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Unicuspid Aortic Valve! *Not So Common!*

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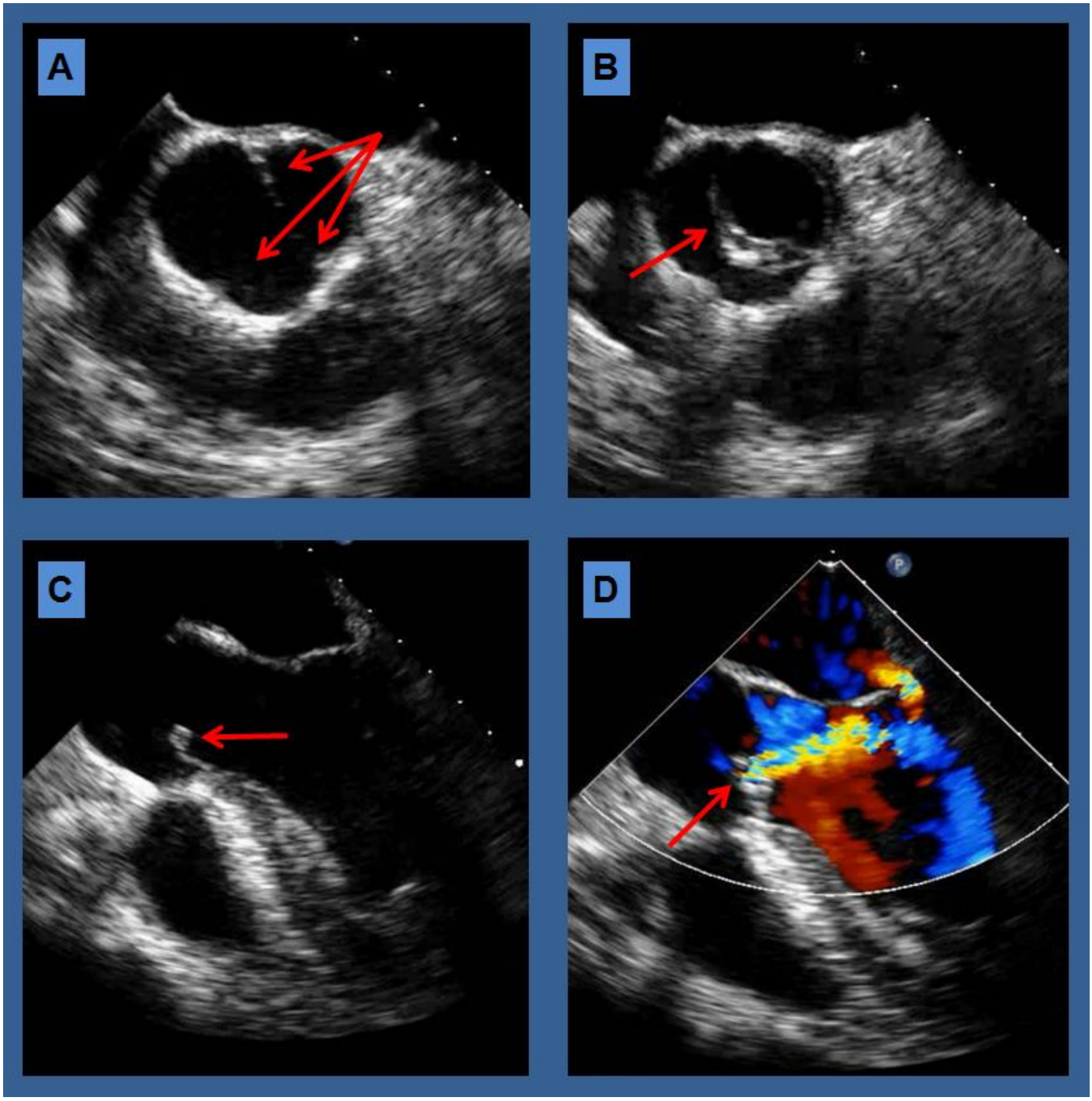

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Description

The transesophageal echocardiography images show a unicuspid aortic valve in the short axis view in closed position (A) with arrows pointing at the fused commissures; and in the open position (B) showing the circular eccentric position of the orifice (arrow). The Long axis views [C] show the valve in open position with doming of the cusps (arrow), and color flow Doppler (D) revealing at least moderate aortic insufficiency (arrow) with a trivial amount of late diastolic mitral regurgitation.

