Patients with hypertrophic cardiomyopathy (HCM) are at increased risk for sudden cardiac death (SCD), mostly caused by ventricular arrhythmias. SCD may occur as the initial presentation of HCM, often in asymptomatic or mildly symptomatic patients. In fact, HCM is the most frequent cause of SCD in young people, including trained athletes. Implantable cardioverter defibrillator (ICD) therapy may effectively terminate potentially life-threatening ventricular arrhythmias, thereby preventing SCD and prolonging life. Still, ICD therapy is not without risk, because inappropriate interventions and device-related complications may occur.

**Background**—Previous observational studies demonstrated that patients with hypertrophic cardiomyopathy at risk for sudden cardiac death (SCD) may benefit from implantable cardioverter defibrillator (ICD) therapy. A complete overview of outcome and complications after ICD therapy is currently not available. This study pools data from published studies on outcome and complications after ICD therapy in patients with hypertrophic cardiomyopathy.

**Methods and Results**—A PubMed database search returned 27 studies on 16 cohorts reporting outcome and complications after ICD therapy in patients with hypertrophic cardiomyopathy. In case of >1 publications on a particular cohort, the publication with the largest number of patients was included in the meta-analysis. ICD interventions, complications, and mortality rates were extracted, pooled, and analyzed. There were 2190 patients (mean age, 42 years; 38% women), most of whom (83%) received an ICD for primary prevention of SCD. Risk factors for SCD were left ventricular wall thickness ≥30 mm (20%), family history of SCD (43%), nonsustained ventricular tachycardia (46%), syncope (41%), and abnormal blood pressure response (25%). During the 3.7-year follow-up, the annualized cardiac mortality rate was 0.6%, the noncardiac mortality rate was 0.4%, and the appropriate ICD intervention rate was 3.3%. The annualized inappropriate ICD intervention rate was 4.8% and the annualized ICD-related complication rate was 3.4%.

**Conclusions**—This meta-analysis demonstrates a low cardiac and noncardiac mortality rate after ICD therapy in patients with hypertrophic cardiomyopathy. Appropriate ICD intervention occurred at a rate of 3.3%/year, thereby, most probably, preventing SCD. Inappropriate ICD intervention and complications are not uncommon.

**Key Words:** hypertrophic cardiomyopathy implantable cardioverter defibrillator sudden cardiac death prognosis complications

**Clinical Perspective on p 559**

Previous observational studies have reported on the use of ICD therapy for primary and secondary preventions of SCD in HCM. A complete overview of outcome and complications after ICD therapy in patients with HCM at risk for SCD is currently not available. The goal of this analysis was to pool the individual studies in an effort to examine the precise rate of cardiac and noncardiac mortality, appropriate and inappropriate interventions, and complications. This knowledge may aid clinical decision making and counseling in patients with HCM at increased risk for SCD considered for ICD therapy.

**Methods**

**Study Design**

This systematic review and meta-analysis included all available original studies reporting clinical outcome and complications in patients with HCM who underwent ICD implantation. Studies that did not provide data on outcome or complications and review manuscripts were excluded. Studies focusing on SCD in patients with HCM without ICD were excluded.

**Literature Search**

The online MEDLINE database was searched for literature in March 2012 using PubMed (National Center for Biotechnology Information, US National Library of Medicine, Bethesda, MD). The search strategy...
was hypertrophic cardiomyopathy and defibrillator. No time restriction for publication dates was used. All titles and abstracts of the articles were evaluated. After exclusion based on the title and abstract, full articles were evaluated, and articles meeting the inclusion criteria were identified. In addition, a manual search of the reference lists of the identified studies was performed, and references were evaluated using the same inclusion and exclusion criteria.

Data Extraction
Selected studies were reviewed and relevant patient characteristics, known risk factors for SCD, and follow-up duration were registered. Extracted outcome parameters were as follows: cardiac mortality, noncardiac mortality, heart transplant, appropriate ICD intervention, inappropriate ICD intervention, and complications, including lead malfunction, infection, lead displacement, psychological complication, and total complications. The outcome parameter total complications included all reported ICD-related complications, except inappropriate ICD intervention; this parameter was registered separately. No time restriction for complications was used; both early and late complications were included in the analysis. Studies with overlapping data were identified, and in cases of apparent serial reporting of a particular patient cohort, only the publication with the largest number of patients was included in the meta-analysis. However, all serial publications on a particular cohort were registered and tabulated.

