Should patients with congenital heart disease and a systemic ventricular ejection fraction less than 30% undergo prophylactic implantation of an ICD?

Patients With Congenital Heart Disease and a Systemic Ventricular Ejection Fraction Less Than 30% Should Undergo Prophylactic Implantation of an Implantable Cardioverter Defibrillator

Michael J. Silka, MD; Yaniv Bar-Cohen, MD

A reduced left ventricular (LV) ejection fraction (EF) is established as one of the strongest risk factors for sudden and total cardiac mortality in adults with ischemic and nonischemic heart disease.1,2 Based on the Multicenter Automatic Defibrillator Implantation Trial II and Sudden Cardiac Death in Heart Failure Trial (SCD-HeFT) clinical trials, patients with an LVEF <30% are at a significantly increased risk for sudden cardiac death (SCD) and thus warrant an implantable cardioverter defibrillator (ICD).3,4 At the same time, increasing numbers of patients with congenital heart disease (CHD) continue to survive complex repairs or palliations of their heart defects, allowing survival beyond adolescence and into adulthood. However, it is uncertain whether the risks of systemic ventricular dysfunction and the benefits of primary prevention ICDs demonstrated in adult clinical trials extend to patients with CHD. Clarification regarding this issue is important because in the current era, the majority of ICD implantations in patients with CHD are for primary prevention of SCD.5

**Background**

Several major randomized clinical trials have been reported regarding the efficacy of ICDs for the primary prevention of SCD in adults with ischemic and nonischemic heart disease and have been the subject of several meta-analyses and...
These studies include a total of 7501 patients, with a mean LVEF between 21% and 28%. The use of ICDs reduced all-cause mortality in patients with LV systolic dysfunction by 19%, primarily because of a 54% relative reduction in SCD. Based on the consistency of the data, the following class I recommendations were made in the 2008 American College of Cardiology/American Heart Association/Heart Rhythm Society guidelines on device-based therapy:

- ICD therapy is indicated in patients with a LVEF <35% due to prior myocardial infarction who are ≥40 days post–myocardial infarction and in New York Heart Association (NYHA) functional class II or III.
- ICD therapy is indicated in patients with a LVEF <35% due to nonischemic dilated cardiomyopathy who are in NYHA functional class II or III.
- ICD therapy is indicated in patients with a LVEF <30% due to prior myocardial infarction who are ≥40 days post–myocardial infarction and in NYHA functional class I.

Therefore, based on the data from the aforementioned clinical trials and guidelines, an EF <30% will be considered the crucial threshold for ICD implantation in patients with CHD.

Systemic Ventricular Dysfunction and CHD

As a point of departure, it is crucial to emphasize that given the heterogeneity of CHD, “systemic” does not necessarily equate to “left” ventricular dysfunction. This is particularly relevant for transposition of the great arteries (TGA) complexes, either for patients with d-TGA who have undergone Mustard or Senning atrial redirection procedures or patients with discordant atrioventricular and ventriculo-arterial connections, commonly referred to as congenitally corrected TGA (CC-TGA). Further complexities arise in patients with functional single ventricle physiology, where the ventricular morphology may be left, right, or indeterminate. Therefore, the term “systemic” rather than “left” ventricle will be used.

Evaluation of ventricular function in CHD has proven a complex challenge because of multiple factors: ventricular morphological abnormalities due to the basic congenital heart defect, abnormal ventricular geometry due to circulatory shunting, patch closure or augmentation of the ventricular septum, regional dysynchrony due to conduction delay or heart block after surgery, and atrioventricular valve insufficiency. Therefore, surrogate measures of systemic ventricular function are often used, including the echocardiographic cross-sectional shortening fraction, systemic ventricular end-diastolic pressure, and overt clinical evidence of heart failure. When possible, however, reference in this study will be made to angiographic or cardiac magnetic resonance estimates of global ventricular EF.

