Πως προλαμβάνω τον αιφνίδιο καρδιακό θάνατο στις διαυλοπάθειες

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Επικ. Επιμελητής Καρδιολογίας

EHRA accredited in EP and Devices (Step 2)
No conflict of interest
- Long QT
- Short QT
- Brugada
- CPVT
- Early repolarisation
**HRS/EHRA Expert Consensus Statement on the State of Genetic Testing for the Channelopathies and Cardiomyopathies**

<table>
<thead>
<tr>
<th>Section # – Disease</th>
<th>Diagnostic</th>
<th>Prognostic</th>
<th>Therapeutic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Section I – LQTS</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
</tr>
<tr>
<td>Section II – CPVT</td>
<td>+++</td>
<td>+</td>
<td>–</td>
</tr>
<tr>
<td>Section III – BrS</td>
<td>+</td>
<td>+</td>
<td>–</td>
</tr>
</tbody>
</table>

Ackerman et al

Europace (2011) 13, 1077–1109
doi:10.1093/europace/eur245
Causes of sudden cardiac death (SCD)

- **Valvular heart disease**
  - 1-5%

- **Inherited arrhythmia syndrome**
  - (LQTS, BrS, CPVT, ERS, etc)
  - 1-2% in Western countries
  - 10% in Japan

- **Cardiomyopathies**
  - (NIDCM, HCM, ARVC, etc)
  - 10-15% in Western countries
  - 30-35% in Japan

- **Coronary heart disease**
  - ~75% in Western countries (~50% in women and blacks)
  - 50-60% in Japan

- **Substrates in myocardium**
  - Fibrosis
  - Hypertrophy
  - Ion channel functional modification
  - Abnormal calcium handling

- **Triggers**
  - Heart failure / Stretch
  - Ischemia
  - Myocardial inflammation
  - Sympathetic nerve surge
  - Electrolyte abnormality
  - Environmental stress
  - Psychological stress / Depression

**SCD causes and rates**

**Predisposing factors and risks**
- J wave / Repolarization abnormality
- Parasympathetic nerve dysfunction
- Male sex
- African descent
- Diabetes
- Smoking

**Genetics (Family history)**
- Shortage of N-3 PUFA
- Atrial fibrillation
- Chronic kidney disease
- Obstructive sleep apnea
LONG QT syndrome

Type 2

Type 1

Type 3
Long QT syndrome, genotype subdivision

- Type 1: KCNQ1 (KvLQT1) 50%
- Type 2: KCNH2 (HERG) 35%
- Type 3: SCN5A 8%
- Type 4-13: rare
Schwartz criteria

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>ECG Findings:</strong></td>
<td></td>
</tr>
<tr>
<td>QTC (ms) †</td>
<td></td>
</tr>
<tr>
<td>≥ 480 ms</td>
<td>3</td>
</tr>
<tr>
<td>460 – 469 ms</td>
<td>2</td>
</tr>
<tr>
<td>450-459 ms (in male patient)</td>
<td>1</td>
</tr>
<tr>
<td>Torsade de pointes ‡</td>
<td>2</td>
</tr>
<tr>
<td>T-wave alternans</td>
<td>1</td>
</tr>
<tr>
<td>Notched T wave in 3 leads</td>
<td>1</td>
</tr>
<tr>
<td>Low heart rate for age §</td>
<td>0.5</td>
</tr>
<tr>
<td><strong>Clinical History:</strong></td>
<td></td>
</tr>
<tr>
<td>Syncope ‖</td>
<td></td>
</tr>
<tr>
<td>With stress</td>
<td>2</td>
</tr>
<tr>
<td>Without Stress</td>
<td>1</td>
</tr>
<tr>
<td>Congenital deafness</td>
<td>0.5</td>
</tr>
<tr>
<td><strong>Family History:</strong></td>
<td></td>
</tr>
<tr>
<td>A. Family members with definite LQTS #</td>
<td>1</td>
</tr>
<tr>
<td>B. Unexplained sudden cardiac death &lt;30 y in an immediate family member</td>
<td>0.5</td>
</tr>
</tbody>
</table>
## Expert Consensus Recommendations on LQTS Therapeutic Interventions

