UC San Diego

UC San Diego Previously Published Works

Title

A case report of percutaneous MitraClip implantation in an adult with a double-outlet right ventricle.

Permalink

https://escholarship.org/uc/item/2rw5t8b3

Journal

European heart journal. Case reports, 7(6)

ISSN

2514-2119

Authors

Kumar, Preetham
Gordon, Brent M
Kheiwa, Ahmed
et al.

Publication Date

2023-06-01

DOI

10.1093/ehjcr/ytad247

Peer reviewed

eScholarship.org Powered by the California Digital Library
University of California

ESC European Society of Cardiology

European Heart Journal - Case Reports (2023) **7**, 1–5
https://doi.org/10.1093/ehjcr/ytad247

CASE REPORT
Adult congenital heart disease

A case report of percutaneous MitraClip implantation in an adult with a double-outlet right ventricle

Preetham Kumar [1]*, Brent M. Gordon[2], Ahmed Kheiwa[2], and Islam Abudayyeh[3,4]

[1]Division of Cardiology, Department of Medicine, University of California, Riverside, 900 University Ave, Riverside, CA 92521, USA; [2]Division of Pediatric Cardiology, Department of Pediatrics, Loma Linda University Children's Hospital, Loma Linda, CA, USA; [3]Division of Cardiology, Department of Medicine, Loma Linda University Medical Center, Loma Linda, CA, USA; and [4]Section of Cardiology, Department of Medicine, Veterans Administration Loma Linda Healthcare System, Loma Linda, CA, USA

Received 16 August 2022; first decision 2 November 2022; accepted 17 May 2023; online publish-ahead-of-print 19 May 2023

Background Atrioventricular valve regurgitation (AVVR) is present in up to 75% of Fontan patients, and it is associated with an increased risk of Fontan circulation failure, morbidity, and mortality. Traditional treatment options include surgical repair vs. surgical replacement. We present, to the best of our knowledge, one of the first cases of successful trans-catheter repair of severe common AVVR using the MitraClip device.

Case summary A 20-year-old male with a history of double-outlet right ventricle (DORV) with unbalanced common atrioventricular canal to the right ventricle, severely hypoplastic left ventricle, and total anomalous pulmonary venous return status post-Fontan procedure presented with progressively worsening dyspnoea on exertion. Transoesophageal echocardiogram demonstrated severe common AVVR. After discussion of the case during the adult congenital heart disease multidisciplinary conference, patient underwent successful placement of two MitraClip devices, reducing the regurgitation from torrential to moderate.

Discussion MitraClip therapy can be used to alleviate symptoms in patients deemed as high risk for surgery. However, careful attention must be paid to haemodynamics before and after clip placement, which may predict short-term clinical outcomes.

Keywords Common atrioventricular valve regurgitation • Fontan palliation • MitraClip • Case report

ESC curriculum 9.7 Adult congenital heart disease • 4.9 Multivalvular disease • 2.2 Echocardiography • 6.1 Symptoms and signs of heart failure • 7.1 Haemodynamic instability

Learning points

- Atrioventricular valve regurgitation is common in patients with Fontan circulation, and it can result in detrimental haemodynamic consequences, requiring timely intervention.
- Catheter-based leaflet repair techniques using devices such as the MitraClip can be a feasible strategy to address atrioventricular valve regurgitation in selected cases.
- Once the atrioventricular valve regurgitation is successfully treated with a trans-catheter device, the Fontan baffle puncture–site leak should be cautiously and meticulously assessed to ensure haemodynamics, including blood pressure and oxygen saturation, are stable, and systemic ventricle function is adequate.

* Corresponding author. Tel: +1 951 827 1012, Email: pk.kumarp.316@gmail.com
Handling Editor: Anastasia Egorova
Peer-reviewers: David Blusztein; A. Shaheer Ahmed
Compliance Editor: Sara Monosilio
Supplementary Material Editor: Gonçalo Costa
© The Author(s) 2023. Published by Oxford University Press on behalf of the European Society of Cardiology.
This is an Open Access article distributed under the terms of the Creative Commons Attribution-NonCommercial License (https://creativecommons.org/licenses/by-nc/4.0/), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact journals.permissions@oup.com

Introduction

Atrioventricular valve regurgitation (AVVR) is a common long-term complication in adult patients with Fontan palliation and represents a poor prognostic factor with an increased risk of Fontan circulation failure, morbidity, and mortality.[1] Traditionally, treatment options were limited to surgical repair or replacement in low-risk patients vs. heart transplantation in high-risk patients. We present, to the best of our knowledge, one of the first cases of successful trans-catheter repair of severe common AVVR using the MitraClip device and the associated haemodynamic consequences.

