A Population-Based Prospective Evaluation of Risk of Sudden Cardiac Death After Operation for Common Congenital Heart Defects

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Objectives. This study sought to define 1) the risk of sudden death after operation for common congenital heart defects; and 2) factors associated with an increased risk of sudden death.

Background. Although the prognosis for patients with congenital heart defects is improved by surgical treatment, they remain at a well recognized but poorly defined risk of late sudden death.

Methods. This population-based study evaluated all patients <19 years old undergoing surgical treatment of common forms of congenital heart disease in the state of Oregon between 1958 and 1996. Patients were identified retrospectively through 1958, with prospective biannual follow-up beginning in 1982. The incidence and cause of late sudden death were evaluated for 3,589 patients surviving operation for the following defects: atrial, ventricular and atrioventricular septal defects; patent ductus arteriosus; pulmonary stenosis; aortic stenosis; coarctation of the aorta; tetralogy of Fallot; and d-transposition of the great arteries.

Results. There were 41 unexpected late sudden deaths during 45,857 patient-years of follow-up, an overall event rate of 1/1,118 patient-years. Thirty-seven of the 41 late sudden deaths occurred in patients with aortic stenosis, coarctation, transposition of the great arteries or tetralogy of Fallot, an event rate of 1/454 patient-years. In contrast, only four sudden deaths occurred among these patients during short-term follow-up. Furthermore, it is uncertain whether risk factors proposed for patients with TOF are relevant to postoperative patients with other forms of congenital heart disease.

Conclusions. The risk of late sudden death for patients surviving operation for common congenital heart defects is 25 to 100 times greater than an age-matched control population. This increased risk is primarily represented by patients with cyanotic or left heart obstructive lesions. The risk of sudden death appears to be time dependent, increasing primarily after the second postoperative decade.

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Methods

Patients. The study evaluated all Oregon residents undergoing surgical treatment of congenital heart disease between 1958 and 1996. Patient inclusion criteria were age <19 years at initial operation and survival >30 days after operation. During the study interval, 99.5% of operations for congenital heart disease and the relatively low incidence of sudden death among these patients during short-term follow-up. Furthermore, it is uncertain whether risk factors proposed for patients with TOF are relevant to postoperative patients with other forms of congenital heart disease.

This study of sudden cardiac death (SCD) is part of a prospective population-based study of patients who have undergone surgical treatment of congenital heart disease. The overall 25-year survival of this cohort has been the subject of another report (8). The purpose of the present study was to specifically evaluate the risk of SCD during long-term follow-up after surgical treatment of congenital heart disease. Secondary objectives were to compare the relative risks and causes of sudden death among the various heart defects and determine whether objective factors could be identified that predicted an increased association with late sudden death.

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A registry of all patients with congenital heart disease was initially established in 1982, with retrospective identification of all patients undergoing surgical treatment of congenital heart disease since 1958. Patient accrual has been prospective since that time. After establishment of the registry, biannual follow-up has been determined by clinical evaluation, telephone interview or mailed questionnaire. If no response was obtained, the National Death Index was reviewed for identification of missing patients. Patients in whom earlier follow-up had been obtained, but whose status was not determined in the most recent data cycle, were included in analysis up to the time of most recent follow-up. The most recent data cycle was completed in 1996.

For the purposes of this study, patients with the following congenital heart defects were evaluated: secundum atrial septal defect, ventricular septal defect, atroventricular (AV) septal defect (including primum atrial septal defect), patent ductus arteriosus, pulmonary stenosis, aortic valve stenosis (AS), coarctation of the aorta (CoA), TOF and D-transposition of the great arteries (D-TGA). These defects were selected because they represent all defects that would allow at least 20 years of postsurgical follow-up and adequate numbers to permit inferential statistical analysis. Patients with complex forms of these defects, such as AV septal defect with TOF or D-TGA with right ventricular hypoplasia were excluded from this analysis.

