Ventricular Fibrillation

A Possible Mechanism of Sudden Death in Patients with Wolff-Parkinson-White Syndrome

By Leonard S. Dreifus, M.D., Robert Hiat, M.D., Yoshio Watanabe, M.D., Jaime Arriaga, M.D., and Norman Reitman, M.D.

SUMMARY

A case of Wolff-Parkinson-White syndrome associated with ventricular fibrillation is presented. The control of recurrent ventricular fibrillation with large doses of digitalis and other antiarrhythmic drugs, including propranolol, lidocaine, procainamide, and quinidine, is discussed. As far as we can determine, this is the first human case in which the precise onset of ventricular fibrillation was documented. Possible mechanisms are presented that may produce ventricular fibrillation in patients with Wolff-Parkinson-White syndrome associated with atrial flutter or fibrillation.

Additional Indexing Words:
Pre-excitation Ventricular vulnerability Wolff-Parkinson-White tachycardia Electronic demand pacemaker Atrial fibrillation

The purpose of this paper is to report a case of ventricular fibrillation in a patient with Wolff-Parkinson-White (W-P-W) syndrome. Such an association is rare and has been documented in only six previous observations. It may account for some instances of unexplained sudden deaths in the W-P-W syndrome.

Case Report

A 63-year-old female was admitted to the Hahnemann Hospital on December 1, 1969. She had a history of supraventricular paroxysmal tachycardia with narrow QRS complexes for many years. Numerous electrocardiograms showed type A W-P-W configuration (fig. 1). The patient had been doing well until September, 1969, when she noted recurrent episodes of lightheadedness, without palpitations. She was then admitted to St. Michael’s Hospital in Newark, New Jersey, for implantation of a transvenous demand pacemaker in an attempt to control her recurrent tachycardia. Although the rhythm was moderately well-controlled for three or four weeks, she had to be readmitted to St. Michael’s Hospital in November, 1969, because of the recurrence of persistent tachycardia.

The patient was subsequently transferred to the Hahnemann Hospital for further evaluation. On admission, the physical examination revealed an obese, slightly lethargic white female in no acute distress. Her blood pressure was 100/60 mm Hg, and her pulse was irregularly irregular at 180 beats/min. Neck vein distention was present at 35°. The previously implanted pacemaker was palpated in the left anterior chest wall. All peripheral pulses were palpable. The right calf was tender. Heart tones were distant but no murmur, rub or gallop was present. Decreased breath sounds were noted at the right base. The examination of the other systems was not contributory.

The electrocardiogram taken on admission (fig. 2) revealed atrial fibrillation with an average ventricular response of 190 beats/min. The QRS complexes were wider and more bizarre. Evidence of actual delta waves could not be seen in this tracing although the QRS configuration was similar to figure 1, and type A pre-excitation was suggested. Confirmation of atrial fibrillation was made by a right atrial electrogram (fig. 3).

The chest roentgenogram revealed cardiomegaly and a right pleural effusion which was
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A sinus rhythm is present. Prominent delta waves are seen in this tracing, showing W-P-W type A.

**Figure 1**

ECG taken on admission, showing atrial fibrillation with an irregular ventricular response (average rate of 190 beats/min).

thought to be associated with a pulmonary infarction. The results of all laboratory studies were normal, including electrolytes and serum enzymes. An acute myocardial process was not identified.

**Hospital Course**

Intravenous propranolol at a rate of 2 mg/hour was started, and the patient was given 0.25 mg of digoxin intravenously in an attempt to slow the ventricular rate. Two hours later, a second dose of 0.25 mg of digoxin was administered, and, because of the continued arrhythmia, a third dose of 0.25 mg of digoxin was given two hours later (midnight). At 2 a.m. (December 2, 1969), the patient suddenly lost consciousness. The heart sounds were no longer perceptible. On the electrocardiogram (fig. 4), ventricular fibrillation was identified and was reverted by a direct current precordial shock. Because a rapid ventricular response continued even after the ventricular fibrillation had been reverted, 0.25 mg of digoxin was given again on two occasions. A second episode of ventricular fibrillation occurred at 6 a.m. that morning, and was again reverted by direct current precordial shock. A lidocaine drip
Figure 3
The right atrial electrogram (RAE) shows atrial fibrillation. Arrows indicate pacemaker-induced beats.

