# A diagnostic dilemma: early repolarization syndrome associated with ventricular fibrillation

## Tanısal ikilem; ventrikül fibrilasyonuyla ilişkili erken repolarizasyon sendromu

**Yavuzer Koza, M.D., Zakir Lazoğlu, M.D., Kamuran Kalkan, M.D., Serdar Sevimli, M.D.**

Department of Cardiology, Ataturk University Faculty of Medicine, Erzurum

***Summary–*** An early repolarization (ER) pattern, characterized by J-point elevation, slurring of the terminal part of the QRS and ST-segment elevation, is a common finding on the 12-lead electrocardiogram. It has been suggested that J-point elevation, which was considered benign for many years, may play a critical role in the pathogenesis of idiopathic ventricular fibrillation (VF). Recent studies have shown that an ER pattern in inferior leads or inferolateral leads is associated with increased risk for life-threatening arrhythmias. We report the case of a 52-year-old man with no structural heart disease whose electrocardiogram showed type 2 ER pattern (with evidence of J-point and ST-segment elevation in electrocardiogram leads II, III, and aVF). The patient presented with VF.

***Özet–*** Erken repolarizasyon paterni olarak da isimlendirilen, QRS dalgasının inen kolunda çentiklenme ve ST segment yükselmesi ile karakterize olan J nokta yükselmesi 12 derivasyonlu EKG'de sıklıkla görülen bir bulgudur. Uzun yıllar boyunca iyi huylu olarak yorumlanan J noktası yükselmesinin, idiyopatik ventrikül fibrilasyonu (VF) patogenezinde kritik rolü olabileceği öne sürülmüştür. Son zamanlarda yapılan çalışmalarda, inferiyor ve inferolateral derivasyonlardaki erken repolarizasyon paterninin hayatı tehdit edici aritmilerle ilişkili olduğu gösterildi. Bu yazıda, yüzey elektrokardiyografisinde Tip 2 erken repolarizasyon paterni bulunan, yapısal kalp hastalığı olmayan, takibinde VF gelişen 52 yaşında bir erkek hasta sunuldu.

The early repolarization (ER) pattern is defined as a notch or a slurring of the QRS-ST junction resulting in a so-called "J wave", with an elevation of the J point in the inferior leads or/and the lateral leads above the isoelectric line.[1] This pattern has been recognized for several decades and was accepted as a variant of normal electrocardiogram (ECG), as it was frequently observed in healthy subjects or athletes.[2] However, in recent years, it has been suggested that the ER pattern may not be a benign finding. An association between the ER pattern and idiopathic ventricular fibrillation (VF) has been reported.[3-5] Furthermore, a higher prevalence of the ER pattern has been reported in subjects resuscitated from cardiac arrest due to idiopathic VF. Some population-based studies have shown that the ER pattern is associated with an increased cardiovascular and total mortality in healthy young adults.

## CASE REPORT

A 52-year-old man with no previous history of cardiac disease was admitted to our emergency service with complaints of dyspnea and retrosternal chest pain. His blood pressure was 120/70 mmHg, heart rate 67 beats/minute and body temperature 36.4°C. He lost consciousness suddenly during the examination. An immediate ECG revealed VF. He was defibrillated and was admitted to our intensive care

**Abbreviations:**

*ECG* *Electrocardiogram*
*ER* *Early repolarization*
*ICD* *Implantable cardioverter defibrillator*
*VF* *Ventricular fibrillation*

*Received:* March 15, 2013 *Accepted:* June 27, 2013
*Correspondence:* Dr. Yavuzer Koza. Osman Gazi Mahallesi, 230. Sokak, Taner Apt., B-Blok, Kat: 2, No: 4, Palandöken, Erzurum.
Tel: +90 442 - 231 85 21 e-mail: yavuzerkoza@hotmail.com

unit, where several VF episodes were observed. He had no family history of sudden cardiac death. The ECG showed normal sinus rhythm with normal PR and corrected QT intervals. There was a J-point elevation with slurring of the end point of QRS in the inferior leads (Fig. 1). Cardiac enzyme levels, electrolyte levels, and thyroid function tests were found to be within normal limits. Hematological and serological tests and noninvasive cardiac tests, including echocardiogram, cardiac computed tomography, and cardiac thallium scintigraphy, were normal. Coronary angiography showed normal coronary vessels and left ventricular wall motion (Fig. 2a, b). The patient underwent implantation of a dual-chamber implantable cardioverter defibrillator (ICD) (Medtronic Inc., Minneapolis, MN, USA) and was discharged from the hospital on the seventh day.

