JACC: CASE REPORTS VOL. 28, 2023

CASE REPORT

CLINICAL CASE

Left Atrial Hypertension and Respiratory Failure Requiring Venoarterial ECMO After Transcatheter Pulmonary Valve Replacement

Conor P. O'Halloran, MD, Jeremy Fox, MD, Sandhya R. Ramlogan, MD, Andrew De Freitas, MD, Alan W. Nugent, MBBS, Paul Tannous, MD, PhD

ABSTRACT

A 33-year-old woman with aortic valve stenosis status-post Ross at age 6 years developed symptomatic right heart failure from right ventricle to pulmonary artery conduit stenosis. Conduit rehabilitation and transcatheter pulmonary valve replacement resulted in acute left atrial hypertension and respiratory failure requiring venoarterial extracorporeal membrane oxygenation and atrial septal defect creation as a bridge to recovery. (J Am Coll Cardiol Case Rep 2023;28:102125) © 2023 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/).

HISTORY OF PRESENTATION

A 33-year-old woman with aortic valve stenosis status-post Ross procedure at 6 years of age presented with fulminant right heart failure. Echocardiogram demonstrated depressed right ventricular (RV) function with severe conduit stenosis; left ventricular (LV) size and systolic function were normal, but there were low septal tissue Doppler and an abnormal E/A ratio indicating LV diastolic dysfunction. Computed tomography angiography showed conduit stenosis and severe systemic venous dilatation (**Figure 1A**). She was referred for conduit rehabilitation and transcatheter pulmonary valve replacement (TPVR).

LEARNING OBJECTIVES

- To understand the implications of left heart dysfunction in patients undergoing TPVR.
- To understand the physiological interplay between the right and left heart in patients with a history of bilateral ventricular injury.
- To describe the management strategy for TPVR-induced respiratory failure secondary to left heart diastolic dysfunction.

Baseline physical examination included normal vital signs with blood pressure of 124/80 mm Hg, heart rate of 71 beats/min, respiratory rate of 15 breaths/min, and oxygen saturation of 99% in room air. Physical exam was notable for a 4/6 systolic murmur, no diastolic murmur was appreciated. Baseline hemodynamics demonstrated venous and RV hypertension with a mean right atrial pressure of 20 mm Hg, RV pressure of 98/7 mm Hg, and

From the Division of Pediatric Cardiology, Department of Pediatrics, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, Illinois, USA.

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.

Manuscript received June 28, 2023; revised manuscript received October 12, 2023, accepted October 17, 2023.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2023.102125

ABBREVIATIONS AND ACRONYMS

LA = left atrial

LV = left ventricular

LVEDp = left ventricular end-diastolic pressure

PA = pulmonary artery

RV = right ventricular

TPVR = transcatheter pulmonary valve replacement

VA-ECMO = venoarterial extracorporeal membrane oxygenation

simultaneous aortic pressure of 94/50 mm Hg. A balloon wedge catheter could not be advanced through the stenotic conduit before dilation. Using the superior vena cava as the mixed venous saturation, the cardiac output was 2.7 L/min. The patient developed sustained ventricular tachycardia when the pigtail entered the LV, resolving on withdrawal of the catheter. Thus, preintervention wedge and LV end-diastolic pressures were not measured. Pulmonary artery (PA) pressure after the first conduit dilation was 33/24/26 mm Hg.

Conduit rehabilitation began with a 7-mm balloon dilation. A 4-cm-long covered stent was deployed at 16 mm and dilated sequentially to 24 mm. A bare-metal stent was placed at 24 mm to re-enforce the covered stent. Coronary angiography was performed throughout, and no compression was observed. At this point, the RV pressure was 55/22 mm Hg with aortic pressure of 109/60/80 mm Hg and PA pressure of 47/23/27 mm Hg. A Melody valve was deployed on a 22-mm Ensemble Delivery System (both Medtronic). On reassessment, the aortic pressure was 102/58 mm Hg, whereas RV pressure was 95/22 mm Hg, with PA 92/40/59 mm Hg (3 mm Hg valve gradient) with no insufficiency angiographically (Figures 2A to 2C).

