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Case Report

Severe Aortic Regurgitation and Ascending Aneurysm in a Patient with Pentacuspid Aortic Valve: Case Report and Review

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Abstract

Pentacuspid aortic valve is an exceptionally rare congenital anomaly, often associated with functional deterioration and aortopathy. We report a case of a 39-year-old male presenting with severe aortic regurgitation and ascending aortic aneurysm in the setting of a pentacuspid aortic valve. The patient underwent a successful Bentall and hemiarch replacement using a composite mechanical valved conduit. This case emphasizes the potential association between rare aortic valve morphologies and ascending aortic pathology and includes a brief review of existing literature on pentacuspid aortic valve.

Keywords: pentacuspid aortic valve; aortic regurgitation; ascending aortic aneurysm; Bentall procedure; congenital aortic valve anomaly; case report

1. Introduction

Congenital anomalies of the aortic valve (CAAV) are rare, affecting approximately 1–2% of the general population [1–4]. Pentacuspid aortic valve (PAV), defined by the presence of five aortic cusps, is the rarest form, with estimated prevalence of less than 1 in a million people [2]. Most of very few reported PAV cases are associated with valvular dysfunction, primarily aortic regurgitation (AR), and a subset are complicated by ascending aortic aneurysms [5–17]. The pathophysiological mechanisms linking abnormal cusp morphology and aortopathy remain incompletely understood but may share features with those seen in bicuspid aortic valve (BAV) related aortopathy [3,18,19].

This case report describes a patient with severe AR and ascending aortic aneurysm caused by a functionally pentacuspid valve and discusses its clinical and surgical implications in the context of available literature.

2. Case Presentation

A 39-year-old male presented with symptoms of exertional dyspnea, orthopnea, and episodes of shortness of breath. Ten months prior to admission, he experienced pulmonary edema, which was managed medically at a regional hospital. The patient had no known comorbidities but was an active smoker.

Upon admission, he was normotensive (blood pressure 120/70 mmHg) and receiving the following medications: bisoprolol 2.5 mg once daily, ramipril 2.5 mg once daily, atorvastatin 20 mg once daily, furosemide once daily, and spironolactone 25 mg once daily. Electrocardiogram showed sinus rhythm with a heart rate of 76 beats per minute.

Transthoracic echocardiography (TTE) revealed a left ventricular end-diastolic diameter (LVEDD) of 65 mm and end-systolic diameter (LVESD) of 48 mm, with a preserved ejection fraction

(LVEF) of 56%. Severe aortic regurgitation (grade 4+) was present, with an effective regurgitant orifice area (EROA) of 0.41 cm², regurgitant volume (RV) of 65 mL/beat, and regurgitant fraction (RF) of 52% [20]. The aortic annulus measured 27 mm, the sinotubular junction 40 mm, and the ascending aorta 54 mm. Mild mitral and tricuspid regurgitations (1+) were observed, with a systolic pulmonary artery pressure of 58 mmHg. The aortic valve was functionally described as bicuspid, though leaflet morphology was not clearly defined. Coronary angiography revealed normal coronary arteries.

The patient underwent surgery via antegrade partial cardiopulmonary bypass (CPB). A Bentall procedure with hemiarch replacement was performed using a composite On-X Ascending Aortic Prosthesis (25 mm) with Vascutek Gelweave Valsalva Graft (26 mm) (Terumo Cardiovascular Systems, Ann Arbor, MI). The cardiopulmonary bypass time was 185 minutes, aortic cross-clamp (ACC) time 105 minutes, and circulatory arrest (CA) time 10 minutes (open distal anastomosis) at a core temperature of 19°C and jugular venous oxygen saturation of 100% (Figure 1). Intraoperative transesophageal echocardiography (TEE) showed normal prosthetic valve function and satisfactory hemodynamic performance.

