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AORTIC DILATATION AS A CLUE TO BICUSPID AORTIC VALVE FROM DIAGNOSIS TO TIMELY SURGERY A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Background: Bicuspid aortic valve (BAV) may be associated with progressive aortic enlargement despite preserved valve function. Selecting the appropriate time for prophylactic intervention requires aortic phenotyping and individualized risk assessment. Case presentation: A 56-year-old man without significant medical history or cardiovascular risk factors was evaluated before surgery for lumbar disc herniation with motor-deficit sciatica. He was asymptomatic from a cardiac standpoint; examination revealed no aortic murmur, blood pressure was 120/70 mmHg, and ECG showed sinus rhythm at 75 beats/min without left ventricular hypertrophy. Chest radiography suggested proximal aortic enlargement. Echocardiography revealed type 0 BAV with morphologically altered cusps but neither stenosis nor regurgitation, and an aortic diameter of 42 mm at both the sinuses of Valsalva and sinotubular junction. Cardiac chambers were normal, and laboratory assessment was unremarkable. **Discussion**: BAV-related aortopathy requires evaluation independently of valve dysfunction. Current US and European guidance generally supports imaging surveillance at 42 mm rather than prophylactic surgery, in the absence of additional high-risk findings. **Conclusion**: Comprehensive aortic imaging, familial assessment, and lifelong surveillance are central to management of incidentally identified BAV aortopathy.

KEYWORDS: Bicuspid aortic valve; aortic root dilatation; type 0; aortopathy; surgical threshold; surveillance.

INTRODUCTION

Bicuspid aortic valve is a common congenital valvular anomaly, with heterogeneous phenotypes and long-term risk of aortic stenosis, regurgitation, and proximal aortic enlargement.[1,2] Aortopathy may occur in the presence of normal valve function, and its management must therefore be separated from valve intervention decisions.[2–4] Recent European and North American guidance emphasizes maximal aortic diameter, root versus tubular ascending phenotype, growth rate, associated risk factors, and the expertise of the aortic team.[3–6] We describe a functionally competent type 0 BAV detected incidentally during assessment for noncardiac surgery, and review when prophylactic aortic intervention should be considered.

CASE PRESENTATION

A 56-year-old man with no notable medical or surgical history and no reported similar cases among his siblings was referred by a neurosurgeon for cardiovascular evaluation before an operation for lumbar disc herniation complicated by motor-deficit sciatica. He reported no known conventional cardiovascular risk factors. There were no reported cardiac symptoms. Examination was normal; blood pressure was 120/70 mmHg, peripheral pulses were palpable and symmetric in amplitude, and no systolic or diastolic aortic murmur or clinical sign of heart failure was detected. Physical examination revealed no clinical features suggestive of Marfan syndrome.

The 12-lead ECG showed sinus rhythm at 75 beats/min without signs of left ventricular hypertrophy. *Chest radiography* demonstrated a cardiac silhouette of normal size with an enlarged appearance of the proximal aorta. *Routine laboratory tests* were normal.

Transthoracic echocardiography identified a bicuspid aortic valve without a visible raphe (Sievers type 0), with morphologically altered leaflets but no hemodynamically significant stenosis or aortic regurgitation. Diameters at both the sinuses of Valsalva and the sinotubular junction were reported as 42 mm. The tubular ascending aorta and the aortic arch were not dilated. The mitral valve was morphologically dystrophic without dysfunction. The left ventricle was neither dilated nor hypertrophied; the left atrium and right-sided chambers were not enlarged. There was no pulmonary hypertension or pericardial effusion.

Annual clinical and echocardiographic surveillance was planned to monitor aortic dimensions and aortic valve function.