Timeline

Infancy	Patient was born with a double-outlet right ventricle (DORV) associated with an unbalanced common atrioventricular canal, severely hypoplastic left ventricle, and total anomalous pulmonary venous return (TAPVR). He subsequently underwent pulmonary artery banding, left-sided bidirectional Glenn procedure, and TAPVR repair.
Age 5	Patient underwent right-sided pericardial extracardiac lateral tunnel Fontan completion.
Day 1	A 20-year-old male presented to our adult congenital heart disease (ACHD) centre with progressively worsening dyspnoea on exertion. Transoesophageal echocardiogram (TEE) demonstrated a common atrioventricular valve with annular dilatation and severe regurgitation, a severely enlarged common atrium, and a systemic (right) ventricular ejection fraction of 20%.
Day 2	Patient's case was discussed during the ACHD multidisciplinary conference.
Day 3	Patient underwent successful placement of two MitraClip devices, reducing the severity of AVVR from torrential to moderate.
Day 4	Patient was discharged home.
Day 10	Patient was readmitted for worsening dyspnoea. Following consultation with the ACHD team, patient was urgently taken to the cardiac catheterization laboratory for trans-catheter closure of the Fontan baffle puncture–site leak. However, patient coded following intubation, and he passed away shortly after.

Case presentation

A 20-year-old male with a history of heterotaxy syndrome, dextrocardia, systolic ventricular failure of the systemic ventricle due to DORV associated with an unbalanced common atrioventricular canal, severely hypoplastic left ventricle, and TAPVR presented to our ACHD centre with progressively worsening fatigue and worsening dyspnoea. Of note, he underwent pulmonary artery banding in the 1st month of life followed by left-sided bidirectional Glenn procedure and TAPVR repair at 8 months of age and ultimately right-sided pericardial extracardiac lateral tunnel Fontan completion at 5 years of age. Other relevant history include elevated Fontan pressures, Fontan-associated liver disease, chronic hypoxic respiratory failure, mild-to-moderate intellectual disability, cleft lip and palate status post repair, bilateral conductive hearing loss, asplenia, and obesity.

Vital signs were notable for blood pressure 90–100/60–70 mmHg and SpO_2 80–90% on room air. Physical examination was notable for a 3/6 holosystolic murmur not only heard loudest at the right upper sternal border but also heard all over the precordium and the back. Home medications were notable for aspirin 81 mg, carvedilol 25 mg, chlorothiazide 500 mg, enalapril 10 mg, furosemide 40 mg, sildenafil 20 mg, and spironolactone 25 mg.

Initial evaluation, including transthoracic echocardiogram (TTE) and TEE, demonstrated a common AV valve with annular dilatation and severe regurgitation, a severely enlarged common atrium, and a systemic (right) ventricular ejection fraction of 20%. The AVV leaflets were thickened, with dominant anterior and posterior bridging leaflets and smaller dysplastic leaflets in the medial and lateral commissures (*Figure 1A* and *B* and Supplementary material online, *Videos S1* and *S2*). These leaflets demonstrated excessive mobility and prolapsed into the atrium, resulting in a wide and torrential medioposterior-directed regurgitant jet involving most of the valve and reaching the pulmonary vein inflows. Of note, the Nyquist limit setting on the echocardiogram was 60 cm/s and there were large V-waves.

After a multidisciplinary discussion, surgical repair of the common AVV was deemed high risk due to underlying severely reduced single ventricular systolic function and associated cirrhosis. Similarly, heart transplantation was deemed very high risk in the setting of a poor social support system exacerbated by intellectual disability of the patient. A decision was therefore made to proceed with trans-catheter repair of the common AVV using the MitraClip system (Abbott Cardiovascular, Plymouth, MN, USA). Computed tomography angiography of the chest was obtained for preoperative evaluation (*Figure 2*).

Baseline haemodynamics during cardiac catheterization revealed moderately elevated Fontan pressures of 18 mmHg, mildly elevated pulmonary capillary wedge pressure of 13 mmHg, and pulmonary vascular resistance of 1.5 Wood units. Baseline systemic oxygen saturation was 87% in the setting of multiple veno-venous collaterals.