Statistical Analysis
Statistical analysis was performed using Microsoft Excel 2010 (Microsoft Corporation, Redmond, WA) and SPSS version 15.0 (SPSS Inc, Chicago, IL). Continuous variables were reported as mean, Categorical variables were summarized as percentages. The total number of risk factors for SCD was divided by the total number of patients to assess the average number of risk factors per patient. Heterogeneity among the studies was assessed using the Q test and P index. Random-effects model was used to calculate the summary estimates of the outcome data. From the pooled data, summary estimates of patient characteristics and risk factors for SCD were calculated. Meta-analysis of the outcome data was performed, and weighted event rates and weighted annualized event rates were calculated. Forest plots were constructed using the method of Nefelly et al.11

Results
Search Results
The literature search yielded 469 articles (Figure 1). After review, exclusion, and cross-referencing, a total of 27 observational studies were included in the systematic review (Table 1). Overall, 16 different patient cohorts were identified in these 27 studies.4-30 Because of apparent serial reporting of patient cohorts and to avoid duplicate entering of data, only 1 study per patient cohort was included in the meta-analysis. Hence, the summary estimate of clinical data and outcome is based on 16 studies.3,7,9,12-14,16,18-20,24-28,30 Thirteen (81%) studies reported on a population of patients with HCM with an ICD for primary or secondary prevention of SCD, 1 (8%) study focused on patients with HCM with an ICD for primary prevention of SCD, and 2 (13%) studies reported on patients with hypertrophic obstructive cardiomyopathy who underwent alcohol septal ablation and had received an ICD.

Patient Characteristics
There were 2190 patients (mean age, 42 years; 38% women), most of whom (83%) received an ICD for primary prevention of SCD. Risk factors for SCD were left ventricular wall thickness ≥30 mm (20%), family history of SCD (43%), nonsustained ventricular tachycardia (46%), syncope (41%), and abnormal blood pressure response (25%). Patients had on average 1.8 risk factors for SCD. Hypertrophic obstructive cardiomyopathy was present in 27% of the patients.

ICD Interventions and Outcome
During the 3.7-year follow-up, 311 of 2190 (14%) patients had an appropriate ICD intervention (Table 2). The annualized appropriate ICD intervention rate was 3.3% (Figure 2). Data on inappropriate ICD intervention was available in 13 studies. Inappropriate ICD intervention occurred in 388 of 1966 (20%) patients. The annualized inappropriate ICD intervention rate was 4.8% (Figure 3). Mortality data was reported in 13 studies: there were 53 (3%) cardiac deaths and 49 (2%) noncardiac deaths. The annualized cardiac mortality rate was 0.6%, and the annualized noncardiac mortality rate was 0.4%. Five studies reported follow-up data on heart transplantation; this occurred in 28 of 1214 patients (2%), and the annualized heart transplantation rate was 0.5%.

Complications
Information on ICD-related complications was available in 9 of 16 studies, including a total of 1691 patients (Table 2). Of them, 260 (15%) had any form of ICD-related complications. The most frequently observed complication was lead malfunction in 118 (7%). Other complications were infection in 59 (3%) and lead displacement in 28 (3%). Only 1 study provided information on psychological complications; these occurred in 5 of 132 (4%) patients.

Discussion
This meta-analysis demonstrates a low cardiac and noncardiac mortality rate after ICD therapy in patients with HCM.
Table 1. Summary of the Studies Reporting ICD Therapy in Patients With Hypertrophic Cardiomyopathy