PAST MEDICAL HISTORY

At 6 years of age, the patient underwent Ross procedure with a RV-PA homograft (unknown size and material). There were no other pre- or post-Ross interventions or catheterizations.

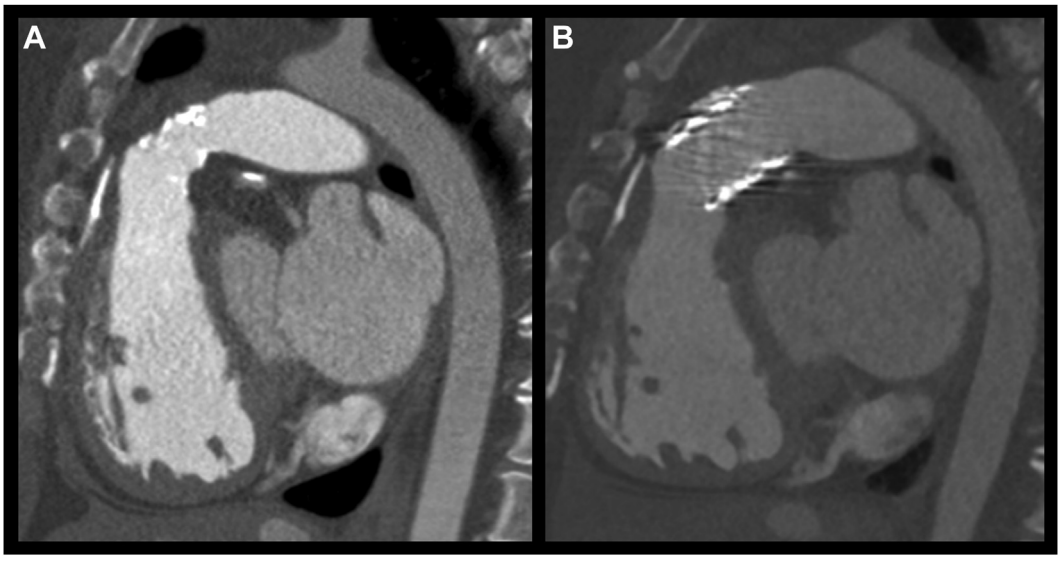
DIFFERENTIAL DIAGNOSIS

The initial differential diagnosis for high PA and RV pressure after TPVR included iatrogenic aortopulmonary window,[1] coronary compression leading to LV failure, and unmasked LV diastolic dysfunction.

