Cardiology in the Young

cambridge.org/cty

An extreme case of cor triatriatum mimicking hypoplastic left heart syndrome and combined pulmonary vein stenosis

Hee Ju Hong[1], Hye Won Kwon[1], Jae Gun Kwak[1], Sang Yun Lee[2] and Yoon Seong Lee[3]

[1]Department of Thoracic and Cardiovascular Surgery, Seoul National University Hospital, Seoul, South Korea; [2]Department of Pediatrics, Seoul National University Hospital, Seoul, South Korea and [3]Department of Radiology, Seoul National University Hospital, Seoul, South Korea

Brief Report

Cite this article: Hong HJ, Kwon HW, Kwak JG, Lee SY, and Lee YS (2024) An extreme case of cor triatriatum mimicking hypoplastic left heart syndrome and combined pulmonary vein stenosis. *Cardiology in the Young* **34**: 205–208. doi: 10.1017/S1047951123003724

Received: 6 August 2023
Revised: 30 September 2023
Accepted: 30 September 2023
First published online: 29 November 2023

Keywords:
Cor triatriatum; pulmonary vein stenosis; angioplasty; balloon

Corresponding author:
Hye Won Kwon; Email: drhwkwon@snu.ac.kr

Abstract

A 65-day-old girl presented to the emergency room with lethargy, requiring emergency venoarterial extracorporeal membrane oxygenation for refractory cardiogenic shock. Initially, hypoplastic left heart syndrome was suspected. However, cor triatriatum with a pinpoint opening on the membrane was diagnosed based on a detailed echocardiographic examination. After membrane resection, the left heart size was restored. However, follow-up echocardiography performed 4 months later showed occlusion of both upper pulmonary veins and stenosis in both lower pulmonary veins. Hybrid balloon angioplasty was performed in all pulmonary veins, and stents were inserted into the right upper and lower pulmonary veins. Despite repeated balloon angioplasty, all pulmonary vein stenosis progressed over 6 months and the patient expired while waiting for a heart-lung transplant. Even after successful repair of cor triatriatum, short-term close follow-up is required for detecting the development of pulmonary vein stenosis.

Case report

A 65-day-old girl visited the emergency room due to poor oral intake and lethargy. Chest radiography revealed cardiomegaly (Fig 1a). Echocardiography revealed a hypoplastic left atrium, left ventricle, aortic valve (6 mm; *Z*-score, −1.88), and mitral valve (8.6 mm; *Z*-score, −2.53) (Fig 2a). Suspecting hypoplastic left heart syndrome, the patient was admitted to the paediatric ICU. On admission, the patient's body weight was 4.5 kg. Initial blood pressure was 65/29 mmHg; heart rate, 162/min; respiratory rate, 62/min; and oxygen saturation, 86%. After intubation, blood pressure decreased to 40/26 mmHg and lactate levels increased from 1.5 to 4.3 mmol/L. Central venoarterial extracorporeal membrane oxygenation support was required because hypotension persisted despite using high doses of inotropes (dopamine, 15 μg/kg/min; dobutamine, 15 μg/kg/min; epinephrine, 0.4 μg/kg/min). After stabilising vital signs using extracorporeal membrane oxygenation, a detailed echocardiography detected a membrane dividing the left atrium into two chambers, indicating cor triatriatum sinister (Fig 2a). Scanty mitral inflow through a 2 mm hole in the membrane resulted in a hypoplastic left ventricle and low cardiac output. Cardiac CT confirmed a dilated proximal chamber and small distal chamber, which was also compressed by an enlarged coronary sinus (Fig 2c and d). After extracorporeal membrane oxygenation support for four days, end-organ functions recovered, and a corrective surgery was performed (Supplementary Fig 1). After complete resection of the membrane in the left atrium cavity, we left a 4–5 mm of fenestration on the atrial septum to prevent pulmonary oedema due to acute volume overload on the small left atrium. The mitral valve was small but showed acceptable leaflet motion and the size of the left heart increased enough to support systemic circulation (Fig 2b). Weaning from cardiopulmonary bypass was uneventful and the patient was transferred to the paediatric ICU without extracorporeal membrane oxygenation support. The patient was extubated on postoperative day 4 and discharged on postoperative day 10. Cardiomegaly improved on pre-discharge chest radiography (Fig 1b), and left ventricle end-diastolic diameter increased from 8.2 mm (*Z*-score, −8) to 19.2 mm (*Z*-score, −0.39) on pre-discharge echocardiography. The brain natriuretic peptide levels decreased from 12,582 pg/mL to 844 pg/mL at discharge. However, the patient visited the outpatient clinic 4 months after discharge, wherein respiratory difficulty was observed. Although chest radiography showed mild pulmonary congestion (Fig 1c), echocardiography revealed severe pulmonary hypertension with multiple pulmonary vein stenosis; both upper pulmonary vein flows were not visible, and the pressure gradient on the right and left lower pulmonary veins was 9.5 mmHg and 3.5 mmHg, respectively. Cardiac CT confirmed total occlusion of both upper pulmonary veins and stenosis in both lower pulmonary

