Hypertrophic cardiomyopathy (HC) has become a contemporary and treatable genetic heart disease, now with disease-related mortality reduced to as low as 0.5% per year, based largely on more effective risk stratification and the use of the implantable cardioverter-defibrillator for primary prevention of sudden death. This paradigm change in the natural history of HC has caused us to reconsider the overall mortality risk in this disease. We interrogated the databases of 2 HC referral centers, Minneapolis Heart Institute and Tufts Medical Center. Of 1,902 consecutive patients evaluated between 1992 and 2013, 1,653 patients (87%) have survived to the end of follow-up and 249 patients (13%) have died. Most deaths (178 of 249; 72%) were unrelated to HC, commonly because of cancer and predominantly in older patients. Non-HC mortality was significantly more common in adults presenting ≥60 years and least common in the youngest patients aged <30 years (p <0.001). Notably, deaths from non-HC causes substantially exceeded HC-related causes by 2.6-fold (p <0.001). In conclusion, only about 25% of patients with HC ultimately died of their disease, including predominantly those who were <30 years of age. These data allow patients with HC to develop a more realistic and reassured perception of their disease. © 2016 Elsevier Inc. All rights reserved. (Am J Cardiol 2016;117:434–435)
The present data, however, clearly dispute such patient intuitions by showing that risk for death in adult patients with HC is significantly greater due to diseases other than HC. Indeed, it is very possible for adult patients in low HC-related risk groups to become distracted by HC and neglect preventive measures for other potentially lethal conditions such as cancer or coronary artery disease. In contrast, not unexpectedly, in children, adolescents, and young adults aged <30 years with HC, mortality is predominantly due to HC, given that deaths from other disorders are rare in this age group.

All-cause mortality in HC exceeds that in the general population likely because deaths due to HC are part of the calculated total mortality. However, adverse interaction between HC and other disease processes that potentially increase lifetime risk cannot be absolutely excluded.

An important message often lost to patients with HC is that their disease is not ultimately lethal, but rather consistent with extended (if not normal) longevity, given that adverse clinical consequences and complications can now be anticipated and accompanied by effective treatment options.

In conclusion, although HC has been cited as a highly visible cause of SD in the young, paradoxically it is uncommonly responsible for mortality in established HC patient cohorts. Only about 25% of patients with HC ultimately die of their disease, and indeed, mortality in adult patients with HC is more frequently due to other organ system diseases. These insights empower many patients to adopt a more realistic and reassured perception of their disease and promote the importance of surveillance and preventive strategies for non-HC conditions which may harbor greater potential for morbidity and mortality.

Table 1
Survival and mortality outcome in 1,902 patients with HC, stratified according to age at presentation

<table>
<thead>
<tr>
<th>Age at Presentation (years)</th>
<th>No. Patients</th>
<th>Survived</th>
<th>All Deaths</th>
<th>Died</th>
<th>Died HC</th>
<th>Ratio†</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;30</td>
<td>474</td>
<td>452 (95%)</td>
<td>22 (5%)</td>
<td>4/22 (18%)</td>
<td>18/22 (82%)</td>
<td>4.6</td>
</tr>
<tr>
<td>30-59</td>
<td>1000</td>
<td>918 (92%)</td>
<td>82 (8%)</td>
<td>42/82 (51%)</td>
<td>40/82 (49%)</td>
<td>1.04</td>
</tr>
<tr>
<td>≥60</td>
<td>428</td>
<td>283 (66%)</td>
<td>145 (34%)</td>
<td>132/145 (92%)</td>
<td>13/145 (8%)</td>
<td>11.5</td>
</tr>
<tr>
<td>Totals</td>
<td>1902</td>
<td>1653 (87%)*</td>
<td>249 (13%)</td>
<td>178/249 (72%)</td>
<td>71/249 (28%)</td>
<td>2.6</td>
</tr>
</tbody>
</table>

* Includes aborted SD and heart failure deaths: by implantable defibrillators (n = 68); heart transplant (n = 31); and out-of-hospital defibrillation/therapeutic hypothermia (n = 29).
† Comparison of HC and non-HC deaths.

The authors have no conflicts of interest to disclose.