Cardiovascular Magnetic Resonance in Hypertrophic Cardiomyopathy

Martin S. Maron, MD, FACC, FHRS
Medical Director
Hypertrophic Cardiomyopathy Center
Tufts Medical Center
Boston, MA

Chanin T. Mast HCM Center
Morristown Medical Center
Morristown, NJ
DISCLOSURE

• Gadolinium is FDA off-label use for CV imaging
Strongest Risk Factors:
- Cardiac arrest/Sus. VT.
- Familial Hx of SD
- Syncope
- Multiple-repetitive NSVT
- ↓BP — exercise
- Massive LVH ≥30 mm

ICD
Challenges of Risk Stratification in HCM

- Three Risk Factors (2%)
- Two Risk Factors (10%)
- One Risk Factor (33%)
- Zero Risk Factors (55%)

~50% of Clinically Identified HCM Pts At Increased Risk For Sudden Death

0.5%/year

The “Grey Area” Of Risk Stratification

~40% of HCM Sudden Deaths
Foci for Ventricular Arrhythmias?
Holter NSVT and Presence of LGE

Adabag et al.  
n=177

Rubinstein et al.  
(Mayo)  
n=220

p<0.001

LGE (+)  
LGE (-)
LGE for Prognosis in HCM Multicenter Study

- Tufts Medical Center, Boston, MA
- Minneapolis Heart Institute
- Toronto General Hospital, Canada
- Azzendia Carregia, Florence, Italy
- Bologna, Italy
- Pisa, Italy
- Rome, Italy
- PERFUSE CMR Core Laboratory
Relation Between Sudden Death and Extent of LGE in 1293 HCM Patients

Freedom from Sudden Death

Follow-up (years)

LGE (-)
LGE < 10%
LGE 10-19%
LGE ≥20%

Chan R, Maron MS et al. in Review
Sudden Death Event Rates in HCM Patients without Conventional Risk Factors

<table>
<thead>
<tr>
<th>% LGE</th>
<th>Adjusted HR</th>
<th>Est. 5-year Event Rate(%)</th>
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</thead>
<tbody>
<tr>
<td>0%</td>
<td>1.0</td>
<td>2.5</td>
</tr>
<tr>
<td>5%</td>
<td>1.3</td>
<td>3.2</td>
</tr>
<tr>
<td>10%</td>
<td>1.6</td>
<td>4.0</td>
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<tr>
<td>15%</td>
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<tr>
<td>20%</td>
<td>2.6</td>
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<td>25%</td>
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<tr>
<td>30%</td>
<td>4.2</td>
<td>10.0</td>
</tr>
<tr>
<td>40%</td>
<td>6.7</td>
<td>15.5</td>
</tr>
</tbody>
</table>

Chan R, Maron MS et al. *in Review*
Improvement of Sudden Death Prediction with Addition of %LGE to Risk Model
When Extensive LGE Counts

32 year-old man with Sudden Death
Asymptomatic (Max LV WT = 26 mm)
Normal Ejection Fraction
No Traditional Risk Factors RF

LGE = 22% of the LV Mass
Strongest 1° Risk Factors:
- Familial Hx of SD
- Syncope
- Multiple-repetitive NSVT
- ↓BP — exercise
- Massive LVH ≥30 mm

Chan R, Maron MS et al. *in Review*
Risk Factors

Risk of SD is about the **Amount** of LGE... **Not** its Pattern or Location
RV Insertion Point LGE in HCM = Expanded Extracellular space

Not Likely Replacement Fibrosis

Roberts, WC AJC 1992
Sudden Death Risk in Patients with RV Insertion Point LGE Only

Freedom from Adverse HCM-related Events

Follow-up (years)

p = 0.7
LV Apical Aneurysm with Thrombus

Maron MS et al. Circulation 2009
Cardiovascular Event Rate = 11%/yr

Patients with LVAA (n=28)

- Alive/ Clinically Stable (n = 16)*
- Adverse Events (n = 12)
  - Sudden Death (2)*
  - Aborted Cardiac Arrest (2)†
  - Progressive Heart Failure/ Death (5)†
  - Appropriate ICD Discharge (3)*

- non-fatal embolic stroke (1)
- non-fatal embolic stroke (1)

Maron MS et al. Circulation 2009
CMR in HCM

- Particularly well suited to characterize the diverse phenotypic expression of this complex disease...

- Superior to echo for HCM diagnosis...ie., anterolateral wall, apex

- Management strategies for invasive septal reduction therapy...ie., mitral valve, anomalous insertion of papillary muscles

- Sudden Death Risk Prediction...Extensive LGE (≥15% of LV) identifying a novel subgroup of HCM patients at increased risk for SD that would not be considered without CMR and may now be candidates for ICD

- Absence of LGE associated with low risk....may serve to influence decision-making against ICD implants in “grey-zone” situations