CASE REPORT

INTERMEDIATE

HEART CARE TEAM/MULTIDISCIPLINARY TEAM LIVE: SPORTS CARDIOLOGY

An Athlete With Bicuspid Aortic Valve Regurgitation and Left Ventricular Dilatation

A Clinical Conundrum

Nikhil Chatrath, MBChB, Raghav Bhatia, MBChB, Saad Fyyaz, MBChB, Hamish Maclachlan, MBChB, Maite Tome, MBBS, MD, Sanjay Sharma, BS, MBChB, MD, Michael Papadakis, MBBS, MD

ABSTRACT

The physiologic cardiac adaptations caused by intensive exercise and the pathophysiologic changes caused by significant regurgitant valvular lesions can be challenging to differentiate. We describe the clinical course of an asymptomatic 31-year-old elite triathlete with a moderately regurgitant bicuspid aortic valve and severe left ventricular and aortic dilatation. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2023;13:101495) © 2023 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/).

A 31-year-old man attended preparticipation cardiovascular screening. He was an elite triathlete, competing in international championships with a weekly training regimen including up to 16 hours of a combination of running, swimming, and cycling. He was completely asymptomatic and had no past medical or family history of note.

LEARNING OBJECTIVES

- To be able to differentiate the cardiac sequelae of moderate or severe aortic valve disease from the effects of cardiac remodeling secondary to athletic adaptation.
- To review current guidelines pertaining to exercise in aortic valve disease and aortopathy.

On physical examination, his height was 183 cm, and his weight was 79 kg. He did not exhibit any features of connective tissue disease. His blood pressure was 120/63 mm Hg, with a heart rate of 40 beats/min. A soft diastolic murmur was heard at the left sternal edge.

INVESTIGATIONS

An electrocardiogram revealed sinus bradycardia and voltage criteria for left ventricular (LV) hypertrophy (**Figure 1**), in keeping with his athletic stature.

A transthoracic echocardiogram (TTE) demonstrated a severely dilated left ventricle with an LV end-diastolic diameter (LVEDD) of 70 mm (**Figure 2**), an LV end-systolic diameter of 40 mm, and an LV

From the Cardiovascular Clinical Academic Group, St George's, University of London, London, United Kingdom.
Eugene Chung, MD, served as the Guest Associate Editor for this paper.
The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received April 4, 2022; revised manuscript received May 2, 2022, accepted May 19, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.05.021

JACC: CASE REPORTS, VOL. 13, 2023
MAY 3, 2023:101495

ABBREVIATIONS AND ACRONYMS

AR = aortic regurgitation

BAV = bicuspid aortic valve

CMR = cardiac magnetic resonance

LV = left ventricular

LVEDD = left ventricular end-diastolic diameter

LVEF = left ventricular ejection fraction

MDT = multidisciplinary team

RV = right ventricular

SOV = sinus of Valsalva

TTE = transthoracic echocardiogram

ejection fraction (LVEF) of 70%. There was biatrial dilatation but normal right ventricular (RV) size and function. The patient had a type 0 bicuspid aortic valve (BAV) with no raphe identified (Figure 3) and a posteriorly directed jet of at least moderate aortic regurgitation (AR), which was challenging to quantify by conventional means because of its highly eccentric nature (Figure 4). There was concurrent aortic dilatation, with a sinus of Valsalva (SOV) diameter of 49 mm (27 mm/m^2) and an ascending aorta of 42 mm (23 mm/m^2) (Figures 5A and 5B).

QUESTION 1: WHAT ARE THE POSSIBLE CAUSES OF LEFT VENTRICULAR DILATATION IN THIS CASE?

In this individual, LV dilatation may have been caused by the AR-induced volume loading on the LV, cardiac adaptation to exercise, or underlying dilated cardiomyopathy.

