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# Bicuspid Aortic Valve and Ascending Aortic Aneurysm Presented With Fatigue And Gastrointestinal Symptoms: Case report

Yorgunluk ve Gastrointestinal Belirtilerle Başvuran Bicuspid Aort Kapağı ve Aort Anevrizması: Olgu Sunumu

Flora Ozkalaycı[1], Oyku Gulmez[1], Bulent Saritas[2]

1- Department of Cardiology, Baskent University Istanbul Medical and Research Center
2- Department of Cardiovascular Surgery, Baskent University Istanbul Medical and Research Center

## Öz

Bicuspid aort kapağı, doğumsal kalp hastalıkları arasında en sık görülen konjenital anomalidir. Ayrıca aortopati eşlik edebilir. Bicuspid aortik kapak sessiz kalabilir ve ekokardiyografik incelemede tesadüfen bulunabilir. Bu olgu sunumunda hastanemize gastrointestinal şikayetleri ile başvuran ve biküspid aort kapağı ve aort anevrizması tanısı ile gelen bir hastayı sunuyoruz.

**Anahtar Kelimeler:** biküspid aort, yükselen aort anevrizması, gastrointestinal şikayetler.

## Abstract

Bicuspid aortic valve is one of the most common congenital anomaly among congenital heart disease. It may also be accompanied by aortopathy. Bicuspid aortic valve may remain silent and may be found incidentally on echocardiographic examination. In this case report, we present a patient who was admited to our hospital with gastrointestinal complaints and come out with the diagnosis of bicuspid aortic valve and aortic aneurysm.

**Keywords:** bicuspid aorta, ascending aortic aneurysm, gastrointestinal complaints.

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Sorumlu Yazar: Flora Ozkalaycı
Baskent University Istanbul Medical and Research Center,
Department of Cardiology,
Istanbul, Turkey
E-mail: floratanie l@yahoo.com

## Introduction

Bicuspid aortic valve is one of the most common congenital anomaly among congenital heart disease. It effects 1% of all population.[1] The ratio between genders is 2-3:1 in favour of male population.[2] It is often sporadic, but there is increasing evidence in regards of its autosomal dominant inheritance.[3] The prevelance of bicuspid aortic valves among the first degree relatives of a subject with bicuspid valve is approximately 9%.[4,5] Therefore, echocardiographic screening is recommended among the first degree relatives of those with bicuspid aortic valve.[6]

Bicuspid aortic valve exists whether alone or accompanied by aortopathy and other congenital heart defects such as aortic coarctation, ventricular septal defect, supra aortic or subaortic stenosis.[7,8] There is also an underlying aortopathy with cystic medial degeneration.[9] Therefore, bicuspid aortic valve should be considered to be a disease of not only aortic valve, but also aorta itself. Additionally, bicuspid aortic valves and aortic aneurysms may be a component of a genetic disorder such as Turner Syndrom, Loeys-Dietz Syndrome and familial thoracic aortic aneurysm.[10,11]

Bicuspid aortic valve may remain silent and may be found incidentally on echocardiographic examination. It may cause aortic regurgitation or stenosis. Those patients without symptoms or with mild symptoms have similar life expectancy with the healthy population if managed appropriate.[13,14] Mortal complication of aortic dilatation is aortic rupture and the most important determinant of aortic rupture is the diameter and velocity of expansion of aorta concomitant bicuspid aortic valves.[7,15-17]

We describe a case of aortic aneurysm in a 36-year-old man without any known cardiovascular disease and presented with nausea which his gastroenterological findings revealed inadequate to explain the symptoms and chest imaging and echocardiographic findings revealed bicuspid aortic valve and aortic aneurysm.

**Case Report**

A 36-year old man with no known cardiac disease was admitted to the hospital with nausea for 1 month. The outpatient evaluation of the patient by the gastroenterologist for nausea was normal (abdominal ultrasonography, abdominal computed tomography, endoscopy, colonoscopy), and he was referred to our clinic as his chest X-ray showed mild mediastinal enlargement (Figure 1).

