Unexpected Cardiac Arrest in Patients after Surgical Correction of Tetralogy of Fallot

By Frederick W. James, M.D., Samuel Kaplan, M.D., and Te-Chuan Chou, M.D.

SUMMARY

Four of 220 patients without bifascicular block (complete right bundle branch block and left anterior hemiblock) or transient complete heart block immediately after surgery had an unexpected cardiac arrest one to 15 years after satisfactory surgical repair of tetralogy of Fallot. The postoperative electrocardiograms (ECG) revealed complete right bundle branch block in two patients and no intraventricular conduction abnormality in two patients. Each of the four patients had premature ventricular contractions on previous postoperative ECG. The cardiac arrest occurred during normal activity in three patients and mild exercise in one. Following the cardiac arrest, three patients died and one patient survived. Eighteen months before the cardiac arrest, the survivor had a stress test which revealed multifocal premature ventricular contractions with short bursts of ventricular tachycardia after exercise. This ventricular arrhythmia was suppressed with quinidine therapy.

Although complete heart block cannot be excluded in these four patients, we reasoned that the cardiac arrests were probably preceded by ventricular tachyarrhythmia. Because of this experience, we believe that any patient who has had intraventricular surgery should be evaluated for ventricular arrhythmia. If frequent premature ventricular contractions or serious ventricular arrhythmias are documented, we seriously consider antiarrhythmic therapy in an attempt to prevent ventricular tachyarrhythmias and sudden death.

The most common conduction abnormality in patients following corrective surgery for tetralogy of Fallot (TOF) is complete right bundle branch block. Retrospectively, this postoperative conduction abnormality has been associated with a favorable prognosis. Permanent or transient complete heart block and bifascicular block (complete right bundle branch block and left anterior hemiblock) in patients after intracardiac repair of TOF are serious conduction abnormalities which have been associated with an increased morbidity and mortality during the early or late postoperative period.

This report describes four patients without bifascicular block or complete heart block (CHB) who had an unexpected cardiac arrest several years after surgical repair of TOF. The case histories suggest an additional mechanism of cardiac arrest or sudden death in patients after intracardiac repair of TOF. The possible preventive management in future patients following intraventricular surgery is discussed.

Patients

At our institution, 225 patients with TOF have undergone total surgical correction between 1957 and 1974 with follow-up intervals varying from one to 17 years. Their preoperative electrocardiograms (ECG) revealed marked right ventricular hypertrophy with or without abnormal P waves. Biventricular hypertrophy was present in those patients who had large pulmonary flows created by surgical anastomosis performed earlier. In each patient the cardiac anomaly consisted of a large subaortic ventricular septal defect and severe right ventricular outflow tract obstruction with equal peak systolic pressures in both ventricles. The right ventricular outflow obstruction was solely infundibular or was combined with valvar pulmonic stenosis with or without hypoplasia of the pulmonary valve ring and arterial trunk. Either bidirectional or exclusively right-to-left shunting across the ventricular septal defect was present.

The severe right ventricular outflow tract obstruction was relieved by infundibulectomy, with or without pulmonary valvotomy in 56% of the patients. A pericardial patch in the right ventricular outflow was used in 36% and an aortic homograft was used to obtain relief of obstruction in 13 patients. The ventricular septal defect was closed with a teflon patch. During the 17 year interval, there were 181 survivors and 44 deaths. The routine ECGs of the 181 patients who are currently being followed were reviewed in order to determine the incidence of conduction and rhythm disturbances following surgical correction of TOF.

In the 181 patients who are currently followed, the postoperative routine ECG revealed complete right bundle branch block (CRBBB) pattern in 145, complete right bundle branch block and left anterior hemiblock in 22, permanent CHB in three, and no intraventricular conduction abnormality in 11 (table 1). An ectopic atrial or junctional pacemaker was present in 6. First degree atrioventricular block was present in two patients, Wolff-Parkinson-White syndrome in one, and occasional premature ventricular contractions (PVC) in four.

Forty patients died within 72 hours after open heart surgery. Death was attributed to low cardiac output and congestive heart failure. Four of the 225 patients had an unexpected cardiac arrest one to 15 years after satisfactory
hemodynamic correction of their cardiac defect. Three of the four patients died following the arrest, and the remaining patient survived after prompt cardiopulmonary resuscitation by an experienced observer.

