changes in acute infarction as a measure of extent and severity of injury, the interpretation of ST changes during exercise or rapid atrial pacing, and the relationship of heart rate and partial coronary flow reduction to subendocardial and epicardial ischemia.

References

Sudden Death after Repair of Tetralogy of Fallot

Electrocardiographic and Electrophysiologic Abnormalities

PAUL C. GILLETTE, M.D., MARK A. YEOMAN, M.D., CHARLES E. MULLINS, M.D., AND DAN G. MCNAMARA, M.D.

SUMMARY

In order to try to determine the mechanism of sudden death in patients after surgical repair of tetralogy of Fallot, electrocardiographic, intracardiac electrophysiologic, and clinical data of 51 children who had postoperative intracardiac electrophysiologic studies were reviewed. Ninety-four percent had developed right bundle branch block (RBBB) and 16 percent had additional left anterior hemiblock (LAH). Two had had transient complete A-V block (CAVB) and one had permanent CAVB. Six had a first degree A-V block and nine had premature ventricular contractions (PVC).

Nine patients were found to have prolonged intra-atrial conduction times, four prolonged A-V nodal conduction, four prolonged His-Purkinje conduction, and five prolonged corrected sinus node recovery times. Patients with first degree A-V block or LAH did not have an increased incidence of abnormalities on electrophysiologic study.

No patient with RBBB and LAH developed complete A-V block or died. Three of the nine patients with PVCs died, one of intractable ventricular fibrillation and two suddenly, presumably of dysrhythmia. All three had significant congestive heart failure.

Although late complete A-V block occurs and should be watched for, ventricular dysrhythmias in patients with PVCs may be the cause of most sudden deaths after tetralogy repair. We currently are treating all of our postoperative tetralogy patients who have PVCs with quinidine or propranolol.

ALTHOUGH SURGICAL REPAIR of tetralogy of Fallot results in an improvement in the duration and quality of life for most patients, late sudden deaths are known to occur.1–8 The cause of these deaths has been thought to be dysrhythmias. There is a question as to whether ventricular tachydysrhythmias, complete atrioventricular block, or sick sinus syndrome is the cause.1–9

Intracardiac repair of tetralogy of Fallot carries the risk of damage to impulse generating or conducting system as a result of surgical incision, cannulation or suture. The object of this investigation was to use surface electrocardiography, His bundle electrography, atrial pacing, and the atrial extrastimulus technique to study electrophysiologic properties of the sinoatrial node, atria, atrioventricular node, His-Purkinje system and ventricles after intracardiac repair of
tetralogy of Fallot, and to compare any electrophysiologic abnormalities with the patient's clinical course.

Subjects and Methods

The subjects of this investigation were 51 children and young adults aged 18 months to 32 years who had undergone intracardiac repair of tetralogy of Fallot using standard surgical and cardiopulmonary bypass techniques. Normothermic ischemic arrest was used in each operation and lasted from 22 to 68 minutes (mean 41 min). All pre and postoperative electrocardiograms were reviewed for rhythm, PR interval, right bundle branch block, left anterior hemiblock, transient complete A-V block and premature beats.

The following electrophysiological studies were performed, as previously described from this laboratory, during follow-up cardiac catheterization four months to fifteen years (mean three years) after intracardiac repair.

All subjects were sedated with meperidine 1 mg/lb, promethazine 1/4 mg/lb, and chlorpromazine 1/4 mg/lb one-half hour before study. All medications were stopped 36 hours before catheterizations. A bipolar or tripolar electrode catheter with an interelectrode distance of either 1 or 10 mm was introduced in each patient through a percutaneously placed sheath in the femoral vein. The catheter was manipulated across the tricuspid valve and withdrawn until a rapid potential was seen between the atrial and ventricular electrograms in a manner similar to that reported by Scherlag. A multichannel junction box was used to connect the catheters to an Electronics for Medicine photographic recorder. The electrograms were recorded at paper speeds of both 100 and 200 mm/sec with one second time lines. By electronic filtration, the frequencies recorded in the intracardiac electrograms were limited to between 40 and 500 Hz, and in the surface electrocardiogram to between 0.1 and 200 Hz.

