Abstract
We report a case of chronic chest pain syndrome over the years along with ST/T changes on his ECG which otherwise appeared to be benign called early repolarization pattern (ERP). The classic teaching has been to pay close attention to any ST/T changes in the setting of chest pains. This case also gave us a chance to review the literature on this phenomenon of ERP.

Keywords: Early Repolarization Pattern (ERP); ECG; Malingering

Introduction
The phenomenon of Early Repolarization Pattern (ERP) was first described in the literature by Shipley and Hallaran when they noted elevated ST segment in lead II in 25% of men and 16% of females out of 200 healthy young people [1]. The term ‘early repolarization’ was coined by Grant, et al. in 1951 [2] and it was called so because premature repolarization was thought to be the underlying mechanism.

Case Report
A 26-year old man brought to the office from a federal prison for evaluation of his chest pain syndrome. He has had these chest pains over the years, tingling like feelings, not associated with any exertion or activities and no related dizziness or diaphoresis or palpitations. No known history of any arrhythmia, coronary artery disease or congenital heart disease.

His BP was 122/76 mm Hg, respiration 16 and cardiopulmonary examination was unremarkable. By all parameters, his chest pain description was atypical and likely non-cardiac; however, his baseline ECG was read as being abnormal and therefore cardiology consult was requested.

Figure 1: Baseline ECG: Normal Sinus Rhythm. ST/T changes suggestive of Early Repolarization Changes.
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His treadmill stress test duration was 7 minutes with 88% of maximum predicted heart rate achieved without any chest pains and no ischemic ST/T changes seen. His 2D-Echocardiography was unremarkable without any wall motion abnormalities, pericardial disease or valvular dysfunction. By ruling out all significant cardiac etiologies, one can come to the conclusion that ST/T changes on his ECG simply represent Early Repolarization Changes as described in depth below. He was advised to continue to walk and exercise without any restrictions. No further cardiac work-up and follow-up were recommended.

Discussion

This finding of early repolarization on ECG is defined as elevation of the QRS-ST junction (J point) in leads other than V1 - V3 on 12-lead ECG [3]. Historically, the term ERP has been described as ST-segment elevation in the absence of chest pains [4]. However, the term ERP can be complicated by the localization and the number of leads it is seen, QRS character and the J point (notching or slurring), the magnitude of J-point elevation, ST-segment elevation etc [5].

Currently, the ERP is referred as non-specific ST-segment elevation in algorithms used in commercial ECG machines [6]. The American Heart Association/American College of Cardiology/Heart Rhythm Society recommendations for the standardization and interpretation of the ECG defines ERP as a normal variant commonly characterized by J-point elevation and rapidly upsloping or normal ST segment [7].

The ERP has been considered ‘normal, a ‘normal variant’ or ‘benign’ all along up until 2000 [8]. However, since then numerous reports/case studies have suggested an increased chance of fatal arrhythmia and sudden cardiac deaths in those with ERP [9-12].

Prevalence

Prevalence of ERP varies from 1% to 18% [13]. One of the first studies to describe ERP was an investigation of 4-lead ECG in 200 healthy people with its incidence of 25% in men and 16% in women [1]. Young individuals, those exposed to vagotonia, males, African Americans and athletes are more likely to have a higher prevalence [14,15] ERP is also noted to be more frequent in cocaine users, hypertrophic cardiomyopathy and people with interventricular septal hypertrophy [16].

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J-wave elevation is found more frequently among patients with idiopathic ventricular fibrillation (VF) than among the healthy subjects [10]. In a case-control study, subjects with idiopathic VF have higher prevalence of ERP (31%) than healthy control subjects (5%) [17]. Even though the ERP is prevalent in general population, the incidence of idiopathic VF is quite low. In asymptomatic individuals before the age of 45 years, the risk of sudden cardiac death (SCD) is 3 per 100,000 and the SCD risk is 11 per 100,000 with J waves and 30 per 100,000 with ST elevation. In a meta-analysis, the estimated absolute risk for arrhythmic death in patients with ERP was 70 per 100,000 [18].

Genetics of ERP

The repolarization hypothesis suggests that ERP reflects regional heterogeneity in the dispersion gradient of myocardial refractoriness and that is manifested by a net outward shift in repolarization current (decreased inward sodium and calcium currents or increased outward potassium current) [19]. ERP is more common in relatives of unexplained arrhythmic sudden cardiac death suggesting a possible pro-arrhythmic inherited mutation [20]. Responsible genetic mutations may include the KCNJ8 and the ABCC9 genes (ATP-sensitive potassium channel) and many more genes [21,22].

Framingham study and the British cohort’s data suggest that there is evidence of heritability of ERP with a 2- to 2.5 times increased risk in siblings and offspring of subjects with inferolateral ERP [23]. In addition, ERP has been observed as a feature of another genetic arrhythmia such as Brugada syndrome [24].

Clinical Features

Most common scenario for people with ERP will be being completely asymptomatic and needless to say, these people will have incidental finding of ERP on their ECGs [17]. This group of people are less likely to have the risk of sudden cardiac death, but it is difficult to differentiate them from those who would have a benign course [25].

The other group of people are those with high risk presenting with syncope and those who are cardiac arrest survivors [26]. ERP was noted in one study in 18.5% patients with syncope compared to 2% in healthy controls [27], and this is almost 10-times more likely risk of syncope in those with ERP.

The 3rd common scenario seen in clinical practice is patients presenting with chest pains mostly atypical and likely non-cardiac and their routine ECGs show ERP.

The other common problem that we face in work-up of these patients is that one third of patients with benign vagal syncope may have ERP on their ECGs [28]. A history of syncope at rest can be strongly associated with ERP and can lead to pause-dependent increase in J waves that can precede episodes of ventricular fibrillation [29].

Management

In people with ERP without any symptoms, there are no current guideline recommendations to undergo any investigations or therapeutic interventions [30]. However, those who survived sudden cardiac death due to idiopathic ventricular fibrillation (VF), the incidence of recurrent VF can range between 22% and 37% at 2 - 4 years [31]. As per the latest guidelines these patients are best treated with an implantable cardioverter-defibrillator (ICD).

Conclusion

The finding of ERP on 12 - lead ECG has traditionally been considered benign and innocent over the years. Lately some case reports on otherwise healthy subjects surviving a cardiac arrest or idiopathic VF have suggested an association between J point elevation/or QRS slurring in the inferior and lateral leads and the risk of VF.

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The significance of ERP in clinical practice remains unclear, undefined and there is a need for prospective and randomized trials to study this issue further.

Bibliography


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