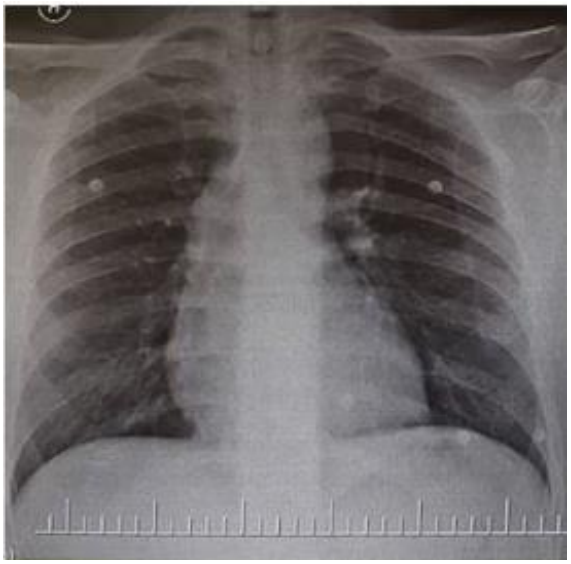

*Figure 1: Chest radiograph on admission revealing enlarged mediastinum*

His blood tests were within normal limits. His physical examination revealed heart rate of 50 beats/ min, blood pressure of 110/80mmHg, respiratory rate of 16 breaths/min, temperature of $37.2^0$C. Electrocardiography showed sinus rhythm with no ischemic changes. Transthoracic echocardiography showed normal size and function of left and right ventricles. However, ascending aorta diameter was measured as 6cm. As aortic valve structure could not be visualised clearly, transesophageal echocardiography was performed. Transesophageal echocardiography revealed bicuspid aortic valve with mild aortic regurgitation (Figure 2). Coronary computed tomography which was normal was performed in order to rule out congenital coronary heart disease. He underwent Bentall operation with the replacement of the aortic root with a graft and aortic valve with a size of number 23 (Medtronic) mechanical valve prosthesis (Figure 3). The postoperative period was uneventful with regressed nausea. He was discharged home on the $9^{th}$ day after admission. The transthoracic echocardiography at discharge revealed normal functioning ventricles and aortic valve with 3,7cm ascending aorta.

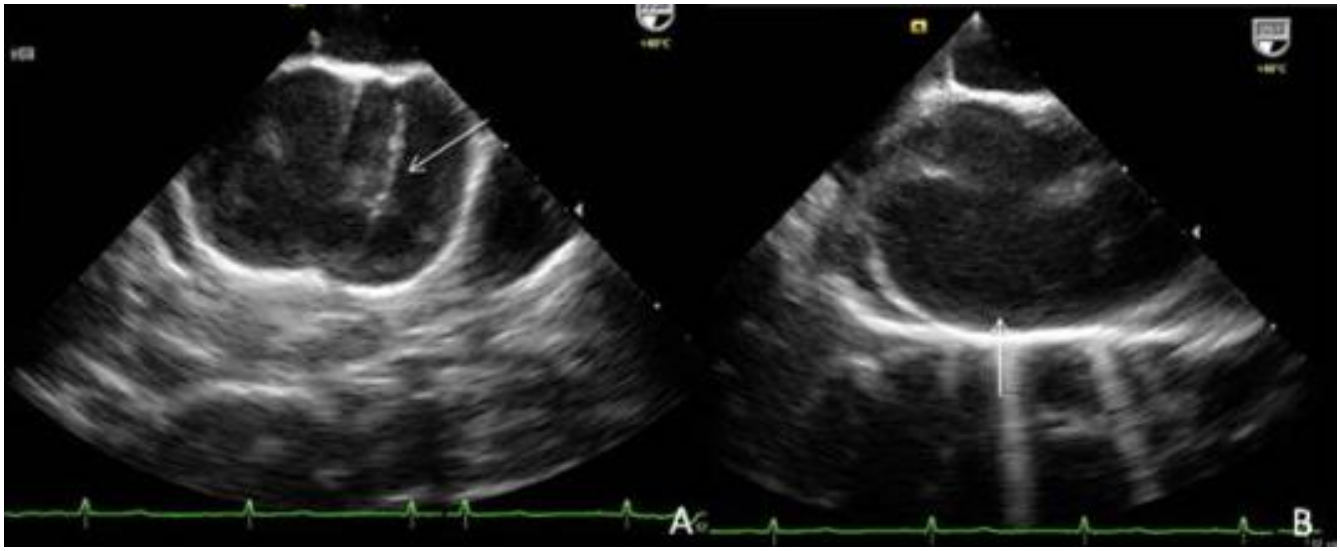


*Figure 2: TEE images of Bicuspid aortic valve (arrow) (A), Dilated aortic rooth and ascending aorta (arrow) (B)*