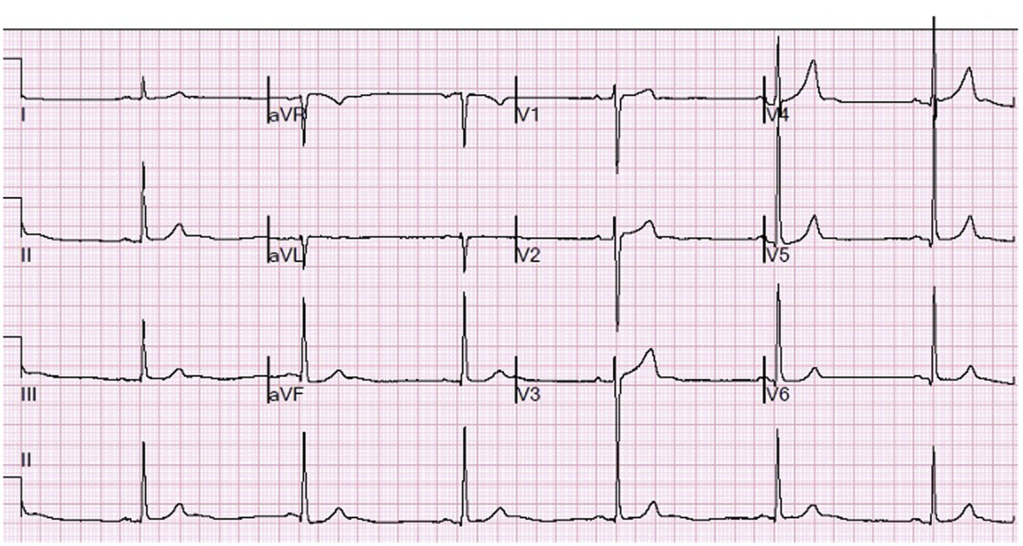
In athletic individuals, an overlap may exist between the cardiac adaptations to exercise and the pathophysiologic effects of AR, and this can present a diagnostic and clinical management dilemma. AR causes volume and pressure loading on the LV, similar to the hemodynamic impact of high-volume, high-endurance exercise. Consequently, one may expect a dilated LV in both scenarios. Furthermore, in athletic individuals, the regurgitant volume may be increased in response to the lengthening of diastole as a result of increased vagal tone and subsequent bradycardia.[1]

An understanding of the common phenotypic features of the “athlete’s heart” is essential to differentiate between the cardiac adaptations to exercise and the pathophysiologic effects of AR. For example, 14% of elite athletes may demonstrate significant LV dilatation (>60 mm). In our patient, the LVEDD was 70 mm, and LV dilatation to this degree, purely resulting from athletic adaptation, is unusual.[2]

Furthermore, in athletes with a dilated LV, other cardiovascular adaptations (eg, proportional RV dilatation) are expected.[3] In our case, the right ventricle remained normal in size, thus further suggesting that the degree of LV dilatation was beyond physiologic cardiac adaptation to exercise.

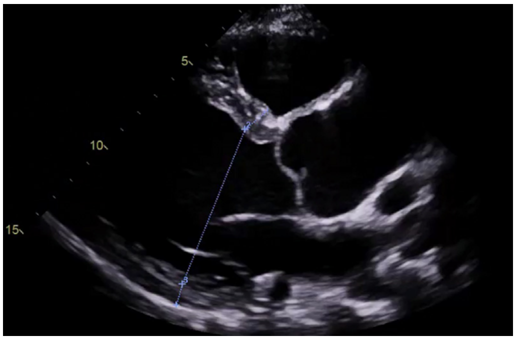
With regard to the aorta, a meta-analysis demonstrated that athletes exhibit 3.2-mm greater dimensions at the SOV compared with sedentary control subjects but with only 1% exceeding conventional upper limits and more commonly seen in athletes participating in isometric exercise.[4] In a more recent study, aortic dilatation, as defined by an aortic dimension of 40 mm or larger, was seen in 21% of competitive masters-level endurance athletes, including 31% of the male athletes.[5]

FIGURE 1 12-Lead Electrocardiogram



A 12-lead electrocardiogram demonstrating sinus bradycardia and voltage criteria for left ventricular hypertrophy.

FIGURE 2 Parasternal Long-Axis Transthoracic Echocardiogram



Parasternal long-axis transthoracic echocardiogram showing severe left ventricular dilatation with an end-diastolic diameter of 70 mm. The interventricular septum measured 10 mm.

Ultimately, although larger aortas may be seen in athletes, the measurement of 49 mm seen in our case was unlikely to be caused by exercise alone and raised further concern about the hemodynamic sequelae of the BAV and AR.

The patient was subsequently referred to our institution for further assessment.

QUESTION 2: WHAT FURTHER INVESTIGATIONS ARE INDICATED AT THIS STAGE?

Current international guidelines recommend an objective assessment of functional capacity. Furthermore, a cardiac magnetic resonance (CMR) study would provide an accurate assessment of LV volume, LVEF, and flow calculations and would detect the presence of myocardial scar.[6,7] The whole aorta can also be visualized for more accurate measurements and to identify conditions known to be associated with a BAV such as coarctation of the aorta.

Exercise echocardiography may provide more information to guide decision making.[1] Athletes with a dilated LV often demonstrate a reduced LVEF at rest that improves on exercise. Conversely, individuals with AR may demonstrate a hyperdynamic LV in the early stages, before systolic function impairment in more advanced, severe AR, with poor contractile reserve demonstrated on exercise testing.