**Definitions.** SCD was defined as acute cardiovascular collapse from which biologic death occurred or from which the patient never regained consciousness. The mechanisms of death were classified according to modification of criteria proposed by the Cardiac Arrhythmia Pilot Study (9): 1) sudden arrhythmic (proven or presumed); 2) sudden nonarrhythmic (i.e., embolism or aneurysm rupture); 3) nonsudden cardiac death (i.e., reoperation or congestive heart failure); and 4) noncardiac (i.e., trauma, malignancy). Patients with non-sudden cardiac death or noncardiac causes of death were included in the cohort until the time of death.

**Statistical analysis.** The statistical methods for this analysis have been previously reported in detail (8). Survival was estimated by the Kaplan-Meier method, defined as the elapsed time from the date of operation to date of death or last known follow-up. The Mantel-Cox statistic was used for comparison of survival between groups of patients.

### Results

**Patients.** According to the aforementioned inclusion criteria, a total of 3,589 patients were identified, from whom a total follow-up of 45,857 patient-years was derived. The distribution of the individual forms of congenital heart disease and follow-up are shown in Table 1. In this registry, 91.5% of patients are non-Hispanic white, 2.4% Asian, 3.2% Hispanic, 2.1% African-American and 0.8% native-American. The gender distribution differs markedly by defect on the basis of natural occurrence; the groups with TOF, D-TGA, AS and CoA are predominately male, whereas those with an atrial or AV septal defect or patent ductus arteriosus are predominately female. Follow-up for survival through 1994 (the beginning of the last completed data cycle) was completed in 90% of the

**Table 1. Specific Congenital Heart Defects and Incidence of Sudden and Nonsudden Cardiac Death**

<table>
<thead>
<tr>
<th>Defect</th>
<th>No. (%) of Pts With Complete Follow-Up</th>
<th>Total Follow-Up (pt-yr)</th>
<th>Sudden Cardiac Death</th>
<th>Non-Glucose Cardiac Death</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>No.</td>
<td>Incidence/1,000 Pts-yr</td>
</tr>
<tr>
<td>ASD</td>
<td>622 (86%)</td>
<td>7,904</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>VSD</td>
<td>527 (87%)</td>
<td>6,354</td>
<td>1</td>
<td>0.2</td>
</tr>
<tr>
<td>AVSD</td>
<td>254 (87%)</td>
<td>2,217</td>
<td>2</td>
<td>0.9</td>
</tr>
<tr>
<td>PDA</td>
<td>623 (82%)</td>
<td>8,753</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>PS</td>
<td>241 (91%)</td>
<td>3,568</td>
<td>1</td>
<td>0.3</td>
</tr>
<tr>
<td>AS</td>
<td>169 (94%)</td>
<td>1,860</td>
<td>10</td>
<td>5.4</td>
</tr>
<tr>
<td>CoA</td>
<td>536 (92%)</td>
<td>6,706</td>
<td>9</td>
<td>1.3</td>
</tr>
<tr>
<td>TOF</td>
<td>445 (91%)</td>
<td>7,082</td>
<td>11</td>
<td>1.5</td>
</tr>
<tr>
<td>D-TGA</td>
<td>172 (95%)</td>
<td>1,413</td>
<td>7</td>
<td>4.9</td>
</tr>
<tr>
<td>Total</td>
<td>3,589</td>
<td>45,857</td>
<td>41</td>
<td>0.9</td>
</tr>
</tbody>
</table>

AS = aortic stenosis; ASD = atrial septal defect; AVSD = atroventricular septal defect; CoA = coarctation of the aorta; PDA = patent ductus arteriosus; PS = pulmonary stenosis; pt-yr = patient-years; Pts = patients; D-TGA = dextro-transposition of the great arteries; TOF = tetralogy of Fallot; VSD = ventricular septal defect.
entire cohort, ranging from 82% in patients with a patent ductus arteriosus to 95% in patients with D-TGA.

Events. There were a total of 41 sudden, unexpected late deaths during the 45,857 patient-years, for an event rate of 0.9/1,000 patient-years, or 1 event/1,118 patient-years. Thirty-seven of the 41 late SCDs occurred in patients with four defects: TOF (n = 11), D-TGA (n = 7), CoA (n = 9) and AS (n = 10). The cumulative event rate for patients with these four defects was 1/454 patient-years, or 2.2 events/1,000 patient years. In comparison, the SCD event rate for patients with left to right shunt lesions or pulmonary stenosis was 1/7,154 patient years, or 0.14/1,000 patient years (p, 0.001). The observed incidence of sudden death ranged from 0 in patients with an atrial septal defect or patent ductus to ~5/1,000 patient-years for patients with AS or D-TGA (Table 1). During this study period, 74 other patients died of non–sudden cardiac causes (reoperation [n = 32], chronic congestive heart failure [n = 22] and pulmonary hypertension [n = 20]), whereas 61 patients died of noncardiac causes.

