Early Repolarization: Innocent or Dangerous?

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Abstract: Electrocardiographic (ECG) findings of wide QRS complexes in right precordial leads with saddle ST elevation in patients with polyarthritis, palpitations and family history of syncope urged us to review early repolarization syndrome (ERS). ERS is commonly seen in young men. The main ECG features are as follows: wide spread concave ST-segment elevation, more in the precordial leads (usually V2–V4); notching or irregular contour of J point and prominent concordant T waves with large amplitude. ERS was historically considered as a benign ECG variant. In recent years, it has emerged as a marker for increased risk of sudden cardiac death. The purpose of this review was to describe the ECG manifestations of this syndrome and to review the literature concerning its arrhythmogenic potential. The authors found it important not only discussing the rate of ERS life threatening but also to reemphasize its differences from other syndromes. Some of the last are much more dangerous: acute myocardial infarction and Brugada syndrome. The tables will be helpful for physicians to distinguish ERS from other syndromes in patients with chest pain and ST elevation.

Key Indexing Terms: Early repolarization; Sudden death; Brugada syndrome; J wave; ST-segment elevation. **[Am J Med Sci 2013;346(3): 226–232.]**

Electrocardiographic (ECG) findings of wide QRS complexes in right precordial leads with saddle ST elevation in patients with polyarthritis, palpitations, personal and family history of syncope urged us to review early repolarization syndrome (ERS).

ERS was historically considered as a benign ECG variant. In the recent years, it has emerged as a marker for increased risk of sudden cardiac death (SCD).[1–7]

Early repolarization (ER) was first described in 1936 by Shipley and Hallaran. They analyzed the ECG of 200 normal persons, aged 20 to 35 years, and noted that a normal variant of ST elevation occurred quite frequently. They observed this in lead II in 25% of the men and 16% of the women.[8] With the advent of the 12-lead ECG, Myers et al[9] pointed out its location in the precordial leads. Grant[10] studied the clinical characteristics of the ST-T vectors and coined the term "early repolarization" for this ECG variant. Tomaszewski[11] described elevated J point in an accidentally frozen human. The point was also known as an Osborn wave after being highlighted by Osborn in hypothermic dogs.[12] ERS is seen in both extracardiac and cardiac disorders: brain injury, hypervagotonia, hypercalcemia, spinal cord injury and vasospastic angina.

Up to 48% of the patients come to emergency department with chest pain, and ECG is performed as the first instrumental investigation.[13] The ECG syndrome of ER is found in 1% to 5% of the population.[2] It is common in young individuals (27.5%) and shows preponderance to men (77%).[14] Sixty thousand ECGs were analyzed for 5 years. Six hundred (1%) revealed ER. Features of ER were compared with race-, age- and sex-matched controls. They found that among patients with ER, 93.5% were Caucasian, 77% were men, 78.3% were younger than 50 years and only 3.5% were older than 70 years.[15] The ERS is often observed in athletes, cocaine users, patients with hypertrophic obstructive cardiomyopathy and in patients with defects and/or hypertrophy of the interventricular septum.[16]

ERS is one of several syndromes producing ECG ST-segment elevation (STE). For decades, the clinical significance of this syndrome has been largely limited to its contribution to the differential diagnosis of ST-elevation myocardial infarction (STEMI), pericarditis and of the highly arrhythmogenic Brugada syndrome (BrS). There are more and more studies suggesting that ERS may be related to electrophysiological instability and even may be associated with vulnerability to ventricular fibrillation (VF).[1–7]

Here, we describe the ECG manifestations of this syndrome. We review the latest literature to understand whether ERS is benign ECG changes or is it really one of the causes of VF and sudden death. We also reemphasize its differences from other common syndromes, some of which are life threatening such as acute myocardial infarction (AMI) and the BrS.

ERS DEFINITION

A common ECG variant characterized by J-point elevation manifested either as terminal QRS slurring (the transition from the QRS segment to the ST segment) or notching (a positive deflection inscribed on terminal QRS complex) associated with concave upward STE and prominent T waves in at least 2 contiguous leads.[17]

ERS ECG SIGNS

ST-Segment Elevation

The ST-segment elevation that begins at the "J point" (the junction between the end of the QRS complex and the beginning of the ST segment). The degree of STE encountered in ERS is usually less than 2 mm (80%–90%) but may approach 5 mm in certain individuals. Only 2% of the cases of ERS manifest STE greater than 5 mm.[13]

ST-Segment Morphology

It appears as if it has been lifted upward evenly from the isoelectric baseline at the J point, preserving the normal concavity of the initial upsloping portion of the ST segment-T wave complex. This STE morphology distinguishes ERS-related STE from that associated with STEMI, which is usually a convex STE (Figure 1).

ST-Segment Distribution

The degree of STE in ERS is usually greatest in leads V2–V5 and less often found in the limb leads. Isolated ERS in the limb leads is a very rare finding. Such isolated STE in the inferior or lateral leads should prompt consideration of another explanation for STE.[13]

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Submitted July 9, 2012; accepted in revised form October 2, 2012.

The authors have no financial or other conflicts of interest to disclose.