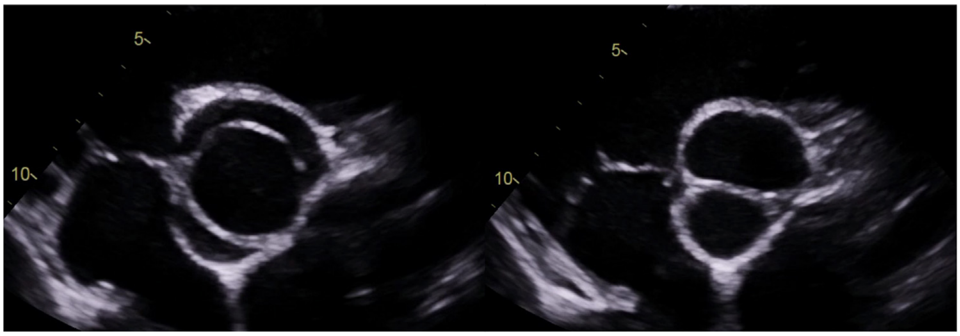
In our patient, further investigation included a cardiopulmonary exercise test using a 36-W ramp protocol on a cycle ergometer, where he achieved a peak oxygen consumption of 75 mL/min/kg, representing 182% of his predicted, with satisfactory blood pressure and heart rate responses and no arrhythmias identified.

CMR confirmed a severely dilated LV (LV end-diastolic volume 340 mL) and an increased stroke volume of 246 mL. The AR was demonstrated with a regurgitant fraction of 43% and regurgitant volume of 84 mL. The aortogram confirmed a dilated aortic root with an SOV of 49 mm and an ascending aorta of 42 mm (**Figure 6**). There was no evidence of myocardial fibrosis.

QUESTION 3: HOW DOES EXERCISE AFFECT THE NATURAL HISTORY OF A BICUSPID AORTIC VALVE?

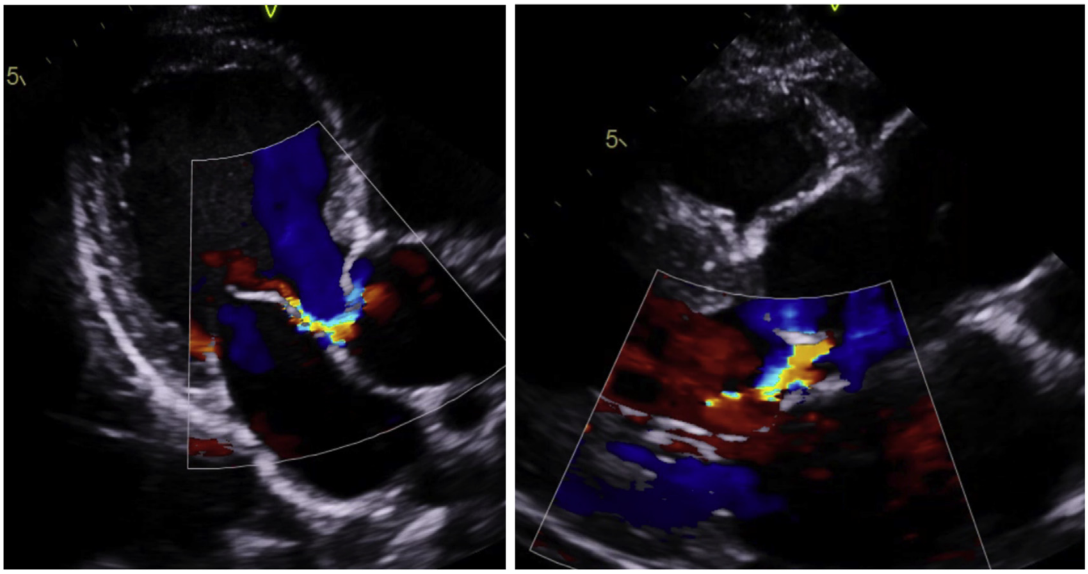
BAV is the most common cardiac defect, with an estimated prevalence of 1% to 2% in the general population.[8] As such, although it is often an