<table>
<thead>
<tr>
<th>Cohort</th>
<th>Region</th>
<th>Author</th>
<th>Year</th>
<th>Population</th>
<th>n</th>
<th>Mean Age, y</th>
<th>Women, %</th>
<th>Primary Prevention, %</th>
<th>Secondary Prevention, %</th>
<th>LW WT ≥ 30 mm, %</th>
<th>Family History of SCD, %</th>
<th>NSVT, %</th>
<th>Syncope, %</th>
<th>Abnormal BP Response, %</th>
<th>HOCM, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aalst, Barcelona</td>
<td>Belgium, Spain</td>
<td>Primo et al⁴</td>
<td>1998</td>
<td>HCM and ICD</td>
<td>13</td>
<td>48</td>
<td>38</td>
<td>77</td>
<td>23</td>
<td>0</td>
<td>NA</td>
<td>NA</td>
<td>46</td>
<td>NA</td>
<td>38</td>
</tr>
<tr>
<td>London</td>
<td>United Kingdom</td>
<td>Elliott et al⁵</td>
<td>1999</td>
<td>HCM with VT/VF</td>
<td>6</td>
<td>19</td>
<td>NA</td>
<td>100</td>
<td>0</td>
<td>NA</td>
<td>17</td>
<td>33</td>
<td>50</td>
<td>67</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Kaski et al⁶</td>
<td>2007</td>
<td>ICD in children with HCM</td>
<td>22</td>
<td>14</td>
<td>41</td>
<td>77</td>
<td>23</td>
<td>50</td>
<td>55</td>
<td>5</td>
<td>32</td>
<td>68</td>
<td>36</td>
</tr>
<tr>
<td>ICD in HCM Registry</td>
<td>United States, Italy</td>
<td>O'Mahony et al⁷</td>
<td>2012</td>
<td>HCM and ICD</td>
<td>334</td>
<td>40</td>
<td>38</td>
<td>92</td>
<td>8</td>
<td>15</td>
<td>51</td>
<td>48</td>
<td>39</td>
<td>33</td>
<td>23</td>
</tr>
<tr>
<td></td>
<td>United States, Europe, Australia</td>
<td>Maron et al⁸</td>
<td>2000</td>
<td>HCM and ICD</td>
<td>128</td>
<td>40</td>
<td>31</td>
<td>66</td>
<td>34</td>
<td>8</td>
<td>30</td>
<td>25</td>
<td>32</td>
<td>NA</td>
<td>18</td>
</tr>
<tr>
<td></td>
<td>United States, Australia</td>
<td>Kaski et al⁹</td>
<td>2007</td>
<td>HCM and ICD</td>
<td>506</td>
<td>42</td>
<td>36</td>
<td>76</td>
<td>24</td>
<td>24</td>
<td>51</td>
<td>46</td>
<td>47</td>
<td>NA</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td>United States, Australia</td>
<td>Maron et al¹⁰</td>
<td>2009</td>
<td>HCM and appropriate ICD intervention</td>
<td>63</td>
<td>43</td>
<td>29</td>
<td>65</td>
<td>35</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>14</td>
</tr>
<tr>
<td></td>
<td>United States, Australia</td>
<td>Sherrid et al¹¹</td>
<td>2009</td>
<td>HCM and ICD</td>
<td>330</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>NIH</td>
<td>United States</td>
<td>Begley et al¹²</td>
<td>2003</td>
<td>HCM and ICD</td>
<td>132</td>
<td>34</td>
<td>39</td>
<td>36</td>
<td>64</td>
<td>23</td>
<td>41</td>
<td>63</td>
<td>39</td>
<td>26</td>
<td>38</td>
</tr>
<tr>
<td>Sydney</td>
<td>Australia</td>
<td>Jayatilleke et al¹³</td>
<td>2004</td>
<td>HCM and ICD</td>
<td>22</td>
<td>NA</td>
<td>NA</td>
<td>82</td>
<td>18</td>
<td>50</td>
<td>36</td>
<td>27</td>
<td>41</td>
<td>23</td>
<td>NA</td>
</tr>
<tr>
<td>Minneapolis</td>
<td>United States</td>
<td>Almqvist et al¹⁴</td>
<td>2005</td>
<td>HCM and ICD</td>
<td>75</td>
<td>37</td>
<td>35</td>
<td>95</td>
<td>5</td>