INVESTIGATIONS

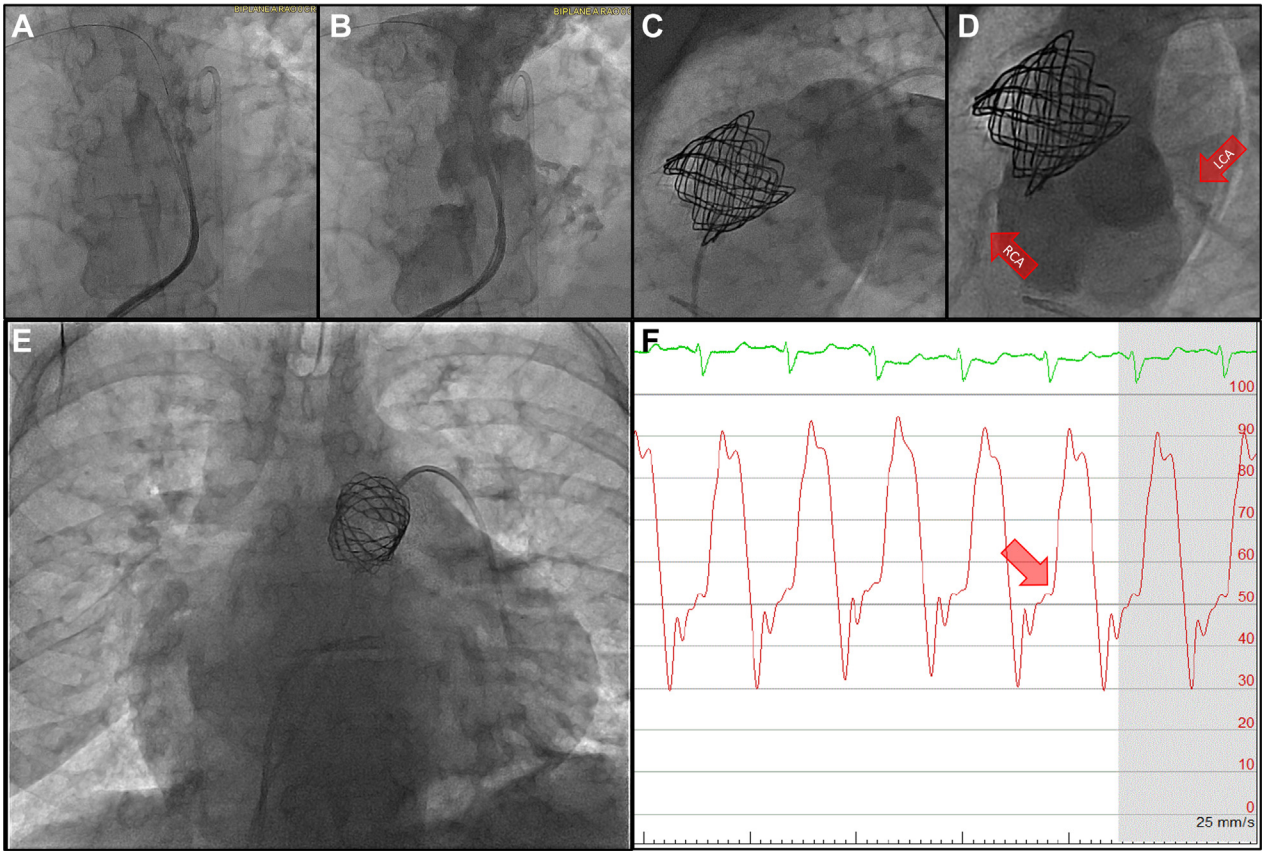
Angiography confirmed coronary artery patency and absence of an iatrogenic aortopulmonary fistula (Figure 2D). The LV was entered, and the LV end-diastolic pressure (LVEDp) was 52 mm Hg (Figure 2F). Cardiac output was 3.8 L/min, 40% higher than baseline.

FIGURE 1 RV Outflow Before and After TPVR



(A) Computed tomography angiography before transcatheter pulmonary valve replacement (TPVR) in the s4785agittal plane shows right ventricular (RV)-pulmonary artery conduit stenosis with calcification. (B) Computed tomography angiography after TPVR in the sagittal plane shows improved diameter of the RV-pulmonary artery conduit with the stents in place within the conduit.

