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CASE REPORT

INTERMEDIATE

CLINICAL CASE

The "Cooing Pigeon"

TAVI for Acute Aortic Regurgitation Secondary to Homograft Degeneration

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ABSTRACT

We present a case of acute aortic homograft regurgitation manifesting as a new-onset "cooing" murmur in a patient with congenital heart disease who did not have signs of clinical decompensation or evidence of infective endocarditis. He underwent successful transcatheter aortic valve implantation following a diagnosis of sterile acute valvular degeneration. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:2162–5) © 2020 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 21-year-old man was resting at home when he suddenly heard a pigeon cooing from somewhere inside his room. After failing to find the bird, the patient paused and felt his chest, timed the sound to his heartbeat, and realized that the "cooing" came from inside him. Assuming that this was a new murmur, he presented to the emergency department.

At presentation, he was asymptomatic, with no change to his exercise tolerance. He denied any recent febrile illness or weight loss. His blood pressure 146/73 mm Hg, and other observations were unremarkable. Examination demonstrated a regular pulse with quiet heart sounds and a loud diastolic decrescendo cooing murmur audible from the bedside, loudest at the left sternal edge, and radiating to the neck and posterior thorax (Video 1). There was a concurrent grade 3 ejection systolic murmur at the left sternal edge, radiating to the carotid arteries. The patient was euvolemic, without signs of pulmonary edema, endocarditis, or embolic phenomena.

LEARNING OBJECTIVES

- To recognize that acute AR does not always manifest with decompensation or the typical features of AR.
- To refer cases of acute homograft degeneration for multidisciplinary team discussion to consider treatment options that include TAVI.

PAST MEDICAL HISTORY

The patient was under regular follow-up by the adult congenital heart disease team at Barts Heart Centre, London, United Kingdom. At 3 years of age he had undergone aortic balloon valvuloplasty for congenital aortic stenosis, with subsequent elective aortic valve (AV) repair at the age of 13 years that was complicated by *Aspergillus fumigatus* infective endocarditis, aortitis with rupture of the ascending aorta, mycotic aneurysm formation, and embolic ischemia to the mesentery and lower limb. He underwent successful

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21-mm homograft aortic root and AV replacement, small bowel resection, and left below-knee amputation with a good recovery. He had since remained well, taking no regular medications and enrolled in full-time education, with serial echocardiography showing a normal left ventricular (LV) ejection fraction (LVEF) and a homograft mean gradient of 22 mm Hg.

DIFFERENTIAL DIAGNOSIS

In the presence of a new murmur on a background of previous cardiac surgery complicated by infective endocarditis and treated with a prosthetic valve, the primary differential diagnosis was of recurrent AV endocarditis with associated aortic regurgitation (AR).

INVESTIGATIONS

Transthoracic echocardiography demonstrated severe transvalvular AR with diastolic flow reversal in the descending aorta (Video 2). The bioprosthesis was well seated, but an oscillating echogenic structure was attached to the AV noncoronary cusp and was suggestive of a flail leaflet or vegetation. There was moderate aortic stenosis (peak gradient 55 mm Hg), normal LVEF (60% to 65%), and normal LV end-systolic volume. Initial blood tests demonstrated a normal C-reactive protein level and white cell count.

The patient was transferred to his adult congenital heart disease center. Here, transesophageal echocardiography confirmed destruction of the noncoronary cusp with mobile structures and suspicion of perforation in the annulus region with gross AR arising from multiple jets (Videos 3 and 4). The left ventricle was hyperdynamic, with an LVEF >70%. Cardiac magnetic resonance showed LV dilation (ratio of right ventricular to LV end-diastolic volume 1:1.85), an LV end-systolic volume index of 27 ml/m^2, LVEF of 68%, severe AR secondary to a coaptation defect and moderate aortic stenosis. A computed tomography-positron emission tomography scan showed no cardiac uptake. Results of serial blood cultures, including mycobacterial and fungal cultures, were negative, and inflammatory markers remained normal. The patient remained well on serial assessment.

MANAGEMENT

Multidisciplinary team consensus was that the presentation was in keeping with rapid degeneration of the homograft without evidence of infective endocarditis. Management with surgical versus transcatheter AV implantation (TAVI) was considered. Overall, it was believed that surgery would be relatively high risk (entailing a third sternotomy) and that TAVI would be preferred given the patient's amenable vascular anatomy. The annular size (**Figure 1**) allowed for implantation of a 26-mm Edwards Sapien 3 (Edwards Lifesciences, Irvine, California) TAVI without complications, and he was discharged.