Mechanisms of unexpected SCD. On the basis of information obtained from witnesses, medical or rescue squad reports and autopsy data, a determination of the actual or probable cause of SCD was made. The causes of the 41 SCDs were arrhythmia in 30 patients, circulatory in 7 (embolic in 5, aneurysm rupture in 2) and acute ventricular failure in 4. The causes of SCD among specific forms of congenital heart disease are illustrated in Figure 1.

Of the 30 patients with an arrhythmic cause of SCD, 14 had ventricular tachycardia or fibrillation documented at time of attempted resuscitation, whereas 2 had complete AV block with an agonal idioventricular rhythm. Of the 14 patients with a presumed arrhythmic cause of SCD, 6 collapsed suddenly during exertion, and 4 died in their sleep; in 4 patients, the circumstances at time of cardiovascular collapse were not clearly recorded.

Temporal analysis of the incidence of SCD. The incidence of late SCD was evaluated as a function of the duration of follow-up, both for the entire cohort and for patients with specific congenital heart defects. For patients with D-TGA, the risk of late SCD was relatively constant, with the incidence of SCD estimated at 4 ± 7% at 10 years and 9 ± 4% at 20 years of follow-up (mean ± SEE). In contrast, the incidence of late sudden death for patients with a previous repair of TOF increased primarily after 20 years of follow-up. At 20 years of follow-up, the incidence of SCD was 2 ± 1%, increasing to 4 ± 1% at 25 years and 6 ± 7% at 30 years (Fig. 2).

An apparent time-dependent incremental risk of late SCD was also observed for patients with left-heart obstructive lesions. The estimated incidence of late SCD for patients with AS was 3 ± 2% at 10 years of follow-up, increasing to 13 ± 5% at 20 years and 20 ± 8% at 30 years. For patients with CoA, the incidence of SCD was 1 ± 0.5% at 20 years of follow-up, increasing to 5 ± 2% at 25 years and 8 ± 3% at 30 years (Fig. 3). Two of the nine patients with CoA who died suddenly had undergone subsequent operation for AS at 14 and 15 years after their initial operation, respectively. However, because the index surgical procedure for entry into this study was for CoA, these patients were included in the analysis of the cohort with this defect.

Causes of late SCD. Several observations were made among the patients who died suddenly: 1) of the seven patients with D-TGA who died suddenly, five were participating in...
The possible associations between surgical era and patient age at definitive operation and the occurrence of late SCD were evaluated. The risk of late SCD by age at operation was not performed for patients with D-TGA because the operation was usually performed within the first 3 months of life. After 15 years of follow-up, no association was observed between age at operation and incidence of late SCD for any defect (Table 3). Additionally, no statistically significant association between SCD and either gender, race or presence of genetic or extracardiac abnormalities was identified.

**Secondary risk factor analysis.** The possible associations between surgical era and patient age at definitive operation and the occurrence of late SCD were evaluated. The risk of late SCD as a function of surgical era was considered a comparison of patients undergoing operation before 1980 with those operated on in 1980 and later. This time frame was chosen because these intervals would allow comparable follow-up of patients with up to 15 years of follow-up data during both periods. This analysis was restricted to patients with TOF, CoA and AS because of the limited number of SCD events among patients with other forms of CHD and because the surgical techniques for D-TGA have evolved from the Mustard and Senning atrial switch procedures to arterial switch methods during the past decades. There was no observed difference in the incidence of late SCD for the three aforementioned defects through 15 years of follow-up, whether operated on before 1980 or later (Table 2). However, these data do not reflect the significant reductions in the perioperative deaths that have occurred over the decades.