After successfully accessing the right and left femoral veins with 8 Fr sheaths and right femoral artery with a 5 Fr sheath and induction of general anaesthesia using a combination of propofol and midazolam, trans-baffle puncture of the Fontan tunnel was performed with fluoroscopy and TEE guidance. Considering underlying dextrocardia and the orientation of the AVV, the puncture was directed counterclockwise and positioned just above the mid-point of the valve on fluoroscopy. Under TEE guidance, the tenting was positioned such that the guide sheath entered the common atrium horizontally and levelled with the centre of the AV valve. A 0.035″ Confida wire was advanced across the baffle and used to facilitate the delivery of a 22 Fr steerable guide catheter and dilator into the atrium. Complicating considerations included the presence of an intra-atrial web, a remnant of the intra-atrial septum, and a left-sided Fontan baffle (*Figure 3*). Additionally, the delivery system needed to be mis-keyed at 180° and the sheath had to be inverted so that it flexed to the patient's right due to the dextrocardia. As the atria and systemic ventricle were horizontal, the system did not require much flexing of the guide catheter. Once a puncture was successfully made, an XTW clip was prepared in the usual manner and introduced via the catheter on the clip delivery system. Once in position above the valve and using the *en-face* view, the orientation of the clip was rotated to intersect the largest of the regurgitation jets. After a couple of attempts and gauging the clip movement in this anatomy, the leaflets were successfully grasped and anchored. This reduced the regurgitation from torrential to severe. A second XTW clip was prepared and advanced close to and slightly lateral to the first clip to better address the valve regurgitation and anchor the first XTW clip given the high mobility of the leaflets (*Figures 1C* and *D* and *4* and Supplementary material online, *Videos S3* and *S4*). This further reduced the regurgitation from severe to moderate. After successful delivery of the clips, Fontan baffle puncture–site closure was considered. However, evaluation with angiography and TEE revealed minimal right-to-left shunting by colour flow, and the

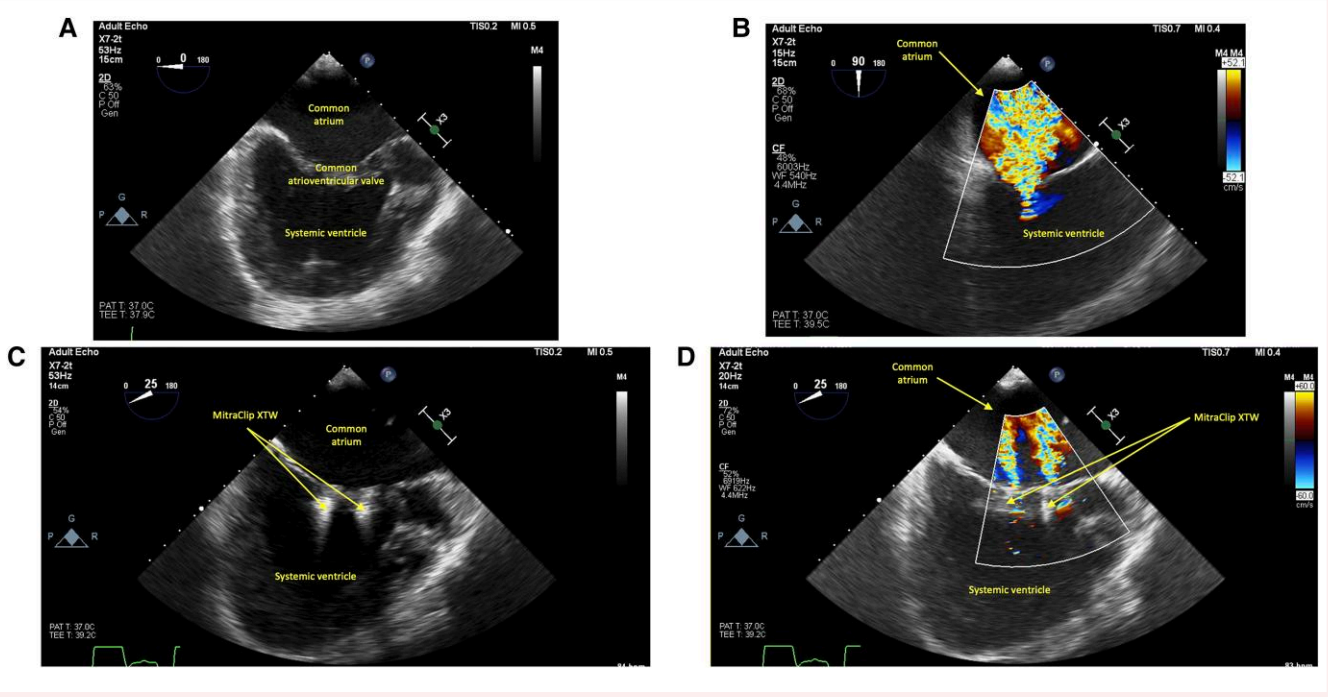

Figure 1 Transoesophageal echocardiogram demonstrating (*A*) an enlarged common atrium, (*B*) a deformed common atrioventricular valve associated with severe regurgitation, (*C*) the two MitraClip XTW, and (*D*) an improvement in regurgitation severity.

