BMC Pediatrics

CASE REPORT Open Access

A case of complete atrioventricular septal defect in which extracorporeal membrane oxygenation could be removed after performing the bilateral Glenn procedure for severe cardiac dysfunction after tricuspid valve replacement: a case report

Hiroki Ishii[1,3*], Yu Matsumura[1*], Yuji Hamamichi[1], Yuya Komori[2*], Naoki Wada[2*] and Tadahiro Yoshikawa[1*]

Abstract

Background Complete atrioventricular septal defect (AVSD) is a congenital heart disease (CHD) in which patients require surgery in early infancy. Tricuspid valve regurgitation (TR) is a complication that can occur a long time after intracardial repair for AVSD, and surgical intervention is occasionally necessary. However, to date, there have been no reports of TR occurring in the acute phase after surgery for AVSD in infancy. In addition, the mortality rate is high for patients with severe symptomatic TR who undergo surgical intervention to correct the tricuspid valve position.

Case presentation The patient was a 17-year-old adolescent girl with severe scoliosis, who was diagnosed as having complete AVSD in the neonatal period owing to a heart murmur detected after birth. The 2-patch repair method was performed at 2-months old, but severe TR was presented from an early phase after the operation. In addition, myxomatous degeneration of the atrioventricular valve was observed as an intraoperative finding. Subsequently, the patient was admitted to our hospital owing to chest discomfort at 17-years old, and tricuspid valve replacement (TVR) was performed. As substantial deterioration of cardiac contraction was observed after the TVR, the patient was placed on extracorporeal membrane oxygenation (ECMO). However, because it was subsequently difficult to remove the patient from ECMO, the bilateral Glenn procedure was performed to increase right ventricular (RV) protection. After the bilateral Glenn procedure, the patient's cardiac contractile function improved, and she could be taken off ECMO.

*Correspondence:
Hiroki Ishii
h.ishii19810814@gmail.com
Yuya Komori
kkomori@shi.heart.or.jp
Naoki Wada
nwada@shi.heart.or.jp
Tadahiro Yoshikawa
tayoshi@shi.heart.or.jp

Full list of author information is available at the end of the article

The patient began treatment with 2new types of therapeutic agents for heart failure, and was discharged from our hospital on the 305th hospital day.

Conclusions If severe TR appears in a patient, it is important to intervene surgically at the asymptomatic phase with no dilatation of the RA. However, in cases of severe symptomatic TR in patients with CHD, the Glenn procedure might be a useful treatment strategy to increase RV protection.

Clinical trial number Not applicable.

Keywords Complete atrioventricular septal defect, Severe scoliosis, Valvular myxomatous degeneration, Severe tricuspid regurgitation, Atrial fibrillation, Tricuspid valve replacement, Extracorporeal membrane oxygenation, Bilateral Glenn procedure

Introduction

Complete atrioventricular septal defect (AVSD) is considered to be mainly caused by a fusion defect of the endocardium during fetal heart development. Complete AVSD accounts for 1.8% of cases of CHD and these patients are considered to be candidates for surgery, except for those with Eisenmenger syndrome [1]. As an intracardiac repair technique, patch closure of the AVSD and division of the common atrioventricular valve, referred to as the 2-patch repair method, are performed. Atrioventricular valve regurgitation (AVVR) is known to be a problem that occurs a long time after the operation, and atrial dilatation and arrhythmia also sometimes occur [2]. However, if the valve itself had been congenitally degenerated as in this case, AVVR might be occurred at an early phase after complete AVSD repair. As atrial dilatation and arrhythmia are caused by severe AVVR, valvuloplasty or valve replacement surgery is sometimes needed. In the present case, the patient was put on extracorporeal membrane oxygenation (ECMO) because substantial deterioration of cardiac contraction was observed after TVR. However, it was difficult to take the patient off ECMO, and hence the bilateral Glenn procedure was performed, and the patient could finally be taken off ECMO. To date, there are no reports on cases similar our present case, and thus we report the clinical course and images of this patient, together with a literature review.

