Sudden Death After Repair of Double-Outlet Right Ventricle

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The outlook for patients with double-outlet right ventricle has improved since the development of corrective operations. Late arrhythmic deaths after successful procedures have been reported; however, the magnitude remains unknown. This study was undertaken to identify the magnitude of late sudden death and the significant factors associated with it. From 1965 through 1985, 118 patients underwent corrective operation for double-outlet right ventricle; of these, 23 died in the hospital and six were lost to follow-up. The 89 remaining patients (52 male and 37 female) made up the study population. Their mean age (±SD) was 10.3±7.8 years at the time of repair. The mean duration of follow-up was 82 months. Of the 22 late deaths, 16 (73%) were sudden. Eight (50%) of the sudden deaths occurred within 1 year of operation. Cox proportional hazards multivariate analysis revealed the following significant risk factors for late sudden death: older age at the time of operation, perioperative or postoperative ventricular tachyarrhythmias, and third-degree atrioventricular block. Factors not associated with late sudden death included year of operation, sex, type and number of associated cardiac anomalies, preoperative functional class, previous palliative procedures, surgical technique, perioperative or postoperative single premature ventricular contractions, and postoperative left or right bundle branch block with or without fascicular block. We conclude that the incidence of late sudden death after successful surgical repair of double-outlet right ventricle is very high. Complete corrective operation at an early age and aggressive diagnosis and treatment of arrhythmias and conduction defects after operation are warranted. (Circulation 1990;81:128–136)

Double-outlet right ventricle (DORV) is a rare congenital cardiac anomaly that includes a diverse group of malformations sharing the common feature that both great arteries arise primarily from the right ventricle.1,2 The natural history and the clinical spectrum vary depending on the location of the ventricular septal defect (VSD), the presence or absence of pulmonary stenosis, and other associated cardiac anomalies.3,4 The overall outlook for patients with DORV has improved greatly since the development of corrective procedures. The surgical mortality has continued to decrease5–12; however, a distressingly high incidence of late deaths, some sudden, has been reported.9,10 The magnitude of this problem and associated causative factors are unknown. Therefore, we undertook a retrospective study to identify patients at risk for late death with special attention to the occurrence of sudden death.

Methods

Patient Selection

The clinical records of all patients who underwent surgical correction for DORV at the Mayo Clinic between January 1965 and December 1985 were reviewed. The Mayo Clinic served as a referral center for all patients reported in this study except for one native resident of Rochester, Minnesota. DORV was defined as both great arteries arising completely or almost completely (more than 50%) from the morphological right ventricle; mitral-aortic or mitral-pulmonic continuity may or may not have been present.5,13–15 The diagnosis was made by angiography, echocardiography, or both and confirmed at operation for all patients. Patients with other cardiac anomalies seriously altering the hemodynamic and surgical characteristics of DORV were excluded from the study. These conditions included atrioventricular discordance, atrioventricular canal, univentricular heart, and DORV with subpulmonic VSD (Taussig-Bing anomaly).

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**Table I. Risk Factors Analyzed for Late Sudden Death in Patients With Double-Outlet Right Ventricle**