FIGURE 2 **Conduit Rehabilitation and TPVR**



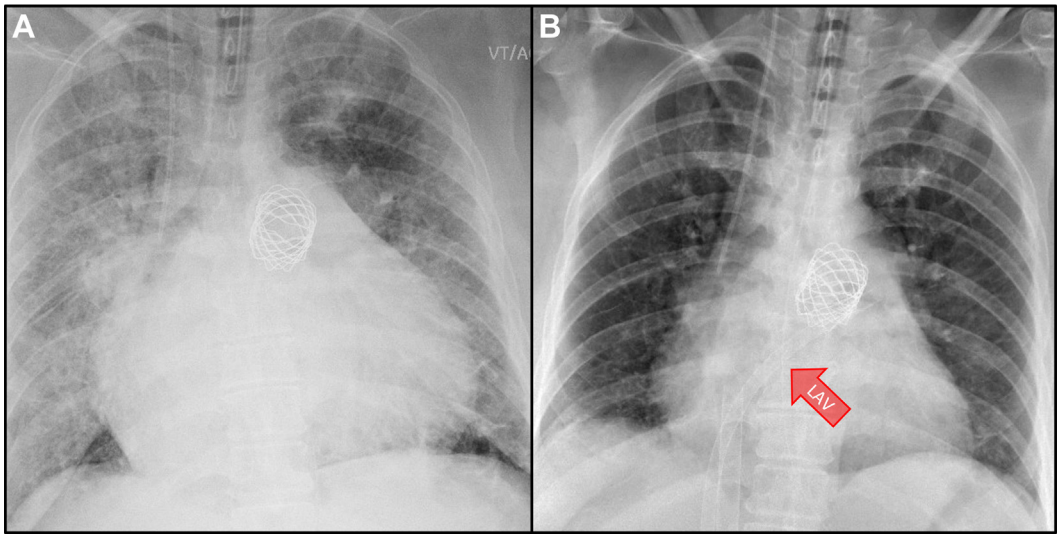
(A and B) Baseline angiography demonstrating conduit calcification. (C) Pulmonary artery angiogram after valve implantation showing no insufficiency. (D) Aortic root angiogram demonstrating coronary artery patency and no iatrogenic aorto-pulmonary window. (E) Interval development of pulmonary edema. (F) Left ventricular pressure waveform after valve implantation showing an end-diastolic pressure of 52 mm Hg (arrow). LCA = left coronary artery; RCA = right coronary artery; TPVR = transcatheter pulmonary valve replacement.

MANAGEMENT

The patient developed acute pulmonary edema (Figure 2E). High positive end-expiratory pressure ventilation, intravenous diuresis, and afterload reduction with intravenous milrinone were initiated. After 30 minutes, the RV pressure was 78% systemic. The patient was transferred to the cardiac intensive care unit with good perfusion and acceptable oxygenation. Limited echocardiography showed normal LV systolic function; the RV could not be well seen. The mixed venous saturation was 76%, indicating adequate cardiac output. Over 3 hours, she developed refractory respiratory failure and pulmonary opacification on chest radiograph (Figure 3A). Given the acuity of her respiratory failure, it was felt that there was inadequate time to return to the catheterization laboratory for intracardiac percutaneous support placement, such as Impella (Abiomed) or ProtekDuo (LivaNova). She was urgently placed on percutaneous venoarterial extracorporeal membrane oxygenation (VA-ECMO) in the cardiac intensive care unit and then transported back to the catheterization laboratory for transseptal puncture and left atrial (LA) vent placement. She remained on full support for 2 days with rapid lung recovery (Figure 3B).

On return to the catheterization laboratory, lung function and cardiac output were acceptable during VA-ECMO wean. Due to her LV diastolic dysfunction and risk of recurrent pulmonary edema, an atrial communication was created. To do this, the LA vent was exchanged for a 16-F sheath, and the atrial