Case presentation

The patient was a 17-year-old adolescent girl with complete AVSD, which was not diagnosed during the fetal period. She had a history of severe scoliosis, and no notable family history. Transthoracic echocardiography (TTE) was performed because a heart murmur was detected immediately after birth, and complete AVSD of Rastelli classification type A and persistent left superior vena cava were confirmed. The 2-patch repair method was performed at 2-months old. Myxomatous degeneration was identified as an intraoperative finding of the atrioventricular valve. Severe tricuspid valve (TR) and moderate mitral valve regurgitation (MR) were observed from an early phase after the operation, but the patient's parents did not consent to reoperation. Therefore, the policy was to continue medical treatment. Subsequently, MR was gradually improved. However, owing to worsening of the TR, cardiac catheterization was performed at 15 years and 2 months. As a result, the TR was grade IV on the sellers classification, which is a scales to evaluate the severity of valve regurgitation, and the patient's central vein pressure (CVP) was 12 mmHg (data not shown. An additional movie file shows this in more detail [see Additional files 1, 2]). Subsequently, the patient noticed palpitations, and presented to our hospital at 17 years and 2-months old. Atrial tachycardia was displayed on 12-lead electrocardiography (ECG), and the patient was admitted to our hospital for electrical defibrillation (DC). Transesophageal echocardiography was performed before DC. There were no thrombi inside the heart. The tricuspid annulus diameter was 30 mm (Z score: −0.33 standard deviation (SD)) × 23 mm (Z score: −3.65 SD), and severe TR was observed. Tethering was observed owing to shortening of the tricuspid valve's septal cusp. There was prominent degeneration of the tricuspid valve, and rupture of the chordae posterolateral to the posterior leaflet. Shortening of the septal cusp of the tricuspid valve was also observed. After performing DC, atrial fibrillation was persistent without returning to sinus rhythm, and thus she began treatment with an antiarrhythmic agent. However, the patient was admitted to our hospital because the patient noticed a feeling of tightness in her chest at 17 years and 5-months old.

The patient's height was 131.6 cm, and weight was 30.5 kg and body surface area was 1.06 m^2. Her body temperature was 36.7 °C, respiratory rate was 18 breaths per minute, heart rate (HR) was 56 beats per minute, and blood pressure was 86/40 mmHg. Distension of the bilateral jugular veins appeared in the sitting position. On physical examination, diastolic rumble and systolic murmur were heard, which was strongest at the apex of the heart, and breathing sounds were normal in both lungs. On abdominal palpation, margins of the liver were located 3-cm below the right costal arch. The patient's medication before operation was furosemide,