Quantification and grading of ventricular systolic dysfunction in CHD has been variably defined, often with discordant results when different methods are used. However, based on available data, the following grading system will be used:

- Good function: ≥50% EF
- Mild impairment: 40% to 49% EF
- Moderate impairment: 30% to 39% EF
- Severe impairment: <30% EF

For purposes of this analysis, patients classified as having mild to moderate ventricular dysfunction are divided equally between the 2 groups.

Systemic Right Ventricular Dysfunction and Sudden Death

The long-term prognosis for patients with a systemic right ventricle (RV) after Mustard or Senning repairs of simple d-TGA is a topic of concern, as these patients are now entering the fourth and fifth decades of life. Failure of the systemic morphological RV has been considered by some to be inevitable, often accompanied by the onset of progressive tricuspid insufficiency. Total cardiac mortality is markedly increased in patients with severe impairment of ventricular function and progressively increases in patients with moderate dysfunction after 20 years of age (Figure 1).

In a prospective study of systemic ventricular function after the Mustard procedure, Roos-Hesselink et al reported a striking decline in function with extended follow-up. This study evaluated 47 patients, with good function reported in all patients 14 years after repair; however, at a median follow-up of 25 years in the same cohort, moderate-to-severe dysfunction had developed in 61% of patients, mild dysfunction in 33%, and normal systemic RV function in only 6%. Although the basis for progressive dysfunction may be multifactorial, myocardial perfusion defects and impaired coronary flow reserve have been reported in these patients.

The incidence of SCD in d-TGA patients demonstrates a time-dependent course, parallel to the progressive deterioration of RV function. In the largest series of SCD after Mustard or Senning procedures, Kammeraad et al reported that of 19 cases of SCD with recent evaluation before their event, RV function was normal in only 5 patients, mild to moderately impaired in 9 patients, and severely impaired in 5 patients.
patients. In comparison, only 1 of 31 age-matched controls had severely impaired function, with mild to moderate impairment in 14 patients, and normal function in 16 patients. Several other studies have also noted correlation between RV dysfunction and mortality in these patients (Table 1).16,17 The data suggest that with 11 to 25 years follow-up, the incidence of severe impairment of systemic RV dysfunction after Mustard or Senning procedures is 6% and the incidence of SCD is 7.6%.

The cause and effect relationship between ventricular dysfunction and arrhythmias in d-TGA patients is an area of active investigation. Gatzoulis et al17 have described an association between the incidence of atrial fibrillation or intra-atrial reentrant tachycardia and reduced ventricular function after the Mustard procedure; a mechano-electric interaction was proposed with ventricular dysfunction predisposing to the development of atrial arrhythmias. Conversely, Kammeraad et al15 reported documented ventricular tachycardia with a rapid ventricular response. Although a complex issue, it is logical that intrinsic impairment of myocardial flow reserve resulting in ischemia or intra-atrial reentrant tachycardia with a rapid ventricular response. Although a complex issue, it is logical that intrinsic impairment of systemic ventricular dysfunction with compromised coronary reserve will result in an ischemic substrate predisposing to the development of life-threatening ventricular arrhythmias.

Congenitally corrected transposition (CC-TGA) is another form of CHD in which progressive deterioration of systemic ventricular function is well documented and is associated with a high rate of sudden and total cardiac mortality. Presbitero et al18 reported the clinical course of 18 CC-TGA patients aged 16 to 61 years with no other associated defects. Patients were rarely symptomatic until the fourth or fifth decade of life, when rhythm disturbances, systemic atrioventricular valve insufficiency, and impaired systemic ventricular function resulted in congestive heart failure in the majority of patients. Connelly et al19 serially evaluated 52 adults with CC-TGA, aged ≥ 18 years, with 13 cardiac deaths, 5 due to heart failure, and 4 sudden deaths during follow-up. Arrhythmias were documented in 9 of 13 patients before death, with a mean age of 38±12 years at death. Systemic RV failure was reported in 8 of 13 patients who died, compared with only 3 of 39 survivors. Similar to d-TGA patients, the potential role of myocardial perfusion defects (fixed or reversible) was considered possibly related to progressive ventricular dysfunction. Oechslin et al20 reported 6 sudden deaths in 57 patients with CC-TGA at a mean age of 37±13 years; all events were in patients with impaired systemic RV function, with overt heart failure in 2 patients. In this series, the overall mortality rate (26%) for CC-TGA patients was the highest for any form of CHD.

