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Case Report

Transcatheter bicuspid aortic valve replacement in Turner syndrome: A unique experience of interventional cardiologist

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ABSTRACT

A 69-year-old short-statured Turner syndrome (TS) patient with a history of poliomyelitis in childhood and moderate bicuspid aortic stenosis (BAS) reported worsening dyspnea and fatigue over six months. A repeat transthoracic echocardiogram revealed progression to severe aortic stenosis with dilated ascending aorta (AA). As part of the work-up for aortic valve replacement, the patient underwent cardiac catheterization, which revealed a severely calcified AV with an area of 0.5 sq. cm and a mean gradient of 37 mmHg. On coronary angiography, there was 70% stenosis of the proximal left anterior descending artery (LAD). Due to poor rehabilitation potential, she was deemed high-risk for surgical aortic valve replacement. A recommendation for transcatheter aortic valve replacement (TAVR) with stenting of the proximal LAD was made. Dilated AA was managed conservatively with serial noninvasive imaging. The patient underwent TAVR with Edwards-Sapien valve (23 mm S3) and stenting of proximal LAD. The procedure was successful without complications. To our knowledge, our patient is the first case of TAVR in BAS with aortopathy in TS.

<**Learning objective:** Therapeutically, transcatheter aortic valve replacement is an off-label indication for bicuspid aortic stenosis. In real-life practice, many of these patients are poor surgical candidates and demand careful and judicious decision-making in the presence of diverse clinical scenarios. In such circumstances, a multidisciplinary approach with shared decision-making is required to recommend best possible therapeutic solution in the presence of limited data.>

Introduction

Turner syndrome (TS) patients bear several cardiovascular malformations such as bicuspid aortic valve (BAV), dilatation or dissection of ascending aorta (AA), and coarctation of the aorta [1], which can make cardiac interventions challenging. Consequently, severe aortic stenosis (AS) with bicuspid morphology in TS offers a unique therapeutic challenge to the interventional cardiologist and portends an additional risk compared to non-Turner patients due to coexisting aortopathy and coarctation. Furthermore, the aortopathy has variable severity ranging from mild to aneurysmal dilation or dissection with consequent rupture, which needs to be considered when intervention is required [2,3].

Surgical replacement, although the first line in bicuspid aortic stenosis (BAS), is not feasible in many real-life scenarios due to the complexity of clinical picture, comorbidities, and operative risk. In these circumstances, a high surgical-risk candidate might require a nonsurgical intervention such as transcatheter aortic valve replacement (TAVR). In the recent past, the performance of TAVR in tricuspid aortic valve stenosis (TAS) was considered a novel technology in poor surgical candidates. However, a rapidly-proliferating body of evidence supported its safety and efficacy and made TAVR the standard of care in inoperable cases. Until recently, published data regarding TAVR in BAS were insufficient, relying mainly on a few case series. However now, more comparative data are becoming available. In a recent study, the outcomes of TAVR in BAS versus TAS were compared. The data

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demonstrated TAVR as an effective and safe option in BAS. The overall prognosis and all-cause mortality after two years were similar in both the groups with fewer device failure rates in TAS [4].

Although this evidence could be convincing for using TAVR in BAS, the unique clinical context of TS with existing aortopathy and comorbidities makes therapeutic decision-making challenging. To our knowledge, there are no cases reported on TAVR in TS patients with BAS. We present a case of TS with BAS who underwent TAVR being a poor surgical candidate.

Case report

A 69-year-old short-statured (height: 1.4 m, body mass index: 36.94) female with TS was being followed as an outpatient for her moderate BAS. She had a prior history of hypertension, diabetes mellitus, obstructive sleep apnea, and hyperlipidemia. Over a six-month period, the patient reported progressive exertional dyspnea and extreme fatigue, which was interfering with her daily activities. She had poliomyelitis in her childhood, which led to the paralysis of her left leg requiring a knee brace. She had difficulty in walking at baseline due to her asymmetric gait and needed a walker for ambulation. Later, she developed significant hip arthritis requiring hip joint replacement. She had a history of palpitations, which on evaluation with a Holter monitor showed frequent premature atrial and ventricular complexes in addition to one brief episode of supraventricular tachycardia. There was no prior history of other arrhythmias including atrial fibrillation. Physical examination showed a grade II/VI soft ejection systolic murmur at the aortic area with a soft second heart sound; cardiac rhythm was regular. Neurologically, her left lower extremity was paralyzed due to poliomyelitis. On transthoracic echocardiogram (TTE), the patient had progressed to severe AS with a valve area of 0.5 sq. cm, AV velocity of 4.1 m/s, and a mean gradient of 39 mmHg. Ejection fraction was 60% with grade II diastolic dysfunction. Additionally, there was enlargement of the aortic root (4.3 cm), severe bi-atrial enlargement, and mild pulmonary hypertension with a right ventricular systolic pressure of 40 mmHg. There was mild mitral and tricuspid regurgitation. No specific cause of severe bi-atrial enlargement could be ascertained apart from diastolic dysfunction and pulmonary hypertension.