ABBREVIATIONS AND ACRONYMS

AR = aortic regurgitation

AV = aortic valve

LV = left ventricular

LVEF = left ventricular ejection fraction

TAVI = transcatheter aortic valve implantation

DISCUSSION

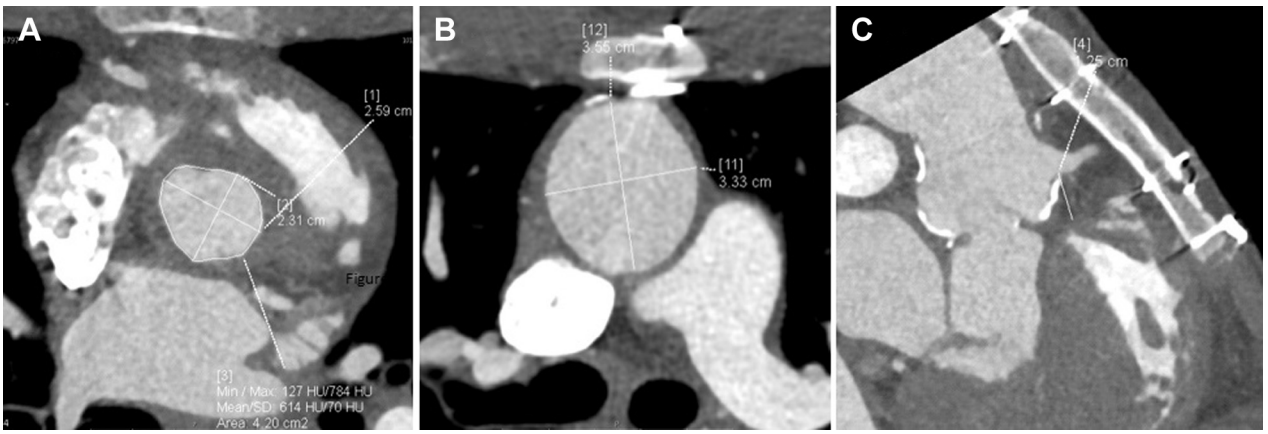
Acute severe AR often manifests as a medical emergency and requires prompt investigation and treatment because, if untreated, it can lead to advanced heart failure and death (1). In chronic AR, the left ventricle gradually dilates over time to maintain cardiac output. In acute AR, however, the left ventricle does not have time to adapt to the sudden increase in end-diastolic volume. This leads to a sudden fall in cardiac output that can manifest as hypotension, cardiogenic shock, and acute pulmonary edema (1). We hypothesize that in this case, the ventricle had been exposed to differing loading conditions from childhood and therefore was adaptable to tolerating acute AR because it was previously remodeled.

The most common causes of acute AR in native valves are infective endocarditis and aortic dissection (1). Other causes include trauma, rupture of congenital malformations, and iatrogenic causes including balloon valvuloplasty (1,2). Acute AR in a prosthetic valve is usually secondary to either endocarditis destroying valve integrity or valve degeneration (1,2). In mechanical valves, regurgitation can also result from pannus or thrombus formation (1).

The murmur in acute AR is normally a low-pitched, short, early diastolic murmur reflecting the sudden equilibration of LV and aortic diastolic pressures (2), rather than the loud decrescendo murmur of chronic AR. The peripheral manifestations of chronic AR that develop secondary to increased stroke volume and wide pulse pressure are typically not seen in acute AR (2).

A "dove-cooing" murmur, as described in our patient, has been reported in only a handful of cases of AR (3,4). Mondillo et al. (3) described a "seagull's cry murmur" occurring in several valvular diseases including severe AR, particularly in the presence of higher-velocity regurgitant flow. Kohno et Al. (4) also described a patient presenting with exertional dyspnea who had a grade 5 diastolic murmur resembling a dove coo. These investigators suggested that the

FIGURE 1 Thoracic CT Images Demonstrating Aortic Root Measurements



(A) Aortic annulus: 2.59 × 2.31cm, area 4.2 cm^2. **(B)** Ascending aorta: 3.33 × 3.55 cm. **(C)** Axial view of the aortic root. CT = computed tomography.

murmur occurred as a result of fluttering of both the AV and the posterior aortic wall, rather than just the valve itself, with induced resonance of the aortic wall facilitated by the mitral valve (4).

TAVI has historically been used in patients with degenerative calcific aortic stenosis, but there has been increasing use in regurgitant native or bioprosthetic AVs. TAVI in patients with native AV regurgitation is technically more challenging than in stenosis, first because of the absence of annular calcification and second because of coexistent dilatation of the aortic root and ascending aorta (5). Data from an international registry study confirmed the early safety and clinical efficacy of TAVI in the bioprosthetic valve group over the native valve group (6). This procedure appears to be a particularly favorable option in patients with congenital AV disease who have already undergone numerous open surgical procedures (7).

FOLLOW-UP

At 5-month follow-up, the patient was asymptomatic, Post-TAVI echocardiography showed a well-seated valve with mobile leaflets, with a peak velocity of 2.4 m/s, peak gradient of 23 mm Hg, and a mean gradient of 15 mm Hg. No significant leak was observed. The LVEF was 60%. He remains under surveillance.

CONCLUSIONS

Our case is interesting for several reasons. First, this patient presented in an unusual way with his new, cooing murmur. A few investigators have previously reported murmurs resembling a range of bird calls, although not in the setting of acute AR secondary to homograft degeneration in the absence of signs of hemodynamic compromise.

Second, this was the second episode of acute AR in this patient, the first instance being an early postoperative fungal infection. Third, this patient underwent a successful TAVI-in-valve procedure for AR to avoid a third sternotomy at the age of 21 years, a rare indication for this intervention.

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AUTHOR DISCLOSURES

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

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KEY WORDS aortic valve, congenital heart defect, transcatheter aortic valve implantation, treatment, valve replacement

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