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BRUGADA SYNDROME

- J point ≥ 2mm
- Coved type ST segment elevation
- Negative T wave

20,387 children (9.7±3.2 years old) during a school health examination
→ Brugada ECG prevalence was 0.05% (Adults 0.12-0.8%)

Sidik et al. Europace 2009; 11: 650-6
Heart Rhythm 2016; 13: 1274-1282

BRUGADA SYNDROME in CHILDREN

20,387 children (9.7±3.2 years old) during a school health examination
→ Brugada ECG prevalence was 0.05% (Adults 0.12-0.8%)

2014-2017

54 pts < 18 yrs at the diagnosis
→ Mean age 13 ± 5 years old, 42 (76%) male

Predictors of events during f-up:
- Spontaneous Brugada type I ECG pattern
- Symptoms at diagnosis

47% genotype positive
9/10 pts with life-threatening arrhythmias were genotype +

31/54 (57%) pts with spontaneous Brugada type I ECG
→ 11 (35%) during fever

<table>
<thead>
<tr>
<th>ECG</th>
<th>VT / SVT (%)</th>
<th>Syncope (%)</th>
<th>Asymptomatic / (≥ 1) (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I BrECG</td>
<td>31/3 (100%)</td>
<td>9/9 (100%)</td>
<td>23/3 (86%)</td>
</tr>
<tr>
<td>During fever</td>
<td>1 (3%)</td>
<td>9 (30%)</td>
<td>7 (26%)</td>
</tr>
<tr>
<td>PR (sec) (mean ± sd)</td>
<td>160 ± 23</td>
<td>147 ± 24</td>
<td>138 ± 17</td>
</tr>
<tr>
<td>RR (sec)</td>
<td>1</td>
<td>8</td>
<td>7</td>
</tr>
<tr>
<td>QTc V5 (ms) (mean ± sd)</td>
<td>454 ± 4</td>
<td>423 ± 36</td>
<td>418 ± 35</td>
</tr>
<tr>
<td>QTc V2 (ms) (mean ± sd)</td>
<td>454 ± 4</td>
<td>423 ± 36</td>
<td>418 ± 35</td>
</tr>
<tr>
<td>fQRS</td>
<td>31/3 (100%)</td>
<td>1 (3%)</td>
<td>30 (97%)</td>
</tr>
<tr>
<td>Type I in peripheral leads</td>
<td>27/3 (91%)</td>
<td>8</td>
<td>0</td>
</tr>
</tbody>
</table>

106 patients < 19 yrs at diagnosis
→ 16 European Hospitals

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→ 16 European Hospitals

2001-2017

The BRUGADA SYNDROME in CHILDREN study

Mean age 13 ± 5 years old, 42 (76%) male

Symptoms at presentation:
- Sudden death
- Sustained ventricular tachycardia
- Syncope
- Asymptomatic

28% familial screening
58% casual ECG (sport participation)
14% ECG during fever

ECG: fQRS

ECG: VT / SVT (%) Syncope (%) Asymptomatic / (≥ 1) (%)
**ECG**

31/54 (56%) pts with spontaneous Brugada type I ECG → 11 (35%) only during fever

<table>
<thead>
<tr>
<th>ECG</th>
<th>Type I BrEcg</th>
<th>Type II/III BrEcg</th>
<th>Asymptomatic BrEcg</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type I BrEcg</td>
<td>3/3 (100%)</td>
<td>2/8 (25%)</td>
<td>7/28 (26%)</td>
<td></td>
</tr>
<tr>
<td>During fever</td>
<td>1/3 (33%)</td>
<td>2/8 (25%)</td>
<td>7/28 (26%)</td>
<td></td>
</tr>
<tr>
<td>PR ms (mean ± s.d.)</td>
<td>142 ± 23</td>
<td>142 ± 23</td>
<td>139 ± 17</td>
<td>NS</td>
</tr>
<tr>
<td>RR ms</td>
<td>1</td>
<td>2</td>
<td>7</td>
<td>NS</td>
</tr>
<tr>
<td>QTc V5 ms (mean ± s.d.)</td>
<td>415 ± 36</td>
<td>417 ± 33</td>
<td>417 ± 33</td>
<td>NS</td>
</tr>
<tr>
<td>QTc V2 ms (mean ± s.d.)</td>
<td>423 ± 36</td>
<td>418 ± 33</td>
<td>418 ± 33</td>
<td>NS</td>
</tr>
<tr>
<td>fQRS</td>
<td>2/3 (66%)*</td>
<td>0/8 (0%)</td>
<td>1/43 (2%)*</td>
<td>p&lt;0.05</td>
</tr>
</tbody>
</table>

Type 1 in peripheral leads 2/3 (66%)*

31/54 (56%) pts with spontaneous Brugada type I ECG → 11 (35%) only during fever

**GENETIC TEST**

Genetic test performed in 33/54 pts → positive in 17 (52%)

<table>
<thead>
<tr>
<th>GENETIC TEST</th>
<th>SD / VT (3 pts)</th>
<th>Syncope (8 pts)</th>
<th>Asymptomatic (43 pts)</th>
</tr>
</thead>
<tbody>
<tr>
<td>positive</td>
<td>3/3 (100%)</td>
<td>2/8 (25%)</td>
<td>12/24 (50%)</td>
</tr>
<tr>
<td>SCN5A</td>
<td>1 homozygous</td>
<td>2</td>
<td>11</td>
</tr>
<tr>
<td>SCN10A</td>
<td>1 homozygous</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

**F-UP**

Mean f-up of 49±56 months

1 pt VT → LOOP RECORDER + HQ → no ventricular events during f-up

8 pts syncopés → 4 HQ treatment → no recurrences

43 asymptomatic pts → 3 had syncopal episodes: 2 vaso-vagal 1 during fever → HQ

**Take home messages...**

1. The prevalence of Brugada ECG in children is much lower than in adult
2. The 1st manifestation of Brugada in children can be a ventricular arrhythmia
3. Red flags: symptoms at diagnosis type 1 Brugada ECG, fQRS, Type 1 in peripheral leads.
4. Genetic analysis positive in 47-50% (in adults 21%)
5. Prevention: promptly fever treatment, avoid drugs with potential antiarrhythmic effect
Thank you!