FIGURE 3 Short-Axis Transthoracic Echocardiogram View



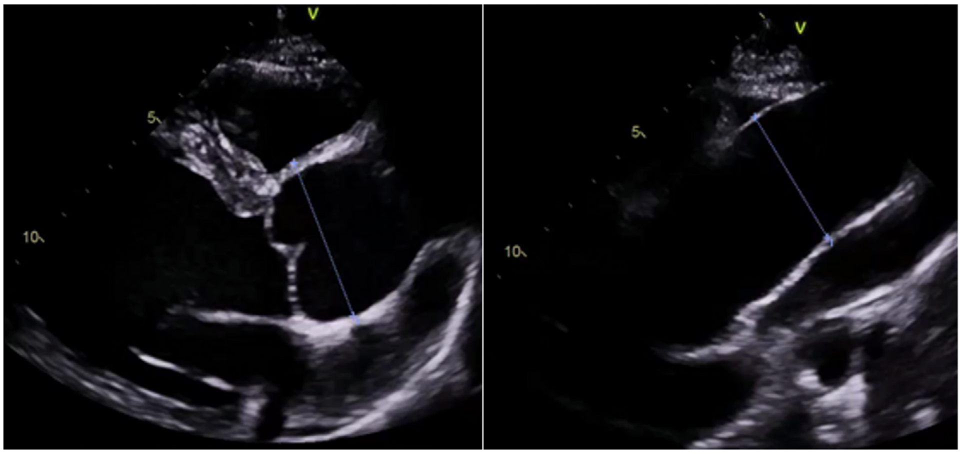
Short-axis transthoracic echocardiogram view at the level of the aortic valve. Two cusps are demonstrated with no discernible raphe (type 0 bicuspid aortic valve). The leaflets are freely mobile, and no significant calcification is seen.

FIGURE 4 Transthoracic Echocardiogram



Transthoracic echocardiogram demonstrating a posteriorly directed, highly eccentric jet of at least moderate aortic regurgitation in the **(left)** apical 3-chamber and **(right)** parasternal long-axis view. The quantitative assessment of aortic regurgitation severity may be underestimated by the transthoracic echocardiogram because of the eccentric nature of the jet and the hyperdynamic left ventricular function. Subsequent cardiac magnetic resonance demonstrated a regurgitant fraction of 43% and a regurgitant volume of 84 mL.

FIGURE 5 Transthoracic Echocardiogram



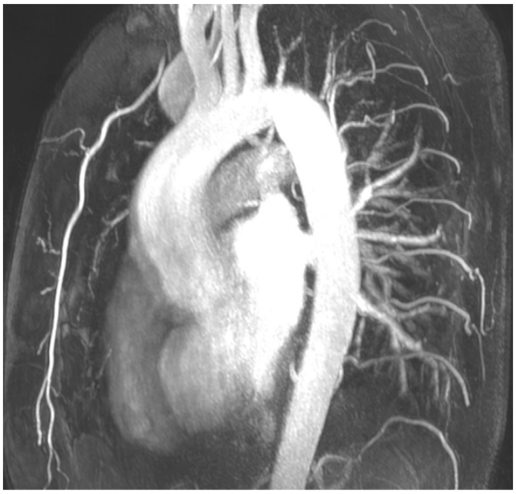
Transthoracic echocardiogram views of the aorta demonstrating **(A)** a sinus of Valsalva diameter of 49 mm and **(B)** an ascending aorta diameter of 42 mm.

incidental finding, BAV is commonly encountered by sports cardiologists and is the most common cause of primary AR in young athletes.

However, the natural history of BAV and the effect of long-term, intense exercise are not fully understood. In a study of athletes with BAV who were matched with sedentary individuals with BAV and athletes with a trileaflet aortic valve, progression of valve disease was not demonstrated over a 3-year follow-up period.[9] In longer-term follow-up studies, the majority of athletes with BAV had a benign clinical course, and progression of root dilatation, valve stenosis, or AR likely occurred independently of athletic activity.[10]

Regardless of the potential impact of exercise, AR or aortic stenosis may develop by the fifth decade, and up to 40% of these patients may develop aortopathy; therefore, regular imaging surveillance of individuals with BAV is recommended.[4]

FIGURE 6 Magnetic Resonance Aortogram



Magnetic resonance aortogram demonstrating severe dilatation of the aortic root and no associated coarctation.

QUESTION 4: WHAT ARE THE MANAGEMENT OPTIONS, AND WHO SHOULD BE INVOLVED IN DECISION MAKING?

There are several management options at this stage: continue to monitor the athlete with repeat imaging; consider a period of detraining to reassess the LV dimensions; or consider surgical treatment, given the degree of aortic root dilatation and AR. Unless hypertension is present, there are limited medical strategies indicated at this stage.

This case highlights the importance of shared decision making to devise a management plan that balances expected outcomes with patient preferences and values. The patient, physician, surgeon, and the patient's family and trainers are key stakeholders in this process.