During surgery, a transverse incision was made into the right ventricle of the four patients who had an unexpected cardiac arrest. An extensive amount of muscle tissue was resected from the infundibular area in order to relieve the obstruction. The pulmonary orifice was adequate in three patients but the remaining one required a valvotomy because of an obstructive bicuspid pulmonary valve. A pericardial patch was not used in any of the four patients. The large subaortic ventricular septal defect was closed with a teflon patch.

Case Reports

Case 1 (D.C.)
In this patient, the diagnosis of TOF was made at two years of age. Because of progressive cyanosis, a left Blalock anastomosis was done. Prior to surgical correction, the ECG revealed a P-R interval of 0.16 sec, diphasic T waves in leads I and V₆, and right ventricular hypertrophy. At 18 years of age, complete surgical correction was performed, and the immediate postoperative period was uneventful. The P-R interval was 0.24 sec on the routine postoperative ECG. Both the ECG and vectorcardiogram were compatible with an anterosetal scar and probable conduction abnormality in the right ventricle, but a typical CRBBB pattern was not present (fig. 1). Occasional unifocal PVC were recorded on previous electrocardiographic tracings.

One year after surgical correction, this patient died unexpectedly while at home alone. An autopsy was not performed. The clinical examination which was performed 3.5 months prior to the sudden event was compatible with a satisfactory hemodynamic result.

Case 2 (T.W.)
The diagnosis of TOF was made at 13 months of age. A Potts' anastomosis was performed at the age of 26 months. Nine years after the shunt procedure the patient was hospitalized because of subacute bacterial endocarditis due to streptococcus viridans. He was successfully treated with penicillin and streptomycin. Prior to surgical correction, the ECG was typical for right ventricular hypertrophy. At 16 years of age, the Potts' anastomosis was closed, and the TOF complex was corrected. The immediate postoperative course was uneventful except for one episode of paroxysmal atrial tachycardia which responded to countershock. Seven years after open heart surgery, the routine ECG revealed a normal QRS interval and T wave inversion in leads I, aV₁, V₄₋₆ (fig. 2). On several subsequent routine ECG, frequent unifocal PVC were recorded.

Eight years after surgical correction this patient died unexpectedly in his sleep. The patient had been seen in the cardiac clinic three months prior to the sudden event and had physical findings of mild residual pulmonic stenosis.

Table 1

<table>
<thead>
<tr>
<th>Electrocardiograms in 181 Patients after Surgical Correction of TOF</th>
<th>No.</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>CRBBB</td>
<td>145</td>
<td>80.5</td>
</tr>
<tr>
<td>CRBBB + LAH</td>
<td>22</td>
<td>12</td>
</tr>
<tr>
<td>CHB</td>
<td>3</td>
<td>1.5</td>
</tr>
<tr>
<td>Normal QRS interval</td>
<td>11</td>
<td>6</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>13</td>
<td>7</td>
</tr>
</tbody>
</table>

Abbreviations: CHB = complete heart block; CRBBB = complete right bundle branch block; LAH = left anterior hemiblock; TOF = Tetralogy of Fallot.

Figure 1

Electrocardiogram (ECG) and vectorcardiogram (VCG) in an 18-year-old male six months after intracardiac repair of tetralogy of Fallot. Both studies are compatible with an anterosetal scar and probable conduction abnormality in the right ventricle. Complete right bundle branch block is not present. The P-R interval is 0.24 sec. On the VCG, the QRS loop is inscribed counterclockwise in the horizontal (H) plane and clockwise in the sagittal (S) and frontal (F) planes.
Autopsy findings revealed an intact ventricular septum, normal aortic valve, and slight atherosclerosis of the coronary arteries on microscopy.

Case 3 (N.C.)

This patient was 11 years of age when the diagnosis of TOF was made. At 12 years of age, a Brock procedure was done because of progressive cyanosis and exercise intolerance. Prior to surgical correction, an ectopic atrial pacemaker, junctional rhythm, and right ventricular hypertrophy were recorded on the routine ECG. At 22 years of age, complete surgical correction for TOF was performed. Five months after open-heart surgery, the physical examination revealed mild residual right ventricular outflow obstruction. Complete right bundle branch block and occasional premature ventricular contractions were present on a routine ECG. One year after open-heart surgery, this patient died while swimming in a pool. An autopsy was not performed.

Case 4 (J.D.)

