

## Aorto-ventricular tunnel mimicking rheumatic aortic insufficiency

*Romatizmal aort yetersizliğini taklit eden aortoventriküler tünel*

Mustafa Paç,<sup>1</sup> Şevket Ballı,<sup>2</sup> Ayşenur Paç,<sup>2</sup> Mehmet Burhan Oflaz<sup>2</sup>

<sup>1</sup>Department of Cardiovascular Surgery, Türkiye Yüksek İhtisas Hospital, Ankara, Turkey;

<sup>2</sup>Department of Pediatric Cardiology, Türkiye Yüksek İhtisas Hospital, Ankara, Turkey

Aorto-ventricular tunnel is a rare congenital heart disease. The disease should be considered in infants with clinical signs of aortic regurgitation. In this article, we present a five-year-old male case of aorto-ventricular tunnel, who was misdiagnosed as aortic regurgitation due to rheumatic involvement, then diagnosed with aortoventricular tunnel. The differential diagnosis and surgical treatment of aorto-left ventricular tunnel were discussed.

**Key words:** Aortic insufficiency; aorto-ventricular tunnel; rheumatic fever.

Aorto-left ventricular tunnel (ALVT) is an abnormal congenital communication that connects the ascending aorta above the sinotubular junction to the cavity of the left or right ventricle. It should be considered in the diagnosis of infants with clinical signs of aortic regurgitation.<sup>[1,2]</sup> Occasionally, clinicians may fail to identify the defect in some cases due to the following: severe aortic valvular regurgitation in association with a ventricular septal defect (VSD), sinus of Valsalva fistula, common arterial trunk with valvular regurgitation, aorto-pulmonary window, VSD with aortic regurgitation, persistent patency of the arterial duct, coronary-cameral fistula, valvular aortic stenosis and regurgitation, or cerebral arteriovenous malformation.<sup>[1-3]</sup>

In this report, we present a case of aorto-ventricular tunnel that was misdiagnosed as aortic regurgitation attributed to cardiac involvement by rheumatic fever.

### CASE REPORT

A five-year-old boy was referred to our pediatric cardiology department with the diagnosis of severe aortic valvular regurgitation due to rheumatic fever.

Aortoventriküler tünel, nadir görülen doğuştan kalp hastalığıdır. Aort yetersizliği bulguları olan infantlarda bu tanı düşünülmelidir. Bu yazıda, romatizmal kalp hastalığına bağlı aort yetersizliği yanlış tanısıyla gelen, sonrasında aortoventriküler tünel tanısı konulan beş yaşında erkek bir olgu sunuldu. Aorto-sol ventriküler tünelin ayırıcı tanısı ve cerrahi tedavisi tartışıldı.

**Anahtar sözcükler:** Aort yetersizliği; aorto-ventriküler tünel; romatizmal ateş.

His past clinical history was unremarkable, and no clinical sign of acute rheumatic fever was present. On physical examination, he had a low-intensity systolic murmur followed by a decrescendo high-frequency diastolic murmur over his left sternal border. A 12-lead electrocardiogram showed normal sinus rhythm and left ventricular hypertrophy while chest radiography demonstrated mild cardiomegaly. Transthoracic echocardiography revealed aneurysmal dilatation above the aortic valve annulus and moderate left ventricular enlargement. In addition, color Doppler echocardiography demonstrated a significant eccentric regurgitant jet close to the right coronary cusp directed toward the anterior leaflet of the mitral valve (Figure 1). The eccentric nature of the regurgitant jet resembled that of aortic involvement in rheumatic fever in this setting because it was so close to the edge of the aortic annulus and beneath the aortic valve. After a slightly leftward and superior angulation of the transducer toward the left shoulder, we realized an abnormal turbulent flow originating from immediately above the right coronary artery (RCA) and suspected a diagnosis of ALVT (Figure 1). Cardiac catheterization was

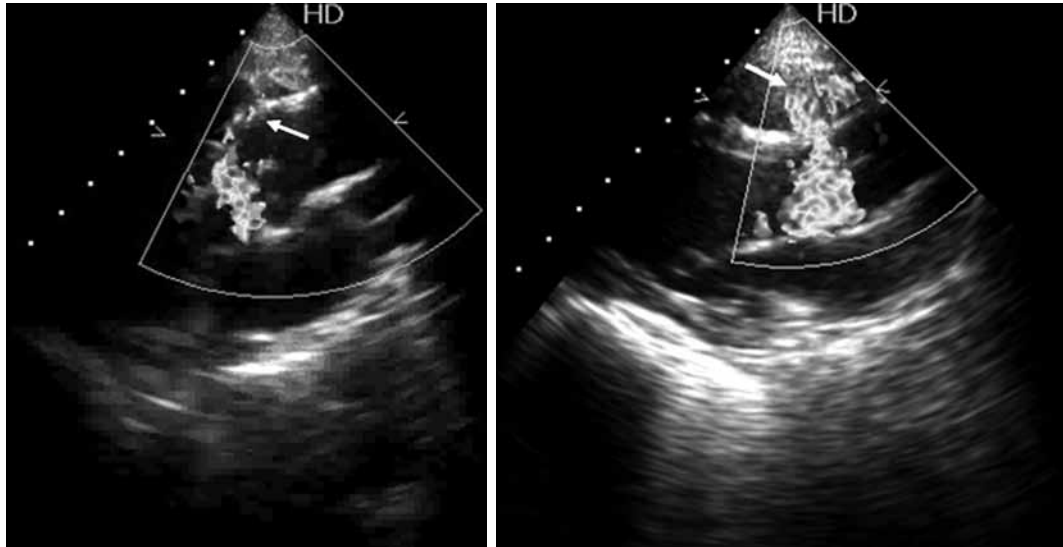


Available online at  
www.tgkdc.dergisi.org  
doi: 10.5606/tgkdc.dergisi.2013.4428  
QR (Quick Response) Code

Received: April 9, 2010 Accepted: October 4, 2010

Correspondence: Ayşenur Paç, M.D. Türkiye Yüksek İhtisas Hastanesi Çocuk Kardiyolojisi Kliniği, 06100 Sıhhiye, Ankara, Turkey.

