

# Unicuspid Aortic Valve in an Elderly Male: Preoperative Transthoracic and Transesophageal Echocardiographic Appraisal

## Case Report

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### Abstract

We present a case of unicuspid aortic valve in an elderly patient who had aortic stenosis, aortic regurgitation and dilated aortic root. Transthoracic and transesophageal echocardiographic profile of this rare entity is being discussed.

**Keywords:** Unicuspid aortic valve; Unicommissural; Acommissural; Aortic stenosis; Aortic regurgitation

**Abbreviations:** TTE: Transthoracic Echo; VTI: Velocity Time Integral; TEE: Transesophageal Echocardiography

### Introduction

Unicuspid aortic valve is a rare congenital cardiac anomaly in adult population. It usually results in a combination of aortic stenosis with aortic regurgitation by 3<sup>rd</sup> to 5<sup>th</sup> decade of life. Diagnosis is commonly made on operative table or during autopsy [1].

### Case Details

The patient was a male of 63 years of age who presented at outdoor clinic with complaints of breathlessness on exertion for last 3 months. On clinical examination there was a grade 4/6 mid systolic murmur at right upper parasternal area which was radiating well to carotids. There was a long diastolic murmur at left parasternal area. Transthoracic echo revealed aortic valve stenosis with a mean gradient of 38.24 mmHg and VTI ratio of 0.29 (Figures 1 and 2). There was moderate aortic regurgitation with pan-diastolic reversal of flow in descending thoracic aorta with end-diastolic velocity of

0.25 mmHg (Figure 3, Video1). There was dilatation of aortic root at sinus level (41 mm) and that of ascending aorta (55 mm) (Figures 4 and 5). Parasternal long axis view revealed thickened and calcified aortic valve which was prolapsing in left ventricular outflow tract during diastole and two separate cusps, as normally visible in this view, were not being seen (Video 2). There was suspicion of bicuspid aortic valve on short axis view with heavily calcified edges (Video 3). Transesophageal echocardiography was performed for further evaluation. TEE revealed a unicommissural unicuspid aortic valve with a slit like opening and calcified edges (Videos 4 and 5).

### Discussion

Unicuspid aortic valve is a rare congenital cardiac anomaly in adult population. In a systemic review, only 231 adult patients with this anomaly could be identified over a period of 42 years [1]. In another single center study, the incidence was found to 0.02% in

adult population [2]. However, in a recent single center study, 42 adult cases of the entity were found over a period of 5 years [3]. It is predominantly found in male patients [2]. Age of presentation is typically 3<sup>rd</sup> to 5<sup>th</sup> decade of life [1]. However, our patient presented in seventh decade of life.

Morphologically there are two varieties of unicuspid aortic valve. It may be a commissural or unicommisural. A commissural unicuspid aortic valve has a central tiny opening (pin hole). It has no

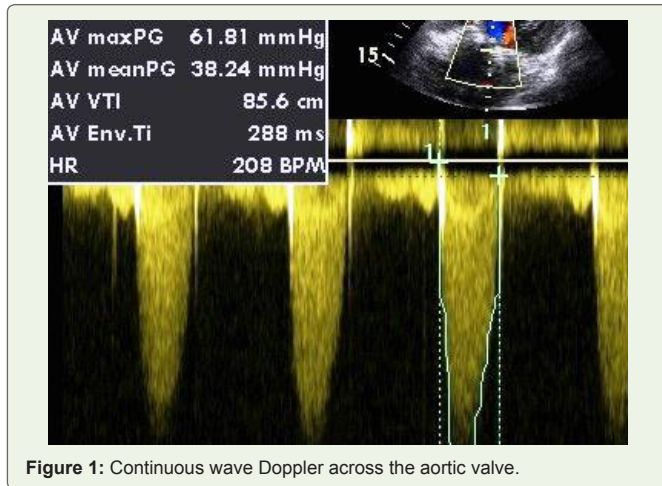


Figure 1: Continuous wave Doppler across the aortic valve.

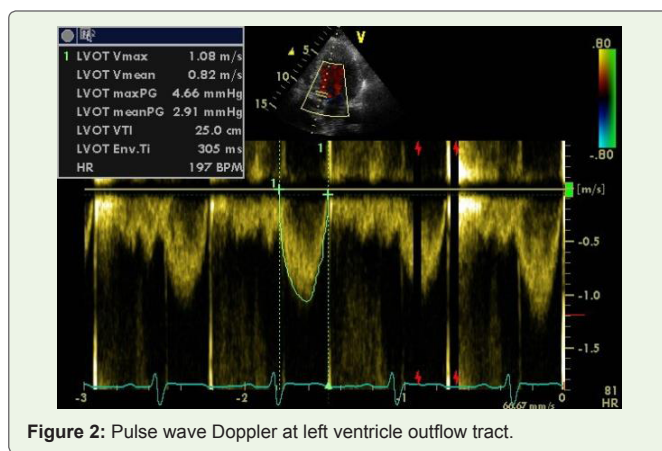


Figure 2: Pulse wave Doppler at left ventricle outflow tract.

