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CASE REPORT

CLINICAL CASE: TECHNICAL CORNER

Transaortic TAVR and Mitral Repair Under Deep Hypothermic Circulatory Arrest in a Porcelain Aorta Patient

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ABSTRACT

Vascular and valvular calcifications, commonly seen in renal patients, increase operative mortality and can preclude conventional valvular management. We show a novel approach to treat aortic stenosis and degenerative mitral regurgitation under hypothermic circulatory arrest in a hemodialysis patient with aortic, mitral disease and porcelain aorta with surgical and transcatheter contraindications. (J Am Coll Cardiol Case Rep 2024;29:102144) © 2024 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/).

INTRODUCTION

Aortic stenosis and mitral regurgitation are the 2 most common valvular pathologies in the United States,[1] which occur frequently in patients with end-stage renal disease (ESRD).[2] Extensive circumferential aortic calcification, known as porcelain aorta, is present in up to 34% of patients with ESRD.[3] Porcelain aorta carries increased operative mortality because manipulation of calcified aortic segments can lead to catastrophic aortic dissection, stroke, and uncontrollable bleeding, and porcelain aorta can be resistant to conventional surgical management.[3] One option can be replacement of the ascending aorta, but this is dependent on an aortic arch that is amenable to hemiarch implantation. As a result, surgical correction of aortic stenosis is considered very high risk in patients with porcelain aorta and transcatheter aortic valve replacement (TAVR) is indicated,[3] although individualized strategies have also been suggested.[4] Patients with ESRD also have higher rates of mitral valve disease including mitral annular calcification (MAC).[5] Surgical repair of the mitral valve with MAC is a formidable technical challenge that is independently associated with poorer outcomes and higher operative mortality,[6] including devastating atrioventricular groove disruption.

In this report, we describe an ESRD patient with severe aortic stenosis and porcelain aorta who was not a TAVR candidate due to leaflet calcium

LEARNING OBJECTIVES

- Vascular and valvular calcifications can preclude conventional management of the cardiac patient.
- We demonstrate a hybrid surgical-TAVR approach to manage a 'technically inoperable' patient with concurrent porcelain aorta and coronary cusp calcifications.

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ABBREVIATIONS AND ACRONYMS

CPB = cardiopulmonary bypass

ESRD = end-stage renal disease

MAC = mitral annulus calcification

TAVR = transcatheter aortic valve replacement

immediately adjacent to the left main coronary artery. This patient also had severe mitral regurgitation and MAC. Here we describe a novel approach of direct TAVR and mitral repair under deep hypothermic circulatory arrest.

CASE PRESENTATION

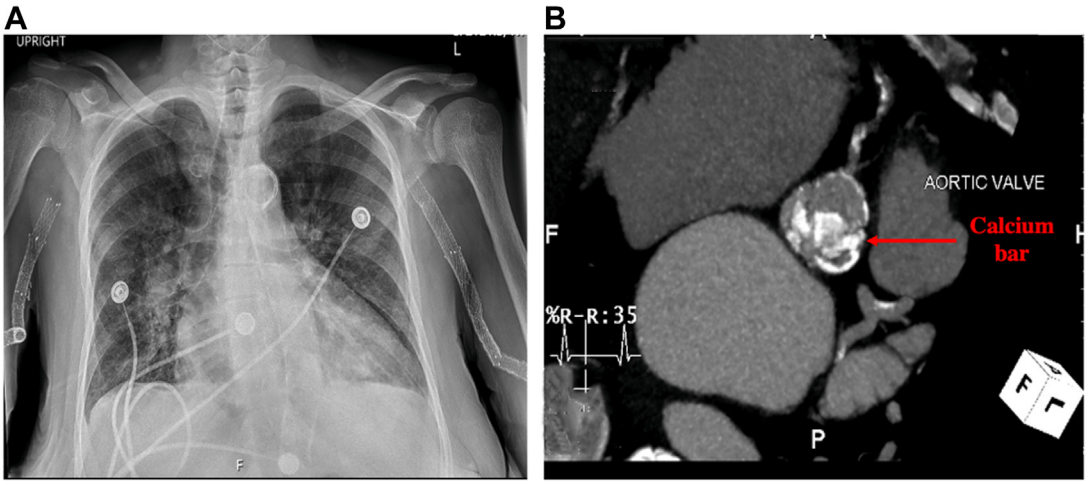
A 64-year-old female patient was referred to us with severe aortic stenosis, an ejection fraction of 50%, and a past medical history of hemodialysis for ESRD. Preoperative imaging revealed porcelain aorta from aortic root to arch (**Figure 1A**, Video 1). A calcified 2-cm nodule on the left coronary cusp of the aortic valve was prominent (**Figure 1B**). This prohibited safe peripherally implanted TAVR owing to concerns for acute left main coronary artery obstruction. Preoperative transthoracic echocardiography computed tomography imaging suggested moderate mitral regurgitation with severe trigone to trigone MAC (**Figure 2**).