<table>
<thead>
<tr>
<th>Clinical</th>
<th>Hemodynamic</th>
<th>Anatomic</th>
<th>Surgical</th>
<th>Electrocardiographic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>Mean right atrial pressure</td>
<td>Ventricular septal defect</td>
<td>Type of ventriculotomy</td>
<td>Supraventricular arrhythmia</td>
</tr>
<tr>
<td>Sex</td>
<td>Mean pulmonary arterial pressure</td>
<td>Atrial septal defect</td>
<td>Ventricular septal defect enlargement</td>
<td>Paroxysmal supraventricular tachycardia</td>
</tr>
<tr>
<td>Functional class</td>
<td></td>
<td>Pulmonary stenosis</td>
<td>Infundibular septal excision</td>
<td>Atrial fibrillation/atrial flutter</td>
</tr>
<tr>
<td>Cardiac size</td>
<td></td>
<td>Patent ductus arteriosus</td>
<td>Atrial septal defect closure</td>
<td>Ventricular arrhythmia</td>
</tr>
<tr>
<td>Pulmonary vasculature</td>
<td></td>
<td>Pulmonary venous anomaly</td>
<td>Right ventricular outflow tract</td>
<td>Premature ventricular contractions</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Bilateral superior vena cava</td>
<td>reconstruction</td>
<td>Couplets</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Anomalous coronary artery</td>
<td>Previous palliative operation</td>
<td>Multiform premature ventricular contractions</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Juxtaposed atrial appendages</td>
<td>Year operation was performed</td>
<td>Ventricular tachycardia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Aortopulmonary artery interrelationship (malposition)</td>
<td></td>
<td>Sick sinus syndrome</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Conduction defect</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>First-degree block</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Second-degree block</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>(type I and type II)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Third-degree block</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Left bundle branch block</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Right bundle branch block</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Right bundle branch block + fascicular block</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Ventricular hypertrophy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Left ventricular</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Right ventricular</td>
</tr>
</tbody>
</table>

**Surgical Procedure**

The operative techniques have been described in detail previously. The aim of operation is to establish an unobstructed outflow tract from the left ventricle to the aorta through the VSD. This left ventricular outflow tract consists of a tunnel leading from the VSD to the aortic annulus; the tunnel is constructed using a patch of prosthetic material. If the VSD is restrictive, a wedge of septum is excised from its anterosuperior aspect to enlarge the VSD without injury to the conduction tissue. Pulmonary stenosis can be corrected by any of the following: 1) infundibular resection and pulmonary valvotomy, 2) patch enlargement of the right ventricular outflow tract, which may or may not extend across the pulmonary annulus into the pulmonary artery, or 3) placement of a valved external conduit. The respective indications for each type of reconstruction will not be discussed here.

**Data Collection**

For purposes of analysis, the variables to be studied were categorized into the following five major groups (Table 1): clinical, hemodynamic, anatomic, surgical, and electrocardiographic. These were further divided into four time periods: preoperative, the time before operation; perioperative, within 24 hours of operation (including intraoperative time); postoperative, before hospital dismissal; and follow-up, after hospital dismissal. The patient's preoperative functional status was defined according to the New York Heart Association classification. Cardiac size (cardiothoracic ratio) and pulmonary vasculature (normal, increased, or decreased) were assessed from chest radiographs. The mean right atrial pressure (normal, less than 8 mm Hg; abnormal, 8 mm Hg or more) and the mean pulmonary arterial pressure (normal, less than 20 mm Hg; abnormal, 20 mm Hg or more) were measured preoperatively at cardiac catheterization. In addition, the type of VSD was defined as being restrictive or nonrestrictive; the location of the VSD was identified as subaortic, uncommitted, or doubly committed. Ventricular arrhythmias were defined as nonsustained ventricular tachycardia (3 beats to 30 seconds in duration and self-terminating) or sustained ventricular tachycardia (longer than 30 seconds in duration and not self-terminating). Ventricular fibrillation during reconstitution of coronary perfusion after aortic cross-clamping and hypothermia were excluded from the group with perioperative ventricular tachyarrhythmias.

Follow-up information was obtained by sending a questionnaire to each patient and to the referring physician if the patient had not been seen at the Mayo Clinic for more than 6 months. The questions asked were date of last medical checkup, functional status, electrocardiographic and symptomatic evidence for an arrhythmia, and treatment for arrhythmia (none, antiarrhythmic medication, pacemaker, or both medication and a pacemaker). If the patient had died, death was defined as sudden or nonsudden; sudden death was defined as death occurring within 1 hour of the onset of symptoms or as sudden unexplained death when the death was not witnessed. Additional information was sought individually from a patient's family and physician regarding the specific circumstances surrounding a death. Autopsy reports were requested when available.