### Functional Univentricular Hearts

Patients with univentricular physiology present unique challenges regarding both the evaluation of ventricular function as well as assessment for risk of SCD. Patients with “uncorrected” physiology have a poor prognosis, with a 70% mortality rate before age 16 years for patients with a single LV.22 The prognosis is far worse for patients with an uncorrected single RV, with only 50% survival 4 years after diagnosis. The most common causes of death are heart failure, documented arrhythmias, and unexplained SCD (presumably arrhythmic).

In contrast, the benefits of the Fontan operation are well established, with an actuarial survival rate for operative survivors of 90% at 10 years and 83% at 20 years.23 Unfortunately, late complications are numerous and include arrhythmias, thromboembolism, and progressive deterioration of ventricular function. Eicken et al24 performed MRIs on 23 Fontan patients and reported a median single ventricle EF of 49%, compared with a 65% EF in normal controls. Fogel et al25 also evaluated Fontan patients using MRI and showed a decrease in all indices of ventricular performance.

Although some degree of dysfunction is common, severe ventricular dysfunction seems relatively uncommon. Of 36 late Fontan failures in 410 single ventricle patients reported by Gentles et al,26 only 11 (2.7%) had systemic ventricular failure.

Although SCD is responsible for 9% to 16% of deaths in Fontan patients,23,27,28 the limited numbers of patients precludes definitive analysis of risk factors. Khairy et al23 studied 261 Fontan patients, 76 of whom died, including 7 sudden deaths (9.2% of all deaths). No independent predictors for SCD were identified in this study. Bernstein et al27

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**Table 1. Prevalence of Ventricular Dysfunction and SCD in d-TGA**

<table>
<thead>
<tr>
<th>References</th>
<th>No. of Patients</th>
<th>Mean Years Postoperation</th>
<th>&gt;50% EF</th>
<th>40–50% EF</th>
<th>31–40% EF</th>
<th>&lt;30% EF</th>
<th>Mortality/Transplant</th>
<th>SCD</th>
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<tr>
<td>Gelatt et al11</td>
<td>543</td>
<td>11.5</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>23</td>
<td>77</td>
</tr>
<tr>
<td>Kammeraad et al15</td>
<td>50</td>
<td>12.3</td>
<td>21</td>
<td>12</td>
<td>11</td>
<td>6</td>
<td>19</td>
<td>19</td>
</tr>
<tr>
<td>Puley et al16</td>
<td>81</td>
<td>23</td>
<td>28</td>
<td>27</td>
<td>21</td>
<td>5</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>Gatzoulis et al17</td>
<td>51</td>
<td>23.4</td>
<td>23</td>
<td>13</td>
<td>13</td>
<td>2</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>Roos-Hesselink et al12</td>
<td>47</td>
<td>25</td>
<td>2</td>
<td>16</td>
<td>18</td>
<td>11</td>
<td>23</td>
<td>7</td>
</tr>
<tr>
<td>Total, n (%)</td>
<td>772</td>
<td></td>
<td></td>
<td>47 (6)</td>
<td></td>
<td></td>
<td>59 (7.6)</td>
<td></td>
</tr>
</tbody>
</table>

Values refer to numbers of patients, except where noted.
evaluated 97 patients <18 years of age with failure of the Fontan circulation awaiting heart transplantation. Fifteen patients died awaiting transplant, including 2 sudden deaths. Gentles et al reported 11 late deaths in Fontan failures due to ventricular dysfunction, including 3 sudden deaths among 7 patients who had intact Fontan physiology, with 4 other deaths after Fontan takedown.