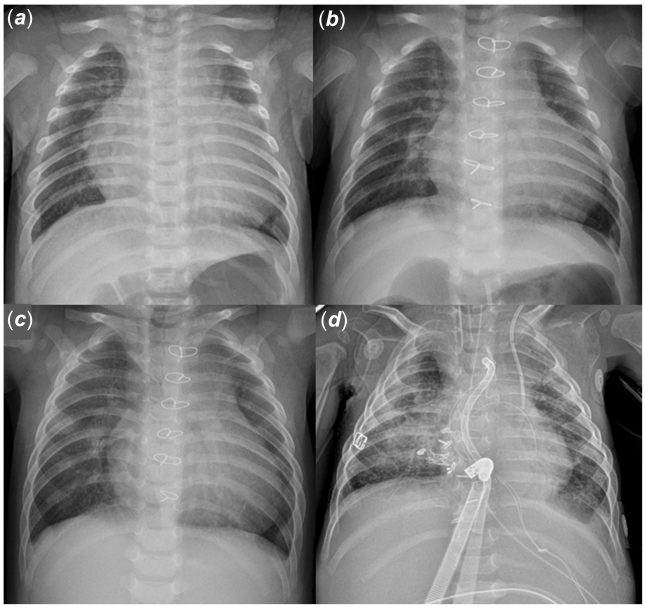

Figure 1. Chest radiography. (***a***) Initial chest radiography showing severe cardiomegaly, with a cardiothoracic ratio of 74.2%. (***b***) At discharge, the cardiothoracic ratio decreased to 63.3%. (***c***) At the 4-month follow-up, pulmonary congestion develops. (***d***) The last chest radiography showing aggravated pulmonary congestion on central venoarterial extracorporeal membrane oxygenation. The distal part of stent on the right upper pulmonary vein is not fully dilated, and the vein is finally occluded. The right lower pulmonary vein stent is patent. However, there is severe in-stent stenosis on the last echocardiography due to intimal proliferation.

veins (Fig 2e and f). Percutaneous balloon angioplasty was impossible because a previously left atrial septal defect was spontaneously closed. We decided to perform a hybrid pulmonary vein angioplasty, comprising a surgical approach to the left atrium and subsequent ballooning or stenting in the stenotic pulmonary veins under direct vision through the open left atrium. In the operative field, most of the left atrial endocardium showed fibrotic changes. Additionally, the openings of both upper pulmonary veins appeared almost closed (Supplementary Fig 2). The smallest Bowman probe (1 mm) could barely enter the upper pulmonary vein openings. Once the probe penetrated, we could make the opening larger step-by-step with one size larger probe. Then, we surgically managed the opening and extended the incision until we reached the relatively larger lumen. However, the stenotic portion of pulmonary vein was significantly longer than anticipated and it seemed almost reached the lung parenchyma. The vessel wall of pulmonary vein was also severely thickened. Following the surgical venous opening, we decided to add stents as a primary treatment option because we thought surgical widening alone would not be effective. After balloon angioplasty using a 4 mm-sized Sterling™ Balloon Dilatation Catheter (Boston Scientific Corporation, MA, USA) in the right upper pulmonary vein, we placed a 4.5 mm diameter and 12 mm length Resolute Onyx Zotarolimus-Eluting Coronary Stent (Medtronic Cardiovascular Ltd., CA, USA). Ballooning and stenting in the left upper pulmonary vein were performed in the same manner. However, ballooning in the left lower pulmonary vein with a 7 mm-sized Sterling™ Balloon Catheter crushed the stent in the left upper pulmonary vein, for which we had to remove that crushed stent. Finally, we performed ballooning in the right lower pulmonary vein and made a fenestration in the atrial septum. Postoperative chest radiography showed that the distal portion of the stent in the right upper pulmonary vein was not fully dilated. Therefore, we attempted transcatheter balloon angioplasty for further dilatation of the stent. We administered sirolimus (0.8 mg/m^2) to prevent proliferation of fibroblasts.[1] However, the pulmonary vein stenosis progressed despite repeated balloon angioplasty every month. Three months after the first hybrid procedure, we performed a second hybrid procedure for stenting in the right lower pulmonary vein with a 7 mm diameter and 15 mm length stent (PALMAZ BLUE, Cordis, Bridgewater, NJ, USA) and ballooning in the left pulmonary veins. Even with this procedure, extracorporeal membrane oxygenation support was required due to severe hypoxaemia and right ventricular failure (Fig 1d). We registered the patient for heart-lung transplantation due to the intractable pulmonary vein stenosis and severe fibrotic endocardial change in the left atrium. She had experienced many complications associated with extracorporeal membrane oxygenation for 3 months and eventually expired of uncontrolled gastrointestinal haemorrhage.