<table>
<thead>
<tr>
<th>Class</th>
<th>Recommendation</th>
</tr>
</thead>
</table>
| I     | The following lifestyle changes are recommended in all patients with a diagnosis of LQTS:  
|       | a. Avoidance of QT-prolonging drugs (www.qtdrugs.org)  
|       | b. Identification and correction of electrolyte abnormalities that may occur during diarrhea, vomiting, metabolic conditions, or imbalanced diets for weight loss.  
|       | 2. Beta-blockers are recommended in patients with a diagnosis of LQTS who are:  
|       | a. Asymptomatic with QTc ≥ 470 ms and/or  
|       | b. Symptomatic for syncope or documented ventricular tachycardia/ventricular fibrillation (VT/VF).  
|       | 3. Left cardiac sympathetic denervation (LCSD) is recommended in high-risk patients with a diagnosis of LQTS in whom:  
|       | a. Implantable cardioverter-defibrillator (ICD) therapy is contraindicated or refused and/or  
|       | b. Beta-blockers are either not effective in preventing syncope/arrhythmias, not tolerated, not accepted or contraindicated.  
|       | 4. ICD implantation is recommended in patients with a diagnosis of LQTS who are survivors of a cardiac arrest.  
|       | 5. All LQTS patients who wish to engage in competitive sports should be referred to a clinical expert for the evaluation of risk.  
| IIa   | Beta-blockers can be useful in patients with a diagnosis of LQTS who are asymptomatic with QTc ≤ 470 ms.  
|       | 7. ICD implantation can be useful in patients with a diagnosis of LQTS who experience recurrent syncopal events while on beta-blocker therapy.  
|       | 8. LCSD can be useful in patients with a diagnosis of LQTS who experience breakthrough events while on therapy with beta-blockers/ICD.  
|       | 9. Sodium channel blockers can be useful, as add-on therapy, for LQT3 patients with a QTc > 500 ms who shorten their QTc by > 40 ms following an acute oral drug test with one of these compounds.  
| III   | 10. Except under special circumstances, ICD implantation is not indicated in asymptomatic LQTS patients who have not been tried on beta-blocker therapy.  |
Genotype - phenotype, LQT registry (Schwartz e.a. 2001)
Not All Beta-Blockers Are Equal in the Management of Long QT Syndrome Types 1 and 2
Higher Recurrence of Events Under Metoprolol
Ranolazine for Congenital Long-QT Syndrome Type III
Experimental and Long-Term Clinical Data

Ehud Chorin, MD, PhD; Dan Hu, MD, PhD*; Charles Antzelevitch, PhD;
Aviram Hochstadt, MD; Luiz Belardinelli, MD; David Zeltser, MD; Hector Barajas-Martinez, PhD;
Uri Rozovski, MD; Raphael Rosso, MD; Arnon Adler, MD; Jesaia Benhorin, MD; Sami Viskin, MD

![ECG Comparison](image)
Long-term flecainide therapy in type 3 long QT syndrome

Ehud Chorin¹, Rivki Taub², Aron Medina², Nir Flint¹, Sami Viskin¹, and Jesaia Benhorin¹*

**Table 2** Cardiac events after stopping flecainide

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Gender</th>
<th>Time on flecainide (months)¹</th>
<th>Time to cardiac event (months)²</th>
<th>Cardiac event</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>89</td>
<td>1</td>
<td>ACA</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>122</td>
<td>1</td>
<td>ACA</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>84</td>
<td>11</td>
<td>Syncope</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>37</td>
<td>4</td>
<td>ACA</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>1</td>
<td>1</td>
<td>SCD</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>35</td>
<td>1</td>
<td>TDP</td>
</tr>
</tbody>
</table>

ACA: aborted cardiac arrest; SCD, sudden cardiac death; TDP, torsade de points.
¹Without symptoms.
²After stopping flecainide.
High risk carriers
• Timothy
• Jervel – Lange – Nielsen
• QTc > 500 (esp > 600) msec
• T wave alternans
Nocturnal bradycardia

Heart rate 34/min, QTc 566 msec

Heart rate 32/min, QTc 477 msec

? B blocker
Clinical Aspects of Type 3 Long-QT Syndrome
An International Multicenter Study

logrank $p<0.001$

Cumulative Probability of Cardiac Event vs. Age

- Sync/ACA/SCD
- ACA/SCD
- SCD
QTc > 500

QTc > 470
S-ICD screening failure occurs in up to 13% of patients with inherited primary arrhythmia syndromes.