Age at primary operation was the second variable that was evaluated in relation to the risk of late SCD. This analysis compared patients undergoing surgical repair of their congenital heart disease at age <2 years compared with those undergoing operation at ≥2 years of age. Analysis of the risk of late SCD by age at operation was not performed for patients with D-TGA because the operation was usually performed within the first 3 months of life. After 15 years of follow-up, no association was observed between age at operation and incidence of late SCD for any defect (Table 3). Additionally, no statistically significant association between SCD and either gender, race or presence of genetic or extracardiac abnormalities was identified.

**Discussion**

The unexpected SCD of a young person from cardiovascular disease is a tragic, but fortunately, extremely uncommon event in the general population. Several population-based studies (10–12) have evaluated the risk of SCD in young patients (<20 years old), with the reported incidence between 1/25,000 and 1/100,000 patient-years. The incidence of unexpected SCD has also been the subject of study among selected subpopulations of young patients, particularly young athletes or patients with known cardiovascular diagnoses, such as

**Table 2.** Probability of Sudden Death-Free Survival by Surgical Era and Duration of Follow-Up

<table>
<thead>
<tr>
<th>Surgical Era</th>
<th>No. of Pts</th>
<th>Postoperative Follow-Up Duration (mean ± SEE)</th>
<th>5 Years</th>
<th>10 Years</th>
<th>15 Years</th>
</tr>
</thead>
<tbody>
<tr>
<td>TOF</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;1980</td>
<td>265</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td></td>
</tr>
<tr>
<td>≥1980</td>
<td>180</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td></td>
</tr>
<tr>
<td>AS</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;1980</td>
<td>63</td>
<td>0.98 ± 0.02</td>
<td>0.97 ± 0.02</td>
<td>0.95 ± 0.03</td>
<td></td>
</tr>
<tr>
<td>≥1980</td>
<td>106</td>
<td>0.97 ± 0.02</td>
<td>0.97 ± 0.02</td>
<td>0.88 ± 0.09</td>
<td></td>
</tr>
<tr>
<td>CoA</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;1980</td>
<td>236</td>
<td>1.0</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td></td>
</tr>
<tr>
<td>≥1980</td>
<td>300</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations as in Table 1.

**Table 3.** Probability of Sudden Death-Free Survival by Age at Operation and Duration of Postoperative Follow-Up

<table>
<thead>
<tr>
<th>Age at Operation (yr)</th>
<th>No. of Postop Survivors</th>
<th>Postop Follow-Up Duration (mean ± SEE)</th>
<th>5 Years</th>
<th>10 Years</th>
<th>15 Years</th>
<th>20 Years</th>
<th>25 Years</th>
</tr>
</thead>
<tbody>
<tr>
<td>TOF</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;2</td>
<td>236</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td>0.95 ± 0.04</td>
<td></td>
</tr>
<tr>
<td>≥2</td>
<td>209</td>
<td>0.98 ± 0.01</td>
<td>0.98 ± 0.01</td>
<td>0.98 ± 0.01</td>
<td>0.97 ± 0.01</td>
<td>0.96 ± 0.02</td>
<td></td>
</tr>
<tr>
<td>AS</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;2</td>
<td>59</td>
<td>0.98 ± 0.02</td>
<td>0.98 ± 0.02</td>
<td>0.98 ± 0.02</td>
<td>0.98 ± 0.02</td>
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<tr>
<td>≥2</td>
<td>110</td>
<td>0.98 ± 0.02</td>
<td>0.96 ± 0.07</td>
<td>0.93 ± 0.03</td>
<td>0.85 ± 0.05</td>
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<tr>
<td>CoA</td>
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<tr>
<td>&lt;2</td>
<td>262</td>
<td>0.98 ± 0.01</td>
<td>0.98 ± 0.01</td>
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<tr>
<td>≥2</td>
<td>274</td>
<td>1.0</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td>0.99 ± 0.01</td>
<td>0.94 ± 0.03</td>
<td></td>
</tr>
</tbody>
</table>

Postop = postoperative; other abbreviations as in Table 1.
Wolf-Parkinson-White syndrome or the congenital long QT syndromes (13–15). Most of these studies have excluded patients with previous surgical treatment for congenital heart disease because of a presumed higher risk of late SCD.