Figure 4
Direct current defibrillation terminated the ventricular fibrillation. (A.) Onset of ventricular fibrillation. (B.) Ventricular fibrillation. (C.) Following direct current shock, electronic pacemaker controls the heart. (D.) 15 minutes later, supraventricular tachycardia.
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was started but quickly discontinued due to the onset of extreme drowsiness and confusion. Intravenous quinidine was given at a rate of 100 mg every 2 hours, as well as intravenous procaainamide which was given at an average rate of 2 mg/min. Digoxin (0.25 mg intramuscular) was continued daily. These drugs were alternately titrated during the first week in order to achieve an appropriate regimen which would control the patient's heart rate without engendering hypotension. A therapeutic dose of $^{131}$I (18 mCi) was administered on December 9, 1969. On this regimen, no further evidence of tachycardia was noted, and sinus rhythm was predominantly present. Slowing of the ventricular rate below 58 beats/min allowed the demand pacemaker to escape and control the ventricular rate (fig. 5).

The patient stayed in the hospital for 27 days, and the remainder of her course was uneventful. She was maintained on intravenous heparin because of the possible presence of a pulmonary infarction. During the second week, the following antiarrhythmic drugs were administered orally: propranolol (20 mg), quinidine (200 mg), procaainamide (250 mg, four times daily), and digoxin (0.25 mg daily). The patient was discharged (December 27, 1969) on this regimen, and was free from both paroxysmal tachycardia and heart failure until August, 1970.

Discussion

Cardiac arrhythmias, especially supraventricular tachycardias, are well known to occur in patients with W-P-W syndrome.1-5

Reports of ventricular tachycardia in patients with W-P-W syndrome have been reviewed by Langendorf et al.,2 Giraud et al.,3 and Newman et al.4 According to Langendorf et al., no unequivocal case of ventricular tachycardia could be demonstrated, as independent atrial activity could not be identified. Furthermore, in their review they said: "It is likely that these so-called ventricular tachycardias were in fact, supra-ventricular tachycardias with widened and slurred ventricular complexes."2

Although the prognosis of W-P-W syndrome is usually said to be benign in the absence of underlying cardiopathy, several sudden and unexpected deaths in these patients have been previously reported. Okel6 reviewed 22 patients and reported an additional case of sudden death associated with W-P-W syndrome. Ten other previously reported cases were identified during the preparation of this manuscript.5,7-14 The

Figure 5

The ventricles are under the control of a demand type pacemaker with retrograde conduction to the atria.
mechanism of sudden death has not been demonstrated with accuracy. Ventricular fibrillation as a possible mechanism of sudden death in patients with W-P-W syndrome has been observed in only six other instances. (Table 1).

The two observations of ventricular fibrillation in W-P-W syndrome reported by Schwartz and Jezer (1934) and Gould and Mundal (1951), and mentioned by Prinzmetal et al., are questionable because neither the presence of W-P-W complexes nor ventricular fibrillation was clearly identified. Fox et al. reported in 1952 the first documented case of W-P-W syndrome associated with ventricular fibrillation. In this observation, ventricular fibrillation occurred 4 hours after the intravenous administration of 500 mg of procainamide.

Five other cases have been reported subsequently. In one of these patients, an autopsy showed myocarditis, interatrial lipomatous hypertrophy, and a prominent right moderator band. All of the other patients were successfully resuscitated, usually by one or two direct current precordial shocks. It is noteworthy that all patients presented had a previous history of palpitations or proved paroxysmal tachycardias, and in three cases, including this present report, atrial fibrillation was present at the time ventricular fibrillation occurred.

