

# A Young Female Patient with Corrected Transposition of the Great Arteries and Ebstein-Like Anomaly

## BÜYÜK ARTERLERİN DÜZELTİLMİŞ TRANSPOZİSYONU VE EBSTEİN ANOMALİLİ GENÇ BİR BAYAN OLGU

Recep DEMİRBAĞ, MD,<sup>a</sup> Ali YILDIZ, MD,<sup>a</sup> Remzi YILMAZ MD,<sup>a</sup>  
Cüneyt SELLİ, MD,<sup>b</sup> Mustafa KÖSECİK MD,<sup>c</sup> Halit ANDAÇ, MD<sup>b</sup>

Departments of <sup>a</sup>Cardiology, <sup>b</sup>Cardiovascular Surgery, <sup>c</sup>Pediatric Cardiology, Harran University, Faculty of Medicine, ŞANLIURFA

### Abstract

An 18-years-old-female patient with corrected transposition of the great arteries and severe systemic atrioventricular valve regurgitation due to Ebstein-like anomaly was reported. She was referred to our hospital for evaluation of dyspnea on exercise. On auscultation, a grade 3/6 holosystolic murmur was detected. The chest X-ray showed cardiomegaly, and echocardiographic findings established the diagnosis of Ebstein anomaly with corrected transposition of the great arteries. She underwent left sided atrioventricular valve replacement using a bio-prosthetic valve due to severe tricuspid insufficiency due to Ebstein-like anomaly.

**Key Words:** Tricuspid valve insufficiency; arteries; ebstein's anomaly

### Özet

Ebstein anomalisi nedeniyle ciddi atriyoventriküler kapak yetersizliğinin eşlik ettiği büyük arterlerin düzeltilmiş transpozisyonuna sahip 18 yaşında genç bayan olgu sunuldu. Egzersizle gelen nefes darlığı sebebinin araştırılması için hastanemize gönderilen olgunun oskültasyonunda 3/6 pansistolik üfürüm tespit edildi. Telesinde kalp de büyüme ve ekokardiyografik bulgular büyük arter düzeltilmiş transpozisyonu ile birlikte olan Ebstein anomalisi izlendi. Ebstein anomalisi nedeniyle ciddi triküspit yetersizliği olan düzeltilmiş transpozisyonlu olguya biyoprotez kapak kullanarak kapak replasmanı yapıldı.

**Anahtar Kelimeler:** Triküspit kapak yetersizliği; arterler; ebstein anomalisi

Türkiye Klinikleri J Cardiovasc Sci 2007, 19:201-203

**C**ongenitally corrected transposition of the great arteries (CCTGA) is a rare congenital heart disease in which atrioventricular and ventriculoarterial discordance exist concomitantly.<sup>1</sup> Prevalence of this anomaly is approximately 0.5% to 1.4% of all the congenital heart diseases.<sup>2</sup> In patients with CCTGA some anomalies such as pulmonary stenosis and ventricular septal defect, or systemic AV valve (anatomical tricuspid) regurgitation and subsequent development of systemic (morphological right) ventricular dysfunction or heart block

accompany to the disease.<sup>2,3</sup> The presence of associated congenital defects, like development of total atrioventricular block or right ventricular dysfunction, limits the survival in patients with CCTGA.<sup>1</sup> The patients with CCTGA with several anomalies were reported in the literature.<sup>3</sup> However, the patient with corrected transposition of the great arteries associated with severe tricuspid insufficiency due to Ebstein-like anomaly was rarely reported.<sup>4,5</sup>

We report a case of successfully operated by valve replacement in a young female patient with Ebstein's anomaly with severe left-sided tricuspid valve incompetence accompanying CCTGA.