As the general population of patients surviving operation for congenital heart defects continues to achieve greater longevity, questions regarding the long-term prognosis for these patients become increasingly pertinent (16). These include the relative risk of late SCD among patients with diverse forms of congenital heart disease, potential identification of risk factors for late SCD and the requirement for ongoing medical evaluation decades after ostensibly corrective cardiac surgery. The purpose of the present study was to address these issues in a population-based study of patients after operation for common forms of congenital heart disease, in whom long-term survival would be anticipated.

Late SCD, years or decades after operation for congenital heart disease, has been the subject of several reports (1–6). However, most reports have focused on one specific defect, such as TOF or d-TGA, in which there is a relatively high incidence of late SCD (17,18). In contrast, one purpose of the present population-based study was to evaluate the comparative risk of late SCD among patients with various common defects, given the long-term follow-up, high rates of patient follow-up and unbiased population study design.

Factors associated with increased risk of late SCD. The two principal findings of this study were the marked variance in the relative risks for late SCD among patients with various defects and the apparent time-dependent nature of this risk for certain defects. Overall, the incidence of late SCD in the entire cohort was 0.9/1,000 patient-years, representing an increased risk of between 25 and 100 times that for the general population. However, the incidence of late SCD of 1/7,154 patient-years for patients with left to right intracardiac shunt lesions or pulmonary stenosis approaches that of the general population (1/25,000 to 100,000 patient years). In contrast, the incidence of late SCD was markedly increased in patients with left heart obstructive lesions and cyanotic defects, with the estimated risk between 50 and 200 times that of the general population. The marked increase in the incidence of late SCD was primarily accounted for by patients with AS and d-TGA, in whom the incidence of late SCD was ~1/200 patient-years.

Late SCD after repair of d-TGA. Further evaluation of the incidence of SCD for specific defects demonstrated an early onset and relatively constant risk for patients undergoing the Mustard or Senning procedures for d-TGA, with estimated SCD-free survival rates of 96% at 10 years and 91% at 20 years of postoperative follow-up. The risk of late SCD in patients with d-TGA in this series is similar to that reported in separate series by Gewillig et al. (6) and Gelatt et al. (18), although those two studies primarily evaluated total actuarial survival rather than late SCD. In those two studies, actuarial total cardiac death-free survival rates of 67% and 76% at 20 years of postsurgical follow-up were reported. By comparison, the total cardiac death-free survival rate for patients in the present study with d-TGA at 20 years was 71%.

Late SCD after repair of TOF. The incidence and risk factors for late SCD after operation for TOF have both been the subject of multiple studies that have involved single institutions as well as collaborative efforts (1,17,19,20). Although comparison of those studies is limited by differences in design, definition and length of follow-up, a meta-analysis of 39 studies (21) reported a 1.8% incidence of late SCD among 4,583 patients with an average of 8.3 years of follow-up after surgical treatment of TOF. This relatively low incidence of SCD is consistent with the findings of the present study, where the incidence of late SCD at 10 years of follow-up was 1.2%. A relatively low incidence of late SCD after surgical treatment of TOF was also observed in our study during the second postoperative decade, with a cumulative 20-year actuarial incidence of late SCD of 2.2%. However, incremental increases in the incidence of SCD to 4% at 25 years and 6% at 30 years of follow-up suggest a time-dependent risk of late death.

Murphy et al. (22) also reported a 6% incidence of late SCD among patients with 30 years of follow-up after surgical treatment of TOF (10 deaths among 163 patients). However, although identical rates of late SCD during equivalent follow-up are reported in that (22) and our study, a late or time-dependent risk for late SCD was not observed by Murphy et al. In light of the small numbers of events in both series, the need for cautious interpretation of data would seem prudent. Conversely, it would seem reasonable to conclude that the earlier estimates of a 5% to 10% risk of late SCD during adolescence after repair of TOF have not been substantiated by long-term follow-up.

