A hypertrophic cardiomyopathy patient with intermediate sudden cardiac death risk: What do the Guidelines say?

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Mannheim

Buenos Aires - Argentina
ESC at the 41st Congress of Argentine Society of Cardiology
Thursday 15 - Saturday 17 October 2015
HCM with intermediate SCD

42 yr old man
Abnormal ECG
No FH of HCM / sudden death
asymptomatic
HCM with intermediate SCD
### HCM with intermediate SCD

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Exercise test:</td>
<td>normal BP response</td>
</tr>
<tr>
<td>48 h Holter monitoring:</td>
<td>no VBP, no AF</td>
</tr>
<tr>
<td>Echo:</td>
<td>normal LV-function</td>
</tr>
<tr>
<td></td>
<td>septum 13 mm</td>
</tr>
<tr>
<td></td>
<td>LA size 46 mm</td>
</tr>
</tbody>
</table>
HCM with intermediate SCD

What is your therapeutic advice?

A. No specific therapy – yearly follow-up
B. Betablocker and follow-up
C. Prescription of NOAC - follow-up
D. Prophylactic treatment prescribing amiodarone
E. Implant an ICD for primary prevention
HCM with intermediate SCD

Further risk stratification?

A. Use the HCM risk prediction model?
B. Genetic testing?
C. MRI imaging?
HCM with intermediate SCD

HCM Risk-SCD Calculator

Age [ ] Years

Maximum LV wall thickness [ ] mm

Left atrial size [ ] mm

Max LVOT gradient [ ] mmHg

Family History of SCD

- No
- Yes

Non-sustained VT

- No
- Yes

Unexplained syncope

- No
- Yes

Risk of SCD at 5 years (%): [ ]

ESC recommendation: [ ]

http://doc2do.com/hcm/webHCM.html
## HCM Risk-SCD Calculator

### Age
- **42** Years
  - Age at evaluation

### Maximum LV wall thickness
- **13** mm
  - Transthoracic Echocardiographic measurement

### Left atrial size
- **46** mm
  - Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation

### Max LVOT gradient
- **2** mmHg
  - The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernoulli equation: Gradient= $4V^2$, where $V$ is the peak aortic outflow velocity

### Family History of SCD
- **No**
- **Yes**
  - History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).

### Non-sustained VT
- **No**
- **Yes**
  - 3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.

### Unexplained syncope
- **No**
- **Yes**
  - History of unexplained syncope at or prior to evaluation.

### Risk of SCD at 5 years (%): **1.5**

**ESC recommendation:** ICD generally not indicated **

**ICD not recommended unless there other clinical features that are of potential prognostic importance and when the likely benefit is greater than the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.
HCM with intermediate SCD
HCM with intermediate SCD
HCM with intermediate SCD

Do you these MRI findings alter your treatment strategy?

A. Yes
B. No
HCM with intermediate SCD

Would you implant an ICD?

A. Yes
B. No
Cardiomyopathy

- Sudden death
- Heart Failure
- AF Thromboembolism
A novel clinical risk prediction model for sudden cardiac death in hypertrophic cardiomyopathy (HCM Risk-SCD)

Constantinos O’Mahony¹, Fatima Jichi², Menelaos Pavlou⁸, Lorenzo Monserrat³, Aristides Anastasakis⁴, Claudio Rapezzi⁵, Elena Biagini⁵, Juan Ramon Gimeno⁶, Giuseppe Limongelli⁷, William J. McKenna¹, Rumana Z. Omar²⁸ and Perry M. Elliott¹*, for the Hypertrophic Cardiomyopathy Outcomes Investigators

Eur Heart J. 2014; 35: 2010-2020
HCM Risk-SCD Calculator

Age [Years] Age at evaluation

Maximum LV wall thickness [mm] Transthoracic Echocardiographic measurement

Left atrial size [mm] Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation

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Risk of SCD at 5 years (%)

Recommendations

Reset
# A novel clinical risk prediction model for sudden cardiac death in hypertrophic cardiomyopathy (HCM Risk-SCD)

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<table>
<thead>
<tr>
<th></th>
<th>N</th>
<th>SCD</th>
<th>%</th>
<th>Annual SCD</th>
</tr>
</thead>
<tbody>
<tr>
<td>≥ 2 RF</td>
<td>412.0</td>
<td>32.0</td>
<td>7.8</td>
<td>1.6</td>
</tr>
<tr>
<td>1 RF</td>
<td>1074.0</td>
<td>27.0</td>
<td>2.5</td>
<td>0.5</td>
</tr>
<tr>
<td>0 RF</td>
<td>1580.0</td>
<td>25.0</td>
<td>1.6</td>
<td>0.3</td>
</tr>
</tbody>
</table>
Prevention of Sudden Cardiac Death

Recommendations for ICD in each risk category take into account not only the absolute statistical risk, but also the age and general health of the patient, socio-economic factors and the psychological impact of therapy.
Take Home Messages

• Sudden Death is uncommon in patients with HCM
• The risk of SCD can be estimated using simple non-invasive assessment
• Decisions on treatment are determined by absolute risk but also societal and economic considerations
• Future refinement of risk prediction should be based on robust modelling and not “expert opinion” alone.