### Case Report

An 18-year-old female patient complaining dispnea for over 6 months which evolved into disp-

Geliş Tarihi/Received: 11.04.2007 Kabul Tarihi/Accepted: 18.05.2007

**Yazışma Adresi/Correspondence:** Recep DEMİRBAĞ, MD  
Harran University Faculty of Medicine  
Department of Cardiology, ŞANLIURFA  
rdemirbag@yahoo.com

Copyright © 2007 by Türkiye Klinikleri

nea on moderate exertion admitted to our clinic. On physical examination, the patient was non-cyanotic; her blood pressure was 110/60 mmHg, and her heart rate was 86 beats per minute. On cardiac auscultation, she had an accentuated second heart sound best heard at the upper left sternal border. A grade 3/6 holosystolic murmur was heard at the left lower sternal border. The patient's functional capacity was class III according to the classification of New York Heart Association.

The electrocardiogram showed sinus rhythm, left ventricular hypertrophy, P mitrale in leads V1, and absence of Q waves in lateral leads (I, AVL, V5-V6). The chest X-ray showed cardiomegaly (cardio thoracic index >0.60) with significant left atrial enlargement. Holter electrocardiogram showed no evidence of A-V block.

Transthoracic echocardiography revealed that the cardiac apex was displaced to the rightwards with ventricular inversion. The right atrium was connected through the mitral valve to the anatomically left ventricle, located to the right. The left atrium was connected through the tricuspid valve to the morphologically right ventricle, located to the left. The tricuspid septal leaflet was displaced 13 mm below the atrioventricular annulus (Figure 1) and posterior leaflet not seen. The left-sided (morphological right) ventricular wall hypertrophy, increased trabeculation and diffuse hypokinesis (ejection fraction=40%) were



**Figure 1.** Apical four-chamber view showing the apical displacement of the tricuspid valve (TV) and normal position of mitral valve (MV). RA= right atrium; LV= left ventricle; RV= right ventricle; LA= left atrium.



**Figure 2.** Two-dimensional echocardiographic short-axis scan demonstrates pulmonary artery (PA) and aorta (AO) relations. LA= left atrium. Coronary A= coronary artery.

detected in echocardiography. The tricuspid insufficiency was severe. The pulmonary artery emerged from the left ventricle, which was located to the right, and the aorta emerged from the right ventricle, located to the left (Figure 2). We did not detect a ventricular septal defect, patent ductus arteriosus, atrial septal defect or pulmonary stenosis.

Since the echocardiographic findings, which established the diagnosis of Ebstein anomaly with CCTGA were sufficient cardiac catheterization was not indicated. After the patient was operated for bioprosthetic tricuspid valve replacement, she had prominent clinical improvement, and was compensated during the discharge from the hospital.

## Discussion

Corrected transposition of the great arteries without associated cardiac anomalies is a rare cardiac malformation.<sup>2</sup> The most common associated defects are ventricular septal defect (80%) and pulmonary stenosis (57-70%).<sup>2</sup> The patients with CCTGA with several anomalies were reported in the literature. However, the patient with CCTGA associated with severe tricuspid insufficiency due to Ebstein-like anomaly was rarely reported.<sup>3-5</sup>

The natural history of this heart defect is not very well defined, and it depends on the associated defects and their repercussions,<sup>2</sup> as well as on the capacity of the right ventricle to preserve its func-

tion.<sup>1</sup> Survival in cases of CCTGA without associated cardiac anomaly is longer. Dysfunction of the right ventricle, from which the aorta emerges, is one of the most investigated and debated points in this heart disease. For many years, the cause of the progressive failure of right ventricle was believed to be its ejection against the systemic territory, which resulted in having to support an elevated pressure to which it was not adapted.<sup>6</sup> In this patient, systemic (morphologic right) ventricular failure was the cause of exertional dyspnea. Adult patients with CCTGA do not have a benign condition, and increasing systemic A-V valve regurgitation and complete heart block are known as deteriorating factors of systemic ventricular function.<sup>7</sup> Our patient was unlucky since she was asymptomatic only until 18 years old. This case is a rare entity because it was associated with Ebstein-like anomaly. Later, the patient evolved systolic dysfunction of the systemic ventricle and severe regurgitation of the left atrioventricular valve. Despite all these associated anomalies, the patient had not manifested significant cardiac decompensation with functional class III at the age of 18.

