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# An Abnormal Left Main Coronary Artery Origin, Which is Rare in A Young Athlete with A Bicuspid Aorta

## Genç Bir Atletde Konjenital Biküspit Aort Kapağı ile İlişkili Sol Ana Koroner Arterin Nadir Anormal Çıkışı Vakası

**CASE REPORT**
OLGU SUNUMU

**ABSTRACT**

A bicuspid aortic valve (BAV) is one of the most congenital anomalies of the heart in adults. It is also associated with a higher-than-expected incidence of coronary artery anomalies. We present a rare case of congenital BAV associated with anomalous origin of the left main coronary artery (LMCA) from the posterior left coronary sinus of Valsalva in young symptomatic athlete.

**Keywords:** Angiography, bicuspid aorta, coronary artery anomaly

**ÖZET**

Biküspit aort kapağı erişkinlerde kalbin en sık görülen konjenital anomalilerinden biridir. Aynı zamanda koroner arter anomalilerinin daha yüksek insidansı ile ilişkilidir. Semptomatik genç bir sporcuda, sol ana koroner arterin sol arka koroner sinüs Valsalva'dan anormal çıkışı ile ilişkili nadir bir konjenital biküspit aort kapağı vakasını sunuyoruz.

**Anahtar Kelimeler:** Anjiyografi, biküspit aort, koroner arter anomalisi

A bicuspid aortic valve (BAV) is one of the most common congenital anomalies of the heart in adults. Although in majority of patients it remains as an isolated defect, it is also associated with a higher-than-expected incidence of coronary artery anomalies.[1] Coronary anomalies are classified into abnormalities of origin, distribution, and association with fistulae. The anomalous aortic origin of the coronary arteries occurs in up to 0.7% of the general population and is the second most common cardiac cause of death in young athletes.[2] The origin of the left main coronary artery (LMCA) from the posterior aspect of the left coronary sinus associated with BAV is an extremely rare condition and only a few cases are presented in the literature. In this case, we report a rare association of congenital BAV with an anomalous origin of the LMCA from the posterior aspect of the left coronary sinus of Valsalva in the young athlete.

## Case Report

A 17-year-old athlete girl with congenial bicuspid aortic valve (BAV) presented with an atypical retrosternal chest pain beginning after hard physical stress. She had no risk factors and a family history of cardiovascular disease. Electrocardiography was without pathology. Trans-thoracic echocardiography revealed presence of BAV without transaortic gradient. Biventricular functions were normal without regional wall motion abnormalities. For the clarification of abnormal flow detected in parasternal short-axis view during trans-thoracic echocardiography, transesophageal echocardiography was accomplished. In transesophageal echocardiography, it was noted that the LMCA was abnormally originated from

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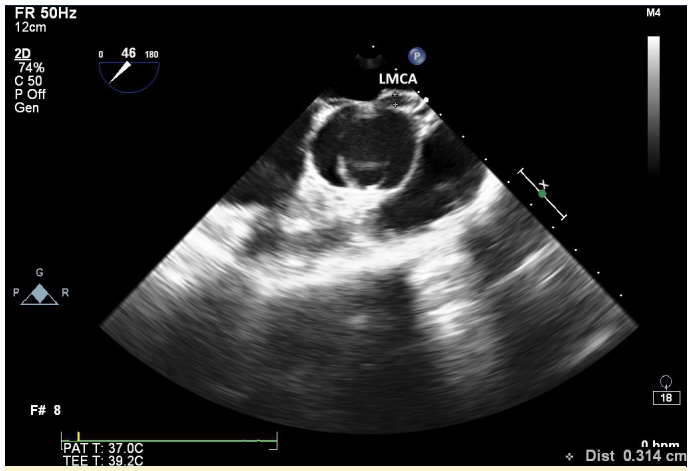


**Figure 1. Transesophageal echocardiography of the patient. LMCA: Left main coronary artery.**

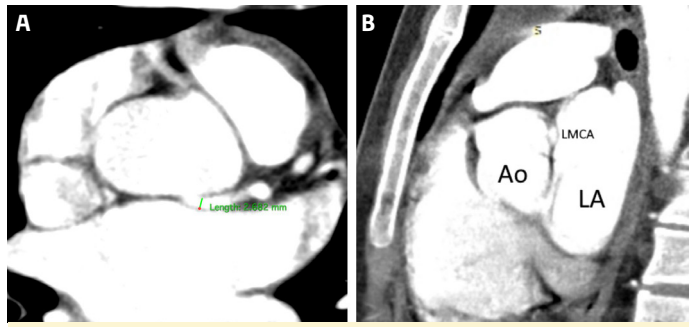


**Figure 2. Cardiac CT angiogram of the patient in axial (A) and sagittal (B) plane. LMCA diameter was measured 2.7 mm in systole. Ao: Aorta; LMCA: Left main coronary artery; LA: Left atrium.**