**Statistical Analysis**

The observed proportions of discrete events in two or more groups were compared with \( \chi^2 \) tests. Signif-
tricular block. Postoperatively, the patient had a junctional escape rhythm without any symptoms; a permanent pacemaker was not implanted. She died 4 months after operation. One patient (case 10) developed postoperative ventricular tachycardia, which was effectively treated with quinidine. The quinidine therapy was discontinued 2 years after operation, at which time he was completely asymptomatic; however, he died suddenly 2 weeks after quinidine was discontinued. One patient (case 11) was reported to have paroxysmal supraventricular tachycardia unresponsive to digoxin and propranolol; she died in the emergency room during tachycardia. The remaining three patients (cases 8, 9, and 14) of the nine patients who died during routine activities died suddenly at home while walking or watching television.

Survival analysis of late sudden death beginning after hospital dismissal and ending at mean follow-up (7 years) is shown in Figure 2. The cumulative survival probability with respect to late sudden death (excluding the six nonsudden deaths) was 91% at 1 year and 80% at 7 years. The mean survival for the 16 patients who died suddenly was 26 months (range, 1 month to 7.5 years). Of the 16 patients who died suddenly, eight (50%) did so within 1 year of operation.

Univariate analysis of risk factors for late sudden death after successful repair of DORV showed the following significant factors: 1) older age at the time of operation, 2) elevated preoperative mean right atrial pressure and mean pulmonary arterial pressure, 3) preoperative supraventricular tachyarrhythmias, 4) perioperative and postoperative ventricular tachycardia or ventricular fibrillation, 5) perioperative and postoperative third-degree atrioventricular block, and 6) perioperative and postoperative atrial fibrillation or flutter.

The effect of age at operation on subsequent survival is shown in Figure 3. Of the 55 patients who were younger than 12 years at operation, five died suddenly. Among the 34 patients who were 12 years old or older, 11 died suddenly. The difference in survival probabilities (88% vs. 67% at 7 years) was significant ($p<0.04$).

Figure 4 shows the significant preoperative risk factors. The mean right atrial pressure was obtained in 81 patients; 12 of the 45 patients who had mean pressures of 8 mm Hg or more died suddenly (7-year survival, 70%), and only two of the 36 patients with a preoperative mean pressure less than 8 mm Hg died suddenly (7-year survival, 94%; $p<0.02$) (Figure 4A). Patients with mean pulmonary arterial pressures of 20 mm Hg or more had a higher incidence of sudden death (7-year survival, 76%) than those with mean pressures of less than 20 mm Hg (7-year survival, 92%; $p<0.04$) (Figure 4B). The difference in survival between patients with preoperative supraventricular tachyarrhythmia (7-year survival, 20%) and those without it (7-year survival, 85%) was highly significant ($p<0.01$) (Figure 4C). However, the number of patients in this group was small; among the 89
TABLE 2. Characteristics of 16 Patients Dying Suddenly After Repair of Double-Outlet Right Ventricle