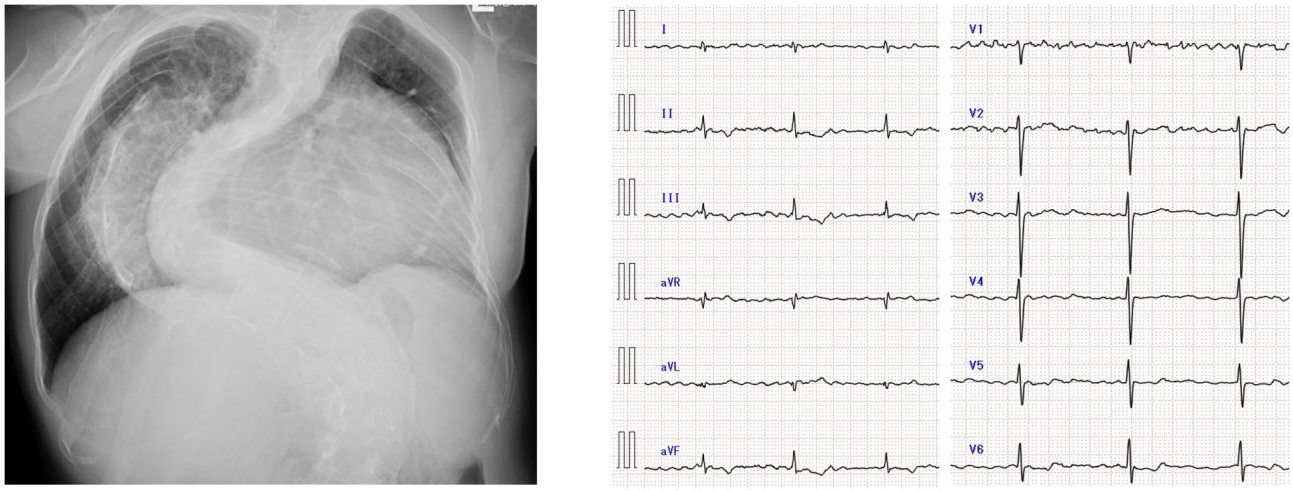

Fig. 1 Chest X-ray and 12-lead ECG findings of the patient before TVR. (**A**) Chest X-ray on admission displaying severe scoliosis. (**B**) The 12-lead ECG on admission, displaying atrial fibrillation. The patient's heart rate was 50 beats per minute

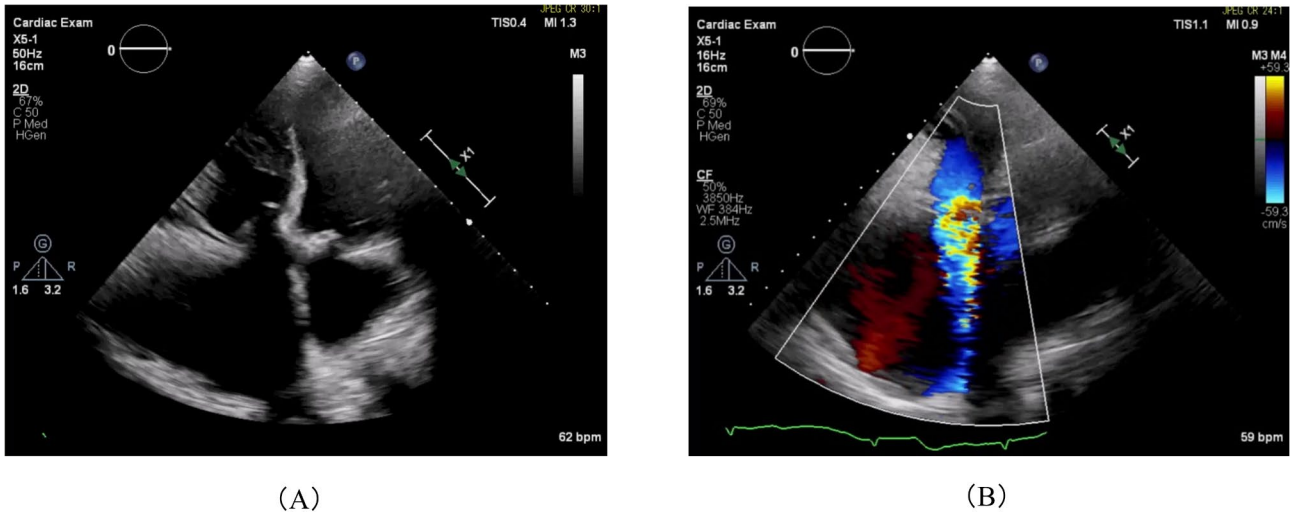

Fig. 2 Transthoracic echocardiogram findings of the patient before tricuspid TVR. (**A**) Four-chamber view displaying RA dilatation. (**B**) Color doppler imaging displaying severe tricuspid regurgitation

spironolactone, enalapril maleate, carvedilol, digoxin and warfarin potassium.

The patient had severe scoliosis, her cardiothoracic ratio was 72%, and there was no pleural effusion on chest X-ray at the time of admission (Fig. 1A). Laboratory analysis demonstrated a brain natriuretic peptide level of 885 pg/mL, but other laboratory data were normal. Her HR was 50 beats per minute, and atrial fibrillation was presented on 12-lead ECG at the time of admission (Fig. 1B). On TTE, left ventricular ejection fraction and right ventricular fractional area change were 57.4% and 42.6%, and tricuspid annular plane systolic excursion was 18 mm, respectively. Substantial dilatation of the right atrium (RA), and severe tricuspid valve regurgitation was observed (Fig. 2A, B). As consent for surgery was obtained, TVR and pulmonary vein isolation were performed.