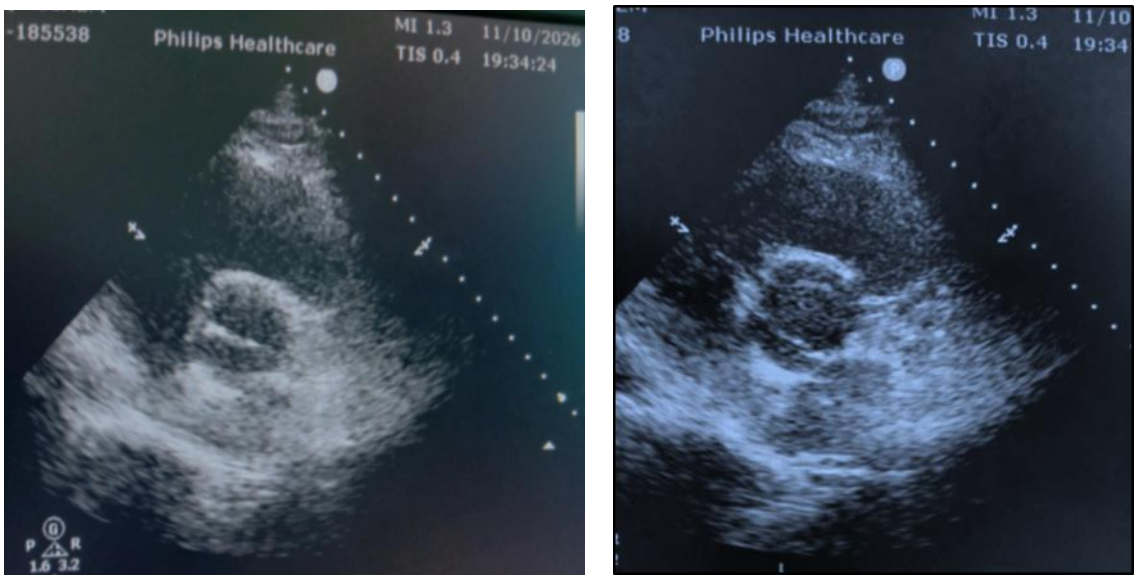

Figure 1: Parasternal short-axis imaging demonstrated a bicuspid aortic valve with two cusps and two commissures, without an identifiable raphe, consistent with Sievers type 0 morphology. The valve exhibited an oval-shaped systolic opening, with no aortic stenosis or regurgitation on Doppler assessment.

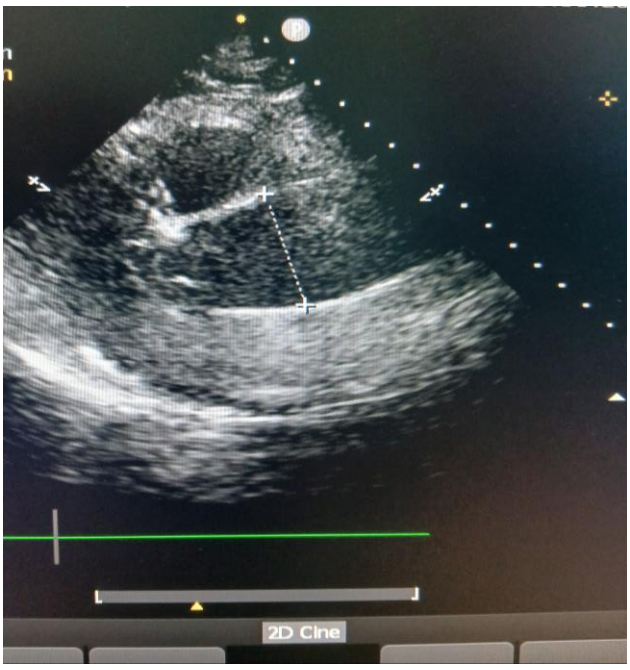

Figure 2: Parasternal long-axis imaging showed proximal aortic dilatation, measuring 42 mm at both the sinuses of Valsalva and the sinotubular junction."

DISCUSSION

A clinically silent valve–aorta disorder

Normal auscultation does not exclude BAV when no significant stenosis or regurgitation is present. In this patient, the abnormal proximal aortic contour on chest radiography was the only extracardiographic clue. BAV is best regarded as a valve–aorta disorder: a competent valve may coexist with clinically relevant aortic enlargement.[1,2] Longitudinal cohorts show a substantial lifetime burden of valve interventions and aortic complications, although the absolute risk differs by phenotype and clinical setting.[7,8]

Morphology and phenotype

The classical Sievers designation "type 0" denotes absence of a raphe. The international consensus classification additionally distinguishes fused, two-sinus and partial-fusion phenotypes.[1] The echocardiographic description is compatible with two functional cusps without a visible raphe, but definitive phenotyping requires careful review of valve and sinus anatomy. BAV aortopathy may be predominantly root-related, tubular ascending, or extensive. Because the present observation supplies the diameters of the sinuses and sinotubular junction only, labeling the condition as definitively "root phenotype" would be premature until the entire ascending aorta is assessed.

Why can the aorta dilate without valve dysfunction?

Both intrinsic wall susceptibility and abnormal aortic flow contribute to bicuspid-associated aortopathy. Genetic and developmental influences on vascular smooth-muscle cells and extracellular matrix coexist with eccentric flow, altered wall shear stress, and regionally heterogeneous remodeling.[1,2] Accordingly, aortic surveillance cannot be based solely on transvalvular gradients or the degree of regurgitation.

When should prophylactic aortic surgery be considered?