The diagnosis of TOF was made at 23 years of age. Sinus rhythm and right ventricular hypertrophy were present on the ECG. Two years later complete surgical correction was performed. The postoperative course was uneventful. The postoperative examination revealed mild residual pulmonic stenosis and insufficiency and/or small residual ventricular septal defect. Complete right bundle branch block was present after open-heart surgery (fig. 3). During a 15-year interval after open-heart surgery, this patient successfully completed three normal pregnancies and performed her routine duties at home. She complained on a few occasions of palpitations, but her routine ECG revealed only occasional unifocal PVC. Because of this history, a maximal exercise test was recommended in an attempt to unmask any serious conduction abnormalities. This maximal exercise test was performed as previously reported from our institution.

Before exercise, sinus rhythm was present, and bigeminy was precipitated by hyperventilation (fig. 3B). During exercise, sinus rhythm dominated. Within two minutes after exercise, multifocal PVC and short bursts of ventricular tachycardia were recorded (fig. 3C).

Eighteen months after the exercise test (15 years postoperative), this patient had a cardiac arrest in a hotel lobby during the family’s vacation. She received immediate cardiopulmonary resuscitation and was taken promptly to an intensive care unit in the local area. Upon arrival, her ECG revealed CRBBB with occasional PVC. She was placed on propranolol (10 mg q.i.d.) and was discharged on the tenth hospital day.

When she returned home, a repeat exercise test was done. The postexercise electrocardiogram was essentially unchanged from the initial study. At that time, her drug therapy was changed from propranolol to quinidine. While on drug treatment, her ECG was recorded on several occasions with a 12-hour ambulatory monitor. The results of the ambulatory monitoring are shown in table 2. During propranolol therapy, frequent PVC with couplets and triplets were recorded. With increasing dosages of

**Table 2**

**Results of Ambulatory Monitoring in Case 4 (JD)**

<table>
<thead>
<tr>
<th>Drugs (Mg Q6Hrs)</th>
<th>PVC/hr</th>
<th>Bigeminy</th>
<th>Couplets</th>
<th>Triplets</th>
</tr>
</thead>
<tbody>
<tr>
<td>Propranolol (10)</td>
<td>TNTC</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Quinidine (200)</td>
<td>TNTC</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Quinidine (300)</td>
<td>8-67</td>
<td>+</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Abbreviations: hr = hour; PVC = premature ventricular contractions; TNTC = too numerous to count; + = Present; 0 = Absent.
quindine, PVC decreased to a range of 8–67 beats/hour, but a few short episodes of bigeminy persisted. Couplets or triplets were not recorded.

The clinical profile of the four patients described in this report are summarized in table 3. Transient CHB immediately after surgery, complete right bundle branch block and left anterior hemiblock or permanent CHB was not documented in any of these patients.

Discussion

Serious conduction abnormalities may develop in patients after open-heart surgery for congenital heart disease. Permanent CHB has occurred after many open-heart procedures and has been associated with a high mortality. If this condition occurs after surgery, it is generally agreed that an artificial pacemaker should be implanted in order to prevent Stokes-Adams attacks and sudden death. The decreased incidence of surgically-induced CHB is due to improved surgical techniques and better understanding of the intracardiac conduction pathways.

In a series of 24 patients with complete right bundle branch block and left anterior hemiblock, Wolff et al. reported five patients who developed permanent CHB during the late postoperative period. In the same series, sudden unexpected death occurred in three patients, and serious ventricular arrhythmias occurred in four. The over-all mortality for the study group was 25%. Late permanent CHB did not develop in their control group, but two patients with CRBBB and occasional PVC died suddenly and unexpectedly.

Godman et al. recorded His bundle electrocardiograms in 22 patients following intracardiac repair of TOF and ventricular septal defect. All patients with prolonged H-V intervals had transient CHB during the immediate postoperative period. The authors suggested that patients who have bifascicular block and transient CHB immediately after surgery are at great risk for developing late permanent CHB.

In contrast, however, Downing et al. reported bifascicular block in 14 of 131 patients after correction

Table 3

Clinical Profiles of the Four Patients Who Had an Unexpected Cardiac Arrest after Surgical Correction of TOF

<table>
<thead>
<tr>
<th>Patients</th>
<th>Age at op (yr)</th>
<th>Yr postop</th>
<th>PS/PI</th>
<th>VSD</th>
<th>Trans CHB</th>
<th>ECG</th>
<th>PVC</th>
</tr>
</thead>
<tbody>
<tr>
<td>TW</td>
<td>16</td>
<td>8</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>T waves</td>
<td>+</td>
</tr>
<tr>
<td>DC</td>
<td>18</td>
<td>1</td>
<td>-</td>
<td>+</td>
<td>ICD</td>
<td>1* HB</td>
<td>+</td>
</tr>
<tr>
<td>NC</td>
<td>22</td>
<td>1</td>
<td>+/+</td>
<td>-</td>
<td>-</td>
<td>CRBBB</td>
<td>+</td>
</tr>
<tr>
<td>JD</td>
<td>25</td>
<td>15</td>
<td>+/+</td>
<td>-</td>
<td>+</td>
<td>CRBBB</td>
<td>+</td>
</tr>
</tbody>
</table>