In 33 of these patients, the catheter was also manipulated to the superior vena cava-right atrial junction for recording high atrial potentials. Atrial pacing was then performed in these patients at the superior vena cava-atrial junction using rates 150 to 250% of the resting rate for two to three minutes. Electrograms from two surface leads were recorded before, during and after the sudden cessation of atrial pacing. Atrioventricular conduction during rapid atrial pacing and the sinus node recovery time after sudden cessation of pacing were evaluated. In an additional seven patients, the His bundle electrograms were recorded during pacing by inserting a second catheter percutaneously into the same or opposite femoral vein. In these seven patients, the effective and functional refractory periods of the conduction system were determined by the introduction of coupled premature atrial stimuli at increasingly shorter intervals until block occurred. Femoral artery pressure was monitored continuously by means of an indwelling plastic cannula.

Definition of Terms

The following conduction intervals were measured: 1) high to low right atrium (representing sinoatrial to atrioventricular nodal time), 2) low right atrium to His potential (A-V node conduction time), and 3) His potential to ventricle (His-Purkinje conduction time) (HV). All intervals were measured from the first rapid deflection of one electrogram.
Electrophysiological Findings (fig. 1)

The values for the study group are listed in table 3.

Intra-atrial (HRA-LRA) Conduction (table 4)

Internodal conduction was determined in 30 patients by recording electrograms from both the high and low right atrium sequentially and the HRA-LRA interval calculated.
Eight patients had slightly prolonged internodal conduction (40-50 msec) while 21 were normal. One patient had a markedly prolonged HRA-LRA interval (58 msec). Of the five patients with RBBB and LAH that were evaluated, two had prolonged intra-atrial conduction. Each of the three patients with ventricular extrasystoles that were evaluated had normal intra-atrial conduction.

**Atrioventricular Nodal and His-Purkinje Conduction**

Four of the 51 patients had prolonged atrioventricular nodal conduction (124, 131, 140 and 170 msec), 45 patients had normal A-V nodal conduction. The complete A-V block in patient 41 was found to be in the A-V node. Only four of the 51 patients had a prolonged HV interval (60, 70, 74, and 75 msec). Many of the other 38 had intervals at the upper range of normal (50-55 msec).

One of the eight patients with RBBB and LAH had prolonged atrioventricular nodal conduction and seven were normal. One patient of eight with RBBB and LAH had a prolonged HV interval and seven were normal.

None of the nine patients with PVCs had a prolonged LRA-H interval and only one had a prolonged HV interval.

**Effects of Rapid Atrial Pacing and Premature Atrial Stimulation**

Atrial pacing, performed in 28 patients, produced a lengthening of the PR interval and caused a Mobitz I (Wenckebach) atrioventricular block in eight patients at rates between 117 and 170 beats per minute, while the remaining 20 developed this phenomenon at a rate greater than 180. One patient with a partial atrioventricular block at a low rate (117 beats/min) also had prolonged atrioventricular node conduction interval.

Corrected sinus node recovery time was prolonged (320, 330, 350 and 553 msec) in four patients and normal in 25 patients. The sinus node was the site to recover in all subjects.

In three of the seven patients who had the refractory periods of their conduction system determined, the most refractory part of the conduction system was below the recording site of the His bundle potential. In one patient, the atrium had the longest refractory period, and in three the A-V node was the most refractory. The intervals for the refractory periods were normal in all except one subject with prolonged His-Purkinje refractory period (450 msec).

**Follow-up**

Follow-up of these patients found that three had died suddenly, one of documented ventricular fibrillation, the other two suddenly. All three had PVCs and only one a conduction abnormality (a prolonged HV interval of 74 msec) but a normal response to pacing. The PVCs were multifocal in all three. All three had 24 hour ambulatory ECG monitoring and in two this revealed couplets, although none had ventricular tachycardia. None of these patients was performing exercise when he died. Autopsy was performed on the one patient who died in our hospital (with documented fibrillation). The myocardial histology was normal in specimens from both ventricles. No autopsy was done on the other two patients. Digitalis toxicity was not considered a likely cause of death since they were taking a low stable dose at the time of death.