LQTS 5%
<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients who have a history of LQTS-related symptoms (prior cardiac arrest or syncopal event), QTc &gt;470 ms (males) or &gt;480 ms (females) should be limited to Class IA sports.</td>
<td>LQTS symptomatic patients are disqualified from all competitive sports. Recommended using QTc values of &gt;440 ms (males) or &gt;460 ms (females) as a trigger for further evaluation.</td>
<td>LQTS symptomatic patients (corrected QT interval &gt;470 ms in males, or &gt;480 ms in females) should be restricted from competitive sports until a comprehensive evaluation has been completed, a treatment program has been implemented, and the athlete has been asymptomatic on treatment for 3 months.</td>
<td>LQTS symptomatic patients may return to sports (except swimming in a previously symptomatic LQT1) after institution of treatment and appropriate precautionary measures, assuming the athlete has been asymptomatic on treatment for at least 3 months.</td>
</tr>
</tbody>
</table>

Asymptomatic athlete

Patients with phenotype-negative, genotype-positive LQTS are not restricted, but LQT1 patients should refrain from swimming.  

LQTS asymptomatic patients are disqualified from all competitive sports. Recommended using QTc values of >440 ms (males) or >460 ms (females) as a trigger for further evaluation.  

In asymptomatic athletes with genotype-positive/phenotype-negative LQTS, it is reasonable to participate in all competitive sports, with appropriate precautionary measures including:  
1) avoidance of QT-prolonging drugs(http://www.crediblemeds.org)  
2) electrolyte/hydration replenishment and avoidance of dehydration for all  
3) treating hyperthermia or avoidance training-related heat exhaustion or heat stroke  
4) acquisition of automatic external defibrillator  
5) establishing an emergency action plan with an appropriate team
Expert Consensus Recommendations on BrS Diagnosis

1. BrS is diagnosed in patients with ST-segment elevation with type I morphology ≥2 mm in ≥1 lead among the right precordial leads V1, V2 positioned in the 2nd, 3rd, or 4th intercostal space occurring either spontaneously or after provocative drug test with intravenous administration of Class I antiarrhythmic drugs.

2. BrS is diagnosed in patients with type 2 or type 3 ST-segment elevation in ≥1 lead among the right precordial leads V1, V2 positioned in the 2nd, 3rd, or 4th intercostal space when a provocative drug test with intravenous administration of Class I antiarrhythmic drugs induces a type I ECG morphology.

Antzelevich et al Heart Rhythm 2016
BRUGADA
Quinidine Normalizes ST Segment Elevation in Brugada Syndrome Patient

Alings et al. PACE 24:1420-1422, 2001

Before

After
Efficacy of Quinidine in High-Risk Patients With Brugada Syndrome
Bernard Belhassen, MD; Aharon Glick, MD; Sami Viskin, MD

Background—Automatic implantable cardioverter-defibrillator therapy is considered the only effective treatment for high-risk patients with Brugada syndrome. Quinidine depresses $I_{Ca}$ current, which may play an important role in the arrhythmogenesis of this disease.

Methods and Results—The effects of quinidine bisulfate (mean dose, 1483 ± 240 mg) on the prevention of inducible and spontaneous ventricular fibrillation (VF) were prospectively evaluated in 25 patients (24 men, 1 woman; age, 19 to 80 years) with Brugada syndrome. There were 15 symptomatic patients (including 7 cardiac arrest survivors and 7 patients with unexplained syncope) and 10 asymptomatic patients. All 25 patients had inducible VF at baseline electrophysiological study. Quinidine prevented VF induction in 22 of the 25 patients (88%). After a follow-up period of 6 months to 22.2 years, all patients are alive. Nineteen patients were treated with quinidine for 6 to 219 months (mean ± SD, 56 ± 67 months). None had an arrhythmic event, although 2 had non–arrhythmia-related syncope. Administration of quinidine was associated with a 36% incidence of side effects that resolved after drug discontinuation.

Conclusions—Quinidine effectively prevents VF induction in patients with Brugada syndrome. Our data suggest that quinidine also suppresses spontaneous arrhythmias and could prove to be a safe alternative to automatic implantable cardioverter-defibrillator therapy for a substantial proportion of patients with Brugada syndrome. Randomized studies comparing these two therapies seem warranted. (Circulation. 2004;110:1731-1737.)