Late SCD in patients with AS. The highest incidence of late SCD in the present series was observed among patients with AS, with an incidence of 3% at 10 years of follow-up, increasing to 13% at 20 years and 20% at 30 years of follow-up. This finding is somewhat surprising in view of the paucity of published data on SCD in young patients with AS. The issue of late SCD among postsurgical patients with AS was noted by the Second Natural History Study of Congenital Heart Defects (23). In that study, there were 19 late unexpected SCDs (presumably arrhythmic) and 4 deaths from bacterial endocarditis among 240 surgically managed patients. These events occurred over 15 years of follow-up and represent a nearly 10% incidence of late SCD during this interval. A significant incidence of arrhythmic SCD was also observed among young patients with AS in registry data reported for patients who had undergone implantable cardioverter-defibrillator placement after resuscitation from sudden cardiac arrest (24). Also, the risk of late SCD from embolic sequelae of endocarditis may be an underestimated factor in the evaluation of these patients.

Late SCD after repair of CoA. The finding of late SCD among patients with CoA was unanticipated because only 1 of 536 survivors died suddenly during the first 20 years of follow-up. However, recent studies (25) have reported unexplained advanced left ventricular hypertrophy in patients with previous repair of CoA, with the suggestion that the biologic template for advanced ventricular hypertrophy may be estab-
lished at a young age, with complete expression only during early adulthood. This finding emphasizes the need for ongoing follow-up of these patients despite apparent complete relief of aortic obstruction at rest. Similar caution and concern are also warranted for patients after patch aortoplasty repair of CoA, who require long-term follow-up for both the occurrence and significance of aortic aneurysm after this procedure (26).

Factors not associated with increased risk of late SCD. Two additional issues that were examined in our study were impact of the surgical era on late incidence of SCD and whether early repair (age <2 years) was associated with a lower risk of late SCD. With regard to surgical era (before 1980 vs. 1980 and later), no difference in the incidence of late SCD was observed for the same defects with equal duration of follow-up. However, there were distinct differences in the operative mortalities for the two eras, a subject that has been addressed previously (8). Additional follow-up of the cohort through 30 years after operation will be required to determine whether the trend for increasing rates of mortality over a longer postoperative interval will be repeated in the group operated on in the more recent era. However, it would appear that the duration of postsurgical follow-up rather than the surgical era may be the critical factor in defining the risk of late SCD.

With recent advances in surgical techniques, it has been proposed that early primary repair may reduce the risk of late SCD (27). However, others have suggested that longer follow-up will be required to substantiate this observation. The results of the present study are consistent with the latter proposal: Decades rather than years of follow-up may be required to validate a benefit of early primary repair. In our study, comparison of patients undergoing operation at <2 years versus ≥2 years of age failed to demonstrate a significant difference in the incidence of late SCD with up to 25 years of follow-up. Although it is possible that significant differences may emerge with longer follow-up, these differences have yet to be established.

Study limitations. The present population-based study of patients undergoing operation for common congenital heart defects since 1958 included only residents of Oregon. A small number of patients have incomplete follow-up data because of patient relocation or failure to respond to inquiries on status. However, it is unlikely that catastrophic events would not have been detected in light of the multiple methods of investigation and 90% follow-up through 1996. Also, because of the observational study design, there are distinct limitations in defining secondary risk factors for late SCD, such as the lack of consistent objective data regarding measures of ventricular function.

Classification of the exact cause of SCD may be complicated by the coexistence of factors such as venous obstruction and atrial flutter in a patient with a previous Mustard repair of D-TGA. In the present study, the classification of presumed arrhythmic death was made only in the absence of other identifiable causes of SCD. Conversely, arrhythmias were invariably present in patients who died of acute ventricular failure or coronary embolism, although these arrhythmias were interpreted as sequelae rather than causes of SCD.

Conclusions. 1) The risk of late SCD during long-term follow-up is diverse among patients with heterogeneous forms of congenital heart disease. For some, the risk is only slightly higher than the general population; conversely, a much higher risk is present for patients with other defects, such as AS or D-TGA. 2) The risk of late SCD may be time dependent for high risk defects, increasingly primarily ≥20 years after operation. Further follow-up will be required to determine whether this risk is linear or exponential. 3) Most but not all SCD in these patients is presumably due to arrhythmic causes. Future studies should consider whether some late SCDs may be due to circulatory rather than arrhythmic causes.

References