<table>
<thead>
<tr>
<th>Case</th>
<th>Year of operation</th>
<th>Sex</th>
<th>Age at operation (yr)</th>
<th>Survival duration (mo)</th>
<th>Preoperative Arrhythmia</th>
<th>Perioperative Arrhythmia</th>
<th>Postoperative Arrhythmia</th>
<th>Medication at death</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1968</td>
<td>F</td>
<td>14.0</td>
<td>3</td>
<td>...</td>
<td>VF</td>
<td>VF/VT</td>
<td>Quinidine</td>
<td>Roller skating</td>
</tr>
<tr>
<td>2</td>
<td>1971</td>
<td>M</td>
<td>18.0</td>
<td>90</td>
<td>First-degree block</td>
<td>VT</td>
<td>First-degree block, PVC</td>
<td>...</td>
<td>Swimming</td>
</tr>
<tr>
<td>3</td>
<td>1971</td>
<td>M</td>
<td>20.0</td>
<td>8</td>
<td>First-degree block</td>
<td>VT</td>
<td>First-degree block</td>
<td>...</td>
<td>Playing touch football</td>
</tr>
<tr>
<td>4</td>
<td>1973</td>
<td>M</td>
<td>13.0</td>
<td>12</td>
<td>...</td>
<td>...</td>
<td>Digoxin</td>
<td>...</td>
<td>Running from a dog</td>
</tr>
<tr>
<td>5</td>
<td>1974</td>
<td>M</td>
<td>12.5</td>
<td>45</td>
<td>AF/PSVT</td>
<td>AF</td>
<td>...</td>
<td>Digoxin</td>
<td>Playing, running at school</td>
</tr>
<tr>
<td>6</td>
<td>1967</td>
<td>F</td>
<td>7.8</td>
<td>4</td>
<td>...</td>
<td>Third-degree block</td>
<td>Third-degree block</td>
<td>...</td>
<td>Persistent third-degree block without PPM</td>
</tr>
<tr>
<td>7</td>
<td>1971</td>
<td>F</td>
<td>7.8</td>
<td>78</td>
<td>...</td>
<td>VT, third-degree block</td>
<td>Third-degree block</td>
<td>...</td>
<td>Autopsy: fibrosis of penetrating bundle</td>
</tr>
<tr>
<td>8</td>
<td>1974</td>
<td>M</td>
<td>19.5</td>
<td>63</td>
<td>PSVT</td>
<td>AF</td>
<td>AF, first-degree block</td>
<td>Dyazide, propranolol</td>
<td>Walking in the living room</td>
</tr>
<tr>
<td>9</td>
<td>1976</td>
<td>F</td>
<td>4.0</td>
<td>54</td>
<td>...</td>
<td>...</td>
<td>...</td>
<td>...</td>
<td>Suddenly fell down the stairs at home</td>
</tr>
<tr>
<td>10</td>
<td>1977</td>
<td>M</td>
<td>39.5</td>
<td>24</td>
<td>SVT</td>
<td>...</td>
<td>VT</td>
<td>Furosemide, digoxin, quinidine</td>
<td>Recurrent PSVT reported by referral physician; not responsive to resuscitation</td>
</tr>
<tr>
<td>11</td>
<td>1977</td>
<td>F</td>
<td>15.5</td>
<td>15</td>
<td>...</td>
<td>...</td>
<td>...</td>
<td>Furosemide, propranolol, digoxin</td>
<td>Autopsy: recurrent VSD due to ruptured septal aneurysm after infarction</td>
</tr>
<tr>
<td>12</td>
<td>1979</td>
<td>M</td>
<td>2.0</td>
<td>4</td>
<td>...</td>
<td>VT</td>
<td>...</td>
<td>Digoxin</td>
<td>Autopsy: scarring of main His bundle</td>
</tr>
<tr>
<td>13</td>
<td>1979</td>
<td>M</td>
<td>16.5</td>
<td>5</td>
<td>...</td>
<td>VT</td>
<td>...</td>
<td>Dyazide</td>
<td>Sitting and watching television</td>
</tr>
<tr>
<td>14</td>
<td>1983</td>
<td>M</td>
<td>26.3</td>
<td>10</td>
<td>...</td>
<td>Third-degree block/VT</td>
<td>...</td>
<td>Digoxin, captopril, furosemide</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>1968</td>
<td>M</td>
<td>16.8</td>
<td>8</td>
<td>...</td>
<td>Third-degree block/VT</td>
<td>AF/VT</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>16</td>
<td>1976</td>
<td>M</td>
<td>5.5</td>
<td>1</td>
<td>SVT</td>
<td>Third-degree block</td>
<td>AF, first-degree block</td>
<td>...</td>
<td>...</td>
</tr>
</tbody>
</table>

AF, atrial fibrillation/atrial flutter; VF, ventricular fibrillation or flutter; VT, ventricular tachycardia; PVC, premature ventricular contractions; PPM, permanent pacemaker; SVT, supraventricular tachycardia; PSVT, paroxysmal supraventricular tachycardia; VSD, ventricular septal defect.

