Multimodality Imaging of Two Unique Etiologies of Supravalvular Aortic Stenosis

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INTRODUCTION

Supravalvular aortic stenosis (SAS) is an uncommon form of aortic stenosis that occurs distal to the aortic valve (AV). This condition is most recognized as a congenital heart defect that accounts for 8% to 14% of all cases of congenital aortic stenosis. Classically, there are three morphologic subtypes of SAS, including hourglass, membranous, and hypoplasia of the aortic arch.[1] Although congenital SAS is typically associated with genetic syndromes such as Williams-Beuren syndrome, iatrogenic causes are rare and may result from surgical complications. We present two unique cases illustrating the role of multimodality imaging in diagnosing SAS of different etiologies.

CASE PRESENTATIONS

Case 1: Congenital SAS

A 55-year-old woman presented with chronic exertional dyspnea. They had no significant medical history and no known genetic disorders.

Transesophageal echocardiography (TEE) demonstrated normal left ventricular size and function with elevated transaortic velocity and gradients and AV thickening. AV area, by the continuity equation, was 0.68 cm^2, and the AV area index was 0.4 cm^2/m^2. The dimensionless index was 0.25. There was also mild to moderate aortic regurgitation. The aortic root and ascending aorta were normal in size. The ascending aorta diameter was measured to be 3.3 cm (1.9 cm/m^2) (Figure 1A, Video 1).

Cardiac computed tomography (CCT) revealed a calcified trileaflet AV with reduced excursion and a discrete narrowing above the sinotubular junction (i.e., both valvular aortic stenosis and SAS). The minimum diameter and area at the level of the supravalvular aortic ridge were measured to be 11.7 mm and 1.4 cm^2, respectively. The diameter of the sinotubular junction was 16.3 mm. There were no signs of aortic coarctation or other congenital anomalies. The coronary arteries did not demonstrate obstructive pathology (Figure 1B).

The patient was deemed to have a class I indication for AV replacement. However, given the valvular and supravalvular stenoses as well as young age, the patient was not considered a good candidate for transcatheter AV implantation. The Ross procedure and the Bentall procedure were offered instead, and the patient ultimately selected the Ross procedure, which was tolerated well. Postoperative two-dimensional TEE revealed trace aortic regurgitation (pulmonary autograft) and normal function, with a peak velocity of 1.0 m/s and a mean gradient of 4 mm Hg.

Case 2: Iatrogenic SAS After Aortic Repair

A 41-year-old man with a history of coronary artery disease underwent surgical robotic mitral valve repair to treat severe mitral regurgitation due to mitral valve prolapse. At the end of the surgery, closure of the cardioplegia site on the ascending aorta required the placement of multiple purse strings to obtain hemostasis given that the aortic wall was very fragile because of a large layer of fat in the adventitia.

Transthoracic echocardiography (TTE) immediately after the operation did not reveal abnormal perivalvular pathology. However, routine repeat TTE 6 months later demonstrated a luminal narrowing of the ascending aorta approximately 1.90 cm above the sinotubular junction, consistent with the level of the cardioplegia repair site. TEE was pursued for further evaluation, which demonstrated turbulent flow in the ascending aorta with a maximum velocity of 3.3 m/s and peak and mean gradients of 44 and 23 mm Hg. The diameter of the ascending aorta at the site of the narrowing was approximately 1.3 cm. The AV appeared normal, and there was no evidence of valvular stenosis (Figure 2A, Video 2).

The patient subsequently underwent CCT, which demonstrated a localized area of luminal narrowing in the ascending aorta proximal to the aortic arch. The minimal area of this region measured 1.3 cm^2, with linear dimensions of 13.6 mm × 12.1 mm. The sinotubular junction diameter was measured to be 23.8 mm (Figure 2B). Given that the patient was asymptomatic, and the severity of the iatrogenic

VIDEO HIGHLIGHTS

Video 1: Two-dimensional TEE, midesophageal zoomed long-axis (125°) view centered on the AV initially without and then with color flow Doppler of the congenital SAS and valvular aortic stenosis, demonstrates the thick, calcified AV with reduced opening and the accelerated flow across the serial stenoses.

Video 2: Two-dimensional TEE, midesophageal zoomed long-axis view centered the ascending aorta initially without and then with color flow Doppler of the iatrogenic SAS, demonstrates the focal narrowing with turbulent flow in the ascending aorta.