Surgical technique

After general anesthesia, median sternotomy, and establishment of extracorporeal circulation, the RA was opened after aortic occlusion. After identifying and incising the atrial septum, the left atrium (LA) was exposed. Four pulmonary veins (PVs) were returned to the LA. Pulmonary vein (PV) isolation was performed using the cryo-isolation system. In addition to PV isolation, we ablated the cardiac tissue from edges RA incision line to isthmus position of superior and inferior vena cava by using same system. The tricuspid valve's septal cusp was stuck to a patch that had been used to close the

ventricular septum defect. The tricuspid annulus was small, and there was substantial degeneration of the valve leaflets (Fig. 3). We decided to perform valve replacement as valvuloplasty was deemed to be difficult due to substantial degeneration of the valve. After quickly removing the valve leaflets, 14 sutures were placed along the valve ring, and a 23-mm Carpentier-Edwards PERIMOUNT MAGNA (MAGNA) was implanted. After TVR and PV isolation, the atrial septum and RA were closed. We considered removal of the cardiopulmonary bypass, but this was not possible because the patient's cardiac contractility was substantially decreased. Therefore, ECMO was introduced, and the patient was managed in the intensive care unit with thoracotomy. The cannulation sites were left femoral vein and left femoral artery, a diameter of venous and arterial cannula were 18 French and 13 French. The ECMO flow was 2.8 L/min, which was maximal flow rate in the ECMO circuit for the patient's physique. Contentious hemodiafiltration (CHDF) was not introduced during management of ECMO. The extracorporeal circulation time was 315 min, aortic clamping time was 129 min.

Clinical course after surgery

After placing the patient on ECMO, it subsequently became difficult to take her off owing to poor recovery of cardiac contraction. To reduce the volumetric load on the right ventricle, the bilateral Glenn procedure was performed on the 14th hospital day. An end-to-side cavopulmonary anastomosis was created on the beating heart. Cardiac contractility was gradually improved after the bilateral Glenn procedure, and she was taken off ECMO on the 21st hospital day (data not shown. An additional movie file shows this in more detail [see Additional files 3, 4]). After taking the patient off ECMO, it was difficult to extubate her owing to severe respiratory muscle fatigue, and thus tracheostomy was performed on the 34th hospital day. However, cardiac catheterization was performed on the 46th hospital day because the patient's facial edema had gradually worsened. As stenosis was observed upon bilateral Glenn anastomosis, percutaneous angioplasty was performed on the stenosis. However, after the treatment, the patient's CVP was found to be very high. The pressure of the right superior vena cava (RSVC) and inferior vena cava (IVC) were 43 mmHg and 25 mmHg, respectively, and the pressure of the right ventricle (RV) and left ventricle (LV) were 58/ end diastolic pressure (EDP) 27 mmHg and 97/EDP 29 mmHg, respectively. New therapeutic agents for heart failure, namely, an angiotensin receptor neprilysin inhibitor (ARNi) and sodium glucose cotransporter 2 (SGLT2) inhibitors were introduced in addition to diuretic agents. After introducing these agents, the patient remained asymptomatic. The artificial respirator and spiritual tube could be removed on the 156th and 258th hospital day, respectively, and her spontaneous breathing was stable. The patient had no problems regarding activities of daily living, and was discharged from our hospital on the 305th

