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Sequential Transcatheter Valve-in-Valve-in-Valve Implantation Within a Degenerated Bio-Bentall Conduit in a Patient with Marfan Syndrome

INTRODUCTION

The transcatheter aortic valve replacement (TAVR) journey, initiated in 2002, evolved with the first successful Valve-in-Valve (ViV) cases in 2007.[1,2] By 2014, ViV TAVR became established in modern interventional cardiology.[3,4]Today, "valve-in-valve" failures are now manifesting, giving rise to the necessity for Valve-in-Valve-in-Valve (ViViV) interventions. This 38-year-old patient with Marfan syndrome provides significant evidence to the literature regarding the technical feasibility, device mechanics, and technical nuances within constrained graft anatomies of multi-layered ViViV procedures.

CASE REPORT

A 38-year-old female with Marfan syndrome presented with a 2-month history of progressive exertional dyspnea and significant functional limitation, corresponding to New York Heart Association (NYHA) functional class III. Her medical history was notable for Marfan syndrome, which precipitated an ascending aortic aneurysm and severe aortic regurgitation, treated in 2008 via a surgical Bentall procedure utilizing a 25 mm porcine bioprosthetic valved conduit. Bioprosthesis was selected through shared decision-making to facilitate future pregnancy planning and avoid lifelong anticoagulation. Over the following asymptomatic years, she developed accelerated structural valve deterioration (SVD) of the surgical prosthesis, which was managed in 2016 via an initial transfemoral ViV TAVR utilizing a 26 mm Portico self-expanding valve. There was no historical evidence of infective endocarditis, valve thrombosis, or other major chronic comorbidities.

Physical examination revealed a 3/6 systolic–diastolic aortic murmur and bilateral basal crackles on lung auscultation. Other systemic examinations were unremarkable, and vital signs were stable. Laboratory results were unremarkable except for a B-type natriuretic peptide of 121.19 pg/mL. Transthoracic echocardiography (TTE) showed preserved ejection fraction (60%) but severe prosthesis obstruction (peak/mean gradient: 81/46 mm Hg; valve area: 0.7 cm^2). Transesophageal echocardiography (TEE) confirmed SVD, ruling out thrombosis or vegetations (Figure 1). High-risk for a redo-Bentall and refusing open-heart surgery, the Heart Team opted for a ViViV TAVR. Pre-procedural multidetector computed tomography (MDCT) revealed a restricted internal luminal dimension of 25.4 × 17.7 mm, with safe coronary ostial heights (right: 16.6 mm, left: 15.8 mm) (Figure 1).

Under transfemoral access, a 24.5-mm Myval balloon-expandable valve was successfully deployed within the failed Portico prosthesis guided by the precise internal orifice margins on MDCT (Figure 2). No intra-procedural complications