In the case presented here, one might argue that ventricular fibrillation was due to the presence of the previously inserted pacemaker, and represented a complication of the electronic pacing rather than a hazard of W-P-W syndrome. This is unlikely, however, since the pacemaker was of the demand, R-inhibited type (Medtronics 3841), and electrical stimulation occurred only after 1 sec of asystole.

The pathogenesis of ventricular fibrillation associated with W-P-W syndrome is unclear. Previous observations concerning the mechanism of premature atrial and ventricular stimulation, with the production and the interruption of recurrent tachycardia as well as the effective results of surgical ligation of the atrioventricular (A-V) junction or the accessory pathway, may offer several clues to the onset of ventricular fibrillation.

Durrer and Roos indicated that, depending upon the timing of premature atrial systole and the state of refractoriness of the His and Kent bundles, excitation of the ventricles could occur either predominantly through one of these two conduction pathways or through both. On the other hand, they said that if a premature ventricular stimulus was given at a

Table 1

<table>
<thead>
<tr>
<th>Authors</th>
<th>Case no.</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms</th>
<th>VF</th>
<th>Atrial fibrillation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fox et al.</td>
<td>1</td>
<td>1952</td>
<td>70</td>
<td>F</td>
<td>Occasional dizziness &amp; palpitations</td>
<td>Yes</td>
<td>Transient episode 3 months later</td>
</tr>
<tr>
<td>Ahlinder et al.</td>
<td>2</td>
<td>1963</td>
<td>42</td>
<td>M</td>
<td>None</td>
<td>Yes</td>
<td>Concomitant</td>
</tr>
<tr>
<td>Touche et al.</td>
<td>3</td>
<td>1966</td>
<td>21</td>
<td>M</td>
<td>Paroxysmal tachycardia 1 year ago</td>
<td>Yes</td>
<td>None</td>
</tr>
<tr>
<td>Okel</td>
<td>4</td>
<td>1968</td>
<td>33</td>
<td>M</td>
<td>Transient atrial fibrillation</td>
<td>Yes</td>
<td>Concomitant</td>
</tr>
<tr>
<td>Wojtasik et al.</td>
<td>5</td>
<td>May, 1969</td>
<td>18</td>
<td>F</td>
<td>None</td>
<td>Yes</td>
<td>None</td>
</tr>
<tr>
<td>Kaplan et al.</td>
<td>6</td>
<td>Aug., 1969</td>
<td>27</td>
<td>F</td>
<td>Palpitations for 5 years</td>
<td>Yes</td>
<td>None</td>
</tr>
<tr>
<td>Dreifus et al.</td>
<td>7</td>
<td>1970</td>
<td>63</td>
<td>F</td>
<td>Recent palpitations</td>
<td>Yes</td>
<td>Concomitant</td>
</tr>
</tbody>
</table>

Abbreviations: VF = ventricular fibrillation; DCD = direct current defibrillation. *Case reported by the authors in this paper.
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<table>
<thead>
<tr>
<th>Emergency treatment</th>
<th>Immediate evolution</th>
<th>Follow-up</th>
<th>Additional remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>Spontaneous reversion to sinus rhythm</td>
<td>Doing well 3 months later</td>
<td>Ventricular fibrillation occurred after injection of procainamide (Pronestyl) (500 mg)</td>
</tr>
<tr>
<td>1 DCD; 1 DCD</td>
<td>Alive; Alive</td>
<td>Alive</td>
<td></td>
</tr>
<tr>
<td>External massage</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 DCD</td>
<td>Alive</td>
<td>Doing well a few weeks later</td>
<td>Autopsy report</td>
</tr>
<tr>
<td>1 DCD</td>
<td>Died</td>
<td>Sudden death 6 months later</td>
<td>Normal selective coronary arteriography</td>
</tr>
<tr>
<td>DCD</td>
<td>Alive</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 DCD</td>
<td>Alive</td>
<td>Doing well</td>
<td>Myocardial infarction 1965; demand pacemaker inserted 1969</td>
</tr>
<tr>
<td>2 DCD</td>
<td>Alive</td>
<td>Doing well 8 months later</td>
<td></td>
</tr>
</tbody>
</table>

A sharply defined delay interval of about 320 msec, the retrograde P wave was followed by a supraventricular tachycardia with normal excitation of the ventricles.