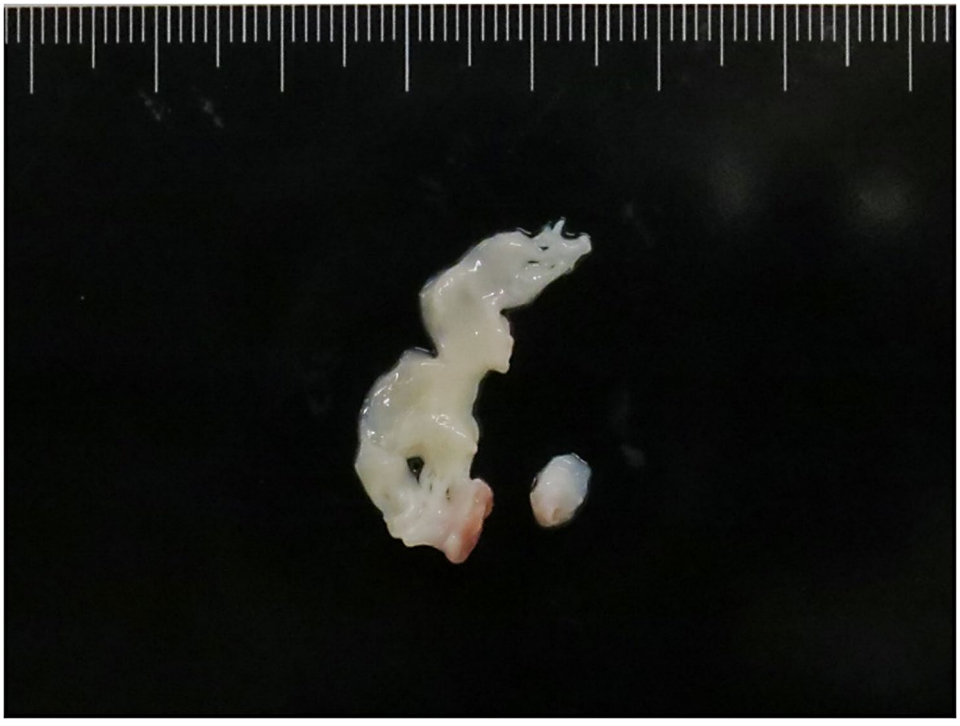

Fig. 3 Resected tricuspid valve specimen. The tricuspid valve shows characteristics of myxomatous degeneration

Table 1 Changes in circulation indicators of the patient upon cardiac catheterization

Circulation indicator	Two years before the operation	Forty-six days after the operation	One year after the operation
RSVC pressure (mmHg)	(11)	(43)	(21)
IVC pressure (mmHg)	(12)	(25)	(11)
mPA pressure (mmHg)	Not performed	43/35 (38)	29/18 (20)
RVpressure (mmHg)	25/EDP 8	58/EDP 27	36/EDP 7
LV pressure (mmHg)	73/EDP 9	97/EDP 29	66/EDP 10
AAo pressure (mmHg)	87/51 (66)	89/63 (76)	63/38 (42)
RVEF (%)	60	Not performed	55
LVEF (%)	59	Not performed	57

RSVC, right superior vena cava; IVC, inferior vena cava; mPA, main pulmonary artery; RV, right ventricle; LV, left ventricle; AAo, ascending aorta; RVEF, right ventricle ejection fraction; LVEF, left ventricle ejection fraction, EDP, end-diastolic pressure

hospital day. The patient's medication at discharge was furosemide, tolvaptan, carvedilol, ARNi, SGLT2 inhibitors, aspirin and warfarin potassium.

Estimation of cardiac catheterization 1 year after TVR and PV isolation demonstrated that the pressure of the RSVC and IVC were 21 mmHg and 11 mmHg, respectively, and RVEDP and LVEDP were 7 mmHg and 10 mmHg, respectively (Table 1).