<td>29</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>20</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Maron et al¹⁵</td>
<td>2009</td>
<td>HCM and VT/VF</td>
<td>39</td>
<td>34</td>
<td>44</td>
<td>46</td>
<td>23</td>
<td>46</td>
<td>41</td>
<td>47</td>
<td>NA</td>
<td>NA</td>
<td>16</td>
</tr>
<tr>
<td>Bielefeld</td>
<td>Germany</td>
<td>Lawrenz et al¹⁶</td>
<td>2005</td>
<td>HOCM, ICD, and ASA</td>
<td>15</td>
<td>53</td>
<td>47</td>
<td>40</td>
<td>60</td>
<td>NA</td>
<td>33</td>
<td>33</td>
<td>33</td>
<td>17</td>
<td>100</td>
</tr>
<tr>
<td>Warsaw</td>
<td>Poland</td>
<td>Przybylski et al¹⁷</td>
<td>2005</td>
<td>HCM and ICD</td>
<td>46</td>
<td>32</td>
<td>59</td>
<td>39</td>
<td>61</td>
<td>17</td>
<td>50</td>
<td>54</td>
<td>63</td>
<td>70</td>
<td>17</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Syska et al¹⁸</td>
<td>2010</td>
<td>HCM and ICD</td>
<td>104</td>
<td>35.6</td>
<td>55</td>
<td>75</td>
<td>25</td>
<td>NA</td>
<td>NA</td>
<td>65</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Alicante, Murcia, A Coruna</td>
<td>Spain</td>
<td>Marin et al¹⁹</td>
<td>2006</td>
<td>HCM and ICD</td>
<td>45</td>
<td>42.8</td>
<td>38</td>
<td>60</td>
<td>40</td>
<td>29</td>
<td>31</td>
<td>64</td>
<td>47</td>
<td>56</td>
<td>44</td>
</tr>
<tr>
<td>Sao Paulo</td>
<td>Brazil</td>
<td>Medeiros et al²⁰</td>
<td>2006</td>
<td>HCM and ICD</td>
<td>26</td>
<td>42.7</td>
<td>54</td>
<td>62</td>
<td>38</td>
<td>19</td>
<td>58</td>
<td>46</td>
<td>77</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Rochester</td>
<td>United States</td>
<td>Cha et al²¹</td>
<td>2007</td>
<td>HCM and ICD for primary prevention</td>
<td>68</td>
<td>43</td>
<td>40</td>
<td>100</td>
<td>0</td>
<td>NA</td>
<td>56</td>
<td>62</td>
<td>29</td>
<td>5</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>McLeod et al²²</td>
<td>2007</td>
<td>HCM and ICD</td>
<td>125</td>
<td>41.0</td>
<td>38</td>
<td>94</td>
<td>6</td>
<td>6</td>
<td>49</td>
<td>38</td>
<td>36</td>
<td>26</td>
<td>56</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Kiernan et al²³</td>
<td>2008</td>
<td>HCM, ICD, and no ASA or myectomy</td>
<td>69</td>
<td>43.5</td>
<td>29</td>
<td>97</td>
<td>3</td>
<td>1</td>
<td>54</td>
<td>46</td>
<td>30</td>
<td>30</td>
<td>20</td>
</tr>
<tr>
<td>Toronto</td>
<td>Canada</td>
<td>Lin et al²⁴</td>
<td>2009</td>
<td>HCM and ICD</td>
<td>181</td>
<td>44</td>
<td>38</td>
<td>86</td>
<td>14</td>
<td>16</td>
<td>56</td>
<td>41</td>
<td>40</td>
<td>1</td>
<td>20</td>
</tr>
<tr>
<td>Charleston</td>
<td>United States</td>
<td>Woo et al²⁵</td>
<td>2007</td>
<td>HCM and ICD</td>
<td>61</td>
<td>46</td>
<td>34</td>
<td>82</td>
<td>18</td>
<td>15</td>
<td>46</td>
<td>5</td>
<td>25</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cuoco et al²⁶</td>
<td>2008</td>
<td>HCM, ASA, and ICD for primary prevention</td>
<td>123</td>
<td>48</td>
<td>34</td>
<td>100</td>
<td>0</td>
<td>11</td>
<td>38</td>
<td>NA</td>
<td>63</td>
<td>34</td>
<td>100</td>
</tr>
</tbody>
</table>