**FIGURE 2.** Graph of survival free from late sudden death after successful surgical correction of double-outlet right ventricle (Kaplan-Meier estimate).

**FIGURE 3.** Graph of survival probability with respect to age at operation (Kaplan-Meier estimate). Heavy line, patients younger than 12 years; thin line, patients 12 years old or older (p<0.04).
patients, only five had preoperative supraventricular tachyarrhythmia, and four of the five died suddenly.

Figure 5 shows the effect of significant perioperative and postoperative arrhythmias on subsequent survival. Twenty-seven patients had ventricular tachyarrhythmias. Of the 27 patients, 25 had ventricular tachycardia (18 nonsustained and seven sustained), seven had ventricular fibrillation, and five had both. Nine patients died suddenly. The survival probabilities at 7 years were 65% and 87% for patients with and without these factors, respectively (p<0.01) (Figure 5A). Of the 14 patients who had third-degree atrioventricular block, five received a permanent pacemaker because of persistent postoperative complete heart block, but none of these five patients died suddenly. The nine remaining patients were not treated with a permanent pacemaker because pacemakers were not available at the time of operation (one patient) or the complete heart block was intermittent and resolved at the time of hospital dismissal (eight patients). Five of the nine patients without a pacemaker died suddenly. The survival probabilities

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**Figure 4.** Graph of survival probability with respect to significant preoperative factors. Panel A: Mean right atrial pressure. Heavy line, patients with pressure less than 8 mm Hg; thin line, patients with pressure 8 mm Hg or more (p<0.02). Panel B: Mean pulmonary arterial pressure. Heavy line, patients with pressure less than 20 mm Hg; thin line, patients with pressure of 20 mm Hg or more (p<0.04). Panel C: Supraventricular tachyarrhythmia. Heavy line, patients without condition; thin line, patients with condition (p<0.01).

**Figure 5.** Graph of survival probability with respect to significant perioperative and postoperative factors. Panel A: Ventricular tachycardia or fibrillation. Heavy line, patients without these factors; thin line, patients with these factors (p<0.01). Panel B: Third-degree atrioventricular block. Heavy line, patients without block; thin line, patients with block (p<0.04). Panel C: Atrial fibrillation or flutter. Heavy line, patients without condition; thin line, patients with condition (p<0.05).
at 7 years were 70% and 86% for patients with and without third-degree atrioventricular block, respectively ($p<0.04$) (Figure 5B). Among the five patients who died suddenly, three also had perioperative or postoperative ventricular tachyarrhythmias. Of the 11 patients who had atrial fibrillation or flutter, four died suddenly. The survival probabilities at 7 years were 41% and 83% for patients with and without atrial fibrillation or flutter, respectively ($p<0.05$) (Figure 5C).

Multivariate analysis showed that the following were significant risk factors for late sudden death: older age at the time of operation, perioperative or postoperative ventricular tachycardia or fibrillation, and third-degree atrioventricular block. The preoperative hemodynamic variables of mean right atrial pressure and mean pulmonary arterial pressure were not independent predictors for late sudden death, nor was perioperative or postoperative atrial fibrillation or flutter. Multivariate analysis was not applied to preoperative supraventricular tachyarrhythmia because of the small number of patients.

The factors that were not significantly associated with late sudden death were year of operation, sex, type and number of anatomic defects, preoperative functional class, prior palliative operation, surgical techniques, single premature ventricular contractions, and postoperative left or right bundle branch block with or without fascicular block. Among the above-mentioned variables, the location of VSD has been of surgical interest. Of the 118 patients undergoing complete repair of DORV, nine had doubly committed VSD, five had noncommitted VSD, and 104 had subaortic VSD. Of the patients with doubly committed and noncommitted VSDs, three of each group died in the hospital before dismissal, and one of each group died suddenly during follow-up.