Discussion and conclusion

As a complications that occur a long time after radical surgery for complete AVSD, mitral valve regurgitation, left ventricular outflow tract obstruction, and residual shunt of VSD are known. However, severe TR was observed at an early phase after the operation in this patient. There are many causes of TR, but valvular myxomatous degeneration is thought to be the main cause [3, 4]. Our present patient had severe TR for 16 years, and thus substantial dilatation of the RA and atrial fibrillation were observed. We decided to perform to TVR because there was severe degeneration of the tricuspid valve. There is insufficient evidence regarding surgical indications for TR. However, Said et al. discussed that a delay in surgical intervention for severe TR might lead to irreversible right heart dysfunction, and affect long-term prognosis. Therefore, they emphasized the importance of surgical intervention before the appearance of signs of right heart failure [5]. There are 2 methods for the surgical intervention of tricuspid valve, namely, plastic and replacement procedures. Despite the findings of previous studies, there is no gold standard for prosthetic TVR. Therefore, the choice between using a biological or a mechanical prosthesis in the tricuspid position should be made on an individual basis according to clinical judgement [6]. In the present patient, we chose a biological valve that is designed to prevent thrombus formation, because the right side of the heart had low pressure and low flow. A biological valve may be more preferable for young women considering pregnancy, because there are teratogenic agents among the anticoagulants that are required to be taken for a prosthetic valve. Biological valves are inferior to mechanical valves in terms of durability, and valve dysfunction has been reported to occur from a few years after the operation [7, 8]. However, the development of biological valves with antithrombotic effects and durability has been progressing in recent years. In the present patient, the biological valve used for the TVR was MAGNA which has been used in Japan from 2008. MAGNA is a biological valve consisting of 3 leaflets made from bovine pericardium and a stent, and its durability is enhanced with calcification prevention treatment. Moreover, MAGNA has a true-supra-annular design, which reduces the differential pressure across the valve by increasing effective orifice area.

Although high-quality bioprosthetic valves have been developed, the mortality rate of patients in the acute phase after the operation is 12–45% regarding surgical intervention in the tricuspid valve position. Therefore, it is important to adequately explain the risks of the surgery to the patient and the patient's parents [9]. Our present patient was considered to be a high-risk surgical case, because of her severe scoliosis, myxomatous degeneration of the tricuspid valve, and substantial dilatation of the RA and atrial fibrillation owing to severe TR. In addition, we proposed 2 factors that might lead to decreased cardiac systolic function after the operation, although it is unclear whether they would do so directly. The first factor is myocardial damage caused by long-term cardiac arrest under extracorporeal circulation. There have been some reports that myocardial ischemia owing to cardiac arrest during a cardiac operation might promote the production of inflammatory cytokines and cardiomyocyte apoptosis, and there hence might be a positive correlation between myocardial ischemia time and degree of myocardial damage [10–12]. The second factor is exponential volume overload on the right ventricle. We considered that the right ventricle myocardium damaged by myocardial ischemia might be unable to withstand a substantial stretch of the cardiac muscle owing to the inability of blood to escape from the RV to the RA after TVR. Hembree et al. reported that the postoperative mortality rate was decreased by introducing ECMO after operations in high-risk patients undergoing tricuspid valve surgery, and considered that the protection of the RV by preload reduction might be involved [13]. However, regarding our present patient, there was insufficient improvement in cardiac contractility despite the introduction of ECMO, and it was hence difficult to take her off ECMO. The primary question in this case report

is why RV volume unloading was not successful despite the establishment of ECMO. The volume of RV, which was 99.7 ml (96.7% of normal), was sufficient in volumetry of cardiac catheterization at 2018. However, the RV cavity seemed small in terms of the patient's physique in an intraoperative finding of Glenn procedure. In fact, the volume of RV was 83.8 ml (77% of normal) in volume of cardiac catheterization at 1 year after TVR. We considered that the narrow RV volume might be listed as a cause which was difficult to withdraw ECMO. Based on this consideration, the bilateral Glenn procedure was performed to further protect the right ventricle, and it was then possible to take her off ECMO. Zhang et al. reported a case of a patient with tricuspid valve stenosis and complete AVSD in whom an atrial septum defect was created and the Glenn procedure was performed in addition to TVR with a mechanical valve. Their results showed that the patient had an uneventful postoperative course during her hospital stay, and her circulation was stable after discharge. They concluded that a pressure load reduction of the right heart system might affect the prognosis of pediatric patients with congenital heart disease who require artificial valve replacement [14]. Therefore, the Glenn procedure might be useful for reducing the load of the right ventricular myocardium in high-risk patients who are expected to experience a sudden increase in pressure load to the right heart system after operation. There is a limitation of this case report. In fact, we had not planned to cardiac catheterization or MRI because the patient was claustrophobic, and had not requested it even after explaining the necessity. However, the RV function and volume should be accurately evaluated preoperatively when considering the surgical procedure. Therefore, cardiac catheterization or MRI should be performed to perform the surgery safety even if persuade the patient and patient's parents. On a preoperative trans thoracic echocardiogram, RV contractility was sufficient. But, bilateral jugular veins distension and liver enlargement were observed, preoperatively. As a cause of physical signs, we should consider to be a decrease of RV diastolic function in addition to severe TR. If cardiac catheterization or MRI could be performed preoperatively, we might be able to have thought carefully whether to add atrial septum defect creation in addition to TVR in surgical procedure.