(Continued)
In patients with HCM with on average 1.8 risk factors for SCD, appropriate ICD intervention occurred at a rate of 3.3%/year, thereby, most probably, preventing SCD. These findings emphasize the importance of ICD therapy in patients with HCM at risk for SCD.

Current American College of Cardiology Foundation/American Heart Association guidelines recommend comprehensive SCD risk stratification at initial evaluation and on a periodic basis (every 12 to 24 months) for patients with HCM. A personal history for ventricular fibrillation, sustained ventricular tachycardia, or SCD is recommended, and established risk factors for SCD should be evaluated. ICD placement is recommended (class I recommendation) for patients with HCM and prior documented cardiac arrest, ventricular fibrillation, or hemodynamically significant ventricular tachycardia. A comparable decision strategy for ICD implantation for secondary prevention of SCD was applied in the studies included in the present analysis. In the pooled analysis, 17% of the patients with HCM received an ICD for secondary prevention of SCD.

For primary prevention of SCD in patients with HCM, the guidelines state that it is reasonable to recommend (class IIa recommendation) an ICD for patients with HCM with SCD presumably related to HCM in ≥1 first-degree relatives, or a maximum left ventricular wall thickness ≥30 mm, or ≥1 recent unexplained syncopal episodes. An ICD can be useful in select patients with nonsustained ventricular tachycardia in the presence of other SCD risk factors or modifiers or with an abnormal blood pressure response to exercise in the presence of other SCD risk factors or modifiers. It is reasonable to recommend an ICD for high-risk children with HCM, based on unexplained syncope, massive left ventricular hypertrophy, or family history of SCD, after taking into account the relatively high complication rate of long-term ICD implantation.

In the present analysis, the majority of studies did not provide clear information on the clinical decision strategy for ICD implantation for primary prevention of SCD. Complete information on all 5 established risk factors for SCD in patients with HCM was available in only 7 of 16 (44%) cohorts. Primary prevention of SCD in patients with HCM depends on the presence of SCD risk factors and modifiers; therefore, complete information on all established risk factors is highly relevant. All reported cohorts were collected before publication of the current practice guideline on HCM, and it is not certain that the results from the pooled analysis also apply to patients with HCM who currently receive an ICD. Nevertheless, the present analysis demonstrates that patients with HCM with an ICD had on average 1.8 risk factors for SCD. Consequently, the population in the pooled analysis was at high risk for SCD and is probably comparable to the population that should be considered for ICD implantation according to the current guideline.

The pooled analysis demonstrates that inappropriate ICD intervention and complications are not uncommon (4.8%/year and 3.4%/year, respectively). Previous studies suggested that patients with HCM are more vulnerable to ICD-related complications and inappropriate ICD therapy because of young age at implant and increased prevalence...
## Table 2. Summary of Clinical Outcome