FIGURE 3 Pre- and Post-VA-ECMO With LAV



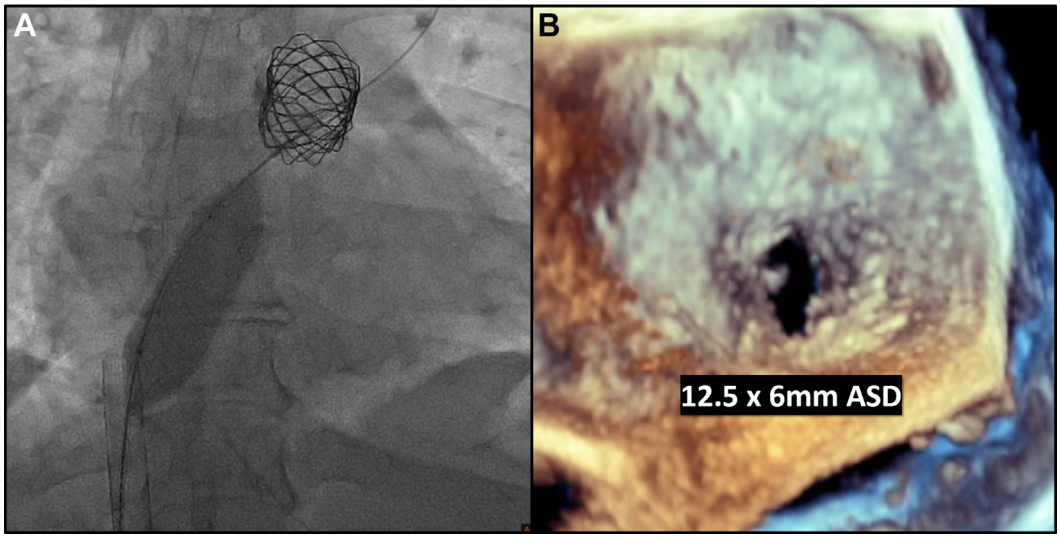
(A) Chest x-ray before initiation of venoarterial extracorporeal membrane oxygenation (VA-ECMO). (B) Significant improvement in the lungs after placement of an left atrial vent (LAV).

septum was dilated using a 20-mm balloon. Three-dimensional transesophageal echocardiogram documented a 12.5 × 6-mm defect (Figure 4), and there was a 1 mm Hg gradient by pullback across the septum (9 to 8 mm Hg).

DISCUSSION

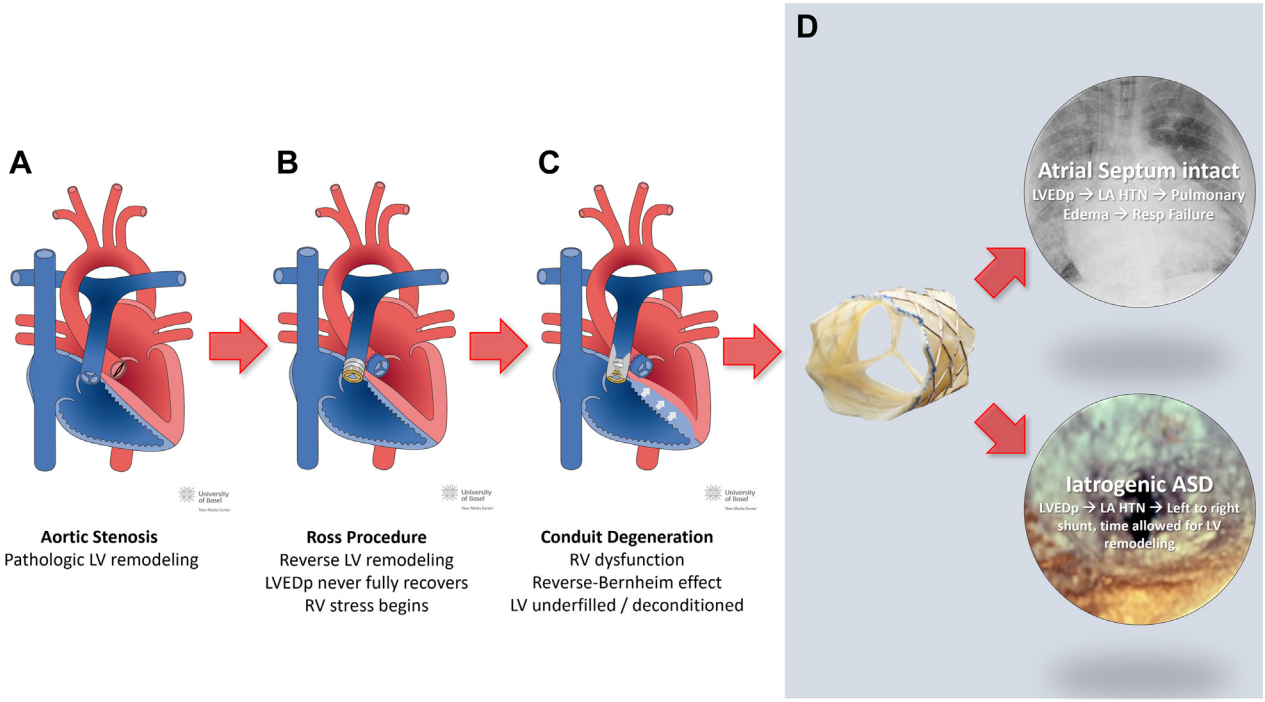
This case describes fulminant respiratory failure secondary to LA hypertension and pulmonary edema following RV-PA conduit rehabilitation and TPVR, 27

