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CASE REPORT

ADVANCED

TCT CONNECT 2021 CASE

Extracorporeal Membrane Oxygenation-Supported Transcatheter Mitral Valve-in-Valve Replacement and Paravalvular Plugging for Critical Prosthetic Mitral Stenosis

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ABSTRACT

We present the case of a patient with rapidly progressive bioprosthetic mitral valve stenosis in the setting of end-stage renal failure. Critical valve stenosis led to cardiogenic shock requiring full hemodynamic support. Emergent extracorporeal membrane oxygenation–supported transcatheter mitral valve-in-valve implantation was successfully undertaken as a life-saving measure with an excellent final result. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:491–496) © 2022 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 57-year-old woman was urgently referred to our institution in 2020 with rapidly progressive dyspnea and cardiogenic shock. She had described a 2-week history of dyspnea before a hypotensive episode during outpatient dialysis that prompted admission to a peripheral hospital. Despite commencing vasopressor support for protracted hypotension, the patient's condition further deteriorated over 48 hours, necessitating medivac retrieval to our center.

LEARNING OBJECTIVES

- To make a differential diagnosis of dyspnea in patients with complex valvular disease.
- To understand the role of valvular calcification in patients with end-stage renal failure.
- To understand the feasibility of transcatheter valve interventions in the emergency setting.

MEDICAL HISTORY

Our patient has a history of end-stage renal failure due to immunoglobulin A nephropathy, maintained on long-term hemodialysis, with associated hyperparathyroidism necessitating parathyroidectomy, renal anemia, and hypertension. In the process of renal transplant suitability assessment in 2017, echocardiography had demonstrated severe mixed valvular heart disease. Specifically, she had severe calcific aortic stenosis with moderate incompetence with a mean transaortic gradient of 48 mm Hg and severe calcific mitral stenosis with a mean

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ABBREVIATIONS AND ACRONYMS

ECMO = extracorporeal membrane oxygenation

inflow gradient of 19 mm Hg. Right heart catheterization at this time demonstrated normal cardiac output with moderate mixed pulmonary hypertension with a mean pulmonary artery pressure of 48 mm Hg and a pulmonary capillary wedge pressure of 32 mm Hg. To mitigate the challenge systemic anticoagulation would pose to the peri-transplantation period if mechanical valves were implanted, she underwent surgical aortic and mitral valve replacement with 23-mm and 25-mm Perimount bioprostheses (Edwards Lifesciences), respectively, in 2017. After the ordeal of cardiac surgery, the patient elected to defer a decision regarding transplantation.

DIFFERENTIAL DIAGNOSIS

Dyspnea in the context of renal failure and mixed valvular disease raises many differentials. Main possibilities include valvular dysfunction, either regurgitation or obstruction, which itself may be degenerative, thrombotic, or infective in etiology. Obstructive coronary artery disease, inflammatory cardiomyopathies, and pulmonary emboli are other important considerations.

INVESTIGATIONS

The patient underwent transthoracic echocardiography, which demonstrated complete immobility of the mitral prosthetic leaflets resulting in critical mitral stenosis with a mean inflow gradient of 30 mm Hg (Video 1, **Figure 1**). She had hyperdynamic left ventricular function with flattening of the interventricular septum throughout the cardiac cycle (Video 2), with severe right ventricular dilatation and impairment in the setting of near-systemic estimated pulmonary artery systolic pressures (85 mm Hg). Coronary angiography did not demonstrate an ischemic culprit.

MANAGEMENT

Despite multiagent inotropic and vasopressor support, the patient's condition continued to deteriorate and she was urgently placed on veno-arterial extracorporeal membrane oxygenation (ECMO),

FIGURE 1 Continuous-Wave Doppler Echocardiography Revealing a Mean Transmitral Gradient of 29 mm Hg