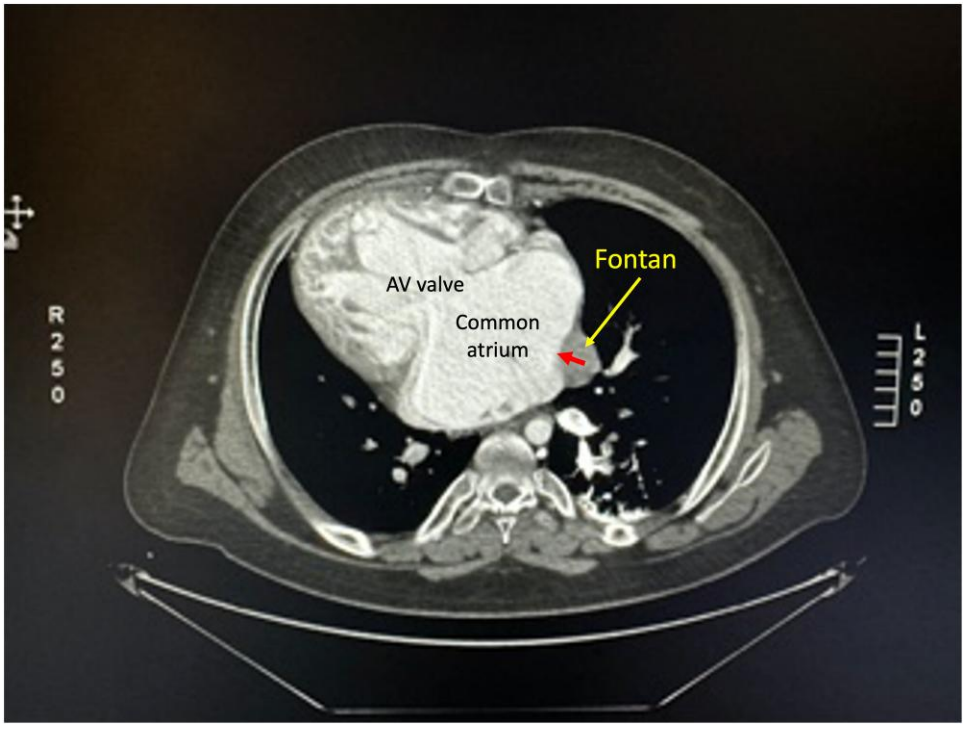

Figure 2 Computed tomography angiography of the chest approximating the Fontan baffle puncture site (black arrow). AV = atrioventricular.

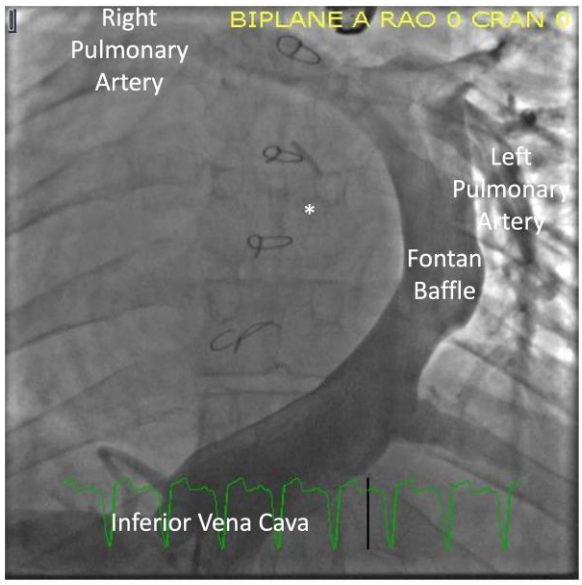

Figure 3 Fluoroscopic image demonstrating the complex anatomy. * denotes the location of the common atrium as a result of dextrocardia.

