Repolarisation précoce – quand s’inquiéter

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Rythmosud - ACCA
Nice, 9 janvier 2018
La repolarisation précoce

- Onde J
- Segment ST

Types de repolarisation:
- Slur
- Notched
- Ascending/up-sloping
- Horizontal
- Descending
C... D..., 57 years-old male

• Sport ++ (bicycle 10 hours /week)

• PMH :
  – Duodenal ulcer

• Risk factors:
  – former smoker (30 P-Y)
  – mother died from MI at 62
  – high cholesterol treated by statins (pravastatin 40 mg/day)
“Early repolarization” is present if **all** of the following criteria are met:

1. **End-QRS notch or slur** on the downslope of R-wave (notch or onset of slur entirely above the baseline)

2. $J_p$ is $\geq 0.1 \text{ mV}$ in 2 or more contiguous leads of the 12-lead ECG, excluding leads V$_1$-V$_3$.

3. **QRS duration is <120 ms.**
# Syndrome de repolarisation précoce (ERS) – diagnostic

<table>
<thead>
<tr>
<th>I. Clinical History</th>
<th>Point:</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Unexplained cardiac arrest, documented VF or polymorphic VT</td>
<td>3</td>
</tr>
<tr>
<td>B. Suspected arrhythmic syncope</td>
<td>2</td>
</tr>
<tr>
<td>C. Syncope of unclear mechanism/unclear etiology</td>
<td>1</td>
</tr>
</tbody>
</table>

*Only award points once for highest score within this category*

<table>
<thead>
<tr>
<th>II. Twelve-Lead ECG</th>
<th>Point:</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. ER ≥0.2 mV in ≥2 inferior and/or lateral ECG leads with horizontal/descending ST segment</td>
<td>2</td>
</tr>
<tr>
<td>B. Dynamic changes in J-point elevation (≥0.1 mV) in ≥2 inferior and/or lateral ECG leads</td>
<td>1.5</td>
</tr>
<tr>
<td>C. ≥0.1 mV J-point elevation in at least 2 inferior and/or lateral ECG leads</td>
<td>1</td>
</tr>
</tbody>
</table>

*Only award points once for highest score within this category*

<table>
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<tr>
<th>III. Ambulatory ECG Monitoring</th>
<th>Point:</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Short-coupled PVCs with R on ascending limb or peak of T wave</td>
<td>2</td>
</tr>
</tbody>
</table>

<table>
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<tr>
<th>IV. Family History</th>
<th>Point:</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Relative with definite ERS</td>
<td>2</td>
</tr>
<tr>
<td>B. ≥2 first-degree relatives with a II.A. ECG pattern</td>
<td>2</td>
</tr>
<tr>
<td>C. First-degree relative with a II.A. ECG pattern</td>
<td>1</td>
</tr>
<tr>
<td>D. Unexplained sudden cardiac death &lt;45 years in a first- or second-degree relative</td>
<td>0.5</td>
</tr>
</tbody>
</table>

*Only award points once for highest score within this category*

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<th>V. Genetic Test Result</th>
<th>Point:</th>
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<td>A. Probable pathogenic ERS susceptibility mutation</td>
<td>0.5</td>
</tr>
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</table>

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Score (requires at least 1 ECG finding)

- ≥5 points: Probable/definite ERS
- 3–4.5 points: Possible ERS
- <3 points: Nondiagnostic

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Antzelevitch C et al, Heart Rhythm 2016
2014: 24 h 12 leads Holter ECG

Predominant morphology (99% of PVC): 805 isolated, 26 doubles, 6 triples
Sudden cardiac death on May 4, 2015

- lost consciousness while working (manual worker)
- 1st defibrillation at 5 minutes
- one VF recurrence while being prepared for angiocoronarography (normal)
- LVEF 55%

- 3 other VF recurrences during the first 24 hours despite profound sedation, therapeutic hypothermia, IV betablocker, lidocaïne and amiodarone
- no significant troponine increase (initial H1<0.05, H8=1.03, H21=0.25, H45=0.05) (not HS troponin!)
- normal blood tests (K=4.12)

