Sudden Death Among Postoperative Patients with Tetralogy of Fallot

A Follow-up Study of 243 Patients for an Average of Twelve Years

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SUMMARY Two hundred and forty-three patients were evaluated following total correction of tetralogy of Fallot with special emphasis on postoperative conduction disturbances and on the occurrence of sudden death. The average follow-up period was 12 years with a range of 6½ to 16½ years.

Sudden death occurred in seven patients. Four deaths were among those with right bundle branch block pattern (RBBB) and three of the four had premature ventricular contractions (PVCs) for more than one month postoperatively. PVCs were documented in ten of the 158 patients with RBBB; sudden death occurred in three (30%). Three of the ten (30%) patients with trifascicular block pattern (TB) died suddenly, while no deaths occurred in 24 patients with bifascicular block pattern (BB). Progression of RBBB to BB and TB occurred in 18 patients from one month to seven years postoperatively (58% of BB and 40% of TB).

The risk of sudden death in patients with RBBB and PVCs following tetralogy repair is high and warrants consideration of suppressive therapy. TB also carries a high risk. The finding that RBBB may progress to BB or TB mandates long-term careful follow-up of all tetralogy patients with postoperative conduction disturbances.

CONDUCTION DISTURBANCES which develop after open repair of tetralogy of Fallot or ventricular septal defect remain a subject of controversy. Published reports of the prognosis of surgical bifascicular block pattern (BB) — that is, right bundle branch block pattern (RBBB) with left anterior hemiblock (LAH) — vary from ominous to good.1 4 We therefore undertook a retrospective analysis of the electrocardiograms (ECGs) of patients who had undergone open repair of tetralogy, with special emphasis on the occurrence of sudden death and the possible relationship of the death to postoperative conduction disturbances.

Materials and Methods

Three hundred and thirty-one patients underwent open repair of tetralogy of Fallot during the ten-year period from 1958 to 1967 at the Johns Hopkins Hospital. We excluded from review all patients 1) who died within one month of surgery; 2) who died from direct complications of surgery such as severe intractable heart failure; 3) who died of known causes, for example auto accidents, carcinoma, and coronary arteriosclerosis; 4) who had life-threatening arrhythmias or persistent complete heart block immediately postoperatively; 5) with frontal plane left axis on preoperative electrocardiograms; and 6) those with preoperative arrhythmias. There remained 248 patients, of whom 243 (98%) had pre or postoperative ECGs available for analysis. These 243 patients are the subject of this report. Their ages ranged from 6 to 30 years, and the majority had had a previous Blalock-Taussig shunt. The follow-up period ranged from 6½ to 16½ years, with an average of 12 years.

The electrocardiograms consisted of routine 12-lead tracings taken during hospitalization or at regular clinic visits.

The following criteria were used: 1) RBBB was diagnosed as an increase in QRS duration equal to or exceeding 0.11 sec with a terminal conduction delay; 2) BB (RBBB with LAH) was diagnosed when, in addition to the pattern of RBBB, the frontal QRS axis was greater than 30° with a Q1S3 counterclockwise pattern; 3) trifascicular block pattern (TB) was diagnosed when, in addition to a pattern of BB, the patient also had A-V block as indicated by a prolonged P-R interval corrected for heart rate (P-R greater than or equal to 0.20 sec); and 4) incomplete RBBB was defined as a QRS duration less than 0.11 sec with a definite terminal delay.

Results

The postoperative electrocardiographic patterns of the 243 patients are presented in table 1.

Sudden death occurred in seven patients (2.9%). The interval between surgery and death varied from 3 months to 5½ years, four of the deaths being more than one year postoperatively. Analysis of the electrocardiographic patterns in these seven patients showed RBBB in four, and TB in three.

More detailed analysis of the four ECGs with RBBB showed that one patient had significant A-V block postoperatively and the other three had premature ventricular contractions (PVC). PVCs occurring more than one month postoperatively were found in 10 of the 158 cases of RBBB (6.3%). Three of these patients (30.0%) died suddenly.

PVCs were present in three of the 24 patients with BB and one of the two with incomplete RBBB and LAH; there were no deaths in either group. None of the 10 patients with TB showed PVCs.

The frequency of PVCs varied from bigeminy to only one or two per 12 lead surface ECG. Frequency of PVCs did not appear greater in the patients who died suddenly than in others. Analysis of the PVC pattern exhibited by the patients with RBBB and sudden death revealed that in each case the PVCs were unifocal, late-coupled, and originating from the right ventricle (fig. 1). No cardiomegaly or right ventricular outflow tract aneurysms were found in patients with PVCs.

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289
In our series there were no deaths among patients with BB, but three of the ten with TB died suddenly. None of our sudden deaths had had transient complete heart block post-operatively. It is also noteworthy that none of the patients who died suddenly were exercising at the time of death. Deaths occurred in three patients while resting in bed, in three others while relaxing at home, and in one patient while walking in the park.

The pattern of BB and TB was noted immediately post-operatively in some patients, but in others developed late (figs. 2, 3). In these patients the postoperative pattern of RBBB did not remain static, but progressed to BB and TB at intervals ranging from one month to seven years, with approximately two-thirds progressing within one year. Thirteen of the 24 BB group (58%) and four of the ten with TB (40%) had delayed shift of their axis leftward. One of the sudden deaths among the TB group occurred in a patient whose electrocardiogram exhibited only RBBB with a normal P-R interval for more than a year postoperatively, at which time his axis shifted to \(-80^\circ\) and his P-R became prolonged (fig. 2). He died suddenly after dinner 4½ years later.