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Keywords: Supravalvular aortic stenosis, Echocardiography, CT angiography, Multimodality imaging

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2468-6441

https://doi.org/10.1016/j.case.2026.02.006

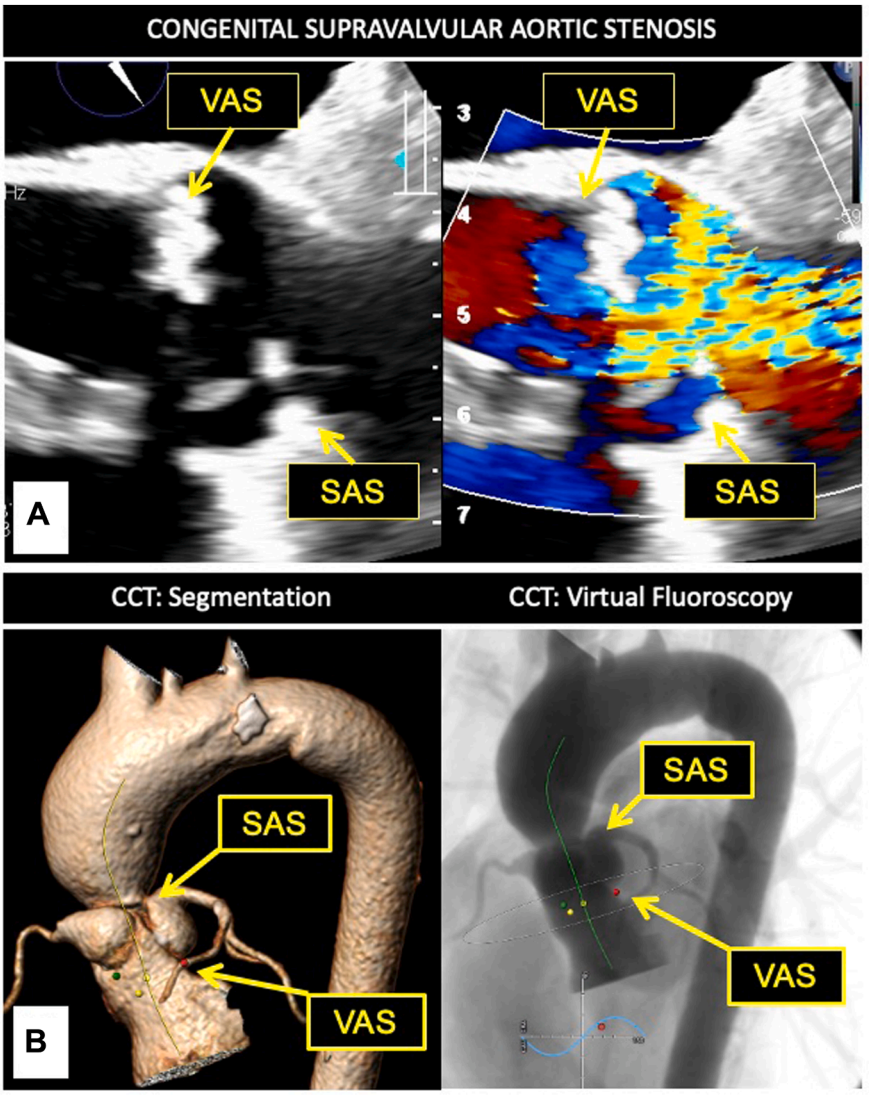

Figure 1 **(A)** Two-dimensional TEE (*top*), midesophageal zoomed long-axis (125°) systolic view centered on the AV without (*left*) and with (*right*) color flow Doppler of the congenital SAS and valvular aortic stenosis (VAS), demonstrates the thick, calcified AV with reduced opening and the accelerated Doppler flow across the serial stenoses. **(B)** CCT (*bottom*), three-dimensional volume-rendered reconstruction (*left*) and virtual fluoroscopy (*right*), sagittal display, demonstrates the VAS and congenital SAS and their relationship to the coronary ostia.

obstruction was moderate, conservative management was recommended, with follow-up TTE in 6 months to ensure a stable supraaortic orifice gradient. Repeat TTE at 6 months, 1 year, and 1.5 years postoperatively continued to demonstrate similar maximal velocity and peak and mean gradients.

DISCUSSION

Although SAS most often presents in patients with congenital diagnoses, iatrogenic causes represent a rare etiology that can present similarly, as demonstrated in this report. The cases presented here highlight the pivotal role of multimodality imaging in the diagnosis and management of SAS regardless of etiology.

In case 1, congenital SAS was identified in an adult presenting with acute decompensated heart failure in the setting of severe perivalvular disease requiring intervention. Consistent with guidelines, multimodality imaging is critical for guiding complex valvular procedures. In this case, imaging confirmed that the patient was a poor candidate for transcatheter AV implantation on the basis of the anatomy in addition to their young age.[2-4] The Ross procedure was considered instead, which the patient ultimately underwent. This approach has been shown to offer distinct advantages for young patients with nonrepairable AV disease, including excellent long-term survival, low reoperation rates, and the avoidance of anticoagulation.[5-9] Following surgery, serial echocardiographic surveillance serves as the cornerstone of long-term follow-up.[10] Although guidelines are limited, prior studies suggest serial follow-up at discharge, 6 months, and annually thereafter.[11]

