Case Report / *Olgu Sunumu*

DOI: 10.4274/haseki.3237
Med Bull Haseki 2017;55:67-9

# Permanent Junctional Reciprocating Tachycardia-induced Dilated Cardiomyopathy: A Case Report

## *Permanent Resiprokan Kavşak Taşikardisine Bağlı Dilate Kardiyomiyopati: Bir Olgu Sunumu*

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**Abstract**

We present a four-year-old girl who was admitted to our hospital with the complaints of dyspnea, tachypnea, cough, excess sweating and fatigue. Electrocardiogram (ECG) in the tachycardic girl showed inverted P waves in leads 2, 3 and aVF along with a P-R interval of 0.16 sec and an R-P interval of 0.28 sec. Transthoracic echocardiography revealed an enlarged and spherical left ventricle with diminished systolic functions. Holter ECG confirmed long R-P tachycardia with a rate of 140-160 beats/minute. She was diagnosed as having permanent junctional reciprocating tachycardia-induced dilated cardiomyopathy and successfully treated with catheter ablation and flecainide.

**Keywords:** Electrocardiography, child, catheter ablation

**Öz**

Bu olgu sunumunda hastanemize dispne, takipne, öksürük, aşırı terleme ve yorgunluk yakınması ile başvuran dört yaşında kız olgu sunduk. Taşikardik olan hastanın elektrokardiyografisinde (EKG) V2, V3 ve aVF derivasyonlarında ters P dalgaları ile birlikte 0,16 sn P-R aralığı ve 0,28 sn R-P aralığı mevcuttu. Transtorasik ekokardiyografide sistolik fonksiyonları azalmış, geniş ve sferik sol ventrikül saptandı. Holter EKG'de kalp hızı 140-160/dk olan uzun R-P intervalli taşikardi konfirme edildi. Hastaya permanent resiprokan kavşak taşikardisine bağlı dilate kardiyomiyopati tanısı kondu ve hasta kateter ablasyonu ve flekainamid tedavisi ile başarılı bir şekilde tedavi edildi.

**Anahtar Sözcükler:** Elektrokardiyografi, çocuk, kateter ablasyonu

### Introduction

Permanent junctional reciprocating tachycardia (PJRT) is a rare form of orthodromic atrioventicular (AV) re-entry tachycardia with antegrade conduction through the AV node and retrograde conduction through a concealed accessory pathway which has very slow retrograde conduction. It is characterized by deeply inverted P waves in lead 2, 3, aVF and left precordial leads (1). It may be diagnosed incidentally, but mostly presents with dilated cardiomyopathy resulting from incessant tachycardia at a rate ranging from 120 to 250 beats/minute. PJRT is usually refractory to medical treatment, thus, early recognition and treatment with non-pharmacological methods, such as surgery or catheter ablation of the disease is essential (2). Herein, we present a four-year-old girl with PJRT-related dilated cardiomyopathy who was successfully treated with catheter ablation and flecainide.

### Case

A 4-year-old girl was admitted to our hospital with the complaints of dyspnea, tachypnea, cough, excess sweating, and fatigue. She had been evaluated at another hospital where the chest X-ray revealed cardiomegaly. On admission to our hospital, physical examination revealed normal blood pressure (100/60 mmHg), tachycardia (180 beats/minute), tachypnea (50 breaths/minute), crackles at the basal areas of the lungs, hepatomegaly and a mild 1-2/6 systolic murmur. The electrocardiogram (ECG) showed inverted P waves in leads 2, 3 and aVF along with a P-R interval of 0.16 sec and an R-P interval of 0.28 sec (Figure 1). Transthoracic echocardiography revealed

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**Received/Geliş Tarihi:** 14 April 2016 **Accepted/Kabul Tarihi:** 06 June 2016



The Turkish abstract was presented as a poster in "14th National Congress of Pediatric Cardiology and Cardiac Surgery" at 15-18 April 2015 in Denizli, Turkey.

an enlarged and spherical left ventricle with diminished systolic functions (ejection fraction: 27%, fractioned shortening: 13%) (Figures 2A and 2B). Biochemical and hematological parameters were within the normal limits. Viral markers were negative. Laboratory tests for inborn errors of metabolism were all negative. Cardiac magnetic resonance imaging was performed to detect possible myocarditis and it was normal. Digoxin, angiotensin-converting enzyme inhibitor and furosemide were started. Holter ECG confirmed long R-P tachycardia with a rate of 140-160 beats/minute. We used amiodarone and propafenone to achieve the sinus rhythm but we failed. Finally, the patient underwent radiofrequency ablation. The accessory pathway was interrupted successfully. No complication developed during the procedure. After two weeks of the procedure, Holter ECG was again positive for PJRT with a rate of 150 beats/minute. Before the second attempt for ablation, oral flecainide was started. After one week of flecainide, the ECG showed a sinus rhythm with a rate of 90-100 beats/minute and Holter was also normal. Additionally, one month after the flecainide treatment, transthoracic echocardiology demonstrated progressive improvement of the ventricular functions (ejection fraction was 57% and fractional shortening was 29%).