Discussion

Cor triatriatum sinister is a rare congenital cardiac anomaly in which the left atrium is divided into two chambers by a membrane that limits blood flow to the left ventricle. Symptoms vary from asymptomatic to severe pulmonary oedema, depending on the degree of intraatrial obstruction and associated congenital anomalies.[2,3] The patient nearly collapsed at the initial presentation, necessitating extracorporeal membrane oxygenation support due to severe restriction of intraatrial communication. Lack of mitral inflow caused the left ventricle to shrink and mimicked the features of a hypoplastic left heart syndrome. After resecting the membrane in the left atrium, the left heart size was restored. Although the patient had a small mitral valve annulus, she had acceptable aortic valve size and a definite apex-forming left ventricle. Thus, we anticipated that biventricular repair might be possible.

Pulmonary vein stenosis is also a rare condition that can result in severe pulmonary hypertension, respiratory failure, and death during infancy.[4,5] In the congenital form, overgrowth of the connective tissue with medial hypertrophy and intimal fibrosis results in vessel obstruction.[5] Acquired pulmonary vein stenosis may occur after cardiovascular surgery, such as total anomalous pulmonary venous return repair.[5] In this case, pulmonary vein stenosis was newly detected 4 months after cor triatriatum repair. Congenital pulmonary vein stenosis might have existed in this patient. However, it could have been masked by the dilated proximal chamber of the left atrium at the initial diagnosis. Otherwise, reactive fibrointimal proliferation of the pulmonary vein might be caused by chronic pressure overload on the proximal chamber or triggered after cor triatriatum repair. This was similar to acquired pulmonary vein stenosis after total anomalous pulmonary venous return repair, because cor triatriatum and total anomalous pulmonary venous return have similar embryological processes.[6] Even though our centre has successful experience with hybrid pulmonary vein stenting for stenosis after total anomalous pulmonary venous return repair,[7] this case showed rapid progression of pulmonary vein stenosis even after repeated catheter interventions. Although lung transplantation is an option in patients with intractable pulmonary vein stenosis,[8] waitlist mortality is high in infants due to the shortage of paediatric

https://doi.org/10.1017/S1047951123003724 Published online by Cambridge University Press