Key Words: antiarrhythmic agents, electrophysiology, tachyarrhythmias
Shock Reduction With Long-Term Quinidine in Patients With Brugada Syndrome and Malignant Ventricular Arrhythmia Episodes

(I Am Coll Cardiol. 2016 Apr 5;67(13):1653-4.)

- 19 patients (66%) remained free of appropriate ICD discharges.

- After quinidine, significant reduction in total number of ICD shocks and in median number of ICD shocks per patient was observed: from 203 to 41 shocks and from 6 shocks per patient IQR (4 to 9) to 0 shocks per patient IQR (0 to 2.5), p < 0.0001, respectively.

The chart shows the median number of implantable cardioverter-defibrillator (ICD) shocks before and after quinidine administration for each patient. The number of shocks was reduced by quinidine use.
## Therapeutic options and recommendations for BrS patients

<table>
<thead>
<tr>
<th>Class</th>
<th>Quinidine Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class IIA</td>
<td>Quinidine <strong>can be useful</strong> in patients with a diagnosis of BrS and history of <strong>arrhythmic storms</strong> defined as more than two episodes of VT/VF in 24 hours.</td>
</tr>
<tr>
<td></td>
<td>Quinidine <strong>can be useful</strong> in patients with a diagnosis of BrS:</td>
</tr>
<tr>
<td></td>
<td>• Who qualify for an ICD but present a <strong>contraindication to the ICD or refuse it</strong>, and/or</td>
</tr>
<tr>
<td></td>
<td>• Have a history of documented <strong>supraventricular arrhythmias</strong> that require treatment.</td>
</tr>
<tr>
<td>Class IIB</td>
<td>Quinidine <strong>may be considered</strong> in asymptomatic patients with a diagnosis of BrS with a spontaneous type 1 ECG.</td>
</tr>
</tbody>
</table>

9. Catheter ablation may be considered in patients with a diagnosis of BrS and history of arrhythmic storms or repeated appropriate ICD shocks.
Risk Stratification in Brugada Syndrome

Results of the PRELUDE (PRogrammed ELectrical stimUlation preDdictive valuE) Registry

Silvia G. Priori, MD, PHD,*†‡ Maurizio Gasparini, MD,§ Carlo Napolitano, MD, PHD,*‡ Paolo Della Bella, MD,‖ Andrea Ghidini Ottonelli, MD,¶ Biagio Sassone, MD,# Umberto Giordano, MD,** Carlo Pappone, MD,†† Giosuè Mascioli, MD,‡‡ Guido Rossetti, MD, §§ Roberto De Nardis MD,||| Mario Colombo, MS¶¶

<table>
<thead>
<tr>
<th>Patient ID #</th>
<th>Sex</th>
<th>Age (yrs)*</th>
<th>Family History of SCD</th>
<th>Spontaneous Type 1 ECG</th>
<th>History of Syncope</th>
<th>Inducibility</th>
<th>VRP &lt;200 ms</th>
<th>QRS-f</th>
<th>SCN5A Mutation</th>
<th>Event Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>4</td>
<td>M</td>
<td>43</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>ICD shock</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>35</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>+</td>
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<td>174</td>
<td>M</td>
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<td>+</td>
<td>+</td>
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<td>+</td>
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<td>+</td>
<td>N.A.</td>
<td>ICD shock</td>
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<td>22</td>
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<td>-</td>
<td>+</td>
<td>-</td>
<td>-</td>
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<td>-</td>
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<td>ICD shock</td>
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<td>ICD shock</td>
</tr>
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<td>58</td>
<td>M</td>
<td>32</td>
<td>-</td>
<td>+</td>
<td>-</td>
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<td>N.A.</td>
<td>ICD shock</td>
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<tr>
<td>63</td>
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<td>+</td>
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<td>-</td>
<td>+</td>
<td>+</td>
<td>N.A.</td>
<td>Resuscitated CA</td>
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<td>F</td>
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<td>+</td>
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<td>+</td>
<td>N.A.</td>
<td>ICD shock</td>
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<td>+</td>
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<td>+</td>
<td>-</td>
<td>ICD shock</td>
</tr>
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<td>+</td>
<td>+</td>
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<td>-</td>
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<td>-</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>ICD shock</td>
</tr>
</tbody>
</table>
Long-Term Prognosis of Patients Diagnosed With Brugada Syndrome
Results From the FINGER Brugada Syndrome Registry

