Relationship between aetiology and left ventricular systolic dysfunction in hypertrophic cardiomyopathy

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Causes of heart failure in AFD

- Cardiomyopathy (diastolic and systolic dysfunction)
- Right ventricular pacing
- Valve disease
- Ischaemic disease (microvascular)
AFD cardiomyopathy on imaging

- Mild-moderate concentric LV hypertrophy
- Hypertrophied papillary muscles
- Hypertrabeculation
- Posterior wall brightness
- Diastolic dysfunction
Cardiac Events Pre ERT

![Bar chart showing the percentage of patients with CV events by age and gender.](chart)

- **Males (N=83)**
- **Females (N=54)**

**Age at First CV Event (years)**
- 0 to <25: n=5 (M), n=1 (F)
- 25 to <35: n=8 (M), n=1 (F)
- 35 to <45: n=21 (M), n=7 (F)
- 45 to <55: n=39 (M), n=19 (F)
- 55 to <65: n=20 (M), n=7 (F)
- >=65: n=6 (M), n=3 (F)

Patel et al. JACC 2011
• Can we predict which patients will develop heart failure and how soon?
Predictors/Associations with cardiac events

- Age
- MSSI
- Renal failure
- QRS duration
- Fractional shortening/Ejection fraction
- Diastolic dysfunction
- Strain
- Late gadolinium
- T1
- BNP
- TnI
- Lyso GB3
- Mutation
Mutations

Arends et al. JASN 2017
LV mass index in untreated patients

Arends et al. JASN 2017
218 patients
7.1 years follow-up

<table>
<thead>
<tr>
<th></th>
<th>&quot;Classical&quot; Mutation</th>
<th>&quot;Cardiac&quot; Mutation</th>
</tr>
</thead>
<tbody>
<tr>
<td>n</td>
<td>149</td>
<td>58</td>
</tr>
<tr>
<td>NYHA 3/4</td>
<td>1.58</td>
<td>1.75</td>
</tr>
<tr>
<td>AF</td>
<td>1.2</td>
<td>0.56</td>
</tr>
<tr>
<td>Device Implant</td>
<td>0.98</td>
<td>1.4</td>
</tr>
<tr>
<td>Cardiac Death</td>
<td>0.5</td>
<td>0.54</td>
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<tr>
<td>Composite</td>
<td>2.23</td>
<td>2.77</td>
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</tbody>
</table>

Incidence per 100 patient-years

No significant differences between Classical and Cardiac mutations
Age & MSSI score independent predictors of heart failure

Patel et al. Heart 2015
Late Gadolinium

LGE may precede LV hypertrophy

Moon et al. EHJ 2003
Medium-term Effects of ERT

Wiedemann et al. Circ 2009
LV Hypertrophy
Fibrosis
Systolic dysfunction
Advanced Heart Failure
T1 mapping

Sado et al. Circ cardiovasc imaging 2013
T1 mapping

Sado et al. Circ cardiovasc imaging 2013
T1 mapping

Sado et al. Circ cardiovasc imaging 2013
Strain
Strain

Gruner et al. Echo 2012
Strain

Gruner et al. Echo 2012
Advanced Heart Failure

T2 Strain

Hypertrophy
Fibrosis
Arrhythmia

Systolic dysfunction

Advanced Heart Failure