J Wave Syndromes

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J Wave Syndromes

Group of electric disorders characterized by > 1 mm elevation of the J point or prominent J wave with or without ST elevation.
Group of electric disorders sharing common mechanisms of arrhythmogenicity, one of which may be variant of the other.

J wave syndromes
Phase 1 of AP (I_{to})

Responsible for the AP notch

Transient outward K current ($I_{to}$)

ATP-sensitive K channel

AC-sensitive K channel

Na current

Delayed rectifier K channel

Na current

Ca^{++}

K^{+}

K^{+}

K^{+}

K^{+}
Normally, AP notch is more prominent in the epicardium than the endocardium, in the RV than the LV, and in males than females.
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J wave syndromes

(> 1 mm elevation of J point + ST elevation)

Duration and magnitude of Endo to epicardial gradient determines the J point/ST segment elevation

Epicardium
Endocardium

ECG
Osborn
Hypothermia
Early repolarization
STEMI
Brugada
Increased AP notch results into failure of L-Ca++ channel activation \(\rightarrow\) regional loss of AP dome

Propagation of the dome \(\rightarrow\) phase 2 reentry

Increased AP notch results into failure of L-Ca\(^{++}\) channel activation $\rightarrow$ regional loss of AP dome

Propagation of the dome $\rightarrow$ phase 2 reentry
Factors that enhance AP notch

Vagal stimulation/bradycardia/pauses

Factors that accentuate AP notch

Magnitude of Na\(^+\) current

Normally

Cell memb

Na\(^+\) channels  L- Ca\(^{++}\) channels
Early closure of Na channels

Slow activation of L-Ca channels $\rightarrow$ prominent notch

Failed activation of L-Ca channels $\rightarrow$ loss of AP dome

SCN5A

Mutation

Na channel blockers

Free radicals

LPC

Cell memb

Na$^+$ channels

L-Ca$^{++}$ channels
Factors that accentuate AP notch

Slow L-Ca\(^{++}\) channel activation

*CACNA1C- CACNB2b* gene mutation

Cell memb

Na\(^{+}\) channels  L- Ca\(^{++}\) channels
Increased transient outward $K^+$ current ($I_{to}$)

$I_{to}$ gain of function mutation

$KCNE3$- $KCND3$ gene mutation

Regional loss of dome


Factors that enhance AP notch

Hypothermia
BS and ERS can be considered to represent a continuous spectrum of the phenotypic expression of gene mutations.
Brugada like or ERS?

J wave Syndrome
Patient with idiopathic VF (the bottom ECG was taken during sleep)
Brugada syndrome causes 4–12% of all SCDs, and up to 20% of SCDs without identifiable structural abnormalities.

Early Repolarization Syndrome

The ER pattern has long been considered to be a “benign” ECG manifestation that is more commonly seen in young healthy men and athletes.

Occurs in 5% of population.
Haissaguerre and co-workers compared 206 subjects with IVF to 412 healthy controls and demonstrated that an ER pattern was in 31% of subjects with IVF vs 5% of controls (P<0.001).

Patients with IVF who had the ER pattern were more likely to experience syncope or cardiac arrest during sleep than those without the ER pattern (hazard ratio, 2.1; 95% confidence interval, 1.2 to 3.5; P=0.008). Additionally, they were able to map the site of origin of ectopic activity in 8 patients and found that the origin of ectopy was consistent with the location of the early repolarization in ECG leads.

In a community-based general population of 10,864 subjects, an ER pattern in the inferior leads was associated with an increased risk of death from cardiac causes. J point elevation of >1 mm in the inferior leads was present in 3.5%, in the lateral leads in 2.4% and in both in 0.1%.

In a study, ERS was found in 60% of patients with IVF (9 out of 15) versus 3.3% of controls adjusted relative risk of death from cardiac causes to 2.98 (P<0.001).

# Early Repolarization Syndrome

<table>
<thead>
<tr>
<th>Type</th>
<th>Leads affected</th>
<th>Gene mutation</th>
<th>VF</th>
</tr>
</thead>
<tbody>
<tr>
<td>ER type 1</td>
<td>Lateral leads</td>
<td>CACNA1C, CACNB2B</td>
<td>Rare</td>
</tr>
<tr>
<td>ER type 2</td>
<td>Inferior or inferolateral</td>
<td>KCNJ8, CACNA1C, CACNB2B</td>
<td>Yes</td>
</tr>
<tr>
<td>ER type 3</td>
<td>Global + Rt precordial leads</td>
<td>CACNA1C</td>
<td>Yes, electric storms</td>
</tr>
</tbody>
</table>

ERS type 4 → Brugada syndrome

Should ERS subjected to risk stratification?

Possible high risk criteria:

- Type 3 ERS
- ≥2mm ST elevation
- Syncope
- FH of SCD
- Inducible VT/VF?
- Short coupled PVCs
Short coupled PVCs characterizing Brugada and ERS
Primary VF during acute STEMI

Different epi and endo response to ischemia

Gilmour and Zipes, 1980; Yan et al. 2004
The concept of phase 2 reentry due to accentuated AP notch during STEMI could answer some questions:

Why women with CHD have only a quarter of the risk of SCD as compared to men? *(AP notch is more prominent in males).*

Why VF is more common with RCA occlusion than LAD occlusion? *(AP notch is more prominent in RV than LV)*
The interplay between hereditary J wave syndrome, hypothermia and myocardial ischemia may perpetuate VF during cardiac arrest.
Management of J wave Syndromes

Since they are sharing common mechanisms, common lines are available for management such as

**Isoprenaline**
- Increase Ca current
- Can terminate electric storms

**Pacing**
- Eliminate pauses
- Reduce AP notch

**Quinidine**
- Inhibits transient outflow K current
- Resume AP dome

**ICD**
- For high risk patients
Management of J wave Syndromes

Since they are sharing common mechanisms, common lines are available for management such as

Future perspectives

- Selective $I_{to}$ blockers
- Selective ATP and AC sensitive $K_{ch}$ blockers for individual cases
- Gene therapy
- Epicardial RV pacing ??? A proposed management that needs to be studied
Epicardial pacing may abolish transmural gradient during AP notch

J wave syndromes are a group of electric disorders sharing the same mechanisms, characterized by transmural voltage gradient manifested as > 1 mm elevation of the J point ± ST elevation featuring BS, ERS, STEMI, and hypothermia.

Regional loss of AP dome can precipitate phase 2 reentry and VT/VF.

Patients with high risk criteria should receive ICD.

Isoprenaline and pacing may stop electric storms.

Quinidine is the only available drug that offers benefit.
Thank You

Osama Diab
Factors that accentuate AP notch

Increased outward ATP sensitive K⁺ current (IK-ATP)

KCNJ8 gene mutation