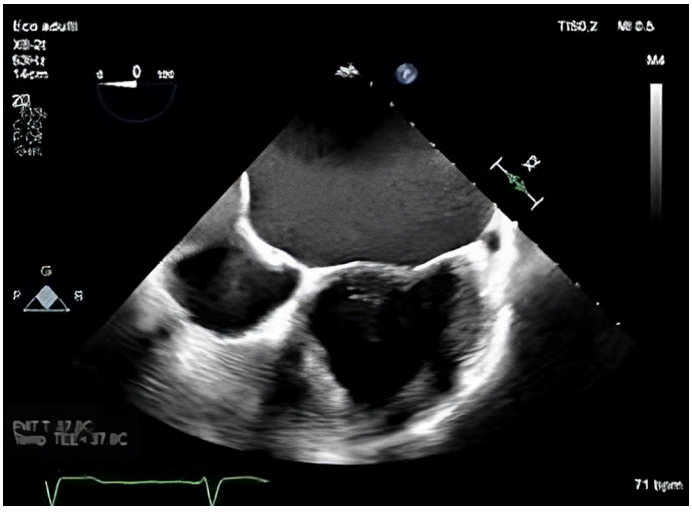


Annular dilatation and complex anatomy of leaflets, with a cleft at P2-P3 and a A2-A3 prolapse causing severe AVV insufficiency. AVV = atrioventricular valve; TEE = transesophageal echocardiography.

and mortality risks (PEACH score, >3 for mitral valve repair and for replacement), history of multiple cardiac surgeries, elevated pulmonary artery and atrial pressures, recurrent episodes of heart failure, atrial fibrillation, and liver and renal dysfunction.

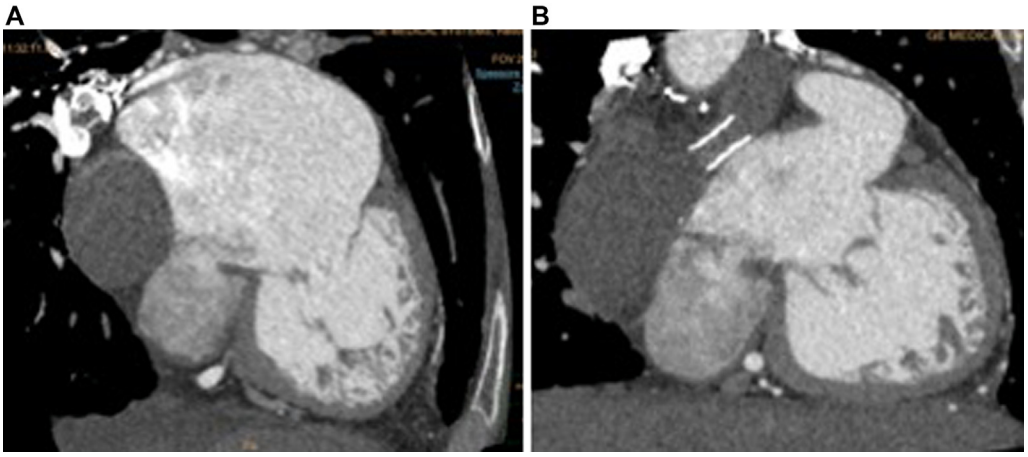
Placement of a MitraClip, a TEER device that mimics the Alfieri stitch used in surgical mitral repair, may offer a lower-risk alternative to surgery by avoiding the need for sternotomy and cardiopulmonary bypass. Given the favorable baseline anatomo-functional characteristics of the mitral valve, the choice of using a MitraClip XTW device was driven by the central location of the main regurgitant jet, by the size of the leaflets, and by the annular dilatation requiring a long-arm mechanical device. A double-clip strategy with converging clips is a common alternative in the case of larger myxomatous valves, but the leaflet sizes in our patient were considered insufficient to consider that strategy because of its intrinsic risk of increasing the transvalvular gradient. Given that the main indication for the procedure is to reduce pulmonary hypertension in order to recover an