In case 2, surgical repair of the ascending aorta resulted in iatrogenic SAS. This case highlights a rare but significant complication of cardiac surgery. As in case 1, multimodality imaging was used in a stepwise fashion (TTE, TEE, and CCT), with each modality providing greater diagnostic clarity. The decision to manage conservatively

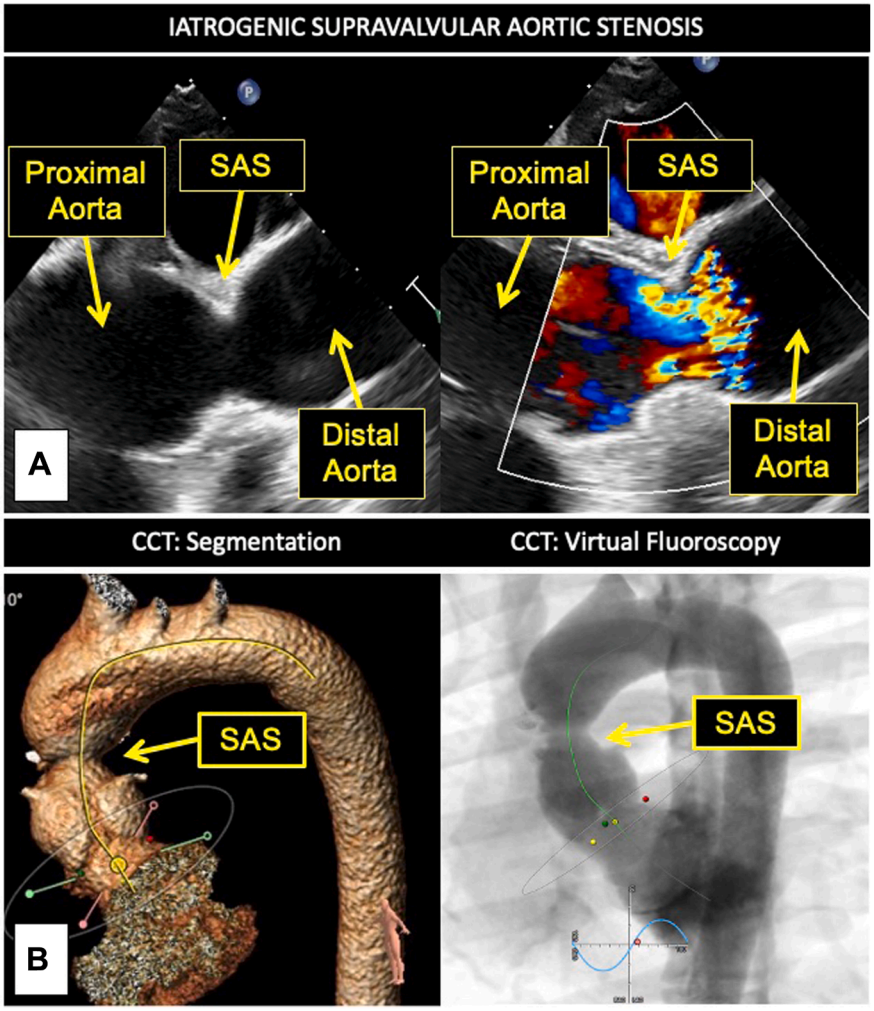

Figure 2 **(A)** Two-dimensional TEE (*top*), midesophageal zoomed long-axis systolic view centered on the ascending aorta without (*left*) and with (*right*) color flow Doppler of the iatrogenic SAS, demonstrates the focal narrowing with the turbulent flow in the ascending aorta. **(B)** CCT (*bottom*), three-dimensional volume-rendered reconstruction (*left*) and virtual fluoroscopy (*right*), sagittal display, demonstrates the iatrogenic SAS.

was based on the patient's overall condition and the moderate severity of the obstruction. Serial echocardiographic surveillance was implemented to ensure stability of the stenosis over time.

These cases underscore the essential role of multimodality imaging in the diagnosis and management of SAS of different etiologies. Conventional and advanced echocardiography provide initial assessment of the severity of SAS. However, given the limitations of Doppler in the case of serial stenoses, complementary imaging modalities are essential.[12,13] CCT offers greater anatomic detail essential for precise diagnosis and management planning. Additionally, cardiovascular magnetic resonance, although less frequently used, serves as an alternative or adjunctive technique to CCT that is noninvasive, is radiation free, and offers high resolution.[14,15] Comprehensive imaging enables timely and accurate identification of SAS and facilitates appropriate interventions and improved patient outcomes.

CONCLUSION

We present multimodality imaging of SAS due to two distinct etiologies, congenital and iatrogenic, using echocardiography and CCT. Both cases demonstrate how multimodality imaging is essential for the accurate diagnosis and optimal management of rare cardiovascular conditions.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a noninterventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient or next of kin under IRB exemption status.

FUNDING STATEMENT

The authors declare that this report did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

DISCLOSURE STATEMENT

The authors report no conflicts of interest.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2026.02.006.

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