Although the number of patients in the current literature is inadequate to statistically establish ventricular dysfunction as a risk factor for SCD, it stands to reason that patients with univentricular hearts are at no less risk for SCD than adults with LV dysfunction. Multiple additional risk factors such as incompetent atrioventricular valves, residual coarctation of the aorta, and abnormalities of coronary circulation and flow reserve must be considered as additive, if not exponential risk factors. Because of the multiple complexities of univentricular physiology, these patients should be presumed to be at even higher risk for SCD in the setting of advanced ventricular dysfunction. However, the decision to proceed with ICD implant in the older Fontan patient must be made with an awareness of the risks of anesthesia and the surgical procedure and after careful consideration of other treatment options, including heart transplantation.

Left Heart Obstructive Lesions
Aortic valve disease and coarctation of the aorta are forms of CHD associated with late development of LV dysfunction. Several series have noted unexpected late SCD decades after surgical treatment of these defects, with most cases involving patients with advanced LV dysfunction. Oechslin et al reported 4 late sudden deaths among 191 survivors of surgical repair of coarctation, with 3 of the 4 victims known to have an EF <40%. Silka et al reported similar findings of 8 late sudden deaths among 536 patients after coarctation repair, with advanced LV dysfunction in 5 of 8 SCD victims. Toro-Salazar et al also reported 7 late sudden deaths among 254 long-term survivors of simple coarctation surgery, with at least 3 victims having a diagnosis of dilated cardiomyopathy. As with most other congenital heart defects, the data are limited but reasonably consistent in that the incidence of late SCD in coarctation patients is approximately 2%, with advanced ventricular dysfunction being the primary risk factor.

Aortic valve disease (stenosis/insufficiency) as well as subaortic stenosis may result in LV dysfunction due to several well-defined pathophysiologic mechanisms. Although most often acquired in older adults, congenital aortic valve disease, at times in association with multiple left heart obstructive lesions, is the most common cause of aortic valve disease in young patients. The Second Natural History Study reported the long-term outcome of 370 young patients with congenital aortic stenosis. The probability of a 25-year survival was 92.4% for patients with an initial peak gradient <50 mm Hg compared with 81.0% for patients with a gradient ≥50 mm Hg. Sudden and unexpected death occurred in 25 patients (6.7%) and was responsible for a majority of patient attrition. Although quantitative ventricular function and gradient were not available for all patients, complex ventricular arrhythmias were associated with elevated LV end-diastolic pressure, aortic insufficiency, and prior aortic valve replacement. A catheter measured LV to peak aortic gradient ≥50 mm Hg predicted a 4-fold increase in the incidence of SCD, ventricular arrhythmias, or other morbid events. Silka et al reported 10 sudden deaths among 169 patients with congenital aortic stenosis during 1860 years of follow-up. These included 4 patients with sudden, unexpected death with poor ventricular function and 2 patients with poor function who experienced acute circulatory collapse.

In general, intervention for relief of aortic stenosis or placement of a competent aortic valve is associated with improvement in LV function and clinical status, with most, if not all, LV remodeling occurring in the first 6 months after surgery. The benefits of aortic valve surgery are established provided that intervention is performed before deterioration in ventricular function (defined as an EF <50%) or marked dilation (LV end-systolic dimension ≥50 mm in adults). The latter 2 factors are predictive of clinical deterioration or sudden death at a rate of 10% to 20% per year. There also remains a subset of patients with persistent poor LV function after relief of stenosis or insufficiency in whom the lack of improvement in EF and functional status has been associated with a significant increase in sudden and total cardiac mortality. Given the known risks of SCD associated with aortic valve disease and lack of other options other than heart transplant, ICD implantation would seem prudent based on criteria established for LV dysfunction in other forms of heart disease.