In conclusion, we presented a case of a high-risk patient with severe scoliosis and valvular myxomatous degeneration, in addition to the development of atrial fibrillation and prominent dilatation of the RA owing to severe TR for 16 years. As previously reported, in patients with severe TR, it is important to intervene surgically at the asymptomatic phase with no dilatation of the RA. However, in patients with severe symptomatic TR, the Glenn procedure might be a useful treatment strategy to protect the RV, because the RV might not be able to withstand a sudden increase in pressure after artificial valve replacement.

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12887-025-05654-0.

Supplementary Material 1: Additional file 1 (mp4). A frontal view of right ventricular angiography (RVG) at 2 years before TVR. The degree of regurgitation was grade IV of sellers classification, which is a scale to evaluate the severity of valve regurgitation. Prominent dilatation of the RA was presented, and the contrast agent was static inside the RA. Moreover, reflux of the contrast agent into the inferior vena cava is shown. There was no stenosis of right pulmonary artery.

Supplementary Material 2: Additional file 2 (mp4). A lateral view of RVG at 2 years before TVR. The degree of regurgitation was grade IV of sellers classification. Prominent dilatation of the RA was presented, and the contrast agent was static inside the RA. RV ejection fraction was 60%. There was no stenosis of RV outflow tract and left pulmonary artery.

Supplementary Material 3: Additional file 3 (mp4). Transthoracic echocardiography with the patient under ECMO after TVR.This is a movie of transthoracic echocardiography before the bilateral Glenn procedure, with the patient under ECMO. The contraction of both ventricles was extremely poor. The tricuspid and mitral valves were not open.

Supplementary Material 4: Additional file 4 (mp4). Transthoracic echocardiography with the patient under ECMO after TVR and bilateral Glenn procedure. The contraction of both ventricles appeared to be improved, and the tricuspid and mitral valves were able to open.

Acknowledgements
Not applicable.

Author contributions
Hiroki Ishii wrote the manuscript. Yu Matsumura, Yuji Hamamichi, Yuya Komori, Naoki Wada, and Tadahiro Yoshikawa participated in clinical practice and provided conceptual advice and edited the manuscript. All authors read and approved the final manuscript.

Funding
No external funding was received for this study.

Data availability
The authors confirm that the data of transthoracic echocardiography and cardiac catheterization supporting the findings of this case report are available within the supplemental materials.

Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
Informed consent to publish this case report was provided in writing by the patient's parents.

Competing interests
The authors declare no competing interests.

Author details
[1]Department of Pediatric Cardiology, Sakakibara Heart Institute, 3-16-1 Asahi-cho, Fuchu-shi, Tokyo 183-0003, Japan
[2]Department of Pediatric Cardiovascular Surgery, Sakakibara Heart Institute, 3-16-1 Asahi-cho, Tokyo 183-0003, Japan

[3]Department of Pediatrics and Adolescent Medicine, Tokyo Medical University Hospital, 6-7-1 Nishishinjuku, Shinjuku-ku, Tokyo 160-0023, Japan

Received: 15 February 2025 / Accepted: 31 March 2025
Published online: 16 April 2025