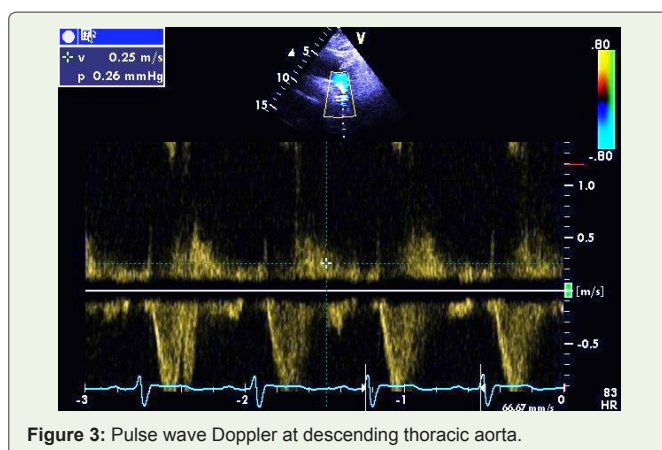


Figure 3: Pulse wave Doppler at descending thoracic aorta.

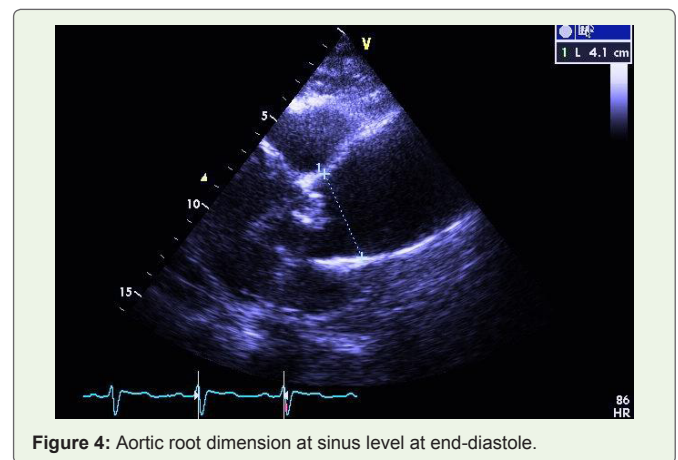


Figure 4: Aortic root dimension at sinus level at end-diastole.

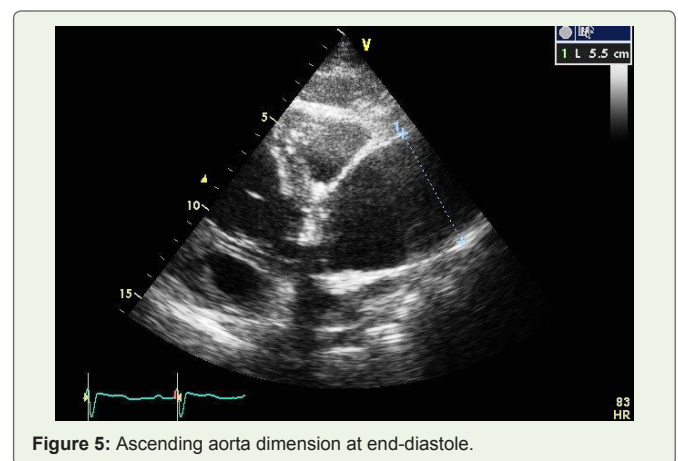


Figure 5: Ascending aorta dimension at end-diastole.

commissure or attachment to the lateral aortic wall at level of orifice [4,5]. It presents early in life typically in infancy. However, in a large surgical study, 4 out of 46 cases of unicuspid aortic valve in adult patients had a commissural valve [6]. Unicommisural unicuspid valve has one commissure and it is attached to the lateral wall of aorta at the level of orifice. It has a slit shaped opening which has been described as “teardrop” shaped orifice. It typically has a lot of calcification which makes its assessment difficult on transthoracic echo. It usually presents as a combination of aortic stenosis with aortic regurgitation. Aortic root dilatation is commonly associated [4,5].

As compared to bicuspid aortic valve disease, patients present early, have predominantly mixed stenotic and regurgitant lesions and have higher incidence of aortic annulus dilatation vis-a-vis ascending aorta dilatation [3].

Commonest presenting feature is breathlessness. Though, patients may present with syncope or angina. Pre-operative diagnosis of this entity is uncommon and as per a systematic review, pre-operative diagnosis could be established only in 40% of the cases [1].

Hence, we present pre-operative echocardiographic profile of a rare adult congenital heart disease which is difficult to diagnose preoperatively and was unusual by the virtue of older age of patient and severe dilatation of ascending aorta.

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