Tetralogy of Fallot
The evaluation of risk factors for SCD in patients with tetralogy of Fallot (TOF) has traditionally focused on RV hemodynamics. This was because of the presumed RV origin of ventricular arrhythmias as well as the hemodynamic burden on the RV due to chronic pressure, volume overload, or both. However, recent studies of TOF have included analysis of LV function and demonstrate a definite association between LV dysfunction and risk for SCD (Table 2). Ghai et al reported SCD in 12 adults with repaired TOF, with moderate or severe LV dysfunction in 42% of the victims compared with advanced LV dysfunction in only 9% of the surviving TOF patients (P < 0.01). Furthermore, in a study by Khairy et al evaluating ICD therapy in TOF patients, an LV end-diastolic pressure ≥12 mm Hg was the most significant risk factor for appropriate ICD discharges in primary prevention patients. When compared with all other variables (QRS duration, inducible sustained VT, nonsustained VT, and RV hemodynamics), an elevated LV end-diastolic pressure was the risk factor most predictive for appropriate ICD therapy by both univariate and multivariate analysis (Figure 2). Knauth et al used MRI to determine risk.
factors for major adverse clinical outcomes in TOF patients. An adverse clinical outcome was seen in 18 of 88 patients studied and was defined as death, sustained VT, or deterioration of NYHA class. By multivariate analysis, an LVEF \( \leq 55\% \) and an RV end-diastolic volume Z score of \( \geq 7 \) were independent predictors of poor outcome. The results in these patients highlight the importance of even mild to moderate impairment of LV function.

Recently, Yap et al.\(^3\) reported the outcomes in 64 CHD adults with ICDs, including 40 patients with TOF. Although no single risk factor was statistically predictive of appropriate ICD therapy, impaired systemic LV function had the highest hazard ratio, with a value of 2.34. Furthermore, several studies have reported a higher degree of ventricular arrhythmias in TOF patients with decreased LV function.\(^4\,^2\,^4\) Similar to the Khairy study, the increased incidence of ventricular arrhythmias with LV dysfunction supports the use of ICDs in the prevention of SCD in TOF patients.

Intrinsic LV dysfunction is far less common than RV dysfunction in TOF patients, with studies suggesting an estimated 12% incidence of significant LV dysfunction in older TOF patients.\(^3\,^7\,^4\) In part, this may reflect interdependence between RV and LV function as has been reported.\(^4\,^5\) Ghai proposed that although abnormal RV hemodynamics and surgical scarring provide a substrate for re-entrant VT, LV function determines the ultimate clinical outcome of arrhythmias. Whether LV compromise increases the risk for arrhythmias, results in poor hemodynamic tolerance of ventricular arrhythmias, or is associated with other confounding factors, TOF patients with severe LV dysfunction are at risk for SCD and warrant consideration for ICD implantation.

### Management of Heart Failure in CHD Patients

Given the complexities of systemic ventricular EF determination in CHD patients, the symptoms of heart failure and functional status have also been studied as risk factors for SCD. One difficulty is that various definitions are used for heart failure, ranging from decreased maximal oxygen uptake and elevated N-terminal probrain natriuretic peptide to patients with clinical findings related to systemic or pulmonary venous congestion and requiring specialized interventions.\(^4\,^6\,^7\)

In most heart failure patients, neurohormonal activation is related to both systemic ventricular function and functional status and is associated with an increased risk of SCD. The degree to which such responses apply in complex CHD patients is uncertain. Bolger et al.\(^8\) reported a significant stepwise increase in probrain natriuretic peptide and norepinephrine related to NYHA functional status and systemic ventricular function in CHD patients. However, no relation-

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**Table 2. LV Dysfunction and Risk of Poor Outcome in TOF**