V. Probst, MD, PhD*; C. Veltmann, MD*; L. Eckardt, MD*; P.G. Meregalli, MD*; F. Gaita, MD; H.L. Tan, MD, PhD; D. Babuty, MD, PhD; F. Sacher, MD; C. Giustetto, MD; E. Schulze-Bahr, MD, PhD; M. Borggrefe, MD, PhD; M. Haissaguerre, MD; P. Mabo, MD, PhD; H. Le Marec, MD, PhD; C. Wolpert, MD, PhD; A.A.M. Wilde, MD, PhD

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**Graph:**
- EPS negative
- EPS positive

**Table:**

<table>
<thead>
<tr>
<th>Follow-up in months</th>
<th>0</th>
<th>12</th>
<th>24</th>
<th>36</th>
<th>48</th>
<th>60</th>
<th>72</th>
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</thead>
<tbody>
<tr>
<td>negative</td>
<td>376</td>
<td>301</td>
<td>237</td>
<td>187</td>
<td>136</td>
<td>94</td>
<td>59</td>
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<tr>
<td>positive</td>
<td>262</td>
<td>212</td>
<td>161</td>
<td>113</td>
<td>81</td>
<td>52</td>
<td>34</td>
</tr>
</tbody>
</table>
- ST augmentation at the early recovery
- Conduction abnormalities
- Inferolateral ER
- Signal averaged ECG
- Large S in I
High rate of subcutaneous implantable cardioverter-defibrillator sensing screening failure in patients with Brugada syndrome: a comparison with other inherited primary arrhythmia syndromes

BrS 18%
LQTS 5%
IVF 7%

Conte G Europace 2017
14.8% των ασθενών με BrS type II ή III και που έκαναν δοκιμασία πρόκλησης με ajmaline απέτυχαν, ενώ στη baseline screening μελέτη είχαν περάσει το test.
Low Prevalence of Inappropriate Shocks in Patients With Inherited Arrhythmia Syndromes With the Subcutaneous Implantable Defibrillator Single Center Experience and Long-Term Follow-Up

Boris Rudic, MD; Erol Tülümen, MD; Veronika Berlin, MD; Susanne Röger, MD; Ksenija Stach, MD; Volker Liebe, MD; Ibrahim El-Battrawy, MD; Christina Dösch, MD; Theano Papavassiliu, MD; Ibrahim Akin, MD; Martin Borggreve, MD; Jürgen Kuschyk, MD
The new kids on the block of arrhythmogenic disorders: Short QT syndrome and early repolarization.

Mazzanti A¹, Underwood K¹, Nevelev D¹, Kofman S¹, Priori SG¹,²,³.
Expert Consensus Recommendations on ER Diagnosis

1. ER syndrome is diagnosed in the presence of J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG in a patient resuscitated from otherwise unexplained VF/polymorphic VT.

2. ER syndrome can be diagnosed in a SCD victim with a negative autopsy and medical chart review, with a previous ECG demonstrating J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG.

3. ER pattern can be diagnosed in the presence of J-point elevation ≥1 mm in ≥2 contiguous inferior and/or lateral leads of a standard 12-lead ECG.

I. Clinical History

A. Unexplained cardiac arrest, documented VF or polymorphic VT 3
B. Suspected arrhythmic syncope 2
C. Syncope of unclear mechanism/unclear etiology 1
*Only award points once for highest score within this category

II. Twelve-Lead ECG

A. ER ≥0.2 mV in ≥2 inferior and/or lateral ECG leads with horizontal/descending ST segment 2
B. Dynamic changes in J-point elevation (≥0.1 mV) in ≥2 inferior and/or lateral ECG leads 1.5
C. ≥0.1 mV J-point elevation in at least 2 inferior and/or lateral ECG leads 1
*Only award points once for highest score within this category

III. Ambulatory ECG Monitoring

A. Short-coupled PVCs with R on ascending limb or peak of T wave 2

IV. Family History

A. Relative with definite ERS 2
B. ≥2 first-degree relatives with a II.A. ECG pattern 2
C. First-degree relative with a II.A. ECG pattern 1
D. Unexplained sudden cardiac death < 45 years in a first- or second-degree relative 0.5
*Only award points once for highest score within this category