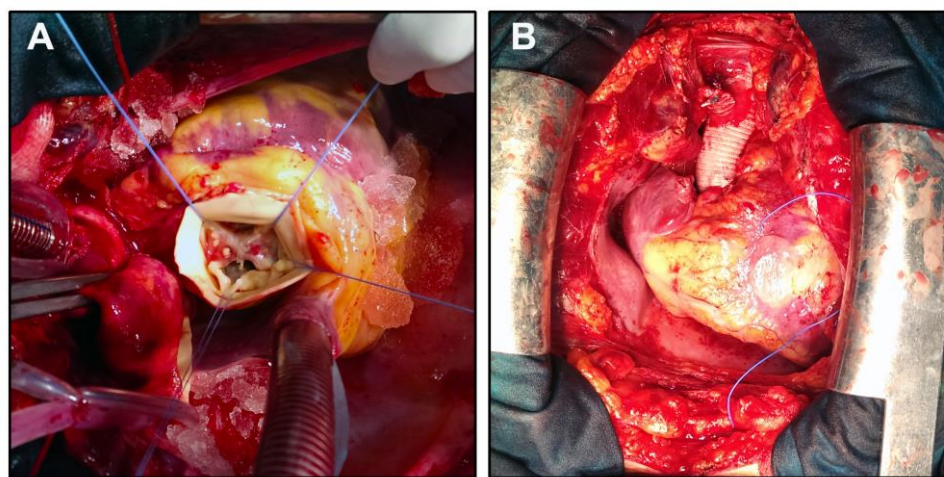


Figure 1. Intraoperative views during surgical correction of a PAV and ascending aortic aneurysm. (A) Intraoperative exposure of a PAV during aortic root replacement. The valve shows five distinct cusps with asymmetric morphology. Stay sutures are placed to retract the aortic wall and expose the valve anatomy. **(B)** Completion of the procedure following valve and ascending aorta replacement with a composite graft. The graft anastomosis and surrounding structures are shown in situ prior to chest closure.

Intraoperative aortic root analysis (Figure 2) revealed a severely dysmorphic PAV with five identifiable cusp regions of irregular sizes and partial fusion predisposing central malcoaptation and eccentric aortic regurgitation. Cusp 1 (green) appears well-developed and free-standing, corresponding with non-coronary cusp of normal, tricuspid valve, with true commissures towards neighboring cusps. Cusp 2 (red) is the smallest accessory cusp. Between cusps 3 (blue), 4 (purple), and 5 (yellow), there appears to be a fusion zone forming raphe-like structures. The commissures between these cusps are asymmetric and seem shallower or less developed, consistent with pseudocommissures [4]. Partial cusp fusion is visible between cusps 2 and 3. Anomalous cord, extending from the raphe of the conjoined cusp, is protruding into excentric orifice [21]. Present morphology with cusp asymmetry and fusion is likely to exhibit bicuspid physiology, as seen on TEE.

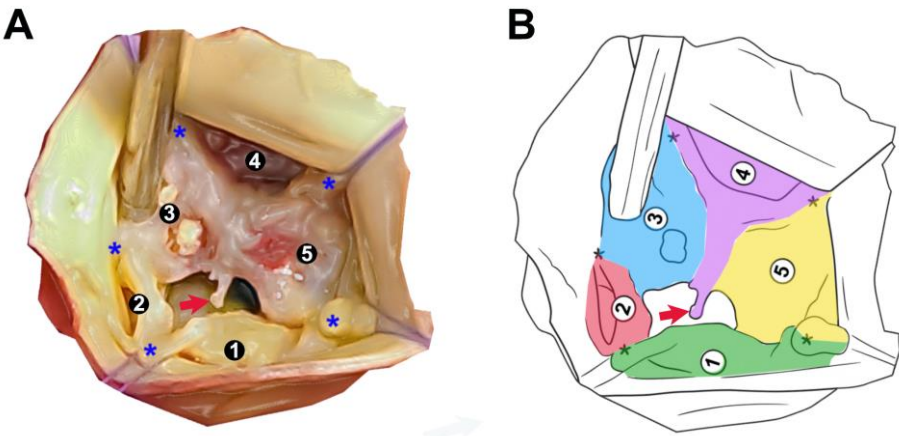


Figure 2. Morphological anatomy of a pentacuspid aortic valve (PAV): surgical and schematic views. (A) Intraoperative photograph demonstrating a severely dysmorphic PAV with five identifiable cusp regions labeled 1–5. Note the irregular cusp sizes, variable commissural spacing, and presence of fused and rudimentary cusp tissue. Blue asterisks (*) denote commissures and pseudocommissures. The red arrow indicates the presence of anomalous cord, extending from the raphe of the conjoined cusp **(B)** Corresponding schematic illustration of the valve morphology shown in (A), highlighting the asymmetry and heterogeneous cusp arrangement. Each cusp is distinctly color-coded (green, red, blue, purple, yellow) and labeled for correlation with panel A. Note the fusion between cusps 3, 4 and 5, as well as the underdeveloped morphology of cusp 2.

The postoperative period was marked by prolonged leukocytosis and neutrophilia (until postoperative day 6), with leukocyte counts to $19 \times 10^9/L$ and neutrophils up to 85%. A de-escalation antibiotic regimen including vancomycin and meropenem was administered. The patient remained afebrile throughout hospitalization. Urine and blood cultures, as well as surgical wound swabs, were negative for infection. Upon normalization of white blood cell counts ($Leu\ 9.1 \times 10^9/L$, $Neu\ 65.7\%$), the patient was discharged on postoperative day 13 in stable hemodynamic condition and without signs of systemic or local infection.