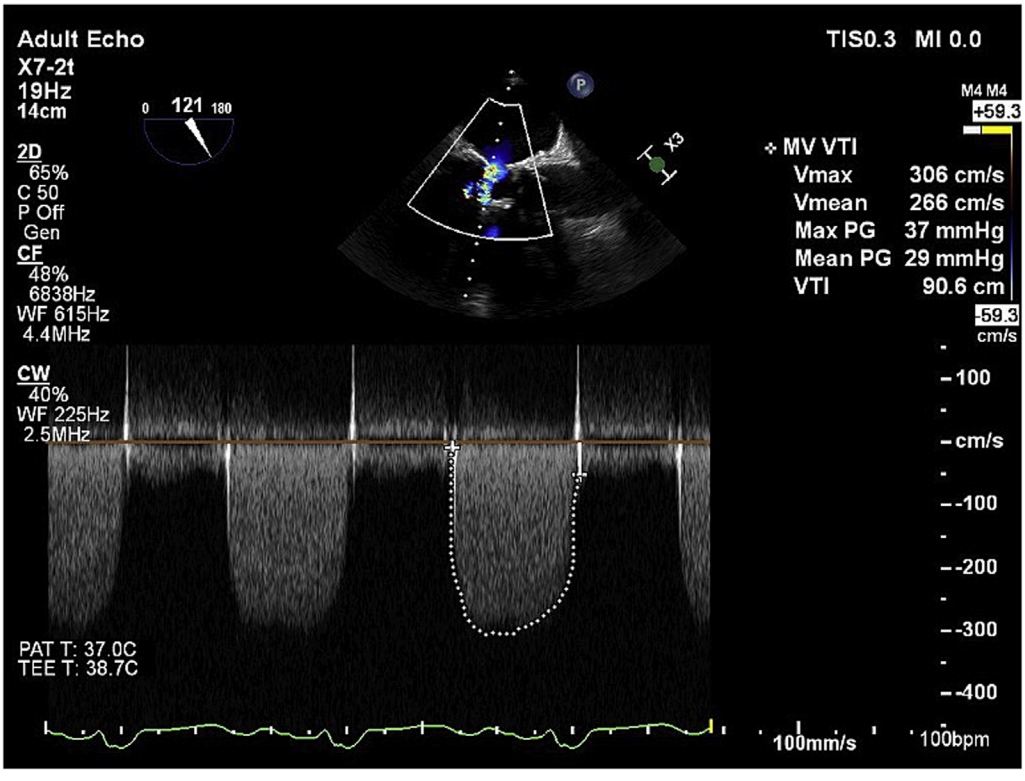


FIGURE 2 A Transeptal Needle is Used to Cross From the Right to the Left Atrium

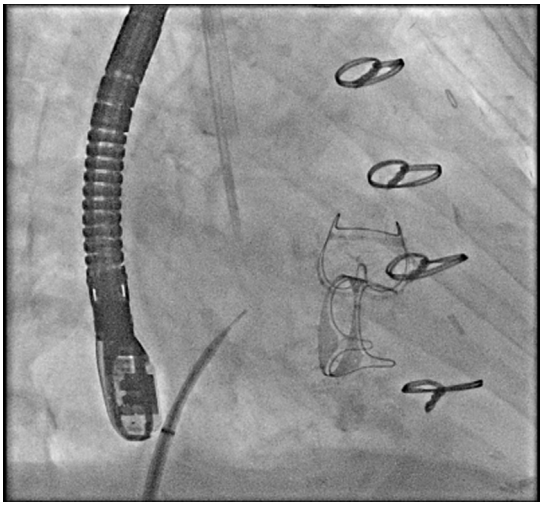
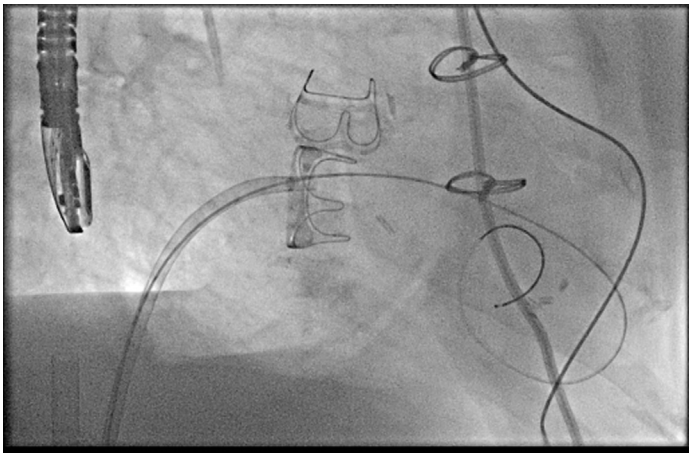