FIGURE 4 ASD Creation



(A) A 20-mm Atlas balloon was used to dilate the septum. (B) Three-dimensional transesophageal echocardiogram demonstrating creation of a 12.5 × 6-mm atrial septal defect (ASD).

FIGURE 5 Summary Model



(A) Aortic valve disease results in pathological left ventricular (LV) remodeling. (B) Ross procedure unloads the LV, but the ventricle never fully recovers. (C) Conduit degeneration causes pathological remodeling of the right ventricle (RV), and as RV pressure rises, the reverse Bernheim effect compromises the LV. (D) TPVR increases cardiac output and LV preload. Without decompression, this can result in pulmonary edema and respiratory failure. An iatrogenic ASD is 1 strategy to mitigate this outcome. HTN = hypertension; LVEDp = left ventricular end-diastolic pressure; other abbreviations as in Figures 1 and 4.

years after Ross procedure. The patient had a history of left heart obstructive disease before Ross, and more recently, long-standing right heart obstructive disease due to conduit degeneration. RV pressure was initially systemic, with only mildly elevated mean PA pressure of 26 mm Hg. After conduit rehabilitation, the RV pressure was half systemic, with trivial conduit gradient, and still only mildly elevated mean PA pressure of 27 mm Hg. Following valve placement, there was immediate LV diastolic failure with LVEDp of 52 mm Hg, resulting in pulmonary hypertension and pulmonary edema. Despite aggressive diuresis, afterload reduction, and high positive end-expiratory pressure ventilation, the patient required VA-ECMO and later atrial septal defect creation as a bridge to recovery.

LV preload increases following TPVR; in many patients, this improves LV function due to optimization of the Frank-Starling relationship.[2] However, in some patients, increased LV preload is not tolerated. Taggart et al[3] reported subacute LV diastolic failure after TPVR in a Ross recipient. In that report, the LVEDp was elevated before valve implantation but increased to a symptomatic level 3 weeks after TPVR. Alsulami et al[4] reported 2 patients who experienced pulmonary edema after TPVR. One Ross recipient with no evidence of LV diastolic dysfunction before or immediately after TPVR developed acute pulmonary edema during recovery. The second patient, with tetralogy of Fallot and unilateral pulmonary vein atresia, also developed pulmonary edema during recovery. These reports all share a predisposition for postcapillary pulmonary hypertension. Three patients had undergone the Ross procedure after prolonged periods of increased LV afterload, and thus are assumed to have abnormal LV diastolic function, and the fourth patient had unilateral vein atresia.