Tel: +90 505 - 316 22 27 e-mail: aysepac@gmail.com



**Figure 1.** Significant color Doppler flow jet imaging between the right aortic coronary cusp and anterior mitral leaflet from parasternal long-axis views.

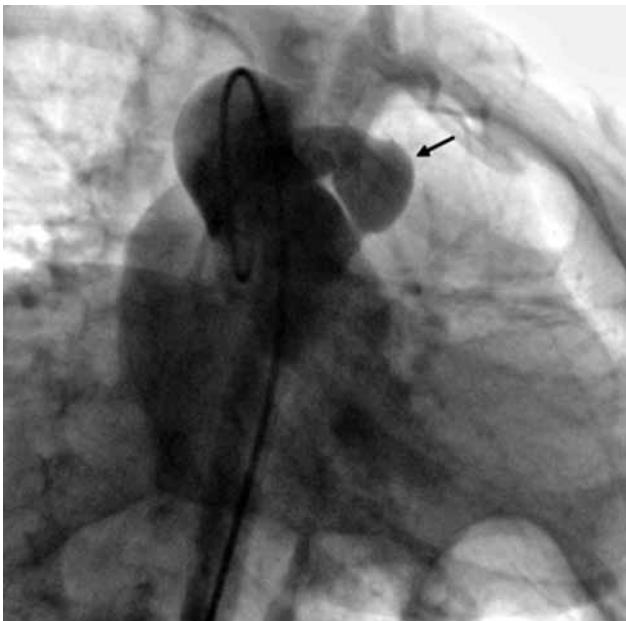
performed to confirm the diagnosis, and this showed the ALVT developing as a large extracardiac aneurysm immediately after the aortic side opening with gradual tapering at its left ventricular side opening beneath the right coronary cusp (Figure 2). Surgical inspection revealed an aneurysmal 3 cm tunnel arising from the ascending aorta just above the right coronary artery. The aortic orifice of the tunnel was situated 2 mm above the ostium of the right coronary artery, and the ventricular orifice was situated under the right-left commissure

of the aortic valve. The aortic end was closed with a single patch, and a second patch was placed through the tunnel to close the ventricular orifice. Postoperative echocardiography confirmed the absence of any residual leak, and the patient was doing well at the six-month follow-up.

## DISCUSSION

Although ALVT is a rare cardiac malformation (occurring in 0.001% of patients with congenital heart disease), it should be considered in cases with continuous systolic diastolic murmur.<sup>[1,2]</sup> Echocardiography is the best diagnostic tool for ALVT since it can reveal the eccentric nature of the regurgitant jet on color-flow Doppler echocardiography close to the right coronary cusp along with the increased left ventricular dimension. The lesion has been misdiagnosed as aortic incompetence in a few cases.

Rheumatic heart disease in our country is a common problem. The Doppler echocardiography findings of our case were interpreted as ALVT; therefore, we wanted to draw attention to this as a possible diagnosis.<sup>[2-4]</sup> Acute rheumatic fever still remains the most common cause of aortic regurgitation in developing countries and is the first suspected diagnosis in patients presenting with left heart valvular involvement.<sup>[3]</sup> The characteristics of the aortic regurgitation jet associated with rheumatic fever are frequently directed toward the anterior leaflet of the mitral valve and may be misdiagnosed as ALVT, as in our case. However, the less intense



**Figure 2.** An aortogram in anteroposterior projection reveals a large extracardiac aneurysm as indicated by the arrow.

but coexisting systolic component of murmur as well as having no previous clinical history of rheumatic fever are also important signs. Although transcatheter closure has been described in one patient with reduced left ventricular function due to coincidental noncompaction of the left ventricle, surgery is the primary mode of treatment. In most cases of ALVT, this is accomplished by transaortic patch closure of the aortic end and placement of a second patch through the tunnel itself to close the ventricular orifice and support the aortic valve.

Closure of the aortic orifice by direct suture also has sometimes produced good results, but more often than not, the tunnel recurs, or progressive aortic regurgitation through an unsupported or distorted right coronary leaflet leads to subsequent valve replacement.<sup>[5]</sup> We thought that the results of transcatheter closure might be similar to the direct suture technique during follow-up; thus, we preferred the aforementioned surgery procedure.

In conclusion, because of the eccentric nature of the regurgitant jet, ALVT can also mimic aortic involvement due to rheumatic fever. A careful clinical examination and review of a patient's clinical history is important to obtain a proper diagnosis. The repair consists of closing the tunnel so that the aortic valve is supported

by the surgical technique in order to avoid unfavorable outcomes during follow-up.

#### **Declaration of conflicting interests**

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

#### **Funding**

The authors received no financial support for the research and/or authorship of this article.

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