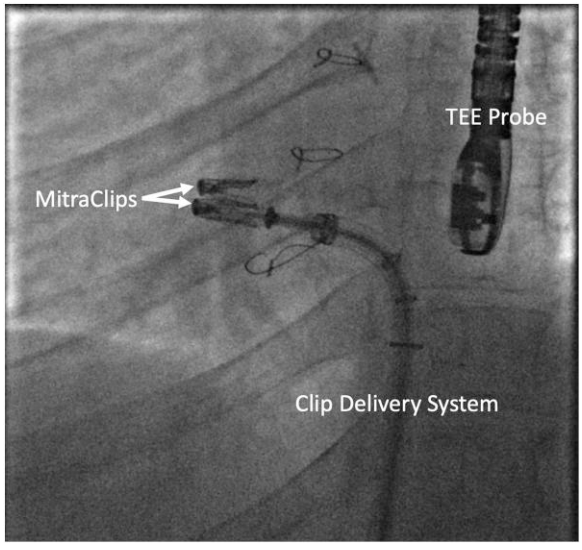

Figure 4 Fluoroscopic image showing the positions of the two MitraClip XTW.

patient maintained stable systemic oxygen saturations at 87%, so it was not performed. Of note, the systemic ventricular ejection fraction remained unchanged at 20%. The patient was discharged home in stable condition.

Six days later, the patient was readmitted for worsening dyspnoea and systemic oxygen desaturations. Chest radiography was negative. Transthoracic echocardiography (TTE) revealed worsening right-to-left shunting through the Fontan baffle puncture site by colour flow and saline contrast study. This was postulated to be due to a decrease in common atrium pressure post successful MitraClip placement, while the Fontan pressure remained constant and elevated. Imaging showed the common atrium had shrunk in size since the MitraClip procedure. A decision was made to close the Fontan baffle puncture site. Unfortunately, induction of anaesthesia was complicated by worsening hypoxemia and cardiac arrest with unsuccessful cardiopulmonary resuscitation.

Discussion

This case report demonstrates that the MitraClip device can be used to successfully treat severe common AVVR in patients with underlying complex congenital heart disease, describes relevant anatomic considerations when seeking such an approach, and highlights the haemodynamic sequelae of the procedure.

Since all patients with Fontan circulation have chronic heart failure, they are often managed similarly to non-Fontan patients with heart failure. However, given the risk of precipitating a low-output state, initiation, and continuation of treatments should be performed with caution and take into account haemodynamic consequences. The heart failure can be worsened by several conditions, including, but not limited to, worsening systemic ventricular function and AVVR. These circumstances yield an elevated common atrial filling pressure, thereby reducing the preload and cardiac index and raising the pulmonary artery pressure, which reflects on the Fontan pressures. In addition, Fontan pressures can also be elevated due to pulmonary vascular changes. For instance, in our patient, after the severe common AVVR was repaired, the common atrial pressure dropped, worsening the right-to-left shunt through the Fontan puncture site, giving rise to hypoxemia.

Patients with congenital heart disease often undergo multiple intricate surgeries to have their complex haemodynamic derangements corrected, increasing the risk for poor outcomes with any further procedure. Percutaneous management can significantly reduce this risk. Furthermore, in cases where the operative risk is particularly high, such as in the one presented here, treatment options are limited, and temporizing the patient's condition with a less risky technique can delay the need for further surgical intervention.

Blusztein *et al.*[2] and Guerin *et al.*[3] recently showed that the MitraClip device can be used to treat severe systemic tricuspid regurgitation (TR) in patients with complex congenital heart disease. Our case report is, to the best of our knowledge, one of the first to report the utilization of the MitraClip device to treat severe common AVVR.

Five main types of AVVR exist are as follows: primary TR, secondary TR, primary mitral regurgitation (MR), secondary MR, and common AVVR. Primary TR and MR occur due to structural abnormalities of the valve apparatus, whereas secondary TR and MR occur secondary to ventricular and annular dilation. Common AVVR, seen in patients with a common AVV, occurs secondary to valve dysplasia, leaflet prolapse, and tethering.[4]

Unrepaired moderate-to-severe AVVR has been shown to worsen outcomes in patients who underwent any form of Fontan surgery in the past.[5] Consequently, treatment of severe AVVR is often promptly sought. Options for treating common AVVR are primarily surgical, and they include repair, closure, and replacement. However, literature regarding non-surgical management of severe common AVVR is scarce.

