From the desk of editor:

Early repolarization syndrome has long been a questionable entity right from the graduation days. Though the prevalence of it is less (around 2%), it always mimics changes of ischemia which is more common. Confusion prevails when patient present with such ECG findings.

This disorder was considered benign till a few years back. But recent studies have poured in new insights and ruled out absolute benign nature of the syndrome.

To improve our understanding, we would like to offer some interesting facts and findings of this unique, but non-benign syndrome “EARLY REPLORIZATION SYNDROME”.

Dr. Joyal Shah

Early Repolarization Syndrome - Current Concepts

Approximately 2-5% of the population demonstrates the early repolarization changes on electrocardiogram; this population mostly consists of men, young adults, athletes, and people of African American heritage. Early repolarization previously was felt to be a rather benign feature; however, experimental studies, isolated case reports and independent case studies have shown its potential arrhythmogenic effects.

The syndrome is characterized by

1. An upward concave elevation of the RS-T segment with distinct or “embryonic” J waves, slurred downstroke of R waves or distinct J points or both.
2. RS-T segment elevation commonly encountered in the precordial leads and more distinct in these leads.
3. Rapid QRS transition in the precordial leads with counterclockwise rotation.
4. Persistence of these characteristics for many years although some intraindividual changes were common. Less commonly found were:
   a. Tall R and T waves in the precordial leads
   b. “Labile” or “juvenile” T wave patterns
   c. “Pseudo-R’” waves
   d. Isolated T negativity syndrome

Differential Diagnosis

- Pericarditis
- Myocardial ischemia
- Left ventricular hypertrophy
- Right bundle branch block
- Brugada syndrome
- Dysplastic ventricle
Clinical Aspects of the Early Repolarization Syndrome

In a normal ECG, the transition of ventricular depolarization into ventricular repolarization corresponds on the surface ECG to this J-point, where an abrupt transition from the QRS complex to the ST-segment occurs. Deviation of the J-point from the isoelectric line leads to the presence of a J-deflection. This typically produces a concave upward curve towards the T wave, helping to differentiate it from the convex, 'tomb-stone' waves seen with infarct. The leads associated with the J wave, commonly shown as ST-segment elevations (a.k.a. J point elevations) typically involve the anterior precordial leads (V1-V3). Recently, data has looked into early repolarization localized to the inferolateral leads.

J-point elevation of more than >0.2 mV carries risk of deaths from arrhythmias, which was statistically significant.

QRS notching is more prevalent in the malignant variants of ER than in the benign cases, findings that could have important implications for risk stratification of patients with ER.

TREATMENT OF ER (Early Repolarization)

While no investigations were carried out on the treatment of ER as long as it was looked upon as a benign phenomenon without clinical significance, the recent information raised interest in a possible drug therapy of this condition.

Recently, idiopathic VF and ER abnormality in the inferolateral leads, concluded that the electrical storms in these patients, all having ICDs implanted, were unresponsive to beta-blockers, lidocaine/mexiletine, and verapamil, while amiodarone was partially effective (in three of 10). Two specific drugs were efficient to abolish and prevent recurrences of VF in these patients: isoproterenol infusion immediately suppressed electrical storms in all patients used, while quinidine was successful, decreasing recurrent VF to nil in all cases used, and also restored a normal ECG (Table 1).

<table>
<thead>
<tr>
<th>Table-1 Treatment of Early Repolarization</th>
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<tbody>
<tr>
<td>Effective</td>
</tr>
<tr>
<td>Beta-blockers</td>
</tr>
<tr>
<td>Lidocaine</td>
</tr>
<tr>
<td>Mexiletine</td>
</tr>
<tr>
<td>Verapamil</td>
</tr>
<tr>
<td>Quinidine</td>
</tr>
<tr>
<td>Isoproterenol IV for electrical storms</td>
</tr>
<tr>
<td>Amiodarone</td>
</tr>
</tbody>
</table>

Implantation of ICDs were mentioned by Benito et al. in their recent review article to be justified in individuals at “very high risk” and pace making to prevent bradycardia.
or increase resting heart rate in some “at-risk” individuals, although no references were provided for the use of these procedures.

**ER: A BRUGADA SYNDROME (BS) VARIANT?**

Although a relatively recent consensus report on BS defined the ST-elevation of these patients strictly to the right precordial leads, it has subsequently been reported that 11% of the classical BS patients exhibit spontaneous ER or coved ≥ 2 mm ST-elevation pattern in the inferior/lateral leads, while in three patients, the coved Brugada pattern was only present in the inferior leads; the authors suggested that such patients should also be diagnosed with BS.

Recently it has been proposed by some to join ER and BS under the name “J-wave syndromes”, as they considered them to represent a continuous spectrum of phenotypic expression.

<table>
<thead>
<tr>
<th>They propose three subtypes:</th>
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<tr>
<td><strong>Type 1</strong></td>
</tr>
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<td><strong>Type 2</strong></td>
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<tr>
<td><strong>Type 3</strong></td>
</tr>
<tr>
<td><strong>Type 4</strong></td>
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</table>

**Diagnosis of Early Repolarization Syndrome and Differential Diagnosis with Brugada Syndrome**

<table>
<thead>
<tr>
<th></th>
<th>ER Syndrome</th>
<th>Brugada Syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right BBB</td>
<td>Not requested</td>
<td>Requested</td>
</tr>
<tr>
<td>ST-elevation in</td>
<td>L1, aVL, V4-6</td>
<td>V1-2</td>
</tr>
<tr>
<td>QRS notch or slurring</td>
<td>Frequent</td>
<td>Absent</td>
</tr>
<tr>
<td>Shape of ST-elevation</td>
<td>Upper</td>
<td>Coved</td>
</tr>
</tbody>
</table>