**Discussion**

This study clearly shows the magnitude of late sudden death after corrective operation for DORV. Among the 89 patients who survived corrective operation, 22 (25%) had late deaths during a mean follow-up of 82 months. Sixteen of the 22 late deaths (73%) were sudden, and eight of the sudden deaths (50%) occurred within 1 year after operation. The most important risk factors for late sudden death in these patients were older age at the time of operation, perioperative or postoperative ventricular tachyarrhythmias, and third-degree atrioventricular block.

Sudden death after repair of congenital heart defects has been reported in patients with atrial and VSD,20 tetralogy of Fallot,21-24 complete atrioventricular canal,25 and transposition of the great arteries.26-30 The incidence of sudden death in these patients ranged from 2% to 8% (2–5% in tetralogy of Fallot,21-24 2–8% in transposition of the great arteries,26-30 and 5% after intracardiac conduit repair of congenital cardiac defects31). The incidence of late sudden death (18% of all the surgical survivors) in patients with DORV is distressingly high, highest among all the previously mentioned postsurgical congenital heart diseases.

Embryologically, DORV arises from the lack of conotruncal inversion and failure of ventricular shift.3,15 It always is associated with other cardiac malformations; hence, intracardiac operation is more complex. Early postoperative mortality is associated with the complexity of other associated anomalies,32,33 surgical techniques,34 and the very young age of the patients.10 Our study reveals the late mortality in these patients.

Older age at the time of operation is a significant risk factor for late sudden death. Other causes of late sudden death are likely to be high-grade conduction defects and malignant ventricular arrhythmias. These causes can be attributable to the long-standing preoperative myocardial changes,35 which are aggravated by incision and suturing of the septum, ventricular wall, and coronary arterial tributaries.36-42 Although early operation may be beneficial from the standpoint of late sudden death, operation during infancy and early childhood for complex anomalies has been associated in former years with high operative mortality.10,33 The issue of preferred age for corrective intervention is complex. The current recommendation of early individualized palliative management and elective corrective operation43-45 is supported by our long-term follow-up findings for patients with DORV.

Univariate analysis revealed that preoperative supraventricular tachycardia is a strong predictor for late sudden death. (Multivariate analysis was not performed for this factor because of the small number of patients.) Only five patients had preoperative supraventricular tachycardia, four of whom died suddenly. Of these four patients, one also had early postoperative ventricular tachycardia, and two were being treated for supraventricular tachyarrhythmia at the time of death. Although the underlying mechanism is not entirely clear, the onset of supraventricular tachyarrhythmia in congenital heart disease is thought to be related to increased atrial pressure resulting in atrial dilatation and myocardial fibrosis, which cause chronic supraventricular tachyarrhythmias.46-48

The presence of ventricular arrhythmias in patients with congenital heart disease is associated with high mortality. An incidence of sudden death of 30–38% has been noted after repair of tetralogy of Fallot in patients who had premature ventricular contractions on the resting, stress, or ambulatory electrocardiogram.22,49,50 Our study has shown that the presence of ventricular tachyarrhythmias during the perioperative and postoperative periods is strongly associated with late sudden death. However, the presence of single premature ventricular contractions during the same period is not associated with an increased risk of late sudden death.

In long-term follow-up studies, significant mortality (60–80%) has been reported in patients with postoperative heart block not treated with pacemakers.51,52 Survival rates are much improved if
pacemakers are implanted in such patients. In our study, five patients had pacemaker implantation for persistent third-degree atrioventricular block; none of them died suddenly. Of the nine patients who had third-degree block without pacemaker implantation (eight had transient block), five died suddenly during follow-up. This result suggests that the presence of perioperative and postoperative transient third-degree block indicates significant trauma to the atrioventricular conduction system, which will eventually result in permanent heart block. In previous reports, right bundle branch block with left anterior hemiblock has been thought to be associated with late sudden death after repair of tetralogy of Fallot, presumably due to progression to complete heart block. However, the prognostic implication of such conduction disturbances is still controversial, possibly because of the inadequacy of the surface electrocardiogram to differentiate a peripheral injury from a central lesion to the atrioventricular conduction system. In our study, we found no association of late sudden death in the presence of postoperative right bundle branch block with left anterior hemiblock.