FIGURE 3 **Univentricular Heart Morphology**



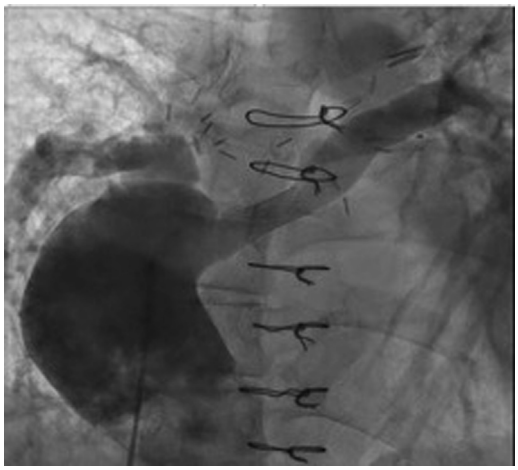
The single functional left ventricle supporting circulation, with a hypoplastic right ventricle characteristic of tricuspid atresia.

FIGURE 4 **Posterior and Anterior Coronal Plane at Heart CT**



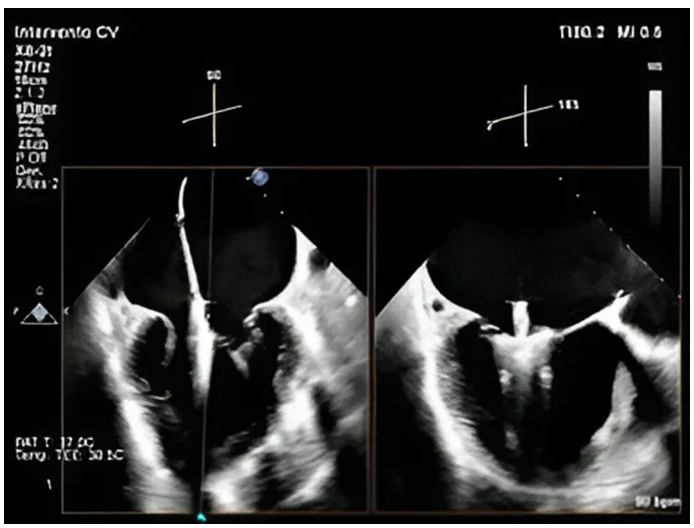
Preoperative cardiac CT, with posterior coronal plane (A) and anterior coronal plane (B), confirmed the severe atrioventricular valve insufficiency, preserved systolic function of main ventricle (left ventricular ejection fraction, 50%), and rudimentary right ventricle connected with a ventricular septal defect to the main chamber (functionally single ventricle). Fontan and drug-eluting stent on left pulmonary artery were patent. CT = computed tomography.

FIGURE 5 Right Heart Catheterizations



Right and left heart catheterization revealed increased pulmonary wedge pressures, with pulmonary artery pressure 25 mm Hg, right atrial pressure 25 mm Hg, and left ventricular end-diastolic pressure 15 mm Hg. The injected contrast medium allowed identification of right atrial enlargement with significant blood stasis, patency of the stent in the left pulmonary artery, and Fontan circulation.

FIGURE 6 Transesophageal Echocardiographic Guidance During MitralClip Deployment



Placement of a single central clip slightly more clockwise from the line of coaptation anterior leaflet to be captured, significantly reducing the prolapse. Posteriorly, an attempt was made to secure both leaflets separated by the cleft.

efficient Fontan circulation, a single-clip strategy was mandatory despite the prolapse or cleft potentially impairing clip placement and stability.[5,6]

A 2022 review by Silini and Iriart[7] analyzed case reports and series on TEER in patients with congenital heart failure, including 5 cases involving the left ventricle. When cases reporting the presence of clefts are analyzed in greater detail, it appears that in expert hands, 2 MitraClips positioned in parallel or, interestingly, in a V configuration, effectively reduce the degree of valvular regurgitation to mild or mild to moderate. Our report describes a novel approach, ie, placement of a single clip, which achieves similar results but reduces the risk of mitral stenosis and preserves the option of a second clip as an additional therapeutic measure should the regurgitation worsen in the future. As in most of the reports analyzed, our patient experienced marked improvement in heart failure symptoms, no decline in systemic ventricular function, and no intraprocedural death.