**Mode of Onset of VF in ER Syndrome versus Brugada Syndrome**

<table>
<thead>
<tr>
<th></th>
<th>ER Syndrome</th>
<th>Brugada Syndrome</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Initiated by PVCs with a short-long short sequence in</td>
<td>72.4%</td>
<td>In 15.1%</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Coupling intervals ms</td>
<td>328</td>
<td>395</td>
<td>&lt;0.01</td>
</tr>
</tbody>
</table>

**ER PATTERN IN ATHLETES**

The first two pathologies, without ST-elevation in the inferior and lateral leads, were significantly more frequently found in these subjects than in control athletes. The authors expressed their opinion that since these changes could reflect an underlying abnormality of repolarization, their myocardium may be more sensitive to various, still not well-defined arrhythmogenic triggers. The incidental finding of a J wave/QRS slurring in a healthy athlete should be considered as a marker that minimally increases the arrhythmic risk. Sport-induced rate-dependent repolarization inhomogeneities also may have been precipitating factors in creating the characteristic ECG patterns, known as ER or “vagotonic heart” in athletes. Bianco and Zeppilli found ER in the ECG of 89% of competitive athletes, none of them has suffered from major ventricular arrhythmias from the time of their study onward; they considered ER to be a complete benign phenomenon, even reversible after a few months of detraining.

**ELECTROPHYSIOLOGICAL OBSERVATIONS**

As noted, for decades the ST-elevation starting at or near the J-point, was unequivocally related to the mechanism of early-phase repolarization and the transmural voltage gradient during the initial phase of the ventricular repolarization was described as the cellular basis of the J wave.

The prognostic significance of the J point pattern in the inferolateral leads which were hypothesized to be
more arrhythmogenic than the more commonly studied anterior precordial leads (leads v1 through v3).

Based on the findings above, early repolarization in the inferior leads appeared to be a strong predictor of death from cardiac causes or from arrhythmia than J-point elevation in the lateral leads. In addition to the location of the early-repolarization pattern, the amplitude of the J-point elevation had great prognostic value. There was a significantly higher risk of death from cardiac causes among subjects with a markedly elevated J point (>0.2 mV) than among those with a more moderate elevation (≥0.1 mV).

In keeping with the study populations demographics and characteristics, case studies found to have the early repolarization pattern were more likely to be of the male sex and have lower baseline heart rates.

What is not clear, however, is the early repolarization pattern localized to the inferior leads increase the risk of death from cardiac and arrhythmic causes.

Hypothesis is the concept that the J-point elevation is a marker of increased transmural heterogeneity of ventricular repolarization. In addition, the left ventricular base defined by the inferolateral leads is an area known to have increased current density. As such, having an episode of early repolarization in an area with known increased current density is what can make the myocardium more vulnerable to ventricular tachyarrhythmias. This vulnerability may be amplified under certain conditions such as a cardiac ischemic event, the use of specific drugs, various levels of autonomic tone, electrolyte disturbances, channelopathies and/or structural cardiac abnormalities. This hypothesis can be further defended by the fact the anti-arrhythmic, quinidine, which is known to restore electrical homogeneity, aborted arrhythmias in a certain number of the patient population studied.

### ERS AND PERICARDITIS

<table>
<thead>
<tr>
<th>Major Differences</th>
<th></th>
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<tbody>
<tr>
<td>ERS</td>
<td>Pericarditis</td>
</tr>
<tr>
<td>Young people</td>
<td>Any age</td>
</tr>
<tr>
<td>STT changes with tall T</td>
<td>Ratio in V6 &gt;0.25</td>
</tr>
<tr>
<td>R/s slurring</td>
<td>ST evolution</td>
</tr>
<tr>
<td>STsegment/T segment ratio in V6&lt;0.25</td>
<td>Ratio in V6 &gt;0.25</td>
</tr>
<tr>
<td>No St evolution</td>
<td>Tallest T in V6</td>
</tr>
<tr>
<td>Tallest R in V6</td>
<td>Tallest T in V6</td>
</tr>
</tbody>
</table>

### Conclusion

Though ERS (Early Repolarization Syndrome) was considered as a benign condition till recently, it is not the same now. There is a risk of sudden cardiac death due to ventricular arrhythmias. They do not carry risk of developing new Myocardial infarction. It is difficult to risk stratify high risk subsets.

Further data is needed to reveal how to identify patients who are at high risk for such arrhythmias and what preventive measures can be done to stop it.

Until then, we recommend a close caution on these asymptomatic patients to keep them just asymptomatic.

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Body Mass Index (BMI)

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<th>Normal</th>
<th>Overweight</th>
<th>Obese</th>
<th>Severe Obesity</th>
<th>Morbid Obesity</th>
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<tr>
<td>18.5 - 24.9</td>
<td>Normal body weight</td>
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<tr>
<td>25 - 29.9</td>
<td>Overweight</td>
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<tr>
<td>30 - 34.9</td>
<td>Obese</td>
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<tr>
<td>35 - 39.9</td>
<td>Severe Obesity</td>
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<td>&gt; 40</td>
<td>Morbid Obesity</td>
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<td>Colorectal</td>
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<td>Gynaecology</td>
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<td>Head &amp; Neck</td>
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<td>Melanoma</td>
<td>Dr. Chaitanya Shroff (M) +91-98240 62590</td>
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<td>Lung</td>
<td>Dr. Hemant Shukla (M) +91-98250 42254</td>
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<td>Lymphoma</td>
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<td>Upper GI (Gastrointestinal)</td>
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