In the operating room, the patient was anesthetized, and transesophageal echocardiography revealed severe mitral insufficiency due to P1 focal flail, as well as moderate to severe aortic insufficiency (Videos 2 and 3). The right axillary artery was exposed and cannulated and used for arterial access of cardiopulmonary bypass (CPB). The patient was placed in the standard supine position and the heart was exposed through a full median sternotomy. Venous access was obtained via bicaval canula. CPB was instituted and the patient was cooled to a core body temperature of 18 °C. Once the desired temperature was reached, and confirmed by suppressed electroencephalogram activity, CPB was stopped, achieving circulatory arrest. Retrograde cerebral perfusion was administered via the superior vena cava targeting a central venous pressure of 20 mm Hg.

First, the left atrium was accessed via a left atriotomy through the interatrial groove of Sondergaard. The mitral annulus was invaded by severe MAC. The flail segment of the mitral valve was stabilized using an Alfieri stitch between A2 and P2 (Video 4).The left atrium was left open to minimize the duration of deep hypothermic circulatory arrest, and we then turned our attention to the aorta.

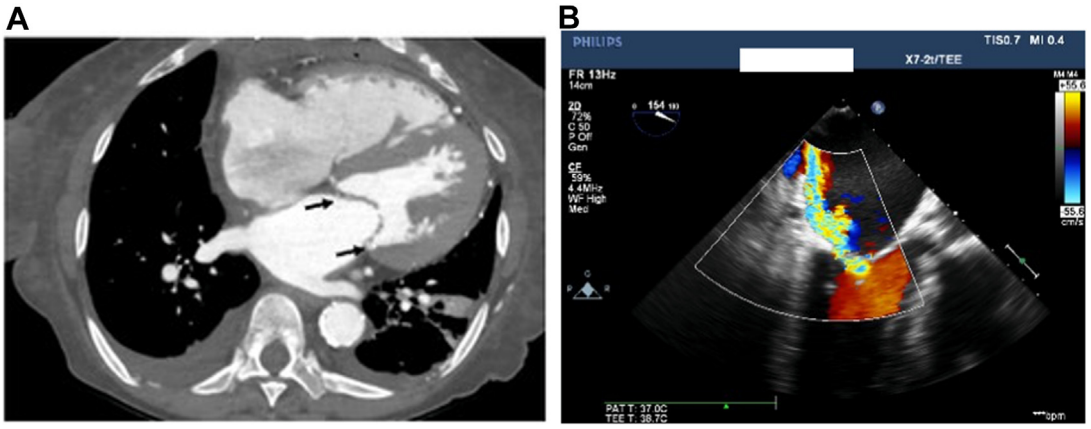
We opened the aorta in a small noncalcified aortic segment (0.5 × 1.5 cm) that was identified by palpation and preoperative planning on computed tomography. A linear arteriotomy was then made on the anterolateral aspect of the aorta ~6 cm distal to the aortic valve (**Figure 3A**).[7] This arteriotomy would not be sufficient for surgical aortic valve replacement. The large nodule of calcium immediately anterior to the left main coronary artery was identified and excised, whereas the remainder of the native valve was left in place to support the TAVR valve. An Edwards Sapien3 23-mm valve (Edwards Lifesciences) was then positioned transaortically through the

FIGURE 1 Extent of Vascular and Valvular Calcifications



(A) Chest x-ray showing porcelain aorta associated with extensive calcifications from ascending to descending aorta. (B) Heavily calcified aortic valve and coronary artery calcifications. Notice labelled (red arrow) leaflet calcifications overlying the ostium projection of the left main coronary artery.