FIGURE 3 Through an Steerable Guide Catheter a Double-Curved Wire is Placed in the Left Ventricle



complicated by transient complete loss of cardiac output. After rapid multidisciplinary team discussions in consultation with the family, we pursued emergent transcatheter mitral valve-in-valve replacement. Via right femoral venous access adjacent to the arterial ECMO inflow cannula, the septum was crossed low in the fossa ovalis with a trans-septal needle (Figure 2). An Agilis (Abbott Laboratories) steerable guide catheter was advanced into the left atrium, and the stenosed prosthesis was crossed with a straight glidewire into the ventricular cavity, then exchanged for a double-curved Lunderquist wire (COOK Medical) through a pigtail catheter (Figure 3). The interatrial septum and then the stenosed valve were sequentially predilated with 9-mm and 14-mm balloons. The guide catheter was then exchanged for the valve delivery sheath through which an Edwards Sapien 3 valve (Edwards Lifesciences) was advanced and deployed under transesophageal echocardiographic guidance (Figures 4 and 5). A 26-mm prosthesis was chosen to accommodate the existing 25-mm surgical bioprosthesis, which has a true internal diameter of 23 mm, in keeping with international and manufacturer recommendations.[1] The valve appeared marginally angulated and seated more ventricular than hoped, particularly at the anterior margin (Figure 5), but was nonetheless stable and functional. Transesophageal echocardiography demonstrated a reduction in the transmitral gradient from 29 to 15 mm Hg with mild-moderate paravalvular leak (Video 3), thought to be due to incomplete device expansion due to a focal ledge of calcification. In light of these improvements, the precarious clinical condition of the patient, and the uncertainty of the reliability of the gradient estimation in the setting of ECMO, the result was accepted lest further instrumentation betray this progress.

Hemodynamic supports were weaned over the next 72 hours, and she was able to be decannulated from ECMO and extubated. She was stepped down from intensive care on single inotropic agent support, however, persistent borderline hypotension

FIGURE 4 A Bioprosthetic Valve is Positioned Within the Previous Surgical Mitral Prosthesis

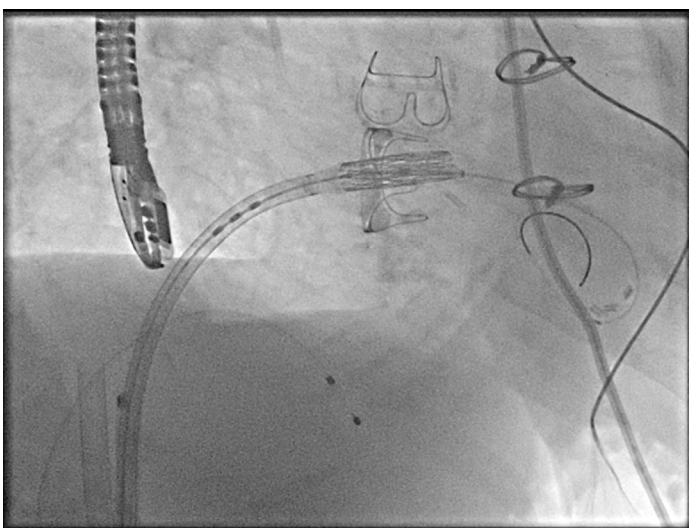


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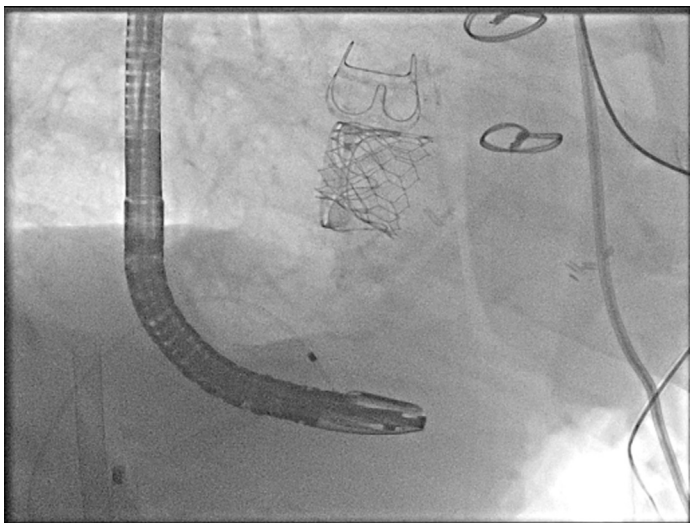
FIGURE 5 Deployed and Released Bioprosthetic Valve-in-Valve