- H24- H48: no VF recurrence under oral atenolol 50 mg
- sedation stopped at H48, complete neurological recovery (Glasgow 15)
- transfer to our center
Continuous 12 lead ECG monitoring
In total:

- 3 VF on May 7, 2015:
  - 02:34
  - 04:05
  - 04:16

- RF ablation on May 7
Spontaneous PVC’s during the whole procedure
LV bipolar voltage
Brief thermal response (same RF)
In the ward

Before ablation

After ablation
Ablation of Left Ventricular Substrate in Early Repolarization Syndrome

DECEBAL GABRIEL LATCU, M.D., SOK-SITHIKUN BUN, M.D., NAIMA ZARQANE, M.D., and NADIR SAOUDI, M.D., Ph.D.

From the Cardiologie, Centre Hospitalier Princesse Grace, Monaco, Monaco (Principality)

Presence of a J wave on the ECG increases the probability of VF from 3.4:100,000 to 11:100,000.


- high-amplitude J wave ≥0.2mV
- horizontal or descending ST segment

- inferior/Inferolateral leads

- Family history of sudden death

- Coexisting Brugada ECG pattern (J waves in V₁–V₃)

- Short QT intervals in subjects with ER

SVP: pas de valeur prédictive

Mahida S, JAm Coll Cardiolog 2015;65:151–159.
ERS – prise en charge consensus 2016

APHRS/EHRA/HRS/SOLACE J-Wave syndromes expert consensus conference report

ER pattern
> 0.1 mV in at least 2 contiguous infero-lateral leads

Symptomatic
- Electrical Storm
  - Isoproterenol (Class IIa) +/- quinidine
  - ICD (Class I) If ICD refused or contraindicated Quinidine
  - Repeated Appropriate Shocks
- Prior Cardiac Arrest
- Sustained VT
- Syncope, Seizure NAR and strong family history of sudden death at young age
  - Presumably Arrhythmic origin

Asymptomatic
- High-risk ER ECG pattern (prominent J-waves, horizontal/descending ST-segment, high dynamicity) and strong family history of unexplained sudden death at young age
  - Yes
    - ICD (Class IIb)
  - No
    - Close Follow-up

ICD (Class IIb) with/without ILR
Quinidine (Class IIa) Cilostazol

Table 1. Features That May Raise Suspicion for a Malignant and Heritable Form of ER

- Family history of sudden cardiac arrest or early unexplained death
- Personal evaluation and workup suggestive of a channelopathy (eg, short-QT syndrome, Brugada syndrome)
- Personal history of unheralded syncope suggestive of an arrhythmogenic pathogenesis (particularly when at rest or recumbent)

ER indicates early repolarization; LOE, Level of Evidence; and VF, ventricular fibrillation.

Table 2. Recommendations for the Management of ER

<table>
<thead>
<tr>
<th>Recommendation</th>
<th>Class</th>
<th>LOE</th>
</tr>
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<tr>
<td>Further evaluation for the incidental findings of an ERP on an ECG in an asymptomatic patient (without family history of sudden cardiac death) is not recommended.</td>
<td>III</td>
<td>C</td>
</tr>
<tr>
<td>In patients with unexplained syncope, incorporation of the presence of most ER electrocardiographic findings into risk stratification is not well established.</td>
<td>IIb</td>
<td>C</td>
</tr>
<tr>
<td>In patients with aborted sudden death or resuscitated VF who have electrocardiographic findings of ER, programmed ventricular stimulation performed on the basis of ER pattern alone is not recommended.</td>
<td>III</td>
<td>B</td>
</tr>
<tr>
<td>In patients with both unexplained syncope and first-degree family history of sudden death, the presence of ER may be considered in overall risk stratification during appropriate evaluation for arrhythmic causes of sudden death.</td>
<td>IIb</td>
<td>C</td>
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