## DISCUSSION

An ER pattern, which is defined as slurring or notching of the terminal part of the QRS complex on the ECG, was first described in 1936.[6] In patients with documented idiopathic VF and a structurally normal heart, the prevalence of the ER pattern is 31%.[1] Prevalence rates up to 60% have been reported in smaller studies.[7] In most of the studies, elevation of the J point and/or ST segment from the baseline by at least 0.1 mV was considered definitive of an ER pattern.[5,7,8] According to 12-lead ECG, it is classified into three subtypes. Type 1, displaying an ER pattern predominantly in the lateral precordial leads, is prevalent among healthy male athletes and rarely seen in VF survivors; type 2, displaying an ER pattern predominantly in the inferior or inferolateral leads, is associated with a higher level of risk; and type 3, displaying an ER pattern globally in the inferior, lateral and right precordial leads, is associated with the highest level of risk for development of malignant arrhythmias and is often associated with VF storm.[9]

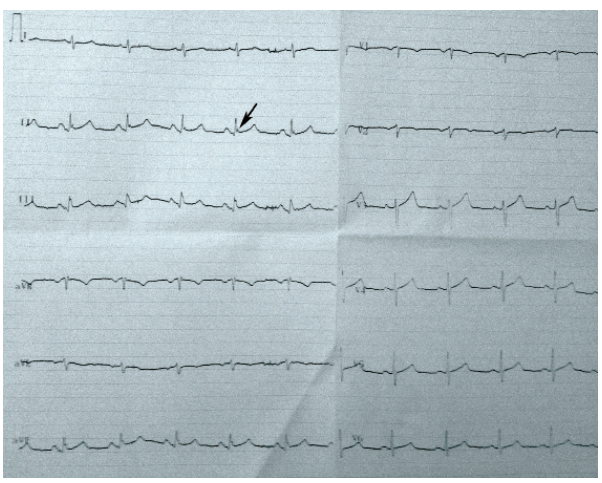

**Figure 1.** Early repolarization pattern. Note the notch with ST-segment elevation in leads II–III and VF (arrow).

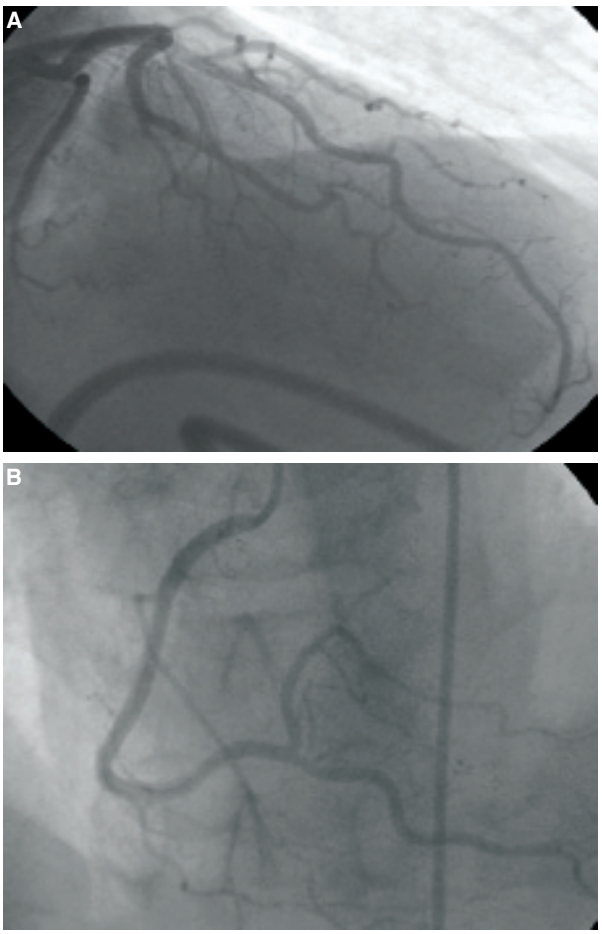


**Figure 2.** Coronary angiography shows normal coronary arteries. **(A)** Left anterior descending and circumflex artery in the right anterior oblique cranial view. **(B)** Right coronary artery in the anterior-posterior cranial view.

During the past decade, a number of clinical reports (mostly from Japan) have described patients who had sudden cardiac arrest and abnormal J waves. The ER was reported as the only "abnormal" finding in patients diagnosed with idiopathic VF.[4,5,8-10] Meanwhile, the potential arrhythmogenicity of the ER pattern was also demonstrated in experimental studies.[10] There are several reports suggesting that the ER pattern is associated with an increased total and cardiovascular mortality.

The ER pattern is common in the young healthy population. Its prevalence varies from less than 1% to 13%, depending on age (predominant in young adults), race (highest among black populations), sex (predominant in males), and the criterion for J-point elevation (0.05 mV vs 0.01 mV).[1,2,8] However, the association of the ER pattern with VF is very rare (1/10,000).[11] ST-segment elevation in inferior leads has been proven to be caused by a G752R mutation in the SCN5A gene. Therefore, it has been speculated that these specific features might be a variant of Brugada syndrome.[12] In asymptomatic subjects with no familial history of sudden death, the prognostic significance of the ER pattern anomaly is still unclear. In our patient, there was no history of unexplained syncope or familial history of sudden death. Inferior J-point elevation may signify peri-infarction block that would indicate latent ischemic heart disease.[5,9] Therefore, this ECG sign should prompt clinicians to consider latent ischemic heart disease before attributing the J-point elevation to a primary electrical abnormality. In our patient, coronary angiography and non-invasive cardiac tests were normal.