V. Genetic Test Result

A. Probable pathogenic ERS susceptibility mutation 0.5

Score (requires at least 1 ECG finding)

≥5 points: Probable/definite ERS
3–4.5 points: Possible ERS
< 3 points: Nondiagnostic
Early repolarization
J Wave Manifestation

**ECG Leads**
- Global (Brugada ECG patterns + high amplitude inferior and lateral J-waves)
- Right Precordial (Brugada ECG Pattern)
- Inferior or Inferior-Lateral
- Lateral

**J Wave Syndromes**

**Clinical and other ECG features**
- Resuscitation from cardiac arrest, documented VF, or documented polymorphic VT
- Positive family history of SCD, arrhythmic syncope; identified gene mutations
- Short-coupled VPEs
- Fragmented QRS
- Co-existing electrical disorder (short QT)
- Dynamic changes in J-wave amplitude
- High amplitude J-waves (>0.2 mV)
- J-waves with horizontal/downsloping ST-segment
- Prolonged Tpeak-Tend interval
- Steep QT/RR
- Tall R-waves, rapidly ascending ST segments

**Arrhythmic Risk**

**Prevalence Log Scale**
Expert Consensus Recommendations on ER Therapeutic Interventions

Class I
1. ICD implantation is recommended in patients with a diagnosis of ER syndrome who have survived a cardiac arrest.

Class IIa
2. Isoproterenol infusion can be useful in suppressing electrical storms in patients with a diagnosis of ER syndrome.
3. Quinidine in addition to an ICD can be useful for secondary prevention of VF in patients with a diagnosis of ER syndrome.

Class IIb
4. ICD implantation may be considered in symptomatic family members of ER syndrome patients with a history of syncope in the presence of ST-segment elevation >1 mm in two or more inferior or lateral leads.
5. ICD implantation may be considered in asymptomatic individuals who demonstrate a high-risk ER ECG pattern (high J-wave amplitude, horizontal/descending ST-segment) in the presence of a strong family history of juvenile unexplained sudden death with or without a pathogenic mutation

Class III
6. ICD implantation is not recommended in asymptomatic patients with an isolated ER ECG pattern.
The genetics underlying idiopathic ventricular fibrillation: A special role for catecholaminergic polymorphic ventricular tachycardia?☆

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2. Beta-blockers are recommended in all symptomatic patients with a diagnosis of CPVT.

3. ICD implantation is recommended in patients with a diagnosis of CPVT who experience cardiac arrest, recurrent syncope or polymorphic/bidirectional VT despite optimal medical management, and/or LCSD.

4. Flecainide can be a useful addition to beta-blockers in patients with a diagnosis of CPVT who experience recurrent syncope or polymorphic/bidirectional VT while on beta-blockers.

5. Beta-blockers can be useful in carriers of a pathogenic CPVT mutation without clinical manifestations of CPVT (concealed mutation-positive patients).

Outcomes of defibrillator therapy in catecholaminergic polymorphic ventricular tachycardia

Ferran Roses-Noguer, MD, Julian W.E. Jarman, MRCP, MD(Res), Jonathan R. Clague, MD, FRCP, Jan Till, MD

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Channelopathies as Causes of Sudden Cardiac Death

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TIME FOR QUESTIONS
The new kids on the block of arrhythmogenic disorders: Short QT syndrome and early repolarization.

Mazzanti A¹, Underwood K¹, Nevelev D¹, Kofman S¹, Priori SG¹.².³.

Expert Consensus Recommendations on SQTS Therapeutic Interventions

Class I
1. ICD implantation is recommended in symptomatic patients with a diagnosis of SQTS who:
   a. Are survivors of a cardiac arrest and/or
   b. Have documented spontaneous sustained VT with or without syncope.

Class IIb
2. ICD implantation may be considered in asymptomatic patients with a diagnosis of SQTS and a family history of SCD.
3. Quinidine may be considered in asymptomatic patients with a diagnosis of SQTS and a family history of SCD.
4. Sotalol may be considered in asymptomatic patients with a diagnosis of SQTS and a family history of SCD.