Brugada syndrome.
20 years of progress.
Genetic background
Brugada P, Brugada J. 1992:

„Right bundle-branch block, persistent ST-segment elevation in the right precordial leads and sudden cardiac death: a distinct clinical and electrocardiographic syndrome“

J Am Coll Cardiol 1992;20:1391
ECG abnormalities constitute the hallmark of Brugada syndrome. They include repolarization and depolarization abnormalities in the absence of identifiable structural cardiac abnormalities or other conditions or agents known to lead to ST-segment elevation in the right precordial leads."
ST-segment elevation
P wave duration ↑
QRS prolongation, RBBB and LFHB
PR prolongation
SN dysfunction
Atrial arrhythmias
Late potentials (SAECG)
Conduction defect and depolarization abnormality
Repolarization abnormality
ST-segment elevation
J wave accentuation
QT prolongation / QT shortening
Ventricular arrhythmias
Spontaneous coved ST-segment elevation is a marker of malignant ventricular arrhythmias and sudden cardiac death
Right bundle-branch block
Variability of the diagnostic ECG pattern

- Transient normalization and/or conversion to saddleback-type pattern occur in > 95% of the patients during long-term follow-up

- 35% of patients diagnosed with Brugada syndrome do not reveal a spontaneous coved-type ECG during 4-years follow-up

- Only every 4th ECG is spontaneously diagnostic and every 2nd ECG does not display any Brugada-type ST elevation

Eur Heart J 2006;27:593
Diagnostic tools to unmask a type I ECG pattern

→ Administration of a class I sodium channel blocker
→ Superior placement of the right precordial leads
→ ECG monitoring during recovery phase of exercise
Class I drugs to unmask Brugada syndrome

- Diagnosis of individuals with normalized or non-diagnostic ECG
- Identification of family members at risk for the disease

<table>
<thead>
<tr>
<th>Class I drug</th>
<th>Dosage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ajmaline</td>
<td>1 mg/kg over 5 min, IV</td>
</tr>
<tr>
<td>Flecaainide</td>
<td>2 mg/kg over 10 min, IV</td>
</tr>
<tr>
<td>Procainamide</td>
<td>10 mg/kg over 10 min, IV</td>
</tr>
<tr>
<td>Pilsicainide</td>
<td>1 mg/kg over 10 min, IV</td>
</tr>
</tbody>
</table>
ajmaline IV
Ajmaline test in children

pre

post

pre

post
Flecainide test in overt Brugada syndrome
affected family member

non-affected family member

Sens  80.0%
Spec  94.4%
PPV   93.3%
NPV   82.9%
ST-segment elevation in lead aVL
after ajmaline IV
after ajmaline IV
“Aizawa pattern“, 1996
Accentuation of the J wave

Lidocaine IV
Electrical cardioversion
Conduction abnormalities in myotonic dystrophy type I
Pectus excavatum

Mediastinal tumor

baseline ajmaline

PACE 1999;22:1264
Conduction abnormalities

- atrial standstill
- sinoatrial conduction time $\uparrow$
- slowed atrial conduction
- infrahisian conduction delay

$\rightarrow$ most frequently observed in patients linked to SCN5A mutations

- SA block III$^\circ$
- SA block II$^\circ$, cSNRT $\uparrow$
- $\rightarrow$ P-wave duration $\uparrow$, PR $\uparrow$
- $\rightarrow$ PR $\uparrow$, QRS duration $\uparrow$, RBBB, LFHB

PR 270 ms
Overlap syndromes

Long-QT 3

Familial conduction disease (Lev-Lenègre disease)

Circ Res 1999;85:1206

Circulation 2001;104:3081
Atrial arrhythmias

- Incidence up to 20%, particularly paroxysmal AFib and AFLutter
- Common cause of frequent inappropriate ICD shocks
- First manifestation of Brugada syndrome
Polymorphic ventricular tachycardia and VF
Monomorphic ventricular tachycardia → rare in Brugada syndrome
Fragmented QRS in Brugada syndrome

A. Filter 0-150 Hz

4 ics
3 ics
2 ics

V1

V2

V3

B. Filter 0-25 Hz
Fragmented QRS in Brugada syndrome

A. f-QRS (+)  LP (+)
B. f-QRS (+)  LP (-)
C. f-QRS (-)  LP (+)
D. f-QRS (-)  LP (-)
Fragmented QRS in Brugada syndrome

A  55 y.o.
V1

B  56 y.o.
V2

C  57 y.o.
V3

D  58 y.o.
Fragmented QRS in Brugada syndrome
Proposed pathophysiological mechanisms of Brugada syndrome.

**Slow conduction**

**Repolarization abnormality**

**Development abnormality**


J Cardiovasc Electrophysiol 2001;12:268

Heart Rhythm 2007;4:359
Management of patients with Brugada syndrome

Coved-type I ECG spontaneously or after class I AAD

<table>
<thead>
<tr>
<th>Aborted SCD or syncope</th>
<th>Asymptomatic</th>
</tr>
</thead>
<tbody>
<tr>
<td>ICD</td>
<td>ICD Follow-up Follow-up</td>
</tr>
</tbody>
</table>

Spontaneous coved-type ECG

EP study

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