over the next few days precluded complete weaning. Repeat transesophageal echocardiography demonstrated now severe paravalvular incompetence (Videos 4 and 5), with a persistent transmitral mean gradient of 15 mm Hg. As such, it was debated whether to return for paravalvular leak plugging and/or postdilatation of the prosthesis. The consensus decision was to attempt both sequentially. Via the right femoral vein, the previously formed atrial septostomy was recrossed without difficulty. First, the paravalvular defect was crossed using a straight glidewire through a steerable guide catheter, which was then exchanged for a delivery catheter and advanced through the defect (**Figure 6**). A 12 × 5 mm AVP III plug (Abbott Laboratories) was deployed with good control of the paravalvular regurgitation, with now a transmitral gradient of 8-9 mm Hg. The valve was then dilated under rapid right ventricular pacing using an overfilled 25-mm balloon achieving satisfactory expansion (**Figure 7**). Final echocardiography demonstrated trivial paravalvular leak with a satisfactory mean gradient of 5 mm Hg (Video 6, **Figure 8**).

FIGURE 6 The Previously Formed Septostomy was Crossed With Ease, and a Vascular Plug was Deployed Across the Paravalvular Defect

DISCUSSION

Left-sided valvular disease occurs more frequently in patients with chronic kidney disease; in those maintained on hemodialysis the prevalence of calcification of the aortic and mitral valves ranges between 45% and 54%.[2] The complex metabolic milieu in patients with renal dysfunction, combined with hemodynamic alterations such as the hyperdynamic circulation associated with arterio-venous fistulae, all contribute to the unifying process that is calcification of the leaflet interstitial cells.[2] Massera et al[3] demonstrated that baseline calcification is positively associated with rate of progression, suggesting that this process, to which chronic kidney disease predisposes, is a self-perpetuating cycle of valve injury, inflammation, and calcification. Mitral valve calcification is associated with valve dysfunction, both regurgitation and stenosis, as well as conduction disease, arrhythmias, and increased mortality.[4] It is, therefore no surprise that the combination of valvular disease in patients with chronic kidney disease is significantly associated with adverse cardiac outcomes and death.[2]

Renal failure is also a risk factor for degeneration of bioprosthetic valves,[5] however, the rapidity of calcific valvular degeneration seen in our case was alarming and highlights the need for close surveillance in this population, in whom prosthesis selection ought to be made judiciously. Arguably, initial implantation of mechanical prostheses would have mitigated this rapid degeneration, however, bioprostheses were chosen at the time after

multidisciplinary team discussion owing to possible impending transplantation listing. It was thought that transplantation would likely occur sooner than bioprosthesis degeneration and would be less complicated in the absence of systemic anticoagulation. Transcatheter mitral valve implantation for both native and prosthetic mitral stenosis has recently become a feasible therapy for select patients.[6] Specifically, patients with a degenerated bioprosthesis demonstrate favorable outcomes after this procedure, compared with patients with native valve disease in whom 1-year mortality rate exceeds 50%, owing to the rigid presized ring into which the valve can be deployed and anchored.[7,8] Adding to the complexity of our case was the influence of ECMO support on intracardiac hemodynamics and the echocardiographic interpretation thereof. We postulate that preload and afterload alterations confounded our initial assessment of paravalvular incompetence, leading to underestimation of severity. Although it was unclear whether balloon postdilatation would further expand the new valve owing to the degree of calcification, it nevertheless proved successful in combination with plugging of the paravalvular defect and, to the best of our knowledge, this has not been reported previously.

FIGURE 7 Satisfactory Expansion of the Valve and Plug Stability Were Achieved Following Postdilation With an Oversized Balloon