**Table 5. Atrioventricular and His-Purkinje Conduction**

<table>
<thead>
<tr>
<th>ECG abnormality</th>
<th>LRA-H interval (msec)</th>
<th>Normal</th>
<th>Increased</th>
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<tbody>
<tr>
<td>RBBB</td>
<td>36</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>RBBB &amp; LAHB</td>
<td>7</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>PVCs</td>
<td>9</td>
<td>0</td>
<td></td>
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<tr>
<td>1° AVB</td>
<td>5</td>
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Discussion

The cause of sudden death in patients who have had intracardiac repair of tetralogy of Fallot has been controversial. Wolff et al. in 1972 reported a series of patients with right bundle branch block and left anterior hemiblock who died suddenly. Although most also had PVCs, it was speculated that they may have developed complete heart block. Moss et al. reported on postoperative patients who developed late onset complete heart block. Half had had tetralogy of Fallot repair and one of these died. Sondheimer et al. found RBBB and LAH in 23% of their postoperative tetralogies, but only one sudden death in this group. They, as well as Moss et al., noted that late CAVB could develop in patients with only RBBB. Steeg et al. found RBBB and LAH in 8.7% of their postoperative tetralogies with no late CAVB or deaths. They speculated, based on the work of Godman et al., that His bundle recordings showing a prolonged HV interval might be of prognostic importance. Godman et al. found a 50% incidence of prolonged His-Purkinje conduction (HV) in patients with RBBB-LAH and also noted that all patients with prolonged HV interval had had transient CAVB at the time of surgery. Several other electrophysiologic studies have been performed in postoperative tetralogy patients. We reported one patient with RBBB-LAH studied two weeks after transient surgical complete A-V block who had a prolonged HV interval. Her study, 18 months postoperatively, is included in the present series. At that time she showed a normal HV interval, but a prolonged refractory period below the His potential.

In the present series, only one of eight patients with RBBB and LAH showed a prolonged HV interval. Only two of the remaining seven had refractory period determination and one of these was prolonged. None of our patients with RBBB and LAH have died or developed late complete A-V block. This was also the experience of Cairns who found no correlation between conduction disturbances and long-term outcome of the patients. In our study, there was no apparent correlation between 1° A-V block, prolonged LRA-H, HRA-LRA, or HV with each other or with a bad long-term outlook. It is of interest that three of six patients with 1° A-V block also had premature ventricular contractions.

Premature ventricular contractions after tetralogy repair have been found to be associated with sudden death and ventricular dysrhythmias by James et al. and Quattlebaum et al. and were considered more important than conduction disturbances by these authors. PVCs may also be induced by exercise in some patients. The significance of PVCs at rest versus exercise in these patients remains controversial. In our group, three of nine (33%) with PVCs at rest have died (patients 22, 30 and 39), one in ventricular fibrillation and two suddenly, apparently of a dysrhythmia. None of these patients had LAH. One (case 30) had prolonged HV interval, but a normal response to pacing indicating a functional normal conduction system. Patient 39 had 1° A-V block and all of his conduction intervals were near the upper limit of normal. Patient 22 had transient CAVB for only one hour after his repeat surgery to relieve persistent pulmonic stenosis. All of these young men had persistent pulmonic stenosis with RV pressures of 75–80% systemic and poorly contracting ventricles. One was being treated with digoxin and propranolol for his congestive heart failure and PVCs, another with digoxin alone for congestive heart failure. The third patient had been treated with quinidine and propranolol, but because of increasing cardiomegaly the propranolol was stopped. He died one month later. He had had multifocal PVCs with R on T which worsened with exercise. The other six patients are all currently being treated with quinidine, although the efficacy of this approach is not proven. Five of these patients also have poorly contractile ventricles. Thus, in these three patients who have died with PVCs after tetralogy repair there was no evidence of significant conduction problems by intracardiac electrophography. Also, in our patients with conduction disturbances there have been no late serious dysrhythmias.