<table>
<thead>
<tr>
<th>Cohort</th>
<th>Author</th>
<th>Year</th>
<th>Follow-Up, y</th>
<th>Appropriate Intervention, %</th>
<th>Inappropriate Intervention, %</th>
<th>Complications, %</th>
<th>Mortality, %</th>
<th>Heart transplant, %</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aalst, Barcelona</td>
<td>Primo et al⁴</td>
<td>1998</td>
<td>2.2</td>
<td>15</td>
<td>23</td>
<td>0</td>
<td>0</td>
<td>8</td>
</tr>
<tr>
<td>London</td>
<td>Elliott et al⁵</td>
<td>1999</td>
<td>6.1</td>
<td>50</td>
<td>NA</td>
<td>8</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Kaski et al⁶</td>
<td>2007</td>
<td>1.7</td>
<td>18</td>
<td>18</td>
<td>5</td>
<td>5</td>
<td>NA</td>
<td>8</td>
</tr>
<tr>
<td>O'Mahony et al⁷</td>
<td>2012</td>
<td>3.6</td>
<td>8</td>
<td>16</td>
<td>5</td>
<td>6</td>
<td>4</td>
<td>18</td>
</tr>
<tr>
<td>ICD in HCM Registry</td>
<td>Maron et al⁸</td>
<td>2000</td>
<td>3.1</td>
<td>23</td>
<td>25</td>
<td>9</td>
<td>2</td>
<td>14</td>
</tr>
<tr>
<td></td>
<td>Maron et al⁹</td>
<td>2007</td>
<td>3.7</td>
<td>20</td>
<td>27</td>
<td>7</td>
<td>4</td>
<td>12</td>
</tr>
<tr>
<td></td>
<td>Maron et al¹⁰</td>
<td>2009</td>
<td>NA</td>
<td>100</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>Sherrid et al¹¹</td>
<td>2009</td>
<td>3.7</td>
<td>17</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>NIH</td>
<td>Begley et al¹²</td>
<td>2003</td>
<td>4.8</td>
<td>20</td>
<td>23</td>
<td>5</td>
<td>3</td>
<td>28</td>
</tr>
<tr>
<td>Sydney</td>
<td>Jayatilleke et al¹³</td>
<td>2004</td>
<td>2.9</td>
<td>32</td>
<td>9</td>
<td>NA</td>
<td>5</td>
<td>NA</td>
</tr>
<tr>
<td>Minneapolis</td>
<td>Alquist et al¹⁴</td>
<td>2005</td>
<td>3.6</td>
<td>7</td>
<td>NA</td>
<td>3</td>
<td>3</td>
<td>8</td>
</tr>
<tr>
<td></td>
<td>Maron et al¹⁵</td>
<td>2009</td>
<td>9.4</td>
<td>41</td>
<td>28</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Bielefeld</td>
<td>Lawrenz et al¹⁶</td>
<td>2005</td>
<td>3.4</td>
<td>27</td>
<td>20</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Warsaw</td>
<td>Przybylski et al¹⁷</td>
<td>2005</td>
<td>2.4</td>
<td>28</td>
<td>30</td>
<td>7</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Sypka et al¹⁸</td>
<td>2010</td>
<td>4.6</td>
<td>26</td>
<td>35</td>
<td>13</td>
<td>2</td>
<td>12</td>
</tr>
<tr>
<td>Alicante</td>
<td>Marin et al¹⁹</td>
<td>2006</td>
<td>2.7</td>
<td>22</td>
<td>29</td>
<td>NA</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Sao Paulo</td>
<td>Medeiros et al²⁰</td>
<td>2006</td>
<td>1.6</td>
<td>15</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Rochester</td>
<td>Cha et al²¹</td>
<td>2007</td>
<td>3.4</td>
<td>13</td>
<td>15</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>McLeod et al²²</td>
<td>2007</td>
<td>4.4</td>
<td>10</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>Kieman et al²³</td>
<td>2008</td>
<td>4.4</td>
<td>17</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td>Lin et al²⁴</td>
<td>2009</td>
<td>4.9</td>
<td>16</td>
<td>23</td>
<td>13</td>
<td>4</td>
<td>26</td>
</tr>
<tr>
<td>Toronto</td>
<td>Woo et al²⁵</td>
<td>2007</td>
<td>3.3</td>
<td>13</td>
<td>33</td>
<td>13</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Charleston</td>
<td>Cooc et al²⁶</td>
<td>2008</td>
<td>2.9</td>
<td>7</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>10 centers</td>
<td>Saumarez et al²⁷</td>
<td>2008</td>
<td>4.3</td>
<td>3</td>
<td>3</td>
<td>NA</td>
<td>NA</td>
<td>1</td>
</tr>
<tr>
<td>7 centers</td>
<td>Hauser et al²⁸</td>
<td>2008</td>
<td>3.3</td>
<td>11</td>
<td>12</td>
<td>5</td>
<td>0.3</td>
<td>3</td>
</tr>
<tr>
<td>Minneapolis, Rochester</td>
<td>Bos et al²⁹</td>
<td>2010</td>
<td>4.6</td>
<td>14</td>
<td>27</td>
<td>NA</td>
<td>NA</td>
<td>3</td>
</tr>
<tr>
<td>Bad Oeynhausen</td>
<td>Prinz et al³⁰</td>
<td>2010</td>
<td>2.0</td>
<td>10</td>
<td>6</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
</tbody>
</table>

**Event rate (95% CI)**
- Bad Oeynhausen: 14.9 (9.9–19.9) (2.2–2.8)
- Aalst, Barcelona: 12.6–25.4 (4.1–8.3)
- Sydney: 12.0–16.9 (0.1–0.8)

**Annualized event rate (95% CI)**
- Bad Oeynhausen: 0.6 (0.1–0.4)
- Aalst, Barcelona: 0.9–1.5
- Sydney: 0.9–1.5

**NA** indicates not available; ICD, implantable cardioverter defibrillator; HCM, hypertrophic cardiomyopathy; NIH, National Institutes of Health.
of atrial fibrillation.\textsuperscript{22} Reports from large ICD registries, including predominantly patients with ischemic heart disease, demonstrate an early complication rate varying from 3.3\% to 11\% during the hospital admission for ICD implantation.\textsuperscript{33,34} Long-term follow-up data on ICD-related complications in general practice are not available, hampering comparison of the inappropriate ICD intervention and ICD-related complication rates observed in patients with HCM. Most patients with HCM who underwent ICD implantation were young (mean age, 42 years), and, therefore, the risk of ICD-related complications should be carefully considered and discussed with the patient during the decision-making process before implantation. This is particularly relevant because of the long periods that young patients will live with the implanted device and leads. Only 3 studies\textsuperscript{6,8,12} reported the occurrence of ICD-related psychological complications. The psychological and behavioral aspects of ICD therapy in patients with HCM should receive more attention because many patients with HCM considered for ICD therapy are otherwise healthy and often asymptomatic young individuals.