Patients who were excluded because of the presence of left axis or PVCs preoperatively were analyzed separately. Of the three patients with left axis prior to open-heart surgery, one developed BB postoperatively and the other two developed TB, with no sudden deaths in either group. There were seven patients with PVCs prior to surgery, and in five these disappeared in the postoperative period. Two patients continued to have PVCs postoperatively, and one died suddenly: this patient also had intermittent WPW syndrome and supraventricular tachycardia.

**Table 1. Study Patients by ECG Pattern**

<table>
<thead>
<tr>
<th>Pattern</th>
<th>Number</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal or incomplete RBBB</td>
<td>49</td>
<td>20.2</td>
</tr>
<tr>
<td>Complete RBBB</td>
<td>158</td>
<td>65.0</td>
</tr>
<tr>
<td>Incomplete RBBB &amp; LAH</td>
<td>2</td>
<td>0.8</td>
</tr>
<tr>
<td>Bifascicular block</td>
<td>24</td>
<td>9.0</td>
</tr>
<tr>
<td>Trifascicular block</td>
<td>10</td>
<td>4.1</td>
</tr>
<tr>
<td>Total</td>
<td>243</td>
<td>100.0</td>
</tr>
</tbody>
</table>

Abbreviations: LAH = left anterior hemiblock; RBBB = right bundle branch block.

Discussion

The overall incidence of sudden death during an average 12-year period of follow-up was 2.9%, mortality being highest in those with RBBB and PVCs (30%) or with TB (30%). The progression of RBBB to BB or TB late postoperatively was another major finding in this series.

The significance of premature ventricular beats depends on the presence or absence of organic heart disease. Their occurrence in healthy young individuals with normal hearts is not uncommon and is usually considered benign. However, premature ventricular beats associated with organic heart disease may have much greater significance.

PVCs that could be considered of minor importance before surgery may assume significance after a ventriculotomy and the development of a block in the conduction system. Postoperative scar tissue in the ventricle, with its altered conduction properties, may result in an increased likelihood of re-entry. This situation, combined with a conduction delay through the right bundle branch, may predispose to re-entry tachycardia or fibrillation.

Sudden unexpected death occurred in three of our ten patients with RBBB and PVCs for more than one month postoperatively. It is of interest that Wolff’s series also showed two sudden deaths in patients with this electrocardiographic combination. The PVCs in our three patients had the characteristics of typical benign PVCs frequently seen in normal hearts, that they were unifocal, late-coupled, and originated from the right ventricle.

It appears that patients with RBBB and PVCs following open repair of tetralogy need close follow-up. Holter monitoring and exercise electrocardiography may prove valuable in deciding on therapy. Attempts to suppress the premature ventricular beats with quinidine appear worthy of study, despite some controversy over the safety of this drug in the presence of bundle branch block. In patients with otherwise normal hearts an increase in PVCs on exercise electrocardiogram is often considered an indication for suppressive therapy; however, since sudden death in our series was not associated with exercise, these criteria may need modifying in this particular group of patients.

The prognostic significance of BB (RBBB with LAH) has been the subject of some controversy. Wolff and Godman

![Figure 1](http://circ.ahajournals.org/)

_Figure 1. The postoperative electrocardiogram of a patient who developed premature ventricular contractions postoperatively. On follow-up she continued to have the pattern of RBBB with unifocal, late-coupled premature ventricular contractions. She died suddenly two years postoperatively while sitting at poolside with her friends._
Figure 2.  Top) The preoperative electrocardiogram of a patient with tetralogy of Fallot. The electrocardiogram shows right axis deviation, a normal QRS duration, and a normal P-R interval. Middle) An ECG taken one month postoperatively shows RBBB with an axis of +180° with a clockwise loop and a normal P-R interval. Bottom) Approximately two years later the patient had developed trifascicular block. The pattern of RBBB is still present, but the axis has shifted to –80° and has developed a prolonged P-R interval. He died after dinner some 5½ years postoperatively.
found an ominous prognosis, especially where BB was associated with transient atrioventricular block in the immediate postoperative period. Downing4 and Pahlajani5 report favorable outcomes over follow-up periods between one and ten years. Since postoperative RBBB may be due to a peripheral block resulting from the ventriculotomy or may develop during repair of the ventricular defect, the two modes of production of RBBB present different theoretical problems in regard to future implications and the possibility of development of late complete block. In our series there were no deaths in the group with BB. However, the group of patients with TB showed a 30% incidence of sudden unexpected death. Patients with an electrocardiographic pattern of TB warrant detailed intracardiac electrophysiologic studies and may benefit from prophylactic pacemaker insertion.

An important finding in this study was the progression of RBBB to BB and TB in 18 patients over periods ranging from one month to seven years postoperatively. Similar progression of conduction disturbances has been reported in ischemic heart disease and various cardiomyopathies. Because of this postoperative progression, careful long-term electrocardiographic follow-up of patients with RBBB is warranted.

The cause of such progression can only be speculated. It may be due to a central lesion surrounded by reactive fibrous tissue gradually involving the anterior and posterior fascicles of the left bundle, resulting in BB and TB. This raises the interesting question of the site of the block in these patients. Although some patients with postoperative RBBB probably do have a distal block related to the ventriculotomy, those who progress to BB and TB must be assumed to have a central block.

Because of the serious prognosis of postoperative TB, the prevention of damage to the conduction system should clearly be a major aim at cardiac surgery. Specialized techniques of intracardiac mapping such as those described by Kaiser may lead to a dramatic reduction in damage to the conducting system and in the occurrence of late sudden deaths in patients with tetralogy who have undergone open repair.

References
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