## Discussion

Tachycardia-induced dilated cardiomyopathy is a relatively rare but treatable type of cardiomyopathy. There are various tachyarrhythmias associated with tachycardia-induced cardiomyopathy, such as atrial fibrillation, atrial flutter, ectopic atrial tachycardia, incessant atrioventricular reciprocating tachycardia/PJRT, and ventricular tachycardia (3). Restoring the sinus rhythm or to slow the ventricular rate may result in an improvement in left ventricular functions.

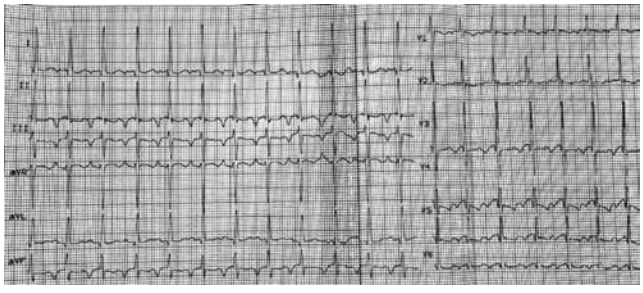

**Figure 1.** The electrocardiogram of the patient

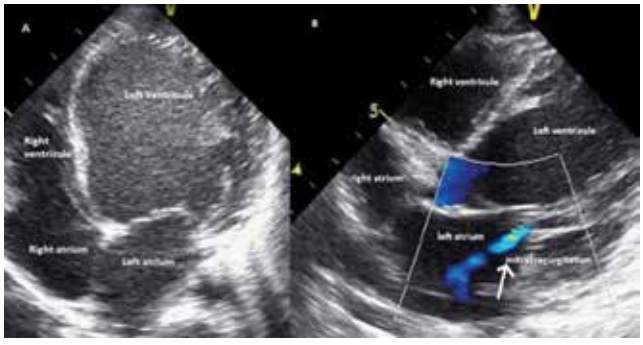

**Figure 2.** A and B; transthoracic echocardiography images of the patient

Although the animal models have showed that rapid pacing produces marked depression of left ventricular ejection fraction, depressed cardiac output and increased systemic vascular resistance, the exact mechanism of tachycardia-induced cardiomyopathy is unclear (4). Depletion of myocardial energy stores and myocardial ischemia, abnormal calcium handling and beta adrenergic responsiveness and increased oxidative stress, and injury are the most studied mechanisms (5-8).

The manifestations of PJRT usually occur in childhood and may not be recognized until adult ages. The most important complication of PJRT is the development of tachycardia-induced cardiomyopathy (9). During PJRT-induced tachycardia the cardiac stimuli conduct antegrade through the AV node and return retrograde through a slowly conducting accessory pathway that is usually located near the ostium of the coronary sinus. It is characterized by deeply inverted P waves in lead 2, 3 and aVF. Our patient had the clinical signs and symptoms of congestive heart failure due to tachycardia-induced cardiomyopathy with specific ECG characteristics.

PJRT is usually resistant to medical treatment. Most of the patients with PJRT undergo various antiarrhythmic regimens to prevent tachycardia-induced cardiomyopathy. Most of the patients finally require catheter ablation procedures. Although there is a disagreement regarding the most useful therapeutic regimen, most of the studies support catheter ablation (9). Because the heart rate of infants and children with PJRT decreases with age, some reports suggest to postpone catheter ablation i older ages and to try medical regimes before ablation (10). Due to the resistance to medical regimen, we preferred ablation in our patient but after unsuccessful ablation, sinus rhythm was achieved with flecainide.

In conclusion, children with dilated cardiomyopathy should undergo a comprehensive evaluation of tachyarrhythmia such as PJRT. Medical regimens should be tried before ablation in small children and infants. If PJRT is considered refractory to medical regimens in the presence of ventricular dysfunction, catheter ablation should be preferred. Recognition and appropriate treatment of PJRT is important to prevent or reduce the complications such as tachycardia-induced cardiomyopathy.

### Ethics

Peer-review: Externally peer-reviewed.

## Authorship Contributions

Concept: Fatih Battal, Şule Yıldırım, Hakan Aylanç, Fatih Köksal Binnetoğlu, Nazan Kaymaz, Celal Akdeniz. Design: Fatih Battal, Şule Yıldırım, Hakan Aylanç, Fatih Köksal Binnetoğlu, Nazan Kaymaz, Celal Akdeniz. Data Collection or Processing: Fatih Battal, Şule Yıldırım, Hakan Aylanç, Fatih Köksal Binnetoğlu, Nazan Kaymaz, Celal Akdeniz. Analysis or Interpretation: Fatih Battal, Şule Yıldırım, Hakan Aylanç, Fatih Köksal Binnetoğlu, Nazan Kaymaz, Celal Akdeniz. Literature Search: Fatih Battal, Şule Yıldırım, Hakan Aylanç, Fatih Köksal Binnetoğlu, Nazan Kaymaz, Celal Akdeniz. Writing: Fatih Battal, Şule Yıldırım, Hakan Aylanç, Fatih Köksal Binnetoğlu, Nazan Kaymaz, Celal Akdeniz.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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