References

1. Burkhart HM, Anderson HN. Commentary: complete atrioventricular septal defect in young infants: the advantages of early repair. J Thorac Cardiovasc Surg. 2021;161(6):2155–6.
2. Muraru D, Badano LP, Hahn RT, Lang RM, Delgado V, Wunderlich NC, Donal E, Taramasso M, Duncan A, Lurz P, De Potter T, Zamorano Gómez JL, Bax JJ, von Bardeleben RS, Enriquez-Sarano M, Maisano F, Praz F, Sitges M. Atrial secondary tricuspid regurgitation: pathophysiology, definition, diagnosis, and treatment. Eur Heart J. 2024;45(11):895–911.
3. He Y, Guo Y, Li Z, Chen J, Kontos MC, Paulsen WH, Han J, Wang L, Gu X, Nixon JV. Echocardiographic determination of the prevalence of primary myxomatous degeneration of the cardiac valves. J Am Soc Echocardiogr. 2011;24(4):399–404.
4. Ma PH, Sachdeva R, Wilson EC, Guzzetta NA. Longitudinal echocardiographic evaluation of an unusual presentation of X-Linked myxomatous valvular dystrophy caused by filamin A mutation. Semin Cardiothorac Vasc Anesth. 2016;20(3):240–5.
5. Said SM, Dearani JA, Burkhart HM, Connolly HM, Eidem B, Stensrud PE, Schaff HV. Management of tricuspid regurgitation in congenital heart disease: is survival better with valve repair? J Thorac Cardiovasc Surg. 2014;147(1):412–7.
6. Liang W, Yue H, Li T, Qin X, Qian Y, Wu Z. The better substitute for tricuspid valve replacement in patients with severe isolated tricuspid regurgitation. Anatol J Cardiol. 2019;22(4):172–6.
7. Kobayashi Y, Nagata S, Ohmori F, Eishi K, Nakano K, Miyatake K. Serial doppler echocardiographic evaluation of bioprosthetic valves in the tricuspid position. J Am Coll Cardiol. 1996;27(7):1693–7.
8. Abdul Qadeer M, Abdullah A, Noorani A, Khan AH, Mustafa MS, Nadeem ZA, Samad S, Siddiq MU, Siddiqui RQU, Said SM. Tricuspid valve replacement with mechanical versus biological prostheses: a systematic review and meta-analysis. J Cardiothorac Surg. 2024;19(1):636.
9. Bleiweis MS, deVirgilio C, Milliken JC, Baumgartner FJ, Sheppard BB, Robertson JM, Nelson RJ. Tricuspid valve surgery: 15-year experience. J Natl Med Assoc. 1996;88(10):645–8.
10. Hasegawa T, Yamaguchi M, Yoshimura N, Okita Y. The dependence of myocardial damage on age and ischemic time in pediatric cardiac surgery. J Thorac Cardiovasc Surg. 2005;129(1):192–8.
11. Modi P, Suleiman MS, Reeves B, Pawade A, Parry AJ, Angelini GD, Caputo M. Myocardial metabolic changes during pediatric cardiac surgery: a randomized study of 3 cardioplegic techniques. J Thorac Cardiovasc Surg. 2004;128(1):67–75.
12. El Feky W, El-Afify D, Abdelhai D, Elkashlan M, Fakhreldin A, El Amrousy D. L-carnitine decreases myocardial injury in children undergoing open-heart surgery: A randomized controlled trial. Eur J Pediatr. 2024;183(6):2783–9.
13. Hembree A, Lawlor M, Nemeth S, Mørk SR, Kaku Y, Spellman J, Miltiades A, Kurlansky P, Takeda K, George I. Effect of venoarterial extracorporeal membrane oxygenation initiation timing on tricuspid valve surgery outcomes. JTCVS Open. 2023;14:171–81.
14. Zhang QL, Chen YK, Lin SH, Du XW, Chen Q, Wang SM, Cao H. Tricuspid mechanical valve replacement for severe tricuspid stenosis in a child who underwent surgical correction of complete atrioventricular septal defect, a rare long-term complications. J Cardiothorac Surg. 2024;19(1):77.

Publisher's note