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CASE REPORT

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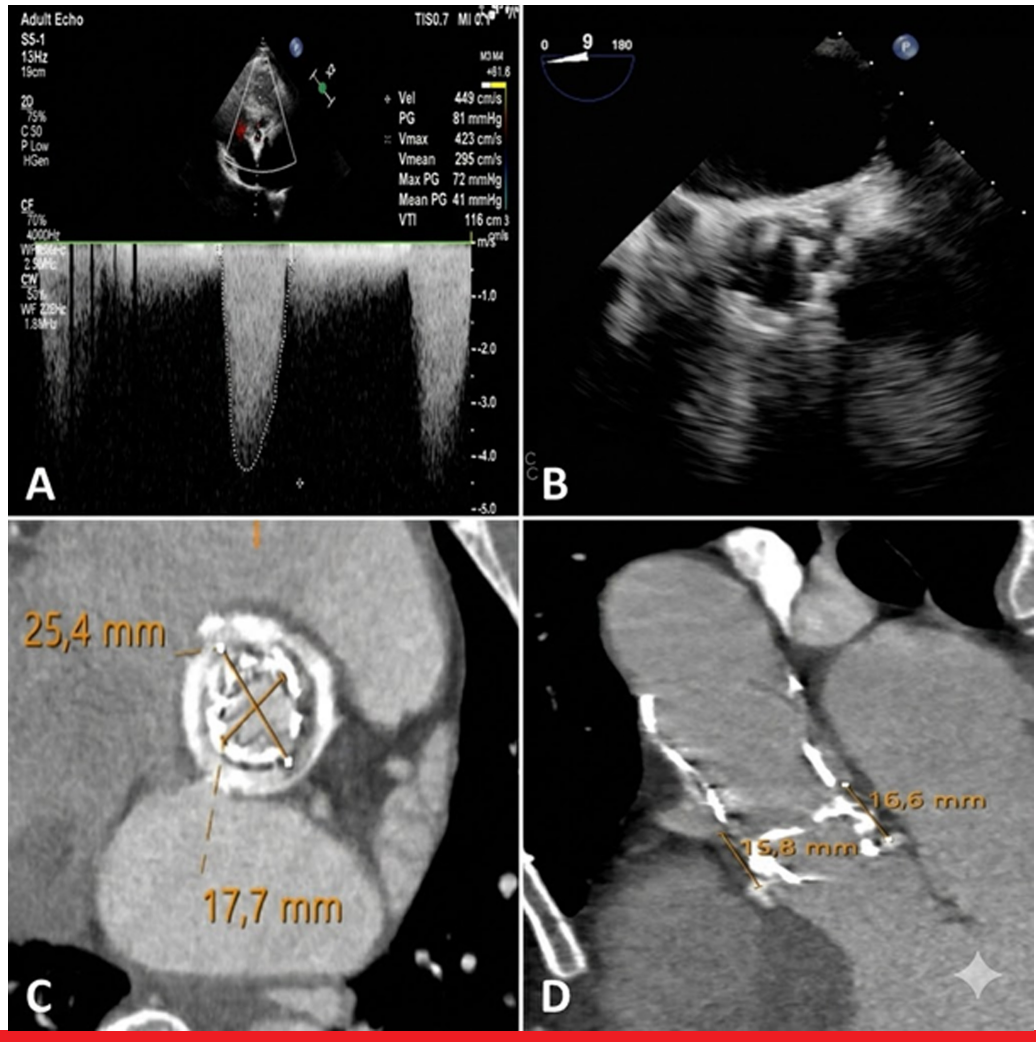


Figure 1. Pre-procedural imaging. (A) TTE CW Doppler (gradients 81/41 mm Hg, Vmax 449 cm/s). (B) TEE (bioprosthetic SVD). (C) Axial MDCT (luminal dimensions 25.4×17.7 mm). (D) Coronal MDCT (ostial clearance: LCA 15.8 mm, RCA 16.6 mm). CW, continuous-wave; LCA, left coronary artery; MDCT, multidetector computed tomography; RCA, right coronary artery; SVD, structural valve deterioration; TEE, transesophageal echocardiography; TTE, transthoracic echocardiography; Vmax, maximum velocity.

occurred. Post-procedural TTE demonstrated a gradient reduction to 26/12 mmHg with minimal residual aortic regurgitation (Figure 2). At the 1-year follow-up, she remained stable with near-complete symptomatic relief.

DISCUSSION

This case underscores the mechanical boundaries and feasibility of a transcatheter ViViV approach for recurrent bioprosthetic failure within a constrained bio-Bentall conduit. While bioprostheses in young patients avoid lifelong anticoagulation, their limited durability triggers a predictable cascade of re-interventions. Thus, prosthesis selection requires meticulous shared decision-making, acknowledging the inevitability of complex downstream transcatheter nesting.

Pathophysiologically, Marfan syndrome involves dysregulated extracellular matrix integrity driven by aberrant transforming growth factor-β signaling.[5,6] This underlying molecular dysregulation, combined with heightened mechanical stress and active calcium metabolism in younger patients, may theoretically accelerate bioprosthetic leaflet calcification and SVD.[7]

Technically, sequential nesting compromises the effective orifice area with each layer—the "matryoshka" effect—elevating baseline gradients and potentially accelerating failure.[3,8] In these scenarios, the primary objective of pre-procedural MDCT is the precise micro-geometric mapping of the true internal diameter to mitigate prosthesis-patient mismatch. Despite extreme structural constraints, strategically deploying a balloon-expandable platform inside a failed self-expanding frame yielded optimal outcomes due to its superior radial force and predictable intra-annular expansion.