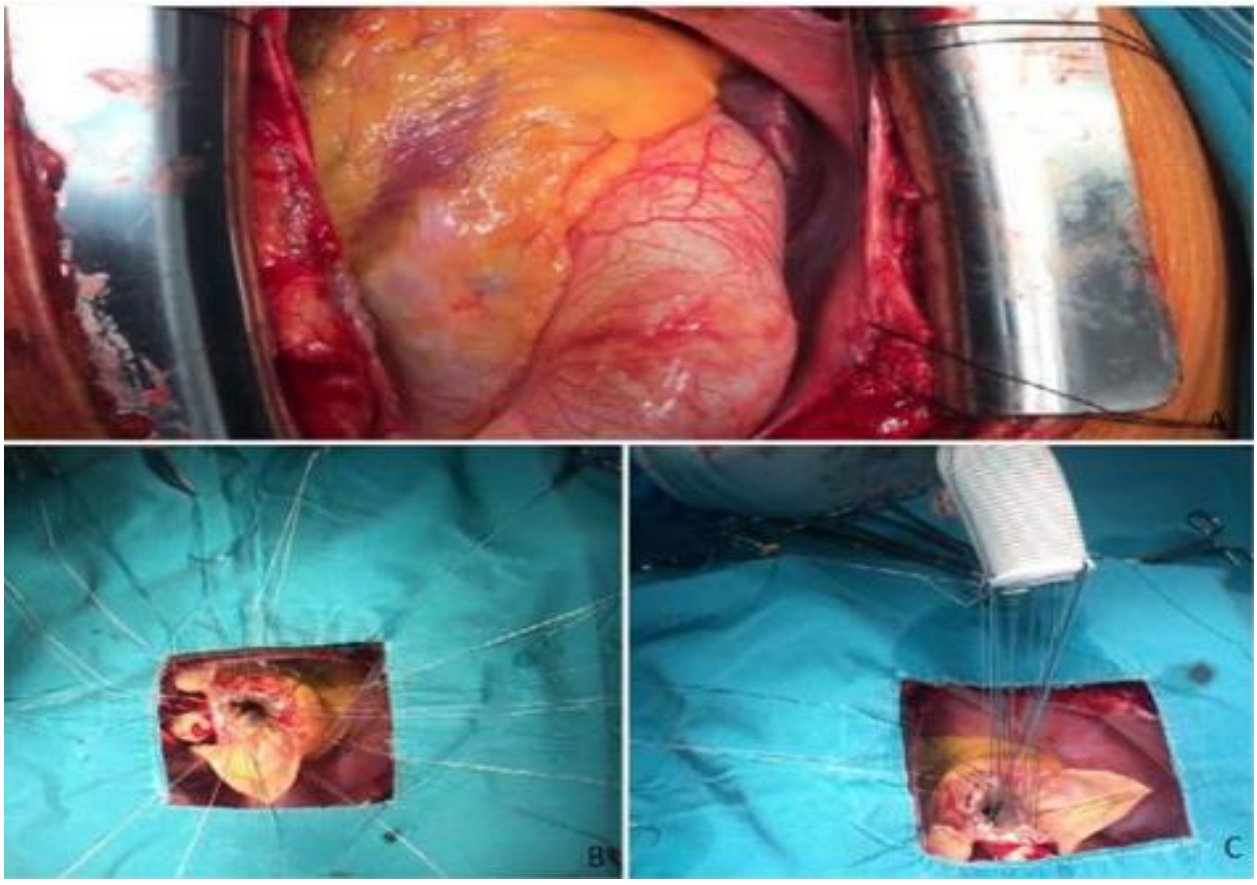


*Figure 3: Intraoperative images of the patient revealing aortic aneurysm(A) and replacement of the aorta and the bicuspid valve*

## Discussion

Patients with bicuspid aortic valves may remain asymptomatic or they may present with the complications of valvular disease and dilated ascending aorta. In this case, we report a patient with atypical symptom of nausea who had bicuspid aortic valve and mild aortic regurgitation with an ascending aorta diameter of 6 cm. His gastrointestinal complaints were not explained by gastroenterological examination but regressed after aortic surgery. This non-specific symptoms might be associated with compression of adjacent structures. Considering that nervus vagus is a component of parasympathetic nervous system vagal compression may result in decreased parasympathetic activity. Consequently, sympathetic system becomes dominant on gastrointestinal motility. Sympathetic system impulses cause symptoms such as nausea, vomiting, sweating, pallor and tachycardia. Vagal nerve compression thereby parasympathetic system repression may be the underlying cause of the nonspecific symptoms in our patient. [18-22]

It was shown that approximately one third of patients with bicuspid aortic valve may develop a complication during their lifetime.[12-14] In patients with aortic aneurysm of >6cm, 5 year rupture risk was reported as 31%, while it was 16 % and none in patients with aortic aneurysm of 4,0-5,9 and <4cm, respectively.[23] In another study, rupture risk was 25% in patients with aortic aneurysm of >5cm, and 37% in patients with aortic aneurysm of >7cm.[24]

Our patient had a aortic diameter of 6cm. According to the ESC guidelines published in 2014, for ascending aortic aneurysm with a diameter of 50 mm associated with bicuspid aortic valves and accompanying risk factors such as hypertension, coarctation of aorta, family history of dissection and pending comorbidity in elderly, surgery is indicated with class 2a indication and level of evidence C.[25] According to the ACC-AHA guidelines operation is

recommended in patients with bicuspid valves with an ascending aorta diameter of 5,5cm without other risk factors, and in patients with 5,0cm aortic dilatation if other risk factors are present with class 1 indication and level of evidence C. [6] Our patient was operated as he had an aortic diameter of 6cm. He had no complication postoperatively, and his atypical symptoms regressed.

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