FIGURE 2 **Calcified Mitral Annulus and Mitral Regurgitation**

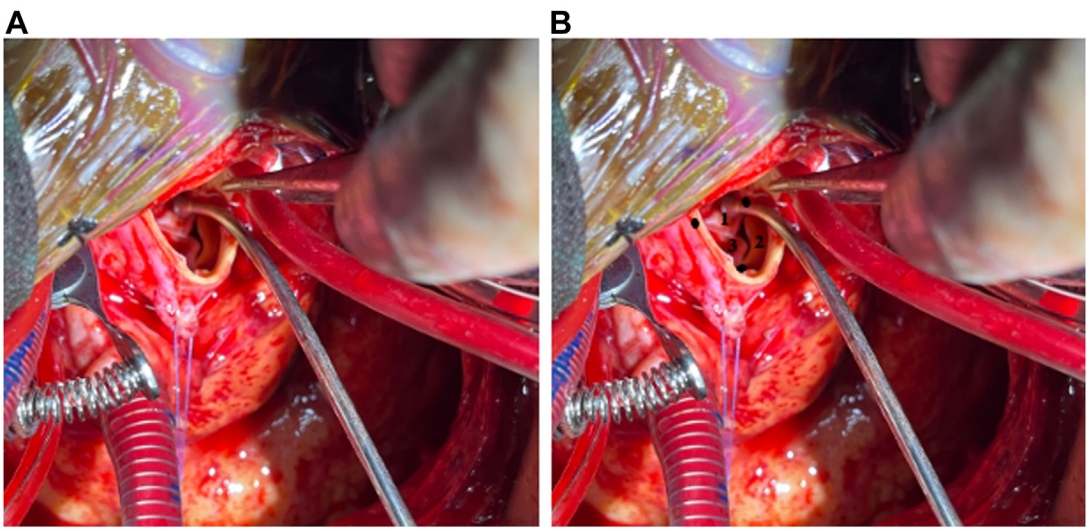


(A) Axial computed tomography scan showing mitral annulus calcification (arrows). (B) Transesophageal echo focal lateral A2 flail and severe posteriorly directed mitral regurgitation.

arteriotomy and deployed under direct visualization (Figure 3B). Detailed description of patient anatomical features is provided in the Supplemental Appendix. Patency of both coronary ostia was confirmed by direct probing. A slightly oversized Dacron patch was sewn along the incision to close the aorta in a tension-free manner.

CPB was restored, the brain was deaired, the left atriotomy was closed, and the patient was rewarmed and weaned off CPB using minimal inotropes. Intraoperative transesophageal echocardiography demonstrated a normal functioning aortic prosthesis, trace mitral regurgitation, and normal left and right ventricular function. Patient recovery was good, and she

FIGURE 3 **Operative View of Aortic Valve**



(A) Calcified aortic valve with severe aortic stenosis. The aortic root is small and calcified. A large bar of calcium threatening the left main was removed, whereas the rest of the valve was left in place to support the valve undergoing transcatheter aortic valve replacement. (B) Layout of the aortic valve commissures and leaflet cusps used for direct transcatheter aortic valve replacement: left coronary cusp, right coronary cusp, and noncoronary cusp.

was discharged to home without any complications. Furthermore, the patient was seen in clinic 18 months after the operation and had a mean mitral valve gradient of 3 mm Hg without insufficiency, normal biventricular function, and an aortic mean gradient of 8 mm Hg without aortic valve regurgitation.

DISCUSSION

The pathophysiology of ESRD promotes the development of cardiovascular calcifications, which can manifest as aortic stenosis and porcelain aorta, as well as MAC.[3]

For management of aortic stenosis, the choice of TAVR vs surgical aortic valve replacement depends on surgical risk and patient-specific anatomic features. Because porcelain aorta can significantly increase risks associated with surgical aortic valve replacement, aortic stenosis in such patients is typically corrected using TAVR.[3,7] However, this case was complicated by the presence of a large calcification bar on the left coronary cusp leaflet immediately adjacent the left main coronary ostia. Although coronary artery obstruction is a relatively rare complication of TAVR, leaflet calcifications can increase the risk of this potentially fatal complication and marks an absolute contraindication for TAVR.[8] This combination of porcelain aorta and cusp calcification made management of the aortic stenosis "technically inoperable." We were able to circumvent this by combining a hybrid surgical-TAVR approach, in which the arteriotomy enabled direct visualization and excision of the calcified bar, whereas the rest of the native valve was left intact to support the TAVR. To the best of our knowledge, this was the first time this approach has been deployed.

Surgical mitral valve repair in MAC can be challenging and can involve either "resection" (en bloc decalcification and annular reconstruction) or "respect" (oversized annuloplasty with minimal decalcification) approaches.[6] In our case, an Alfieri stitch was used to correct the degenerative mitral regurgitation, whereby the flail segments of the anterior and posterior segment are sutured, creating a double orifice mitral valve. The Alfieri stitch allowed us to minimize handling of the calcified annulus.[9] Herein, rapid repair afforded by the Alfieri stitch was desirable given that our patient was under time-sensitive deep hypothermic circulatory arrest and suffered from double valve disease.

We have discussed a complicated case of porcelain aorta, aortic leaflet calcifications, and MAC in an ESRD patient. We demonstrate a novel hybrid surgical-TAVR approach combined with Alfieri stitch that allows effective correction of complex valvular pathology in this patient population.

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Dr Szeto has been an investigator, speaker, and member of an advisory board for Edwards Lifesciences and Medtronic. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS deep hypothermic circulatory arrest, mitral annulus calcification, mitral repair, porcelain aorta, TAVR

APPENDIX For detailed description of patient anatomical features as well as supplemental videos, please see the online version of this paper.