The 2022 ACC/AHA guideline recommends replacement of the aortic root and/or ascending aorta at a maximal diameter of at least 55 mm in BAV.[3] Surgery is reasonable at 50–54 mm with a recognized additional risk factor (family history of dissection, rapid growth ≥3 mm/year, coarctation, or root phenotype aortopathy), and may be reasonable in selected low-risk patients without additional factors. A cross-sectional area/height ratio ≥10 cm²/m is another criterion supporting earlier intervention. In patients already undergoing indicated surgical aortic valve repair or replacement, concomitant aortic replacement is reasonable at ≥45 mm.[3]

European guidance also takes a phenotype-specific approach. The 2024 ESC aortic guideline and the EACTS/STS aortic guideline identify ≥55 mm as a general operative threshold, with lower thresholds for carefully selected BAV patients, particularly root-predominant disease at ≥50 mm and selected ascending phenotypes with increasing risk at approximately 50–52 mm.[4,5] The 2025 ESC/EACTS valvular guideline should be interpreted alongside dedicated aortic guidance when there is an independent indication for valve surgery.[6] Distinct guideline classes, qualifiers, and centre-expertise requirements should not be collapsed into a universal 50-mm rule.

Setting	Guideline-informed implication
BAV aortic diameter ≥55 mm	Elective root/ascending replacement generally indicated.[3–5]
50–54 mm	Individualized consideration in experienced aortic centres; risk modifiers and aortic phenotype matter.[3–5]
BAV root phenotype ≥50 mm	European aortic recommendations support earlier surgery for this phenotype.[4,5]
≥45 mm during indicated surgical aortic valve operation	Consider concomitant aortic replacement; this does not apply to an otherwise competent valve.[3,5,6]
Area/height ≥10 cm²/m	ACC/AHA supports consideration of intervention by a multidisciplinary aortic team.[3]
42 mm with no established high-risk modifier	Baseline tomographic assessment and serial surveillance; no isolated prophylactic operation on diameter alone.[3–6]

For our patient, the documented 42-mm diameter is below usual thresholds for either isolated prophylactic aortic replacement or concomitant replacement during valve surgery. Moreover, no indication for valve surgery exists because the valve is functionally competent. Aortic surgery is therefore not warranted solely by the information currently available. This conclusion should be reassessed if cross-sectional

imaging reveals substantially larger dimensions elsewhere, rapid growth, syndromic disease, or other risk modifiers.

Imaging and surveillance

A baseline ECG-gated CT angiogram or CMR is appropriate to define the sinuses, sinotubular junction, ascending tubular aorta, and arch; assess coarctation; and confirm maximal aortic diameter using standardized measurement methods.[3–6] The 2022 ACC/AHA guideline recommends lifelong serial imaging for BAV patients with aortic root or ascending aortic diameter ≥40 mm, with interval tailored to dimension and rate of growth.[3] In this case, reassessment after approximately 6–12 months could establish growth velocity, followed by individualized monitoring if stable; this is a proposed care plan, not a documented patient outcome. First-degree relatives should be offered echocardiographic screening.[3]

Implications for the planned neurosurgical intervention

Preoperative cardiovascular assessment was requested before planned neurosurgery for lumbar disc herniation complicated by motor-deficit sciatica. A functionally competent bicuspid aortic valve and an aortic diameter of 42 mm do not, in themselves, warrant prophylactic aortic surgery or cancellation of necessary noncardiac surgery. Perioperative management should be individualized through collaboration among the neurosurgical, anesthetic, and cardiology teams, with attention to hemodynamic control and clarification of any uncertainty regarding aortic anatomy.

Strengths and clinical message

The main educational strength of this case is the incidental detection of a type 0 bicuspid aortic valve with proximal aortic dilatation despite an unremarkable cardiovascular examination and preserved valve function. It highlights the importance of carefully assessing aortic valve morphology whenever aortic dilatation is identified. The absence of dilatation of the tubular ascending aorta and aortic arch on echocardiography helps define the distribution of aortic involvement. Annual clinical and echocardiographic surveillance provides an opportunity to detect changes in aortic dimensions or valve function and reassess the timing of intervention. The central message is that recognizing bicuspid aortic valve–associated aortopathy enables structured surveillance and individualized surgical decision-making.

CONCLUSION

A type 0 bicuspid aortic valve can remain clinically silent while proximal aortic enlargement is present. In this asymptomatic 56-year-old man, valve function was preserved and the reported aortic diameter was 42 mm. Current international guidance supports comprehensive baseline imaging and lifelong surveillance rather than surgery based on this diameter alone. Decisions regarding prophylactic aortic replacement should integrate the maximum verified diameter, anatomical phenotype, growth, familial and syndromic risk, body size, concomitant valve pathology, and multidisciplinary expertise.

Declarations

Patient consent *for publication: The patient provided consent for publication of this case report.*

Conflicts of interest*: The authors declare no conflicts of interest.*

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Author contributions*: Professor Annane contributed to the critical revision of the manuscript.*

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