Abbreviations: TOF = tetralogy of Fallot; Op = operation; PI = pulmonary insufficiency (mild); PS = pulmonic stenosis (mild); VSD = ventricular septal defect (small left-to-right shunt); CHB = complete heart block; HB = heart block; ICD = intraventricular conduction defect; CRBBB = complete right bundle branch block; PVC = premature ventricular contractions; - = absent; + = present.
of TOF. Transient CHB occurred in two patients during the immediate postoperative period. Permanent CHB or sudden unexpected death has not occurred in any of these 14 patients in a follow-up period of 1 to 16 years.

In our patients who died suddenly and unexpectedly, CHB was not documented or suspected clinically. Neither bifascicular block nor transient CHB during the immediate postoperative period was documented in any of the four patients. Occasional (PVC) premature ventricular contractions were recorded on the routine postoperative ECG in each patient.

We have observed serious ventricular arrhythmias after exercise in patients who have undergone open-heart surgery for TOF. In the one survivor in this report, multifocal PVC and ventricular tachycardia, which were not suspected from the routine ECG, occurred after exercise. Although CHB with a Stokes-Adams attack remains a possibility in our series, we reasoned that the sudden events were preceded by ventricular tachyarrhythmia which caused sudden death in three patients and a cardiac arrest in one.

We did not find any unusual features that would suggest the cause for the ventricular arrhythmia in the four patients. In three of the four patients, the PVC had a left bundle branch block pattern in several electrocardiographic leads, suggesting that the ectopic focus was in the right ventricle. Because the patients were young adults at the time of their corrective procedures, we have speculated that perhaps they had severe fibrotic changes in the right ventricle which made them more vulnerable to ventricular ectopic rhythms after surgery. Our series is too small to draw any firm conclusions regarding the etiology of the PVC.

Unexpected cardiac arrest and sudden death have occurred in other large series of patients who have had surgical correction of TOF. Table 4 summarizes those series of which unexpected sudden death occurred in 17 patients and a cardiac arrest in one. Because of our experience with the four patients described in this report, we believe that any patient after surgery requiring ventriculotomy who has frequent PVC on the routine ECG or serious ventricular arrhythmia after exercise or during ambulatory monitoring is at risk for sudden death. Therefore, in these patients, the use of antiarrhythmic therapy should be seriously considered in an attempt to prevent ventricular tachyarrhythmia. We are now using quinidine sulfate in three such patients who developed multifocal PVC with couplets and/or triplets after exercise. A history of syncope, palpitation, or cardiac arrest was present in each of the three patients prior to antiarrhythmic therapy. Quinidine has abolished the postexercise arrhythmia in one and markedly diminished the ventricular arrhythmia in the remaining two patients.

We need a longer follow-up period in a larger series before drawing any conclusions about the use of antiarrhythmic therapy in the prevention of sudden death in patients after intraventricular surgery.

### Table 4

<table>
<thead>
<tr>
<th>Authors</th>
<th>Patients SD/CA</th>
<th>Years postop</th>
<th>ECG</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kirklin et al.</td>
<td>337</td>
<td>4/ CA</td>
<td>0.5</td>
</tr>
<tr>
<td>Wolff et al.</td>
<td>291</td>
<td>2/</td>
<td>0.5 CRBBB</td>
</tr>
<tr>
<td>Wolf et al.</td>
<td>146</td>
<td>3/</td>
<td>0.25-2 CRBBB</td>
</tr>
<tr>
<td>James et al.</td>
<td>225</td>
<td>3/</td>
<td>1-15 NQRs Interval CRBBB</td>
</tr>
<tr>
<td>Quattlebaum et al.</td>
<td>251</td>
<td>5/</td>
<td>6.5-16 CRBBB</td>
</tr>
</tbody>
</table>

Abbreviations: TOF = tetralogy of Fallot; SD = sudden death; CA = cardiac arrest; ? = not reported; CRBBB = complete right bundle branch block; PVC = premature ventricular contractions; N = normal.

### References

Unexpected cardiac arrest in patients after surgical correction of tetralogy of Fallot.
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