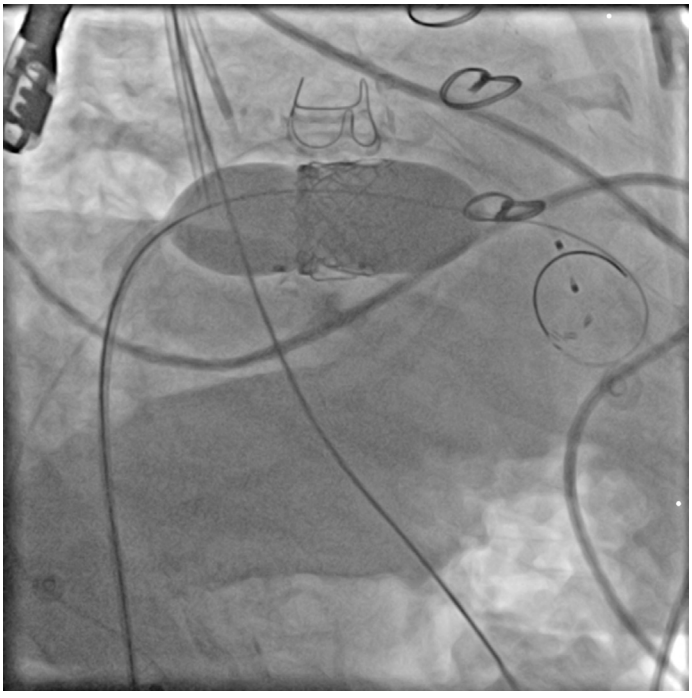
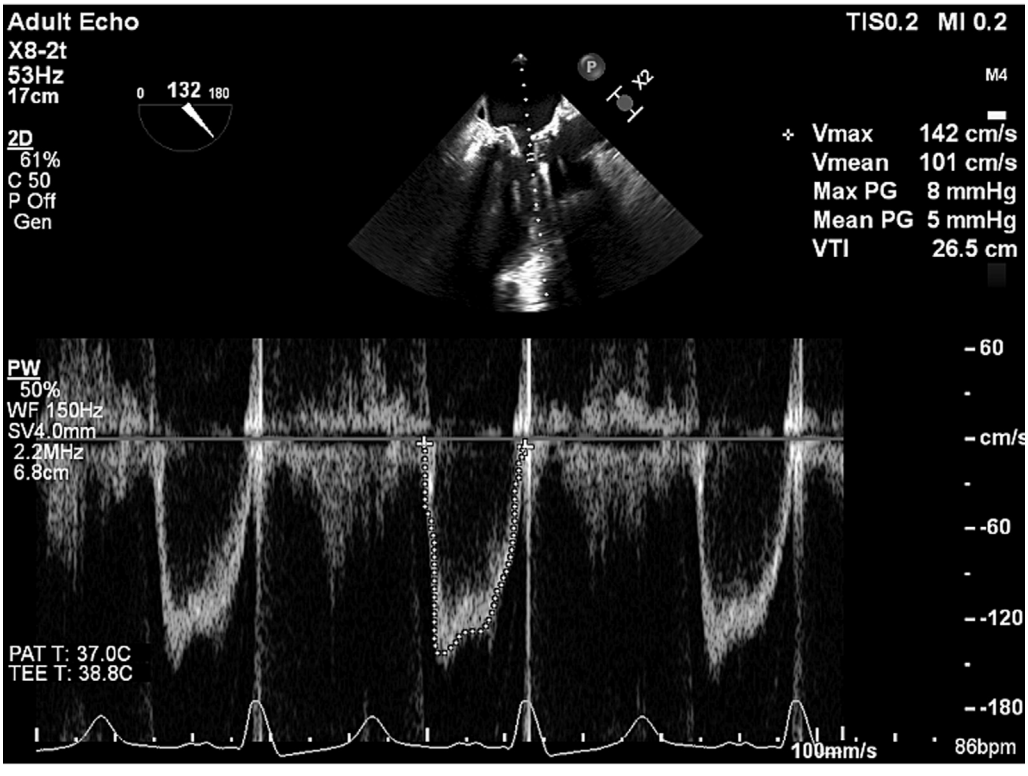


FIGURE 8 Final Echocardiographic Assessment Demonstrating a Satisfactory Mean Transmitral Gradient of 5 mm Hg



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FOLLOW-UP

The patient was weaned from all supports and suffered no neurologic injury. She was shortly thereafter discharged to a rehabilitation facility for reconditioning, ambulating with minimal supervision and assistance.

CONCLUSIONS

Transcatheter mitral valve-in-valve implantation is a recently established procedure for select patients with mitral valve disease. Although routinely performed electively, we successfully performed this procedure emergently on a patient in cardiogenic shock due to critical prosthetic mitral stenosis requiring ECMO support with an excellent clinical outcome. This case highlights the need for rigorous cardiac surveillance for patients with chronic kidney disease and valvular heart conditions.

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

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KEY WORDS echocardiography, hemodynamics, imaging, mitral valve, valve replacement

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