CONCLUSIONS

Our case report adds to the growing body of literature on TEER in patients with complex cyanotic congenital heart disease. To our knowledge, it is the first documented case of successful deployment of a MitraClip device on very challenging AVV anatomy (multiple eccentric jets with concomitant mitral cleft and prolapse) in an older woman with Fontan failure. This highlights a new perspective in the treatment of these highly complex patients.

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Dr Denti has received speaker honoraria from Abbott; and has been a consultant for Approxima, HVR, InnovHeart, Picardia, and Simulands. Dr Maisano has received grant and/or research institutional support from Abbott, Medtronic, Edwards Lifesciences, Biotronik, Boston Scientific Corporation, NVT, Terumo, Venus; consulting fees and honoraria (personal and institutional) from Abbott, Boston Scientific, Medtronic, Edwards Lifesciences, Xeltis, Cardiovalve, Occlufit, Simulands, Mtex, Venus, Squadra, Valgen, croivalve; and royalty income/IP rights from Edwards Lifesciences and is a shareholder (including share options) of Magenta, Transseptalsolutions, and 4Tech. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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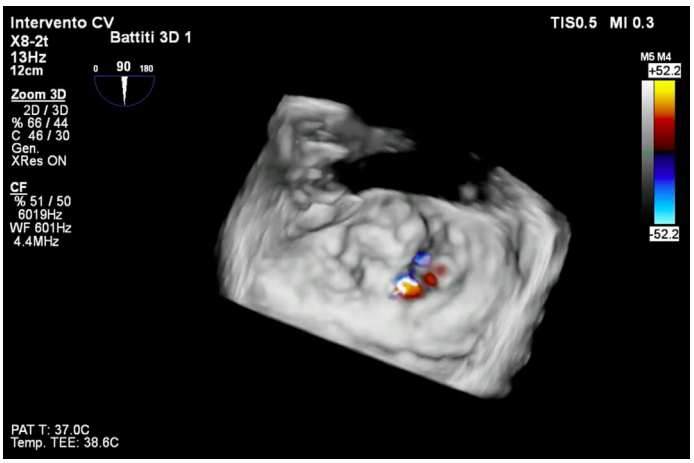
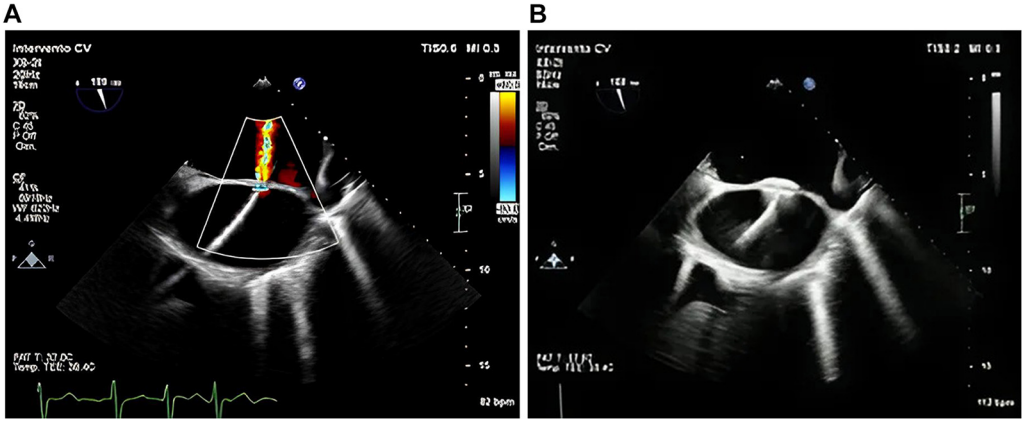
FIGURE 7 **Residual Mild to Moderate Atrioventricular Valve Insufficiency After Final Deployment of MitraClip**