MitraClip, a trans-catheter leaflet edge to edge repair device which reduces mitral valve regurgitation by approximating the anterior and posterior leaflets, has been shown to be safe and effective in patients with primary and secondary MR.[6]

Although treatment of common AVVR with MitraClip is technically challenging, it is feasible, especially with a good understanding of the anatomy, giving physicians a novel non-invasive option for treating this type of valvular disease. The specific access and device approach should be customized to the anatomy of the patient. In our patient, the MitraClip XTW was chosen over MitraClip NTR given the large orifice, flail and redundant tissue, and multi-faceted morphology of the AV valve. XTW clips, compared with NTR clips, are 50% wider at 6 mm and 33% longer at 12 mm, allowing more tissue to be captured and adding fixation to the anchoring, which in turn improves the likelihood of stabilization.

Furthermore, careful considerations to the haemodynamics of the Fontan baffle puncture–site closure at the time of MitralClip placement and of the common atrium and Fontan pressures post-procedure should be made. In our patient, a bubble study following MitraClip placement as a means of identifying any Fontan leak at an early stage could have been considered. However, in retrospect, we noticed colour flow across the Fontan leak, so we did not think a bubble study would have added significant information. We believe that the patient's haemodynamics, over time, progressively changed with the drop in atrial pressures. Similar to atrial septal defect (ASDs), a bubble study, compared with colour flow and haemodynamic assessment, is less of an indicator of a shunt. On a similar note, based on the presence of a significant right-to-left shunt at rest that was noted during our patient's second presentation, we could have considered closing the Fontan baffle puncture site using intra-cardiac echocardiogram under local anaesthesia, thereby avoiding general anaesthesia and the precarious haemodynamic changes. Lastly, these patients should be followed by a physician or team familiar with ACHD management in the outpatient setting.

Conclusions

Reduction of severe common AVVR in Fontan patients is technically feasible with trans-catheter MitraClip therapy. Close evaluation of haemodynamics before and after clip placement is necessary to determine volume and shunt changes, which may predict short-term clinical outcomes.

Lead author biography

Preetham Kumar is currently working as a Clinical Instructor of Medicine at University of California, Riverside. He earned his Bachelor of Science in Biomedical Engineering: Premedical from University of California, Irvine, and his Doctor of Medicine from Virginia Commonwealth University. Upon completion of Internal Medicine Residency at Huntington Hospital, he worked as a clinical research fellow under interventional cardiologist Dr Jonathan Tobis at University of California, Los Angeles, followed by as a cardiac hospitalist at University of Washington. He will be starting a general cardiology fellowship in July 2023 at University of California, Riverside.

Supplementary material

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

Acknowledgment

I am grateful to Drs. Gordon, Kheiwa, and Abudayyeh for this wonderful opportunity and their invaluable patience and support.

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

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

Conflict of interest: None declared.

Funding: None declared.

Data availability

The data underlying this article will be shared upon reasonable request to the corresponding author.

References

1. King G, Ayer J, Celermajer D, Zentner D, Justo R, Disney P, *et al.* Atrioventricular valve failure in Fontan palliation. *J Am Coll Cardiol* 2019;**73**:810–822.
2. Blusztein D, Moore P, Qasim A, Mantri N, Mahadevan VS. Transcatheter edge-to-edge repair of systemic tricuspid valve in extracardiac fontan circulation: first in human. *J Am Coll Cardiol Case Rep* 2022;**4**:221–225.
3. Guerin P, Jalal Z, Le Ruz R, Cueff C, Hascoet S, Bouvaist H, *et al.* Percutaneous edge-to-edge repair for systemic atrioventricular valve regurgitation in patients with congenital heart disease: the first descriptive cohort. *JAHA* 2022;**11**:e025628.
4. Vijarnsorn C, Khoo NS, Tham EB, Colen T, Rebeyka IM, Smallhorn JF. Increased common atrioventricular valve tenting is a risk factor for progression to severe regurgitation in patients with a single ventricle with unbalanced atrioventricular septal defect. *J Thor Cardiovascr Surg* 2014;**148**:2580–2588.
5. Podzolkov VP, Chiaureli MR, Yurlov IA, Zelenikin MM, Kovalev DV, Dontsova VI, *et al.* Results of Fontan operation in patients with atrioventricular valve regurgitation. *Eur J Cardiothorac Surg* 2015;**48**:308–315.
6. Feldman T, Kar S, Rinaldi M, Fail P, Hermiller J, Smalling R, *et al.* Percutaneous mitral repair with the MitraClip system. *J Am Coll Cardiol* 2009;**54**:686–694.