<table>
<thead>
<tr>
<th>Author</th>
<th>No. of Patients</th>
<th>No. With Poor Outcome</th>
<th>Definition of Poor Outcome</th>
<th>Definition of LV Dysfunction</th>
<th>Type of Ratio</th>
<th>Ratio</th>
<th>Range</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ghai et al(^3)</td>
<td>137</td>
<td>12</td>
<td>SCD</td>
<td>LVEF ( \leq 40% )</td>
<td>Estimated odds ratio</td>
<td>7.403</td>
<td>2.120–26.234</td>
<td>0.006</td>
</tr>
<tr>
<td>Geva et al(^3)</td>
<td>100</td>
<td>12</td>
<td>NYHA class III–IV</td>
<td>LVEF (for each 10% decrease)</td>
<td>Odds ratio</td>
<td>3.65</td>
<td>1.71–7.80</td>
<td>0.001</td>
</tr>
<tr>
<td>Yap et al(^3)</td>
<td>40</td>
<td>7</td>
<td>Appropriate shock</td>
<td>Qualitative estimate (echocardiogram)</td>
<td>Hazard ratio</td>
<td>2.34†</td>
<td>0.62–8.87</td>
<td>0.21</td>
</tr>
<tr>
<td>Knauth et al(^4)</td>
<td>88</td>
<td>18</td>
<td>Major adverse outcomes</td>
<td>LVEF (&lt;55% )</td>
<td>Odds ratio</td>
<td>7.13</td>
<td>2.11–24.0</td>
<td>0.002</td>
</tr>
<tr>
<td>Khairy et al(^5)</td>
<td>68</td>
<td>16</td>
<td>Appropriate shock (primary prevention)</td>
<td>LVEDP ( \geq 12 \textrm{ mm Hg} )</td>
<td>Hazard ratio</td>
<td>15.1</td>
<td>1.9–123.7</td>
<td>0.011</td>
</tr>
</tbody>
</table>

\(^*\)Odds ratio calculated using published data.
\(^†\)Hazard ratio for entire group of 64 congenital heart disease patients.
ship was demonstrated between the anatomic type of CHD and neurohormonal activation. Similarly, no demonstrable benefit was reported with the use of angiotensin receptor blockade (losartan) in patients with a systemic RV, attributed to minimal baseline elevation of the renin-angiotensin system.49

The results of cardiac resynchronization therapy for CHD patients with systemic LV dysfunction, particularly when associated with RV pacing, have been encouraging. However, the COMPANION study demonstrated patient survival benefit only when cardiac resynchronization therapy defibrillators study of 1520 class III or IV heart failure patients demonstrated a survival benefit only when cardiac resynchronization was combined with ICD therapy.6 Furthermore, the use of cardiac resynchronization therapy with impaired systemic right or functional univentricular physiology have demonstrated minimal benefit, with only limited data suggesting improvement.50,51 Therefore, medications and pacing interventions commonly used in other forms of heart disease have not demonstrated consistent benefit in patients with CHD, particularly in those with a systemic single or RV. We absolutely endorse the use of “optimal medical therapy” but emphasize the lack of objective benefit in many CHD patients.

ICD Therapy in CHD

The use and outcomes of ICDs in patients with CHD have been reported both as registry reports and single-center studies.5,39,52 These results are summarized in Table 3. The 2 recent studies, which combine primary and secondary prevention ICDs, indicate that ≈25% of patients receive ≥1 appropriate ICD shock, with a higher incidence of therapy in secondary prevention patients. Unfortunately, given the nature of multicenter registry studies, the data are inadequate to allow risk stratification for SCD based on either hemodynamic or electrophysiological profiles. Furthermore, as with all other ICD studies, the use of appropriate ICD shocks as a surrogate for SCD may overestimate the actual benefit of these devices.53

In a multicenter study of 121 TOF patients, Khairy et al41 reported a 23.5% incidence of ICD shocks for primary prevention and 30.2% for secondary prevention during a median follow-up of 3.7 years. As noted earlier, an elevated LV end-diastolic pressure had the most robust predictive value for appropriate ICD therapy. In comparison, the Multicenter Automatic Defibrillator Implantation Trial II, which tested the hypothesis that primary prevention ICDs would reduce mortality for adults with ischemic heart disease and an LVEF <30%, reported a 24% incidence of appropriate ICD therapy. A similar rate of ICD shocks (21%) for primary prevention of SCD in NYHA class II or III heart failure patients with an LVEF <35% was reported by the SCD-HeFT trial.4 Therefore, the incidence of appropriate ICD therapies in CHD patients (the majority of whom receive ICDs for primary prevention) seems comparable to adults with LV dysfunction who receive an ICD for primary prevention.