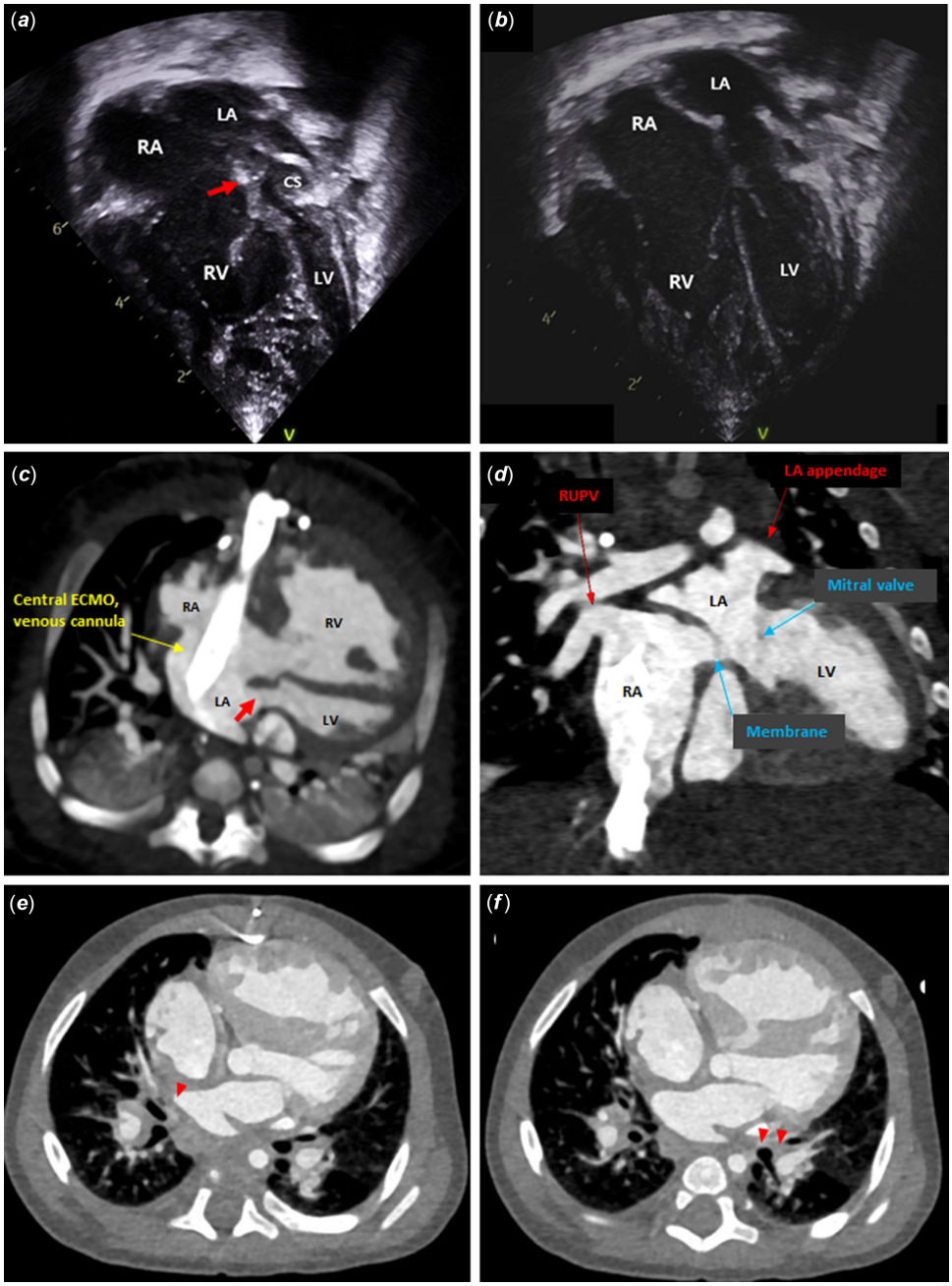

Figure 2. Echocardiography and cardiac CT angiography. (***a***) Apical four-chamber view of echocardiography showing hypoplastic left atrium and ventricle with severe right atrial and ventricular enlargement. The cor triatriatum membrane is visible in the left atrium (red arrow). (***b***) After resecting the cor triatriatum membrane, the size of the left atrium and ventricle increased significantly. (***c***) Preoperative CT angiography showing a membrane in the small left atrium (red arrow). (***d***) The membrane is located distal to the left atrial appendage. (***e***) and (***f***) Postoperative CT shows occlusion of both upper pulmonary veins (red arrowheads). CS, coronary sinus; ECMO, extracorporeal membrane oxygenation; LA, left atrium; LV, left ventricle; RA, right atrium; RUPV, right upper pulmonary vein; RV, right ventricle.

donor lungs. In this patient, the entire left atrium cavity showed severe fibrotic endocardial changes. Therefore, pulmonary vein stenosis could progress even after lung transplantation, even if donor pulmonary veins and the left atrium posterior wall were used. Therefore, heart-lung transplantation was planned. However, receiving a heart-lung transplant was even more difficult.

We report a rare case of cor triatriatum sinister with severe restriction of intraatrial communication and combined pulmonary vein stenosis. Even after successful repair of cor triatriatum, short-term close follow-up should be performed to detect the development of pulmonary vein stenosis at an early stage.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951123003724

Acknowledgements. None.

Financial support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Competing interests. None.

References

1. Patel JD, Briones M, Mandhani M, et al. Systemic sirolimus therapy for infants and children with pulmonary vein stenosis. J Am Coll Cardiol 2021; 77: 2807–2818.
2. Saxena P, Burkhart HM, Schaff HV, Daly R, Joyce LD, Dearani JA. Surgical repair of cor triatriatum sinister: the Mayo Clinic 50-year experience. Ann Thorac Surg 2014; 97: 1659–1663.
3. Kim D, Kwon BS, Kim DH, Choi ES, Yun TJ, Park CS. Surgical outcomes of cor triatriatum sinister: a single-center experience. J Chest Surg 2022; 55: 151–157.
4. Song MK, Bae EJ, Jeong SI, et al. Clinical characteristics and prognostic factors of primary pulmonary vein stenosis or atresia in children. Ann Thorac Surg 2013; 95: 229–234.
5. Pazos-López P, García-Rodríguez C, Guitián-González A, et al. Pulmonary vein stenosis: etiology, diagnosis and management. World J Cardiol 2016; 8: 81–88.
6. Herlong JR, Jaggers JJ, Ungerleider RM. Congenital Heart Surgery Nomenclature and Database Project: pulmonary venous anomalies. Ann Thorac Surg 2000; 69: S56–S69.
7. Yoon JK, Kim GB, Song MK, et al. Hybrid pulmonary vein stenting in infants with refractory to surgical pulmonary vein stenosis repair. Pediatr Cardiol 2018; 39: 1642–1649.
8. Bharat A, Epstein DJ, Grady M, et al. Lung transplant is a viable treatment option for patients with congenital and acquired pulmonary vein stenosis. J Heart Lung Transplant 2013; 32: 621–625.

https://doi.org/10.1017/S1047951123003724 Published online by Cambridge University Press