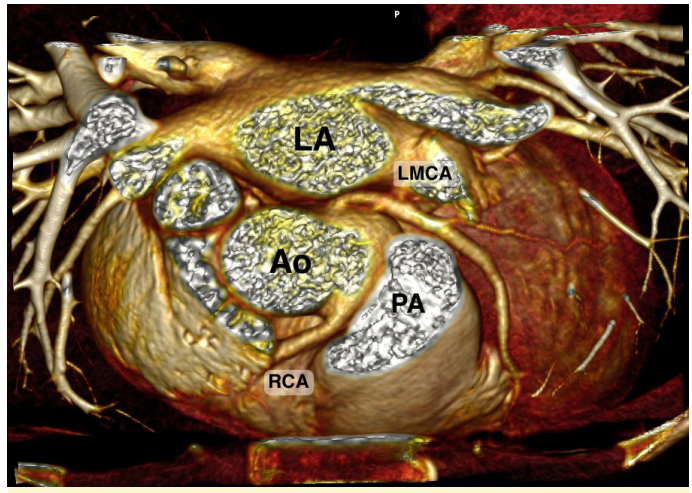


**Figure 3. 3D reconstruction of the Cardiac CT angiogram. Ao: Aorta; LMCA: Left main coronary artery; LA: Left atrium; PA: Pulmonary artery; RCA: Right coronary artery.**

the posterior aspect of the left sinus of Valsalva (Figure 1). A cardiac computed tomographic angiogram showed an anomalous origin of the LMCA from the posterior aspect of the left sinus of Valsalva between the left atrium and aorta with an acute angle takeoff (Figure 2, 3). The narrowest part of LMCA was measured 2.7 mm. Although exercise stress ECG showed no signs of myocardial ischemia, a stress SPECT\CT scan showed signs of mild hypoperfusion in the mid and apical segments of the anteroseptal wall and apex of the left ventricle. The patient was advised to retire from professional sports. Surgical intervention will be discussed if symptoms worsen during close follow-up.

## Discussion

The anomalous aortic origin of the coronary arteries is a rare condition with 0.7% prevalence in the general population. In patients with BAV, the prevalence of this anomaly is higher. Although chest pain and syncope can be the presenting symptoms, sudden cardiac death (SCD) is often the first manifestation of this anomaly. The risk assessment for SCD is complicated because of insufficient data. The reason by which anomalous coronary anomaly causes death is believed to be due to myocardial ischemia secondary to limited coronary flow. Potential mechanisms explaining myocardial ischemia are 1) compression of the vessel between the aorta and pulmonary artery and very rarely between the aorta and enlarged left atrium, 2) kinking due to an acute angle takeoff resulting in a slit-like orifice and of coronary flow, and 3) intramural course of the vessel.

In this patient, coronary arteries were visualized by Coronary CT angiography. In CT images LMCA originated from the posterior aorta, took an acute angle, and pathed between the aorta and the left atrium. The narrowest part was measured 2.7 mm at the section an acute angulation. In the current case, coronary flow limitation can be explained by the kinking and acute angle take-off of the artery resulting in a slit-like orifice. The intramural course of the artery was not detected in CT. Compression of the LMCA between the aorta and the left atrium is a very rare condition and was previously described only in a few cases.[3,4] In all these presented cases left atrium was heavily dilated with increased intra-atrial pressure which caused the compression of the vessel. However, in the current case

### ABBREVIATIONS

| | |
|---|---|
| Ao | Aorta |
| BAV | Bicuspid aortic valve |
| CT | Computed tomography |
| ECG | Electrocardiogram |
| LA | Left atrium |
| LMCA | Left main coronary artery |
| PA | Pulmonary artery |
| RCA | Right coronary artery |
| SPECT\CT | Single-photon emission computerized tomography |

left atrium was not dilated and it is less probably that intra-atrial pressure could exceed the intracoronary pressure to cause compression.

In our case, during the exercise stress test patient had no complications and ECG showed no signs of myocardial ischemia. However, on the 10th minute of the recovery period, the patient began to complain of atypical chest pain. The ECG again was eventless. To confirm the stress test pharmacological stress SPECT was performed. However, the stress SPECT\CT scan showed signs of mild hypoperfusion in mid and apical segments of the anteroseptal wall and apex of the left ventricle, one would expect much significant ischemia in the case of significant LMCA obstruction. Considering that "false-positive" SPECT\CT results are not rare, especially in young and asymptomatic patients, a close follow-up was decided.

In patients with typical angina symptoms who have evidence of stress-induced myocardial ischemia, ESC guidelines recommend a surgical coronary intervention.[5] In asymptomatic patients without evidence of myocardial ischemia, surgery is a class IIa recommendation in the case of LMCA anomaly if high-risk anatomy presents. High-risk anatomy includes high orifice, ostial stenosis, slit-like/fish-mouth-shaped orifice, acute-angle take-off, intramural course, and its length, or interarterial course and hypoplasia of the proximal coronary artery. In our patient, only acute angulation of LMCA with slit-like orifice was present based on CT images. If indicated, a surgical unroofing procedure is the treatment of choice in such anomalies. But there is limited evidence that surgery can provide any benefit in asymptomatic patients. However, it must be kept in mind that, SCD can be the first manifestation of the coronary anomaly and close follow-up of such patient by a cardiologist is mandatory. Unfortunately, there is no guideline recommendations about physical activity in patients with coronary anomaly.

## Conclusion

BAV is sometimes associated with coronary artery anomalies. In young patients who presented with chest pain, especially with BAV coronary arteries should be carefully evaluated.

**Informed Consent:** Written informed consent was obtained from the participants of this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept - U.R., E.K.; Design - E.K.; Supervision - U.R.; Materials - E.K.; Literature Search - E.K., S.M.; Writing Manuscript - E.K.; Critical Review - U.R., S.M., O.K.

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