We acknowledge that there are a number of concerns and technical challenges regarding the use of ICDs in CHD patients. First, although ICD implantation in patients with abnormal venous or ventricular anatomy may require a surgical approach, increasingly innovative methods of implantation continue to be developed.54 Second, device-related complications, specifically lead conductor and insulation defects, occur with unacceptable frequency in all patients, and no data has been published that indicates a greater incidence of lead failure in CHD versus other ICD patients. Third, clinically significant ICD complications, defined as events requiring surgical intervention, hospitalization, or unanticipated therapies, are not unique to CHD patients. Significant complications at the time of implant were reported in 5% of SCD-HeFT patients compared with 12% in the Multicenter Pediatric Registry, with late complications in an additional 9% of patients during the SCD-HeFT trial compared with 24% in the Pediatric Registry. Finally, inappropriate shocks were reported in 83 of 829 (10%) ICD patients in the SCD-HeFT trial compared with 87 of 409 (21%) patients in the pediatric ICD registry. The relevant consideration is that although ICD implantation may be more challenging in CHD patients with complications and inappro-

<table>
<thead>
<tr>
<th>References</th>
<th>Silka et al52</th>
<th>Yap et al39</th>
<th>Berul et al5</th>
<th>Total</th>
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<tbody>
<tr>
<td>No. of CHD ICD patients</td>
<td>22</td>
<td>64</td>
<td>204</td>
<td>290</td>
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<tr>
<td>Mean age at implant, years</td>
<td>14.5</td>
<td>37</td>
<td>16</td>
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<tr>
<td>Type of CHD, n (%)</td>
<td></td>
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<td></td>
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<tr>
<td>d-TGA</td>
<td>9</td>
<td>12</td>
<td>35</td>
<td>56 (19)</td>
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<tr>
<td>TOF</td>
<td>5</td>
<td>40</td>
<td>84</td>
<td>129 (44)</td>
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<tr>
<td>Aortic stenosis/coarctation</td>
<td>5</td>
<td>4</td>
<td>29</td>
<td>38 (13)</td>
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<tr>
<td>Other</td>
<td>3</td>
<td>8</td>
<td>56</td>
<td>67 (23)</td>
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<tr>
<td>Secondary prevention ICD, %</td>
<td>100</td>
<td>61</td>
<td>48</td>
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</tr>
<tr>
<td>Primary prevention ICD, %</td>
<td>0</td>
<td>39</td>
<td>52</td>
<td></td>
</tr>
<tr>
<td>Patients with appropriate ICD therapy, n (%)</td>
<td>15 (68%)</td>
<td>15 (23%)</td>
<td>53 (26%)</td>
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</table>
Applying Existing Guidelines to CHD Patients

SCD is the leading cause of death in adolescents and adults with CHD. On the basis of this review, there is a definite association between advanced systemic ventricular dysfunction and SCD in specific forms of postoperative CHD, which in all probability is relevant for all forms of CHD. In the past decade, there have been significant advances in ICD technology as well as prospective clinical trials in adults with ischemic and nonischemic cardiomyopathy that have consistently demonstrated benefits of ICD therapy in adults with LV dysfunction. In combination, these factors have resulted in general acceptance of ICD implantation in adults with severe ventricular dysfunction.