\section*{Limitations}

This systematic review and meta-analysis has inherent limitations. The data were extracted from observational studies. A potential risk of pooling data from different studies is to mix patients with different clinical characteristics and SCD risk profile. The decision strategy for ICD implantation was not specified in most studies. The currently available studies have reported on outcome and complications after ICD therapy in populations with predominantly adult patients with HCM, except the study by Kaski et al.\textsuperscript{6} More information is desired concerning ICD therapy in children and adolescents with HCM. Data on cycle length of the arrhythmia and type of arrhythmia were not available in the majority of studies. Finally, the first report was from 1998, and over the years significant progress in ICD devices and leads has been made, and experience with implantation and follow-up has increased.

\section*{Future Studies}

ICD therapy has proven benefits in patients with HCM at increased risk for SCD. Future studies on ICD therapy for prevention of SCD in patients with HCM are needed to refine risk stratification for SCD and to define the role of other risk markers, including cardiac magnetic resonance imaging. There are indications that patients with HCM with extensive delayed enhancement on contrast-enhanced cardiac magnetic resonance imaging are at increased risk of ventricular arrhythmias. Efforts to further reduce inappropriate ICD

\begin{table}[h]
\centering
\begin{tabular}{|l|c|c|}
\hline
Author & Rate & 95\% CI \\
\hline
Primo & 7.1 & -7.4 – 21.6 \\
O’Mahony & 2.3 & 0.7 – 4.0 \\
Maron & 5.5 & 3.5 – 7.5 \\
Begley & 4.3 & 0.7 – 7.8 \\
Jayatilleke & 11.0 & -2.9 – 24.8 \\
Almquist & 1.9 & -1.2 – 4.9 \\
Lawrenz & 7.8 & -0.3 – 21.9 \\
Syska & 5.6 & 1.1 – 10.2 \\
Marin & 8.3 & -0.1 – 16.8 \\
Medeiros & 9.7 & -2.3 – 21.7 \\
Lin & 3.3 & 0.6 – 5.9 \\
Woo & 3.9 & -1.0 – 8.9 \\
Cuoco & 2.5 & -0.3 – 5.3 \\
Saumarez & 0.8 & -0.5 – 2.1 \\
Hauser & 3.5 & 1.4 – 5.5 \\
Prinz & 5.0 & -1.2 – 11.2 \\
\hline
\textbf{Summary estimate} & \textbf{3.3} & \textbf{2.2 – 4.4} \\
\hline
\end{tabular}
\caption{Annualized appropriate ICD intervention rate (%/year).

\textit{Note:} Random effects model \( Q = 11.6, i^2 = 0\% \).

Figure 2. Forest plot of annualized appropriate ICD intervention rate (%/year).

\begin{tabular}{|l|c|c|}
\hline
Author & Rate & 95\% CI \\
\hline
Primo & 10.7 & -7.1 – 28.4 \\
O’Mahony & 4.6 & 2.3 – 6.9 \\
Maron & 7.3 & 4.9 – 9.6 \\
Begley & 4.7 & 1.0 – 8.4 \\
Jayatilleke & 3.1 & -4.3 – 10.5 \\
Lawrenz & 5.9 & -6.4 – 18.1 \\
Syska & 7.5 & 2.3 – 12.8 \\
Marin & 10.8 & 1.2 – 20.5 \\
Lin & 4.7 & 1.6 – 7.9 \\
Woo & 9.8 & 2.0 – 17.7 \\
Saumarez & 0.6 & -0.5 – 1.8 \\
Hauser & 3.7 & 1.6 – 5.8 \\
Prinz & 3.0 & -1.8 – 7.8 \\
\hline
\textbf{Summary estimate} & \textbf{4.8} & \textbf{2.9 – 6.7} \\
\hline
\end{tabular}
\caption{Annualized inappropriate ICD intervention rate (%/year).

\textit{Note:} Random effects model \( Q = 7.3, i^2 = 0\% \).

