Healthy Heart

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Honorary Editor : Dr. Joyal Shah

Cardiologists Dr. Anish Chandarana (M) +91-98250 96922 Dr. Ajay Naik (M) +91-98250 82666 Dr. Satva Gupta (M) +91-99250 45780 Dr. Joyal Shah (M) +91-98253 19645 Dr. Ravi Singhvie (M) +91-98251 43975 **Dr. Gunvant Patel** (M) +91-98240 61266 Dr. Keyur Parikh (M) +91-98250 26999 Dr. Milan Chag (M) +91-98240 22107 Dr. Urmil Shah (M) +91-98250 66939 Dr. Hemang Baxi (M) +91-98250 30111

Cardiac Surgeons Dr. Dhiren Shah (M)+91-98255 75933 Dr. Dhaval Naik (M)+91-90991 11133 Dr. Dipesh Shah (M)+91-90990 27945

Pediatric and Adult Cardiac Surgeon Dr. Shaunak Shah (M)+91-98250 44502

Cardiac Anaesthetists Dr. Niren Bhavsar (M)+91-98795 71917 Dr. Hiren Dholakia (M)+91-95863 75818

Pediatric Cardiologists Dr. Kashyap Sheth (M) +91-99246 12288 Dr. Milan Chag (M) +91-98240 22107

Neonatologist and Pediatric Intensivist Dr. Amit Chitaliya (M)+91-90999 87400

Cardiac Electrophysiologist Dr. Ajay Naik (M) +91-98250 82666

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.



Price : ₹ 5/-

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.

Differential Diagnosis

- Pericarditis
- Myocardial ischemia
- Left ventricular hypertrophy
- Right bundle branch block
- Brugada syndrome
- Dysplastic ventricle

The syndrome is characterized by

- 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.
 - Ttall R and T waves in the precordial leads
 - "Labile" or "juvenile" T wave patterns
 - "Pseudo-R'" waves
 - Isolated T negativity syndrome

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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.



Upper panel Holter recording relatively high terminal QRS notch and J-point ST-elevation at heart rate of min Lower panel Holter recording on the same day at heart rate of min notching and Jpoint elevation hardly seen.

Recent studies suggest that the electrocardiographic (ECG) finding of J-point ST-elevation, the early repolarization syndrome, is not as benign as earlier believed. Three important articles published in 2008/2009 suggest that this finding in the inferolateral leads of the ECG may be representing a risk for subsequent ventricular fibrillation

New data on ER syndrome, found that J-point elevation in the inferior leads and in leads I and aVL was more common among the 45 patients with idiopathic VF than among age- and gender-matched control subjects: Jpoint elevation V4-6 occurred with equal frequency among patients and matched control subjects. 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-1 Treatment of Early Repolarization		
	Effective	
Beta-blockers	No	
Lidocaine	No	
Mexiletine	No	
Verapamil	No	
Quinidine	Yes	
Isoproterenol IV for electrical storms	Yes	
Amiodarone	Partially	

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 \geq 2 mm ST-elevation pattern in the inferiorlateral 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.

They propose three subtypes:			
Type 1	ECG pattern predominantly seen in the lateral		
	precordial leads is prevalent among healthy		
	male athletes		
Type 2	ER pattern seen predominantly in the inferior		
	or inferolateral leads and is associated with a		
	higher level of risk		
Type 3	ER pattern is displayed globally in the inferior,		
	lateral, and right precordial leads, is		
	associated with the highest level of risk for		
	development of malignant arrhythmias.		
Type 4	BS		

Diagnosis of Early Repolarization Syndrome and Differential Diagnosis with Brugada Syndrome

	ER Syndrome	Brugada Syndrome
Right BBB	Not requested	Requested
ST-elevationIn	L1, aVL, V4-6	V1-2
QRS notch or slurring	Frequent	Absent
Shape of	Upper	Coved
ST-elevation	Concave	

Mode of Onset of VF in ER Syndrome versus				
	ER	Brugada	P Value	
	Syndrome	Syndrome		
Initiated by PVCs	72.4%	ln 15.1 %	< 0.01	
with a short-long				
short sequenceln				
Coupling intervals ms	328	395	< 0.01	

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 ratedependent repolarization inhomegeneities 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 Jpoint 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 (\geq 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

Major Differences	
ERS	Pericarditis
Young people	Any age
STT changes with tall T	
R/s slurring	
STsegment/T segment ratio in $V_6 < 0.25$	Ratio in $V_6 > 0.25$
No St evolution	ST evolution
Tallest R in V_4	Tallest T in V_s

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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TKR was performed on Hiraben Patel at CIMS by Shree Orthocare team on 24/08/11. Surgery was performed using this patient specific Jig system by Dr Amir Sanghavi & Dr Ateet Sharma, along with Dr Mark Snyder.

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Courtesy :

Dr. Amir Sanghavi (M) +91-98250 66013 Dr. Ateet Sharma Dr. Hemang Ambani (M) +91-98250 20120 Dr. Chirag Patel

Dr. Ateet Sharma (M) +91-98240 61766 Dr. Chirag Patel (M) +91-98250 24473

Bariatric (Obesity) Management





Dr. Digvijaysingh Bedi (M) +91-98240 12582 Dr. Manish Khaitan (M) +91-98252 84543 Dr. Shailesh Shah (M) +91-98240 44443 (Listed alphabetically last name wise)



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CIMS Cardiovascular Team



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