INTRODUCTION

Unicuspid aortic valve [UAV] is an extremely rare congenital anomaly which results from failure of the three aortic cusps to separate before birth [1]. It was first reported by Edwards in 1958 [7]. Its estimated annual incidence is about 0.02% in the adult population [8]. However, its incidence is noted to be higher in patients who undergo surgery for isolated aortic stenosis, up to 4-5%.[1] It is found predominantly in males with male to female ratio of 4:1.

STRUCTURAL FINDINGS

Aortic valve normally consists of 3 cusps with 3 commissures that develop from embryonic tubercules of the aortic trunk. This aberration of unicuspid valve results from failure of the cusps to separate. Unicuspid aortic valves can be further categorized into two types: Acommissural and Unicommissural. In acommissural valves, there are no commissures or lateral attachments to the aorta; the orifice is very small and appears as a pinhole on imaging [6]. These patient have symptomatic severe aortic stenosis at birth or infancy [3]. On the contrary, in unicommissural valves, there is one lateral commissural attachment to the aorta at the level of the orifice which appears as a slit shaped structure. These patients typically present in the 4th to 6th decade of life [1, 2], but can rarely present at infancy [4].

CLINICAL FEATURES

UAV has a bimodal presentation depending upon the type. Accommissural UAV leads to severe aortic stenosis which presents at birth or soon after. Unicommissural UAV has less severe course and presents in late adulthood. Patients may present with dyspnea, angina, or syncope [8]. Isolated aortic stenosis is the most common valvular abnormality associated with UAV. However, it can also be associated with other abnormalities which include aortic aneurysm [5], aortic regurgitation, aortic dissection, coarctation of the aorta, patent ductus arteriosus [2], and aortic dilation. Patients with Aortic stenosis usually become symptomatic once the transvalvular mean gradient exceeds 40 mmHg, the aortic jet velocity is greater than 4 m/s, and the valve area is less than 1 cm^2, irrespective of the commissural type.

MANAGEMENT

UAV can be diagnosed with 2D or 3D TTE or TEE [6], cardiac computed tomography [14], or cardiac magnetic resonance imaging [15]. Transesophageal echocardiography (TEE) is the gold standard for diagnosis with a sensitivity and specificity of 75 and 86% respectively [9]. The UAV has an eccentric "teardrop" opening during systole in a unicommissural UAV [8] due to absence of cusp separation.

Current guidelines from the American College of Cardiology (ACC) and the American Heart Association (AHA) recommend aortic valve replacement (AVR) for symptomatic severe aortic stenosis (Class IB recommendation). However, AVR in patient with UAV is not recommended especially in the young population, due to higher mortality rates when compared to adults. Re-operations due to patient-prosthesis mismatch and structural valve degeneration is also higher [10]. Alternatively, balloon valvuloplasty, surgical valvotomy, or commissurotomy are the initial treatments of

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choice. If AVR is needed, the Ross procedure is recommended, in which aortic valve is replaced with patient's own pulmonic valve, reducing the risk of patient-prosthesis mismatch [11, 12]. The autograft also has some capability to grow along with the patient's heart. The Ross procedure is technically more challenging, with relatively high mortality, but is considered to be safe in experienced hands [10].

CONCLUSION

In Conclusion, unicuspid aortic valve is a rare congenital malformation that often leads to severe aortic stenosis. It is an important clinical entity that should be in the differential diagnosis of younger patients who present with symptoms of heart failure and with a systolic murmur that suggests aortic stenosis. Many cases are diagnosed peri-operatively. However, with evolving imaging technology, a higher incidence of preoperative diagnosis is expected. TEE has a relatively high specificity and sensitivity for diagnosis. Aortic valve repair with balloon valvuloplasty, surgical valvotomy, or commissurotomy are the initial treatments of choice in the young population and AVR is discouraged until patient is fully grown.

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