Patients with the ER pattern who are experiencing VF are candidates for secondary prevention with an ICD. However, VF storms frequently recur in patients with the ER pattern. Haïssaguerre et al.[1] reported that VF survivors presenting the ER pattern showed a higher probability of VF recurrence than VF survivors without ER pattern (43% *vs.* 23%, p<0.001) during five years of follow-up. In a multicenter study including 122 patients with the ER pattern and VF, the authors examined the effect of drug therapy on VF storm. They reported that VF storm was unresponsive to β-blockers, lidocaine, mexiletine, and verapamil, whereas amiodarone was only partially effective. On the contrary, intravenous isoproterenol or deep sedation immediately suppressed VF storm. Quinidine decreased recurrent VF from an average of 33 episodes to none, over more than two years of follow-up. In addition, quinidine restored a normal ECG.[13]

In conclusion, we present a patient with the ER pattern who had syncope due to VF. Idiopathic VF in asymptomatic ER patients is a very rare clinical entity. However, in patients who present with VF with structurally normal hearts, this entity should be kept in mind.

***Conflict-of-interest issues regarding the authorship or article: None declared.***

## REFERENCES


1. Haïssaguerre M, Derval N, Sacher F, Jesel L, Deisenhofer I, de Roy L, et al. Sudden cardiac arrest associated with early repolarization. N Engl J Med 2008;358:2016-23. CrossRef
2. Littmann D. Persistence of the juvenile pattern in the precordial leads of healthy adult Negroes, with report of electrocardiographic survey on three hundred Negro and two hundred white subjects. Am Heart J 1946;32:370-82. CrossRef
3. Aizawa Y, Tamura M, Chinushi M, Naitoh N, Uchiyama H, Kusano Y, et al. Idiopathic ventricular fibrillation and bradycardia-dependent intraventricular block. Am Heart J 1993;126:1473-4. CrossRef
4. Garg A, Finneran W, Feld GK. Familial sudden cardiac death associated with a terminal QRS abnormality on surface 12-lead electrocardiogram in the index case. J Cardiovasc Electrophysiol 1998;9:642-7. CrossRef
5. Takagi M, Aihara N, Takaki H, Taguchi A, Shimizu W, Kurita T, et al. Clinical characteristics of patients with spontaneous or inducible ventricular fibrillation without apparent heart disease presenting with J wave and ST segment elevation in inferior leads. J Cardiovasc Electrophysiol 2000;11:844-8.
6. Shipley RA, Hallaran WR. The four lead electrocardiogram in 200 normal men and women. Am Heart J 1936;11:325-45.
7. Nam GB, Kim YH, Antzelevitch C. Augmentation of J waves and electrical storms in patients with early repolarization. N Engl J Med 2008;358:2078-9. CrossRef
8. Tikkanen JT, Anttonen O, Junttila MJ, Aro AL, Kerola T, Rissanen HA, et al. Long-term outcome associated with early repolarization on electrocardiography. N Engl J Med 2009;361:2529-37. CrossRef
9. Antzelevitch C, Yan GX. J wave syndromes. Heart Rhythm 2010;7:549-58. CrossRef
10. Otto CM, Tauxe RV, Cobb LA, Greene HL, Gross BW, Werner JA, et al. Ventricular fibrillation causes sudden death in Southeast Asian immigrants. Ann Intern Med 1984;101:45-7.
11. Rosso R, Kogan E, Belhassen B, Rozovski U, Scheinman MM, Zeltser D, et al. J-point elevation in survivors of primary ventricular fibrillation and matched control subjects: incidence and clinical significance. J Am Coll Cardiol 2008;52:1231-8.
12. Kalla H, Yan GX, Marinchak R. Ventricular fibrillation in a patient with prominent J (Osborn) waves and ST segment elevation in the inferior electrocardiographic leads: a Brugada syndrome variant? J Cardiovasc Electrophysiol 2000;11:95-8.
13. Haïssaguerre M, Sacher F, Nogami A, Komiya N, Bernard A, Probst V, et al. Characteristics of recurrent ventricular fibrillation associated with inferolateral early repolarization role of drug therapy. J Am Coll Cardiol 2009;53:612-9. CrossRef


*Key words:* Arrhythmias, cardiac/epidemiology; cardiac electrophysiology; electrocardiography; ventricular fibrillation/therapy.

*Anahtar sözcükler:* Aritmi, kardiyak/epidemiyoloji; kardiyak elektrofizyoloji; elektrokardiyografi; ventrikül fibrilasyonu/tedavi.