CONCLUSION

Choosing a bioprosthetic conduit in young patients initiates a predictable cycle of recurrent valve dysfunction. When redo open-heart surgery poses prohibitive risks, the ViViV

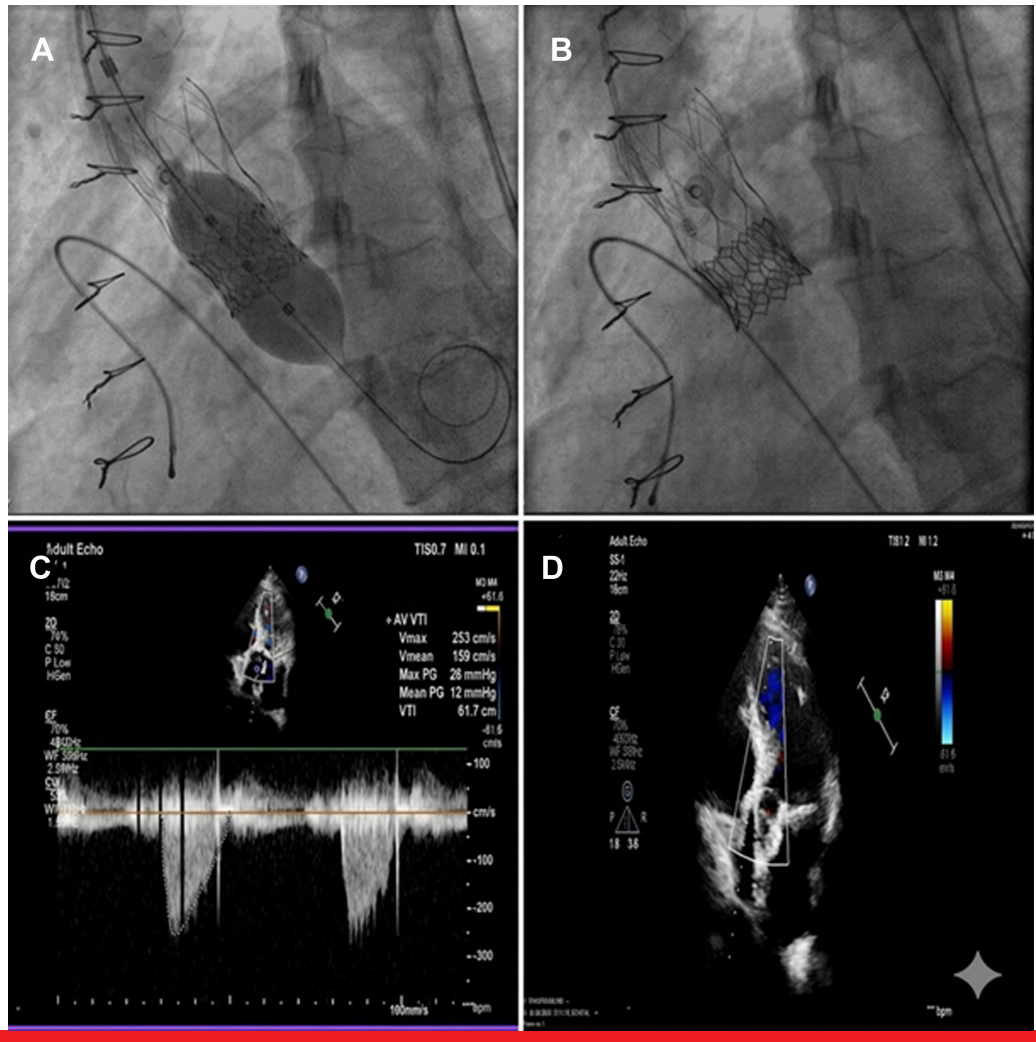


Figure 2. Intra-procedural ViViV execution and post-procedural outcomes. (A, B) Fluoroscopy showing precise positioning and deployment of the 24.5 mm Myval balloon-expandable valve within the 26 mm Portico frame. (C, D) Post-procedural TTE CW Doppler demonstrating gradient reduction to 26/12 mm Hg and minimal residual regurgitation. CW, continuous-wave; TTE, transthoracic echocardiography; ViViV, valve-in-valve-in-valve.

TAVR strategy offers an effective, safe, and reproducible alternative, even under extreme anatomical condtions.

Informed Consent: Written informed consent was obtained from the patient.

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