At 11-year follow-up, the patient remains in good general condition with normal function of the aortic prosthesis and preserved left ventricular systolic function.

3. Discussion

Structural CAAV affect approximately 1–2% of the general population, with BAV being the most common, occurring in 0.5–1.4% of individuals [22,23]. Unicuspid aortic valve (UAV) is considerably rarer, with an estimated prevalence of 0.02% [24], while quadricuspid aortic valve (QAV) is observed in 0.003–0.013% of the population [25]. In contrast, PAV is extraordinarily rare, with only isolated case reports in the literature and no robust epidemiological data, suggesting a prevalence likely well below 0.0001%, or fewer than 1 in 1 million [4]. This extreme rarity has precluded formal population-based prevalence estimates and highlights the need for systematic reporting and surveillance.

Review of 14 reported PAV cases (Supplementary Material: Table S1) reveals a consistent association between the young age (24–39 years), asymmetric cusp morphology, severe eccentric AR, and aortic dilatation, particularly of the root or ascending aorta, in four (28.6%) cases, including the current report [6,9,10]. Of those, three cases underwent root and ascending aorta reconstructions (i.e. Robicsek, Bentall), in addition to AV replacement [9,10]. In our case, the patient underwent a Bentall procedure with hemiarch replacement, which remains the treatment of choice in patients with combined valvular insufficiency and ascending aortic aneurysm [26]. In this case, the decision to use a mechanical composite graft was appropriate given the patient's young age, preserved ventricular function, and the need for durable correction. The uneventful recovery and excel-lent 11-year follow-up outcome reinforce the long-term efficacy of this approach when tailored appropriately.

The observed asymmetry, including a rudimentary cusp (Cusp 2) and raphe formation between cusps 3, 4, and 5 (Figure 2), supports the hypothesis that cusp malformation and fusion contribute to poor coaptation and progressive AR [4]. In addition, the anomalous cord arising from the raphe's free margin, and floating within the regurgitant orifice (Figure 2), adds a unique structural abnormality not previously described with PAV. Similar structure, with its free portion attached to the aorta, has been documented in some BAV cases, contributing to chronic and acute AR [21].

The co-occurrence of root and ascending aortic dilatation in current and previously reported cases, contribute to the hemodynamic burden on the aortic wall, either directly (via eccentric regurgitant jets) or indirectly (through altered flow patterns and wall stress), promoting aneurysmal formation and remodeling [4]. There is also accumulating evidence that CAAV share some genetics and embryological defects involving both valvulogenesis and aortic wall maturation, predisposing aortopathy. Rare histological examination of the resected PAV demonstrated myxomatous degeneration, may support possible underlying connective tissue abnormality [27].

Associated congenital anomalies were reported in two cases: patent foramen ovale [8], and renal arteries dysplasia [13].

Despite the rarity of PAV, this case underscores the importance of considering it in the differential diagnosis of unexplained AR, especially in young people and when associated with ascending aortic dilation. Preoperative recognition remains difficult but may improve with advances in imaging techniques such as 3D echocardiography, MDCT and cardiac MRI, which can better delineate commissural architecture and leaflet morphology [28].

4. Conclusions

Pentacuspid aortic valve (PAV) represents an exceptionally rare congenital anomaly, most often presenting with asymmetric cusp morphology, severe aortic regurgitation, and, in a subset of patients, ascending aortic dilatation. Review of the literature reveals a reproducible pattern of functional insufficiency and structural aortopathy, frequently necessitating combined valvular and aortic surgical intervention. These findings suggest a pathophysiological overlap with BAV-associated aortopathy and support the notion that PAV should be approached as a valvulo-aortic disease entity.

However, current understanding is severely limited by the rarity of PAV, lack of centralized registries, heterogeneous reporting, and scarce histological or genetic characterization. As a result, long-term outcomes, optimal timing of intervention, and risk stratification remain largely undefined.

Future efforts should prioritize the development of multicenter registries and standardized morphological classification schemes. Advanced imaging techniques may enhance preoperative recognition, while systematic histological and molecular investigations could uncover developmental or genetic substrates underlying PAV and its associated aortopathy. Such coordinated strategies are essential to transition PAV from an anecdotal curiosity to a better-understood clinical and surgical entity.

Supplementary Materials: The following supporting information can be downloaded at the website of this paper posted on Preprints.org. Table S1: Literature review of the reported PAV cases.

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Abbreviations

The following abbreviations are used in this manuscript:

MDPI	Multidisciplinary Digital Publishing Institute
DOAJ	Directory of open access journals
TLA	Three letter acronym
LD	Linear dichroism

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