After the Multicenter Automatic Defibrillator Implantation Trial II and SCD-HeFT trials, the relationship between advanced systemic ventricular dysfunction and SCD has become a topic of major relevance in patients with CHD. Unfortunately, the heterogeneity of CHD as well as limited patient numbers in each of these subtypes has limited the ability to statistically prove an exact association between ventricular dysfunction and SCD. However, making decisions based on limited data are a dilemma frequently encountered by those treating patients with CHD, with important decisions made by use of prospective, randomized adult trials in conjunction with the available data in children and adults with CHD. For the current question regarding ICD placement in CHD patients with severe ventricular dysfunction, this extrapolation seems very reasonable. Indeed, it is unrealistic to argue that CHD patients with a systemic LV and an EF <30% should be at lower risk for SCD than adults with (non)ischemic cardiomyopathy with a similar degree of ventricular dysfunction. Another factor that must also be considered is the number of life-years at risk, with most CHD patients with advanced ventricular dysfunction <40 years of age, compared with an average age of 63 years for ICD implantation for adults with ischemic heart disease.7

It is important to emphasize that we are discussing specific CHD patients, those with advanced systemic ventricular dysfunction, which by best estimate, account for <5% of such patients. Using an EF of 30% as a cutoff for ICD placement may be conservative, because this degree of dysfunction is both unusual and often associated with clinical symptoms. The basis for a 30% EF as the critical ICD value is well established by adult studies and seems equally valid for patients with systemic right or single ventricles by this analysis. Given the considerations of impaired coronary perfusion, abnormal RV geometry and hypertrophy, and predisposition to both atrial and ventricular arrhythmias, a 30% EF may be too low of a threshold to consider as the indication for ICD implantation for patients with systemic right or functional single ventricles.

A more difficult decision is presented by the CHD patient with moderate systemic ventricular dysfunction (30% to 40% EF). The use of additional factors such as QRS duration, spontaneous or inducible arrhythmias, and heart failure status in addition to individual judgment is advised. In these more borderline patients, defining when risk for SCD is enough to merit a technically challenging ICD implant may prove far more difficult.

Disclosures

Drs Silka and Bar-Cohen have no disclosures related to industry or other support. Dr Silka was a writing member of the 2006 ACC/AHA/ESC Guidelines Committee on Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death and the 2008 ACC/AHA/HRS Guidelines Committee on Device-Based Therapy of Cardiac Arrhythmias. The topic of this debate was not addressed in either guideline.

References


**KEY WORDS:** death, sudden heart arrest, heart defects, congenital heart failure, defibrillator (ICD)

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**Response to Silka and Bar-Cohen**

*John K. Triedman, MD*

Primary prevention strategies for implantable cardioverter defibrillator (ICD) implantation depend on algorithms that are based on clear, evidence-based criteria to guide patient screening. The utility of left ventricular ejection fraction as a dichotomous variable to evaluate patients with cardiomyopathy is an astonishingly simple, powerful finding, based on well-designed studies that enrolled many thousands of patients. Drs. Silka and Bar-Cohen’s presentation emphasizes the fundamental problem confronting cardiologists who wish to extrapolate these elegant findings to adults with congenital heart disease (CHD). This is a very messy group: the patients are heterogeneous and described by studies that are predominantly retrospective and small in number and size.

The authors suggest that because the relation of ventricular dysfunction and arrhythmia appears to define a final common pathway predisposing to cardiac arrest in acquired cardiomyopathy, it is “reasonable” and “prudent” to infer similar processes—and by extension, outcomes and responses to therapy—in CHD patients. Unfortunately, there is no evidence or a priori reason to believe that this is true at the level of detail necessary to craft recommendations for care guidelines. One might alternatively argue that differences between these populations (eg, ventricular morphology and the causes and time course of myocardial injury and hypertrophy) are more important than their similarities and that observed differences in outcome (lower sudden death rates, higher ICD complication rates) will critically influence imputed risk:benefit ratios. Assessment of ventricular function is of clear importance in risk assessment but should remain just one of many factors considered in therapy decisions for this special group.
Should patients with congenital heart disease and a systemic ventricular ejection fraction less than 30% undergo prophylactic implantation of an ICD?: Patients With Congenital Heart Disease and a Systemic Ventricular Ejection Fraction Less Than 30% Should Undergo Prophylactic Implantation of an Implantable Cardioverter Defibrillator

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