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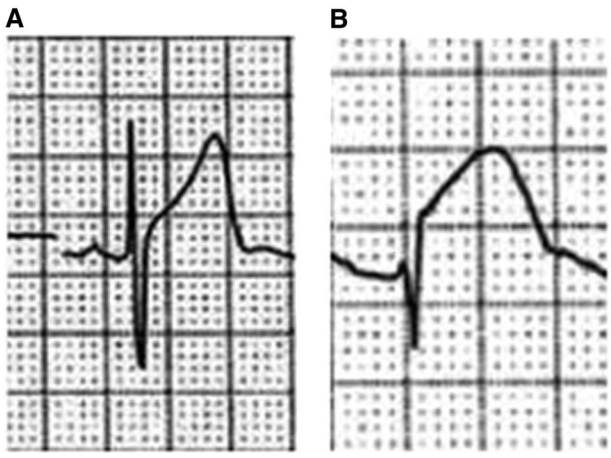

FIGURE 1. (A) Nonacute myocardial infarction cause of ST-segment elevation because of benign early repolarization. (B) ST-elevation myocardial infarction with nonconcave ST-segment morphology.

J-Point Features

There are 2 main types of J waves. The first is a distinct well-defined notch on the down stroke of the QRS complex. The second is slurred down stroke of R wave when the R wave was gradually transformed to the ST segment with upright concavity (Figure 2). An analysis of the ECG of 65 patients with the ERS revealed more than 1 pattern of the J point in the same individual.[18]

QRS Features

1. Rapid QRS transition in the precordial leads with counter clockwise rotation.
2. "Pseudo-R" pattern in lead V1 or V2 that is similar to the pattern of incomplete/complete right bundle branch block (RBBB) but without right axis deviation of the QRS vector (that is seen in RBBB).

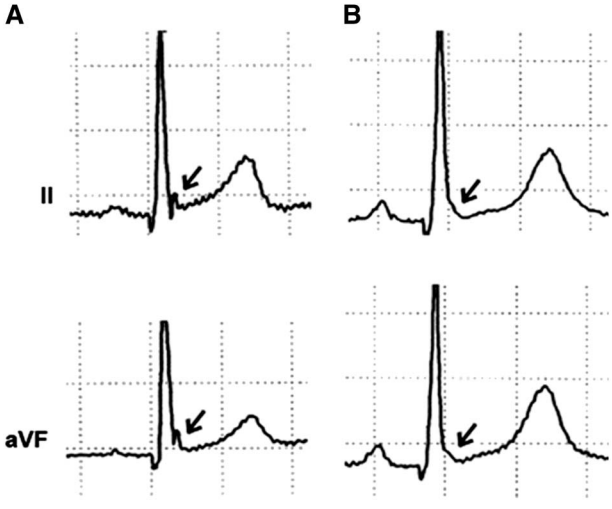

FIGURE 2. Patterns of the J point: (A) a distinct well-defined notch on the down stroke of R wave (B) when the R wave was gradually transformed to the ST segment with upright concavity.

T-Wave Features

Prominent T waves of large amplitude and slightly asymmetric morphology are encountered. The T wave may appear "peaked," suggestive of hyperacute T wave encountered in AMI. The prominent T waves are concordant with the QRS complex and are usually found in precordial leads. The height of the T waves in ERS ranges from ~6.5 mm on the precordial distribution to 5 mm in the limb leads.[13]

Temporal Stability

Long-term follow-up of ECGs (of the same individuals) reveals relatively constant pattern. However, the magnitude of ERS may lessen over time as the patient ages. In 25% to 30% of the patients with ERS previously documented on ECG, a repeat ECG will reveal complete disappearance of the pattern many years later.[14,16] There is also a normalization of the STE with rapid pacing or during exercise.[19] A study that followed athletes during marathon showed that the STE disappeared after exertion in 14 of 20 marathon runners.[20] Table 1 summarizes the ECG manifestations of ERS.

ERS AND ARRHYTHMIA

ER is a common ECG finding that for many decades was considered to be benign. Its potential to cause cardiac arrhythmias has been hypothesized from experimental studies. One study reviewed data from 206 case subjects who were resuscitated after cardiac arrest because of idiopathic VF and assessed the prevalence of ERS. The ERS was found to be more frequent in subjects with idiopathic VF than in control subjects (31% versus 5%, $P < 0.001$). Those subjects with ERS were more likely to have a history of syncope or sudden cardiac arrest during sleep than those without ERS. During a mean follow-up of 61 ± 50 months, defibrillator monitoring showed a higher incidence of recurrent VF in case subjects with a repolarization abnormality than in those without such an abnormality ($P = 0.008$).[1] Another study assessed the prevalence and prognostic significance of ER on 12-lead electrocardiography in a community-based general population of middle-aged subjects —during a mean follow-up of 30 ± 11 years. They found that J-point elevation of at least 0.1 mV in the inferior leads was associated with an increased risk of death from cardiac causes ($P = 0.03$); subjects with J-point elevation of more than 0.2 mV in the inferior leads had a markedly elevated risk of death from cardiac causes ($P < 0.001$) and from arrhythmia ($P = 0.01$). They concluded that an early-repolarization pattern in the inferior leads of a standard ECG is associated with an increased risk of death from cardiac causes in middle-aged subjects.[2]

TABLE 1. ECG characteristics of early repolarization

ECG component	Characteristics
STE	Wide spread STE more in the chest leads (usually V2–V4) than in the limb leads
ST-segment morphology	Concavity of initial upsloping portion of ST segment
J point	Elevation and notching or irregular contour of J point
T waves	Prominent concordant with large amplitude
Influence on ST segment	Reduction in STE with sympathoadrenal factors or with rapid pacing
	Normalizing during exercise and with aging

ECG, electrocardiographic; STE, ST-segment elevation.