Durrer and Roos also suggested that the pre-excitation syndrome could cause a tachycardia resembling a ventricular mechanism by a circus movement in a reverse direction. Basically, the impulse would enter the normal A-V transmission system in a retrograde fashion to activate the atria and then reenter the ventricles through the bypass. Hence, depending on whether the circus movement moved antegrade through the normal A-V conduction system or through the bypass, the resulting tachycardia could resemble either a supraventricular or ventricular mechanism.

It was further noted that the ligation of the A-V node or the accessory pathway could permanently prevent recurrent tachycardia, and that both pathways must be intact for initiation and perpetuation of the W-P-W tachycardia. On the other hand, in the presence of atrial flutter or fibrillation, competition for the normal A-V transmission system and the bypass fibers can occur. If conduction is predominantly through the normal A-V conduction system, the QRS complexes may remain narrow. But if conduction through the bypass fibers occurs in rapid sequence, the...

Figure 6

Atrial fibrillation is present. The ventricular response is rapid and irregular. The second ventricular complex following the long pause occurs on the apex of the preceding T wave. Several multiform beats are then followed by ventricular fibrillation. Note that ventricular fibrillation did not occur with rapidly occurring beats prior to pause (star). The pause may have altered the refractory period of the ventricles.
ventricular rate may become excessive with wide bizarre QRS complexes.

Several of the cases of W-P-W syndrome with "ventricular tachycardia" are, therefore, probably due to either atrial flutter or atrial fibrillation, with the impulse being predominantly conducted through the bypass fibers rather than through the normal A-V transmission system.

It does not seem unreasonable to postulate that in the case reported here, in which the precise moment of the onset of ventricular fibrillation was recorded in the presence of atrial fibrillation, impulses passed through the accessory fibers at a rapid frequency and reached the ventricles during the vulnerable period of ventricular excitability (fig. 6). According to Boineau and Moore,25 who made similar experimental observations, ventricular fibrillation occurred repeatedly in the dog as a direct result of the rapid ventricular rate permitted by the accessory pathway subsequent to induced atrial fibrillation. The mechanism engendering ventricular fibrillation may be similar in this case. This situation is physiologically not possible without accessory pathways because the refractory period of the A-V junctional tissues produces 2:1 or higher conduction ratios, thereby preventing excitation of the ventricles in the vulnerable period.26

Conduction block in the bypass fibers has also been described.27 The fact that fatal ventricular fibrillation does not ensue more often in the presence of atrial fibrillation with pre-excitation is probably explained on this basis.

Although ventricular pacing by the already implanted demand pacemaker was fortuitous in the case presented here, suppression of all A-V conduction and electronic pacing appears to offer a method of therapy in patients with the W-P-W syndrome and persistent atrial fibrillation (fig. 5). Subsequent to this case, one other similar case has been successfully managed in a similar way.

Digitalis may have partly contributed to the onset of ventricular fibrillation in the case currently reported. However, it was continued after successful defibrillation and control of conduction in both the normal and the accessory A-V pathways by appropriate drug administration. Furthermore, it has been clearly shown that ventricular fibrillation can be produced in the absence of digitalis.28 Also, factors favoring a longer vulnerable period of excitability, such as myocardial ischemia, may have contributed to the onset of ventricular fibrillation. However, it should be emphasized that any of these factors in the absence of W-P-W are capable of precipitating ventricular fibrillation. Nevertheless, it is quite clear that the rapid succession of supraventricular impulses with subsequent QRS complexes falling on the T wave of the previous beat may have led to the onset of ventricular fibrillation in the case we have presented here (fig. 6).

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