Figure 3. Forest plot of annualized inappropriate ICD intervention rate (%/year).
intervention and complication rates may have substantial clinical and financial benefits. Authors of future reports on ICD therapy in patients with HCM are encouraged to provide complete information on the clinical characteristics of the study population, established clinical risk factors for SCD, decision strategy for ICD implantation, and device-related complications and outcome (including at least appropriate and inappropriate ICD intervention, and cardiac and noncardiac mortality).

Conclusions
This meta-analysis demonstrates a low cardiac and noncardiac mortality rate after ICD therapy in patients with HCM. Appropriate ICD intervention occurred at a rate of 3.3%/year, thereby, most probably, preventing SCD. Inappropriate ICD intervention and complications are not uncommon (4.8%/year and 3.4%/year, respectively). The benefits and risks of ICD therapy in patients with HCM should be carefully weighted.

Disclosures
None.

References
28. Hauser RG, Katsiyannis WT, Gornick CC, Almquist AK, Kallinen LM. Deaths and cardiovascular injuries due to device-assisted implantable
Patients with hypertrophic cardiomyopathy (HCM) are at increased risk for sudden cardiac death (SCD), most frequently caused by ventricular arrhythmias. Implantable cardioverter defibrillator (ICD) therapy may effectively terminate life-threatening ventricular arrhythmias and thereby prevent SCD. However, ICD therapy is not without risk, because inappropriate ICD interventions and device-related complications may occur. Although previous studies have reported on the use of ICD therapy for prevention of SCD in patients with HCM, a complete overview of outcomes and complications after ICD therapy in patients with HCM is not available. In this meta-analysis, we demonstrate a low cardiac and noncardiac mortality rate after ICD implantation in patients with HCM. Appropriate ICD intervention occurred at a rate of 3.3%/year, thereby, most probably, preventing SCD. Inappropriate ICD intervention and complications occurred at a rate of 4.8%/year and 3.4%/year, respectively, in these patients. The most frequently observed complication was lead malfunction in 7%. Other complications were infection in 3% and lead displacement in 3%. Consideration of these outcome and complication data may help clinicians in decision making and counseling of patients with HCM at increased risk for SCD considered for ICD therapy. Additional research is warranted to further reduce inappropriate ICD intervention and complication rates.

**Clinical Perspective**

Patients with hypertrophic cardiomyopathy (HCM) are at increased risk for sudden cardiac death (SCD), most frequently caused by ventricular arrhythmias. Implantable cardioverter defibrillator (ICD) therapy may effectively terminate life-threatening ventricular arrhythmias and thereby prevent SCD. However, ICD therapy is not without risk, because inappropriate ICD interventions and device-related complications may occur. Although previous studies have reported on the use of ICD therapy for prevention of SCD in patients with HCM, a complete overview of outcomes and complications after ICD therapy in patients with HCM is not available. In this meta-analysis, we demonstrate a low cardiac and noncardiac mortality rate after ICD implantation in patients with HCM. Appropriate ICD intervention occurred at a rate of 3.3%/year, thereby, most probably, preventing SCD. Inappropriate ICD intervention and complications occurred at a rate of 4.8%/year and 3.4%/year, respectively, in these patients. The most frequently observed complication was lead malfunction in 7%. Other complications were infection in 3% and lead displacement in 3%. Consideration of these outcome and complication data may help clinicians in decision making and counseling of patients with HCM at increased risk for SCD considered for ICD therapy. Additional research is warranted to further reduce inappropriate ICD intervention and complication rates.
Outcome and Complications After Implantable Cardioverter Defibrillator Therapy in Hypertrophic Cardiomyopathy: Systematic Review and Meta-Analysis

Arend F.L. Schinkel, Pieter A. Vriesendorp, Eric J.G. Sijbrands, Luc J.L.M. Jordaens, Folkert J. ten Cate and Michelle Michels

_Circ Heart Fail_. 2012;5:552-559; originally published online July 20, 2012; doi: 10.1161/CIRCHEARTFAILURE.112.969626

_Circulation: Heart Failure_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Copyright © 2012 American Heart Association, Inc. All rights reserved.

Print ISSN: 1941-3289. Online ISSN: 1941-3297

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circheartfailure.ahajournals.org/content/5/5/552

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation: Heart Failure_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the _Permissions and Rights Question and Answer_ document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation: Heart Failure_ is online at:
http://circheartfailure.ahajournals.org//subscriptions/