The proposed pathophysiology of TPVR-induced decompensation is demonstrated in the synthesis diagram (Figure 5). It begins with baseline LV diastolic dysfunction. In swine models of pulmonary insufficiency, LV diastolic dysfunction occurs even in the absence of left-sided disease.[5] LV diastolic function is further hindered in patients with a history of aortic

stenosis before the Ross procedure. With RV-PA conduit degeneration, reduced cardiac output and RV hypertension develop. The LV is chronically underfilled and may develop dysfunction due to right-to-left septal shift (reverse Bernheim effect). During conduit rehabilitation, RV afterload is reduced, which allows for adequate preload-flow-afterload balance, but free conduit regurgitation limits the increase in cardiac output, and septal shift is less pronounced. With TPVR, cardiac output and LV preload are acutely increased, thereby unmasking the degree of LV diastolic dysfunction. Acute LA hypertension leads to pulmonary hypertension and pulmonary edema, ultimately resulting in hypoxemic respiratory failure. This physiology is similar to that seen in the right atrium-to-PA VA-ECMO, wherein excessive VA-ECMO flows can lead to pulmonary edema.

FOLLOW-UP

The patient was discharged 10 days after VA-ECMO decannulation. Echocardiogram 3 days after decannulation showed normal biventricular function. At the time of this report, she is now >6 months from the event and doing well. Her symptoms of right heart failure have resolved, and she is maintained on oral diuretics, beta-blocker, and aspirin. Follow-up computed tomography angiography demonstrated a widely patent RV-PA conduit (**Figure 1B**).

CONCLUSIONS

In the vast majority of TPVR cases, the focus is on right heart disease with an assumption that the left heart will easily adapt to the physiological changes. This report, and others, demonstrate that operators must consider the LV capacity to accommodate the acute increase in preload, with proper assessment of LV diastolic dysfunction before TPVR, particularly when there is a history of left heart disease. In this case, early recognition of her LV diastolic dysfunction with elective creation of a small atrial septal defect would have likely mitigated her LA hypertension and ultimate need for VA-ECMO. Alternatively, identifying predictors of flash pulmonary edema after TPVR may allow for pre-emptive staging with conduit rehabilitation alone to relieve obstruction, followed by TPVR months later after allowing time for ventricular remodeling.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Conor O'Halloran, Division of Pediatric Cardiology, Department of Pediatrics, Ann & Robert H. Lurie Children's Hospital of Chicago, 225 East Chicago Avenue, Box 21, Chicago, Illinois 60611, USA. E-mail: cohalloran@luriechildrens.org.

REFERENCES

1. Kenny D, Holoshitz N, Turner D, Hijazi ZM. Aortopulmonary fistula after transcatheter pulmonary valve replacement. *Circ Cardiovasc Interv*. 2013;6(6):e67–e68. https://doi.org/10.1161/CIRCINTERVENTIONS.113.000654

2. Lurz P, Puranik R, Nordmeyer J, et al. Improvement in left ventricular filling properties after relief of right ventricle to pulmonary artery conduit obstruction: contribution of septal motion and interventricular mechanical delay. *Eur Heart J*. 2009;30(18):2266–2274. https://doi.org/10.1093/eurheartj/ehp258

3. Taggart NW, Connolly HM, Hagler DJ. Acute heart failure after percutaneous pulmonary valve (Melody valve) implantation. *Congenit Heart Dis*. 2013;8(2):E61–E63. https://doi.org/10.1111/j.1747-0803.2011.00570.x

4. Alsulami G, Patel M, Malekzadeh-Milani S, Bonnet D, Boudjemline Y. Hyperacute flash pulmonary oedema after transcatheter pulmonary valve implantation: the melody of an overwhelmed left ventricle. *Arch Cardiovasc Dis*. 2014;107(4):219–224. https://doi.org/10.1016/j.acvd.2014.03.007

5. Kuehne T, Saeed M, Gleason K, et al. Effects of pulmonary insufficiency on biventricular function in the developing heart of growing swine. *Circulation*. 2003;108(16):2007–2013. https://doi.org/10.1161/01.CIR.0000092887.84425.09

KEY WORDS diastolic heart failure, pulmonary edema, pulmonic valve, valve replacement