A Rare Case of Atrial Cardiomyopathy with Progressive Cardiac Conduction Defect (PCCD) Successfully Treated with Cardiac Resynchronization Therapy-Defibrillator (CRT-D)

KHRS (June 22, 2019)
Dr. Jemelee Co Hernandez
39/M Caucasian

4 years ago:
- Palpitations, dizziness, near-syncope
- TTE: Normal EF
- CT angiogram: Normal coronaries

Family History
- PPM – Father at 40 y/o
- Cardiomyopathy and Heart transplant – Father at 60 y/o

Figure 1a. Baseline 12 L ECG
39/M Caucasian

4 years ago:
Palpitations, dizziness, near-syncope
TTE: Normal EF
CT angiogram: Normal coronaries

Meds: Bisoprolol, ASA
DC Cardioversion x 2

Figure 1b. 12 L ECG post cardioversion
4 years ago:

EP study: complex fractionated atrial electrograms

s/p RF PV isolation, CFAE ablation, CTI ablation
Amiodarone and Dabigatran

Figure 2. Intervals post ablation
39/M Caucasian
2 years later
Palpitations, decreased effort tolerance

24 Holter
AV dissociation; PVC 2%; NSVT
TTE: Eccentric LVH with global hypokinesia and depressed SF (EF 32%)
Dilated LA (LAVI 52; 5.4 cm) and RA (5.1 cm);
Moderate MR and moderate TR
Coronary Angiogram: Normal

Figure 3. 12 L ECG on follow-up
Figure 6. Cardiac MRI: mid wall late gadolinium enhancement

- Vertical Long axis
- Short axis view
- 4 chamber view
Figure 7a.
Electrophysiologic Study

QRS duration 140 ms
RVSP 680 ms
RVERP 600/300 ms

AH 408 ms
HV 50 ms

AH 534 ms
HV 50 ms

AH 612 ms
HV 50 ms
Figure 7b. Electrophysiologic Study with 1 mg Isuprel
Figure 9. Fluoroscopic views post CRT-D
<table>
<thead>
<tr>
<th>10.23.2018 INTERROGATION</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>A PACING</strong></td>
</tr>
<tr>
<td><strong>V PACING</strong></td>
</tr>
<tr>
<td><strong>PVC</strong></td>
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</tbody>
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<table>
<thead>
<tr>
<th><strong>CAPTURE</strong></th>
<th><strong>SENSE</strong></th>
<th><strong>IMPEDANCE</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>1.25V @ 0.5 ms</td>
<td>-</td>
</tr>
<tr>
<td>RV</td>
<td>1.0V @ 0.5 ms</td>
<td>7.1mV</td>
</tr>
<tr>
<td>LV</td>
<td>1.25V @ 0.5 ms</td>
<td></td>
</tr>
<tr>
<td>HV</td>
<td></td>
<td></td>
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</tbody>
</table>
39/M Caucasian

4 years ago:
Persistent AF
s/p RF PV isolation, CFAE ablation, CTI ablation

2 years later
NICMP (EF 32%, Supraventricular AV block, CLBBB QRS 140 ms) s/p CRT-D

On Followup
2 years post CRT-D:
NYHA FC II
TTE: Normal LV size with mildly impaired SF (EF 48%)
Dilated LA (LAVI 52, 4.8 cm)
and RA
Mild to moderate MR

Figure 10. 12L ECG ON FOLLOWUP (2 YEARS LATER) : LV to RV 30 ms
ATRIAL CARDIOMYOPATHY

Etiological Factors

Mechanical Dysfunction
Procoagulant State

Cardiomyopathic Changes

Electrical Dysfunction
Fibrosis

Strokes

AF Progression

Management Implications

Persistent Atrial Fibrillation
Atrial Cardiomyopathy

35/M

Family History of PPM and Cardiomyopathy requiring Heart Transplant

Supraventricular AV block
LBBB (QRSd 140 ms)
Nonsustained VT

Decline in LVEF (32%)
NICMP
Progressive cardiac conduction disease (PCCD)

- **Diagnosed** in the presence of:
  - unexplained progressive conduction abnormalities in young (<50 years) individuals
  - with structurally normal hearts
  - in the absence of skeletal myopathies
  - especially if there is a family history of PCCD

HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. Europace. 2013
PROGRESSIVE CARDIAC CONDUCTION DISEASE IS A RARE BUT POTENTIALLY LIFE-THREATENING DISORDER!