Sinus node dysfunction has been reported to lead to serious postoperative dysrhythmias after Mustard operation for transposition of the great arteries. Sinus node dysfunction has also been reported in postoperative tetralogies, but has not been correlated with clinical dysrhythmias. Our studies confirm this observation in that four patients were found to have mildly prolonged corrected sinus node recovery times but none have had clinical dysrhythmias referable to the sinus node.

It thus appears that several different mechanisms may be responsible for sudden death in postoperative tetralogy patients. Ventricular dysrhythmias in those with PVCs, particularly with associated heart failure, seems to be one common mechanism. Late onset complete heart block can occur in any setting, but particularly with trifascicular block or trifascicular block after transient complete A-V block. Further follow-up after electrophysiologic studies is needed in this group. Prevention of these deaths may be produced by protection of the myocardium during ischemic arrest and protection of conduction system.

References

Electrophysiologic and Pharmacologic Characteristics of Automatic Ectopic Atrial Tachycardia

PAUL C. GILLETTE, M.D., AND ARTHUR GARSON, JR., M.D.

SUMMARY In seven children six weeks to nine years of age, the diagnosis of chronic atrial tachycardia due to an automatic ectopic focus was established by clinical course, by the recording of intracardiac electrograms, and by atrial stimulation during cardiac catheterization. Both override atrial pacing and programmed premature atrial stimulation failed to influence the tachycardia. Digoxin re-established sinus rhythm in one patient while it slowed the tachycardia rate slightly in six. Propranolol with digoxin was effective in restoring sinus rhythm in three cases, ineffective in one, and slowed the rate in two. Diphenylhydantoin was effective in one of two patients in whom it was used. Reserpine restored sinus rhythm in the one patient to whom it was given. Although automatic ectopic atrial tachycardias are difficult to manage, an aggressive diagnostic and pharmacologic program results in a high degree of control.

NEW ELECTROPHYSIOLOGICAL TECHNIQUES have allowed the classification of atrial tachycardia into two major types: automatic ectopic focus and re-entry. The ectopic focus type is rare in adults but often occurs in children. It is likely that treatment of these two types of tachycardia will be different. It is the purpose of this report to observe the response of ectopic atrial tachycardia to atrial pacing and to long term drug treatment.

Methods Seven patients, six weeks to nine years of age were studied. Each subject was first evaluated by history and physical examination, 15 lead electrocardiography and chest roentgenography. All medications were discontinued 24 hours before the study. All except the youngest were sedated with meperidine 2 mg/kg, chlorpromazine 0.5 mg/kg, and promethazine 0.5 mg/kg, 30 minutes before the study. Two electrode catheters were inserted percutaneously into the femoral veins and positioned in the right heart under fluoroscopic and electrocardiographic control for His bundle potential and atrial recording as previously reported. In one case, an additional quadripolar electrode catheter was positioned in the coronary sinus after percutaneous insertion in a left antecubital vein. Right atrial pacing at a rate slightly faster than the existing tachycardia was carried out in six patients with a Med Data MD-1 rapid stimulator, and recordings were made during abrupt termination of pacing. In five subjects, single, progressively more premature atrial beats were then introduced during existing tachycardia by coupling to the atrial electrogram with a Medtronic 5837 stimulator. External cardioversion with 2 watt seconds/kg of body weight was done in six patients.

In three subjects who had periods of sinus rhythm during the study, premature atrial beats were coupled into their sinus rhythm. In four subjects, propranolol 0.1 mg/kg i.v., over ten minutes was given during the study and in three, xylocaine, 1 mg/kg i.v. bolus was also administered.

In each case, digoxin was selected as the initial drug for chronic treatment of their tachycardia. If digoxin proved ineffective, a second drug was added. Before declaring any drug ineffective, the dose was increased until clinical signs of mild toxicity developed or the serum level was in the upper

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