Late Gadolinium Enhancement and Prognosis of Hypertrophic Cardiomyopathy
Hajime Sakuma, MD, PhD

Hypertrophic cardiomyopathy (HCM) is the most common genetic cardiac disease, with a prevalence of 1:300 in a cohort Japanese workers who underwent echocardiographic screening.1,2 Survival curve for the entire HCM population is not significantly different from that for the general population after adjusting for age and sex.3 However, because of the substantial genetic heterogeneity of this disease, a small but significant number of HCM patients are at risk of sudden cardiac death (SCD) and progressive heart failure (HF). Consequently, accurate identification of patients at higher risk of adverse outcome is required. Currently, several risk factors, including a family history of HCM-related SCD, unexplained syncope, massive LV hypertrophy, nonsustained VT and abnormal blood pressure response during exercise, are used for risk stratification. However, the positive and negative predictive values of the risk assessment using these traditional risk markers for predicting SCD are limited.

Cardiac MR (CMR) has emerged as an important imaging technique that allows for excellent delineation of the ventricular wall in any location of the heart.4 In addition, myocardial fibrosis or expansion of the extracellular volume (ECV) caused by myocyte disarray can be visualized as late gadolinium enhancement (LGE) on contrast-enhanced CMR5 (Figure). There is increasing interest in the ability of LGE to stratify HCM patients according to their risk of SCD and other adverse outcomes.6

In this issue of the Journal, Hen et al investigate the presence and extent of LGE in 345 HCM patients who were followed up for a mean duration of 22 months after CMR for arrhythmic events including SCD, aborted SCD, incessant VT/VF and inappropriate ICD discharge, and for the cardiac failure events of unscheduled hospitalization for HF.7 Cardiovascular events are defined as stroke, acute myocardial infarction and the arrhythmias of hypertrophic cardiomyopathy (HCM) is the most common genetic cardiac disease, with a prevalence of 1:300 in a cohort Japanese workers who underwent echocardiographic screening.1,2 Survival curve for the entire HCM population is not significantly different from that for the general population after adjusting for age and sex.3 However, because of the substantial genetic heterogeneity of this disease, a small but significant number of HCM patients are at risk of sudden cardiac death (SCD) and progressive heart failure (HF). Consequently, accurate identification of patients at higher risk of adverse outcome is required. Currently, several risk factors, including a family history of HCM-related SCD, unexplained syncope, massive LV hypertrophy, nonsustained VT and abnormal blood pressure response during exercise, are used for risk stratification. However, the positive and negative predictive values of the risk assessment using these traditional risk markers for predicting SCD are limited.

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LGE and Prognosis of HCM

Table. Adverse Events in the LGE-Positive and LGE-Negative HCM Groups in 4 Major Previous Studies

<table>
<thead>
<tr>
<th>Study</th>
<th>n</th>
<th>Follow-up (months)</th>
<th>LGE analysis</th>
<th>LGE+ (%)</th>
<th>Cardiac death</th>
<th>SCD/aborted SCD</th>
<th>Heart failure death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maron et al 2008</td>
<td>202</td>
<td>22</td>
<td>&gt;6SD</td>
<td>55%</td>
<td>2/115 (1.7%)</td>
<td>5/429 (2.2%)</td>
<td>3/634 (0.8%)</td>
</tr>
<tr>
<td>Bruder et al 2010</td>
<td>220</td>
<td>36</td>
<td>&gt;2SD</td>
<td>67%</td>
<td>15/148 (10.1%)</td>
<td>10/148 (6.8%)</td>
<td>5/148 (3.4%)</td>
</tr>
<tr>
<td>O'Hanlon et al 2010</td>
<td>217</td>
<td>37</td>
<td>FWHM</td>
<td>62%</td>
<td>8/136 (6.0%)</td>
<td>8/239 (3.3%)</td>
<td>3/136 (2.2%)</td>
</tr>
<tr>
<td>Rubinshtein et al 2010</td>
<td>424</td>
<td>43</td>
<td>Visual</td>
<td>56%</td>
<td>6/239 (2.5%)</td>
<td>8/239 (3.3%)</td>
<td>2/239 (0.8%)</td>
</tr>
<tr>
<td>Pooled data</td>
<td>1,063</td>
<td>37</td>
<td></td>
<td>60%</td>
<td>31/634 (4.9%)</td>
<td>25/634 (3.9%)</td>
<td>13/634 (2.1%)</td>
</tr>
</tbody>
</table>

HCM, hypertrophic cardiomyopathy; LGE, late gadolinium enhancement; SCD, sudden cardiac death.

Table 1. Adverse Events in the LGE-Positive and LGE-Negative HCM Groups in 4 Major Previous Studies

- Table 1 presents data from four major previous studies investigating the prognostic value of late gadolinium enhancement (LGE) in hypertrophic cardiomyopathy (HCM). The table includes studies by Maron et al., Bruder et al., O’Hanlon et al., and Rubinshtein et al., with follow-up periods ranging from 22 to 43 months. The data show the percentage of patients with cardiac death, sudden cardiac death/aborted sudden death, and heart failure death in the LGE-positive and LGE-negative groups.

- The studies include a total of 202 and 220 HCM patients, respectively, with follow-up times of 22 and 36 months. LGE was significant in the LGE-positive group (2.5%/year) and LGE-negative group (0%/year, P=0.037). The difference in the cardiac failure event rate between the two groups did not reach statistical significance. This is the first study to clearly demonstrate the prognostic value of LGE in a large number of Japanese patients with HCM.

- The table also shows that the presence of LGE was significantly associated with SCD or appropriate ICD discharge and progression of HF. LGE was observed in 73% of the HCM patients, with a significantly higher prevalence in the event-positive group (28/29, 96.6%) than in the event-negative group (224/316, 70.9%). These findings support the use of LGE in the assessment of patients with HCM.

- The data from these studies highlight the importance of LGE in predicting future adverse events in HCM, suggesting that the presence of LGE could be a useful tool for risk stratification.

References

5. Moon J, Hong YJ, Kim YJ. Extent of late gadolinium enhancement.

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