The risk of aortic dissection, given the aortic dimension of 49 mm, was a cause for concern in our patient, especially in the context of a high volume and intensity of exercise that could cause repeated and sustained elevation in wall tension and a gradual increase in aortic diameter. The case was discussed with the multidisciplinary team (MDT), and management options were relayed to the patient.

In line with guideline recommendations, the patient was advised to pursue only low-intensity endurance sports.[6] He did not want a period of detraining and desired a longer-term management plan that would not significantly affect his ability to undertake intensive exercise.

Initially, a valve-sparing aortic root replacement was discussed, but in the context of a BAV with already at least moderate AR, the MDT consensus was that the long-term prognosis and freedom from reintervention would be more favorable if both the root and the valve were replaced simultaneously. The risks and benefits of a mechanical valve replacement versus a tissue valve replacement were discussed with the patient, and on the basis of longer-term durability, a metallic valve was selected.

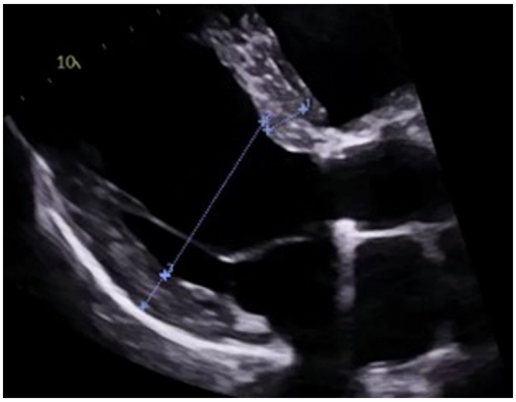
The patient subsequently underwent surgery, with excision of the aneurysmal part of the aorta and replacement of the aortic root and reimplantation of the coronary arteries.

A 25-mm ATS composite mechanical valve (ATS Medical) was implanted, and the patient remains on lifelong anticoagulation.

QUESTION 5: WHAT ADVICE REGARDING EXERCISE PARTICIPATION SHOULD BE GIVEN?

Current international guidelines for sports participation in patients with BAV are identical to guidelines in patients with a trileaflet aortic valve, if the aorta is not dilated.[1,6,7] Individuals with severe AR may participate in low- and moderate-intensity exercise if the LV is not dilated, the LVEF remains >50%, the aortic root is within normal dimensions or is mildly dilated, and exercise stress testing results are normal. Only low-intensity exercise is recommended for those patients with a dilated left ventricle, LVEF <50%, significant aortic root dilatation, or exercise-induced arrhythmias. With regard to the aortic root, individuals with

FIGURE 7 Parasternal Long-Axis Transthoracic Echocardiogram



Postoperative parasternal long-axis transthoracic echocardiogram (TTE) showing a left ventricular end-diastolic diameter of 57 mm. The metallic aortic valve can be seen with the root replacement. The "+" just denotes the start and finish of the measurement lines.

an aorta of >50 mm are considered at high risk, and competitive sports are not recommended.[1,6,7] If mechanical valve replacement is performed, individuals should not engage in contact sports or athletic activities where collision is likely because of the risks of anticoagulation.

In our case, the patient was advised to pursue only low-intensity endurance sports while awaiting surgery, in line with guideline recommendations. Following surgery, the importance of using protective equipment, particularly when cycling, and avoiding steep slopes at a rapid pace has been reiterated to the patient.

FOLLOW-UP

The patient remains clinically well, and although not participating in competitive triathlons, he is still exercising for more than 12 hours per week. He has been advised to avoid contact sports because of the anticoagulation. On serial TTE follow-up, the mechanical aortic valve remains functioning well, and a reduction in the LV size (LVEDD, 57 mm) (**Figure 7**) just 1 year post-surgery has been demonstrated. Genetic testing did not identify a gene associated with a heritable thoracic aortic disease, and on screening his first-degree relatives, no evidence of a BAV or aortopathy was identified.

CONCLUSIONS

The hemodynamic sequelae of AR associated with BAV may overlap with the cardiovascular adaptations to intensive exercise. Thorough evaluation, including multimodality imaging, helps in differentiating between the 2 conditions, and management, including exercise restrictions, is often guided by aortic size.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

Drs Chatrath, Bhatia, Fyyaz have received research grant funding from the charitable organization Cardiac Risk in the Young. Dr Papadakis has received research funding from Cardiac Risk in the Young. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Nikhil Chatrath, Cardiovascular Clinical Academic Group, St George's, University of London, Cranmer Terrace, London SW17 0RE, United Kingdom. E-mail: nchatrat@sgul.ac.uk.

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KEY WORDS aorta, bicuspid aortic valve, echocardiography, exercise, left ventricle