STRUCTURAL

CONGENITAL HEART DISEASE
CARDIOMYOPATHY
EXTRACARDIAC MANIFESTATIONS
FUNCTIONAL OR ISOLATED

LMNA

Differentiation gene expression

Nucleus

NKX2.5

TBX5

HCN4

SCN5A

SCN1B

KCNK17

NCX

Ca²⁺

Na⁺

K⁺

Na⁺

K⁺

Na⁺

K⁺

K⁺

K⁺

Ca²⁺

Ca²⁺

Ca²⁺

Ca²⁺

SR

Ryr2

CASQ2

ANK2

NPPA

KCNJ2

TRPM4

CACNA1D
Dilated cardiomyopathy
Sudden cardiac death
End-stage heart failure
Progressive cardiac conduction disease (PCCD)

• Pacemaker implantation is recommended in:
  a) Intermittent or permanent third-degree or high-grade AV block. (Class I)
  b) Symptomatic Mobitz I or II second-degree AV block. (Class I)

HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. Europace. 2013
Progressive cardiac conduction disease (PCCD)

- **Pacemaker implantation** is recommended in:
  a) presence of bifascicular block with or without first-degree AV block. *(Class IIa)*

- **ICD implantation** can be useful in
  a) with a mutation in the lamin A/C gene with left ventricular dysfunction and/or nonsustained VT. *(Class IIa)*

HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes. Europace. 2013
Table 2. Characteristics of 12-Lead ECGs

<table>
<thead>
<tr>
<th>ECG measures</th>
<th>First ECG</th>
<th>Follow-up ECG</th>
<th>Temporal change*</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart rate (beats/min)</td>
<td>68.6±14.1</td>
<td>66.1±13.1</td>
<td>−2.7±15.5</td>
<td>0.01</td>
</tr>
<tr>
<td>PR interval (ms)</td>
<td>165.0±27.4</td>
<td>178.5±34.0</td>
<td>13.9±20.2</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>QRS duration (ms)</td>
<td>118.9±22.8</td>
<td>142.9±15.4</td>
<td>24.0±22.5</td>
<td>&lt;0.0001</td>
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QRS morphology

<table>
<thead>
<tr>
<th></th>
<th>First ECG</th>
<th>Follow-up ECG</th>
<th>Temporal change*</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal (n, %)</td>
<td>143, 31.2</td>
<td>0, 0</td>
<td>−31.2</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>RBBB (n, %)</td>
<td>186, 40.6</td>
<td>297, 64.8</td>
<td>24.2</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>RBBB with LAH (n, %)</td>
<td>13, 2.8</td>
<td>37, 8.1</td>
<td>5.3</td>
<td>0.0004</td>
</tr>
<tr>
<td>LBBB (n, %)</td>
<td>31, 6.8</td>
<td>47, 10.3</td>
<td>3.5</td>
<td>0.06</td>
</tr>
<tr>
<td>NSIVCD (n, %)</td>
<td>85, 18.6</td>
<td>76, 16.6</td>
<td>−2.0</td>
<td>0.43</td>
</tr>
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Figure 1. Kaplan-Meier survival analysis for a hospitalization for heart failure.

Figure 2. Kaplan-Meier survival analysis for cardiovascular mortality. (A) QRS-duration incremental rate <5 ms/year (Group 1) vs. ≥5 ms/year (Group 2). (B) PR-interval incremental rate <4 ms/year (Group 1) vs. ≥4 ms/year (Group 2). Numbers of subjects at risk are shown under each graph.

Temporal incremental rate of QRS duration was independently associated with increased risk of HF hospitalization and cardiovascular mortality in those with PCCD
2) LBBB with QRS duration 120–150 ms. CRT is recommended in chronic HF patients and LVEF ≤35% who remain in NYHA functional class II, III and ambulatory IV despite adequate medical treatment.
### CRT-D VS CRT-P

<table>
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<tr>
<th>Factors favouring CRT-P</th>
<th>Factors favouring CRT-D</th>
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<tbody>
<tr>
<td>Advanced heart failure</td>
<td>Life expectancy &gt;1 year</td>
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<tr>
<td>Severe renal insufficiency or dialysis</td>
<td>Stable heart failure, NYHA II</td>
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<tr>
<td>Other major co-morbidities</td>
<td>Ischaemic heart disease (low and intermediate MADIT risk score)</td>
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<tr>
<td>Frailty</td>
<td>Lack of comorbidities</td>
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<td>Cachexia</td>
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2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy
CONCLUSION

• A rare disease, which can present with arrhythmias.

• High index of suspicion with regular follow-up are necessary particularly in young patients with a strong family history

• Could develop QRS prolongation and progressive LV dysfunction, for which CRT-D can be the best treatment option
• THANK YOU!