Although Ebstein anomaly is most common congenital cause of tricuspid regurgitation, the others causes include tricuspid valve dysplasia, right ventricular myopathy, rheumatic tricuspid valve disease. Less common causes of acquired TV abnormalities and secondary regurgitation include trauma, endocarditis, RV infarction, carcinoid disease, radiation therapy, and connective tissue disease.<sup>4</sup>

We have diagnosed our case as Ebstein anomaly since she had the following findings: a significant apical displacement of the septal tricuspid valve leaflet ( $\geq 8 \text{ mm/m}^2$ ), a redundant, elongated, anterior tricuspid valve leaflet and right ventricular thinning, enlargement, and dysfunction.<sup>8</sup> Absence of the evidence(s) for alternate reasons of tricuspid insufficiency like history of remote chest trauma, rheumatic fever, intravenous drug abuse, mitral valve involvement, or pulmonary hypertension also supports the diagnosis of Ebstein anomaly.<sup>8</sup>

Two-dimensional echocardiography is a reliable means of diagnosing transposition of the great arteries.<sup>9</sup> Images obtained by transthoracic echocardiography were clear enough to provide useful

information. Since the echocardiographic findings were sufficient, cardiac catheterization was not indicated in our patients, who established the diagnosis of CCTGA with Ebstein-like anomaly.

Surgical results are very poor in patients with impairment of the right ventricle in CCTGA; therefore, surgery must be performed as early as possible, before significant systolic dysfunction.<sup>2,6</sup> Based on this, we choose to treat the patient operatively. The patient underwent valve replacement and discharged in good clinical condition.

In summary, this patient demonstrated that the clinical course with CCTGA varies, depending on the nature and severity of the complicating cardiac anomalies, and that transthoracic echocardiography is useful in revealing associated congenital heart diseases.

## REFERENCES

1. Hornung TS, Bernard EJ, Celermajer DS, Jaeggi E, Howman-Giles RB, Chard RB, et al. Right Ventricular Dysfunction in Congenitally Corrected Transposition of the Great Arteries. *Am J Cardiol* 1999;84:1116-9.
2. Losekoot TG, Becker AE. Discordant atrioventricular connexion and congenitally corrected transposition. In: Anderson RH, Macartney FJ, Shinebourne EA, Tynan M, eds. *Paediatric Cardiology*. Edinburgh: Churchill Livingstone; 1987. p.867-83.
3. Chauvaud S, Berrebi A, d'Attellis N, Mousseaux E, Herzigou A, Carpentier A. Ebstein's anomaly: repair based on functional analysis. *Eur J Cardiothorac Surg* 2003;23:525-31.
4. Attenhofer Jost CH, Connolly HM, Edwards WD, Hayes D, Warnes CA, Danielson GK. Ebstein's anomaly-review of a multifaceted congenital cardiac condition. *Swiss Med Wkly* 2005;135:269-81.
5. Rogers JH Jr, Rao PS. Ebstein's anomaly of the left atrioventricular valve with congenitally corrected transposition of the great arteries. Diagnosis by intracavitary electrocardiography. *Chest* 1977;72:253-6.
6. Dimias AP, Moodie DS, Sterba R, Gill CC. Long-term function of the morphologic right ventricle in adult patients with corrected transposition of the great arteries. *Am Heart J* 1989;118:526-30.
7. Sasaki O, Hamada M, Hiasa G, Ogimoto A, Ohtsuka T, Suzuki M, et al. Congenitally Corrected Transposition of the Great Arteries in a 65-Year-Old Woman. *Jpn Heart J* 2001;42:645-9.
8. Ammash NM, Warnes CA, Connolly HM, Danielson GK, Seward JB. Mimics of Ebstein's anomaly. *Am Heart J* 1997;134:508-13.
9. Daskalopoulos DA, Edwards WD, Driscoll DJ, Seward JB, Tajik AJ, Hagler DJ. Correlation of two-dimensional echocardiographic and autopsy findings in complete transposition of the great arteries. *J Am Coll Cardiol* 1983;2:1151-7.