The high incidence of late sudden death after complete surgical repair of DORV was not the experience in a previous report. Luber et al reported the results of reparative surgery performed between 1973 and 1981 in 57 patients whose mean age was 5.4 years. No late mortality was observed among the 50 surgical survivors (mean duration of follow-up, 3.6 years). Of these 50 patients, 28 had postoperative catheterization, and 15 of these (54%) had significant hemodynamic abnormalities. Of the 15, 12 required repeat operation. Although the exact reasons are not entirely clear, several factors may account for the different observations. The mean age of our patients (10.3 years) at the time of corrective operation was considerably older. Long-standing myocardial changes could have contributed to a higher risk of late sudden death. In addition, in the series of Luber et al., a large number of patients underwent early repeat operation for subclinical hemodynamic abnormalities detected by catheterization; this was not our experience. Most patients referred to our institution came from long distances (some from foreign countries), and routine follow-up, including cardiac catheterization, was not performed. Therefore, patients with subclinical hemodynamic abnormalities leading to sudden death could have been missed.

Also of interest are the factors not associated with increasing risk of late sudden death. Although significant for predicting late sudden death by univariate analysis, elevated preoperative mean right atrial pressure and mean pulmonary artery pressure on multivariate analysis are not independent predictors for late sudden death. In addition, we found no significant differences in survival probabilities for late sudden death with respect to year of operation, sex, number and type of anatomic defects, preoperative functional class, prior palliative operation, and different surgical approaches. An “era effect” has been reported by Piccoli et al when evaluating hospital deaths in patients after repair of DORV. At their institution, there were nine hospital deaths among 23 patients (39%) undergoing operation from 1967 to 1978, but there was only one hospital death among 19 patients (5%) undergoing operation from 1978 to 1982. They attributed the changing and improving results to the more complete preoperative evaluation and more precise and efficient surgical techniques, including the routine use of cold cardioplegic myocardial protection. However, in our study, we have not observed such an “era effect” in the long-term follow-up of late sudden death. Although the reasons are not known, we can only speculate that the mechanisms underlying early hospital deaths and late sudden deaths must differ. Early mortality is believed to be predominantly associated with the complexity of coexisting anatomic defects and the severity of irreversible preoperative physiologic derangement.

The risk factors for late sudden deaths (third-degree atrioventricular block and ventricular tachyarrhythmias, as identified in our present study) can be complications of operation in the presence of preoperative myocardial change, which may manifest itself only by late sudden death during the follow-up period.

Limitations and Clinical Implications

Our study is limited by the retrospective nature of the data collection. Evaluation and monitoring techniques varied during the time that these patients had operation and follow-up. Nevertheless, the incidence of late sudden death after successful repair of DORV is clearly one of the highest among patients with congenital heart disease. Of the total number of late deaths (22 patients), 73% were sudden, presumably due to an arrhythmia. Older age at the time of operation, perioperative and postoperative ventricular tachyarrhythmias, and third-degree atrioventricular block are significant risk factors for predicting late sudden death. These results warrant early repair of DORV, between 2 and 4 years of age, in an effort to minimize the myocardial damage that may predispose to postoperative arrhythmias and sudden death. Complete and early postoperative evaluation, including electrocardiography, treadmill exercise testing, transthoracic monitoring, ambulatory electrocardiography, and possibly programmed stimulation testing, should be considered for the high-risk patient. Patients with transient perioperative and postoperative third-degree atrioventricular block should be monitored closely and considered for pacemaker implantation.

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