Given her severe, symptomatic AS, she was referred for AV replacement.

For further work-up, the patient underwent cardiac catheterization which revealed a severely calcified AV with an area of 0.5 sq. cm and a mean gradient of 37 mmHg; AA was moderately dilated. On coronary angiography, there was a 70% stenosis of the proximal left anterior descending artery (LAD). Computed tomography angiogram (CTA) was performed to delineate the aortic root anatomy and iliofemoral vasculature, which showed a dilated AA measuring 4.3 × 4.3 cm without dissection (Figs. 1 and 2). The aortic valve was bicuspid with significant calcification of one of the leaflets (Fig. 3).

Following cardiac catheterization, the patient's case was presented in the multidisciplinary valve conference. The options of surgical aortic valve replacement (SAVR) with single-vessel bypass and possible aortic root replacement versus TAVR with stenting of the proximal LAD were discussed in detail.

Based on the Society of Thoracic Surgeons (STS) score, the risk of morbidity and mortality for SAVR was 14% in her case. Furthermore, due to short stature, frailty, and physical disability due to polio, her rehabilitation potential was considered poor by a cardiothoracic surgeon, and she was deemed high-risk for SAVR, coronary artery bypass surgery, and aortic root replacement. Therefore, a recommendation was made to stent the proximal LAD and proceed with TAVR for severe AS. The patient agreed and underwent TAVR with a drug-eluting stent to the proximal LAD in the same setting. (Fig. 4).

Her AA, although dilated, was not considered imminently life-threatening and hence a conservative approach with serial noninvasive testing was recommended especially expecting the possibility of aortic root stabilization after AV replacement.

Several anatomic factors were carefully evaluated in pre-procedural planning for TAVR in this patient for reducing the chances of maldeployment, paravalvular leak (PVL), and vascular complications. The factor most technically challenging was a less than 90° angle between the annular plane of the BAV and the long axis of the AA (Fig. 1). Moreover, due to asymmetric dense linear calcifications on one of the leaflets of the BAV, there were increased chances of suboptimal seal and PVL. The aortic arch had a sharp 90° angle at the junction of the arch with the descending aorta, which was also tortuous (Fig. 2). Additionally, the access vessels were small and tortuous with increased chances of bleeding and vascular complications. To facilitate implant planning, sizing and choice of the transcatheter valve, a 3D reconstruction of the aortic root was achieved from CTA gated to electrocardiogram at 41% and 71% phase at a slice thickness of 600 μm using the 3mensio^{R} software (Bilthoven, the Netherlands).

At the time of the procedure, Medtronic's CoreValve (Medtronic Inc., Galway, Ireland) and Edwards-Sapien S3 (Edwards Lifesciences, Irvine, California, USA) were the only two commercially-available valves; new-generation Evolute-R (Medtronic Inc., Galway, Ireland), Evolute-Pro (Medtronic Inc., Galway, Ireland),

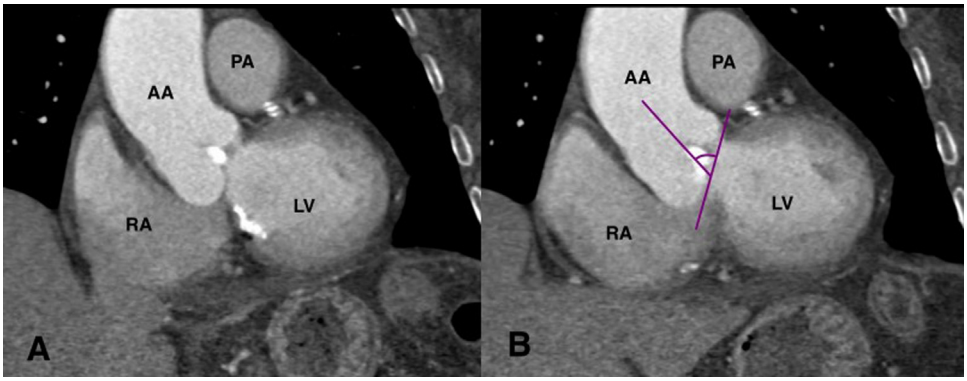

Fig. 1. Computed tomography angiogram displaying double-oblique projections along the aortic root axis from multiplanar reconstruction showing: (A) calcification on the bicuspid aortic valve along with dilated ascending aorta; (B) elucidation of the angle between ascending aorta and the plane of the bicuspid aortic valve. AA, ascending aorta; RA, right atrium; LV, left ventricle; PA, pulmonary artery.

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