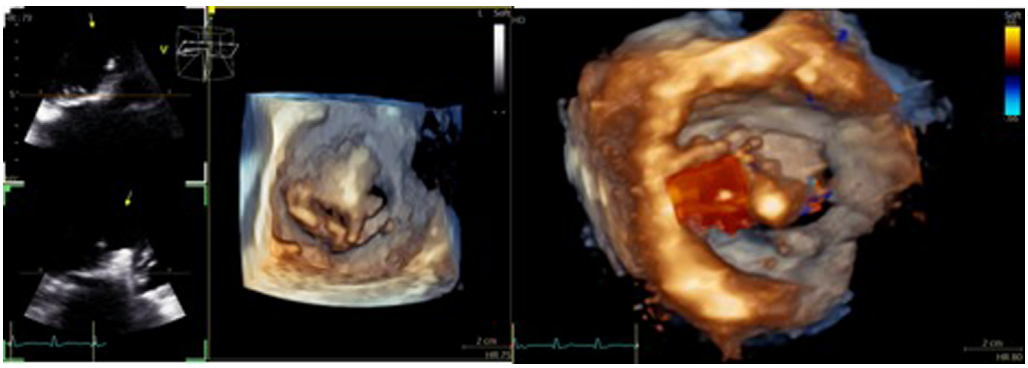
Partially successful deployment, with part of the medial leaflet of the cleft remaining free, resulting in a mild to moderate residual regurgitation and a mean transvalvular gradient of 3 mm Hg.

FIGURE 8 **Deployment of 10-mm Amplatzer Septal Occluder**



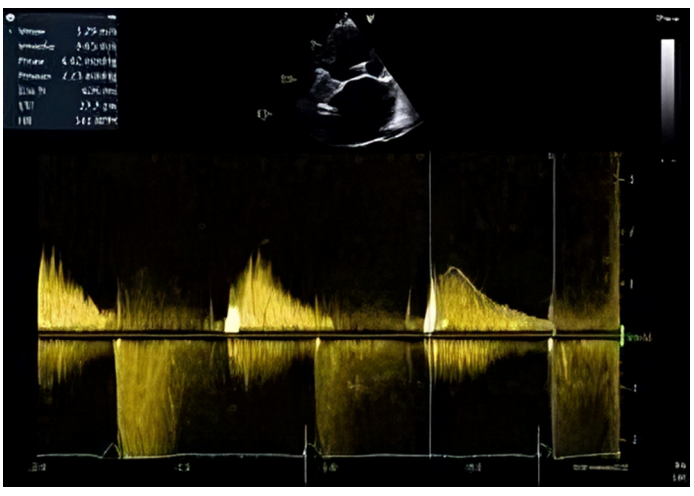
Because of significant oxygen desaturation due to remaining right-to-left shunt (A), a 10-mm Amplatzer septal occluder device was deployed at the site of trans-septal puncture (B).

FIGURE 9 Ventricular and Atrial Views of MitralClip at Transesophageal Echocardiographic Follow-Up



The MitralClip was deployed centrally in a mild clockwise position: the upper arm of the clip grasped the anterior leaflet, and the lower arm grasped the lateral half, successfully grasping both leaflets divided by the cleft. The result was mild to moderate residual regurgitation from the medial side resulting from the partial grasping of the P2 medial cleft.

FIGURE 10 Transvalvular Gradient at TTE Follow-Up



Medium gradient of 2.23 mm Hg was highlighted at TTE at follow-up, confirming the good results of the procedure. TTE = transesophageal echocardiography.

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KEY WORDS **congenital heart disease, Fontan circulation, MitraClip, transcatheter-based treatments, tricuspid atresia, univentricular heart**

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