Ventricular Tachycardia and Ventricular Fibrillation in a Young Population

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SUMMARY In this study, we describe the findings in 18 young patients (age range 4 days to 24 years, mean 16.6 years) who had ventricular tachycardia and/or ventricular fibrillation and were followed for 4–70 months (mean 22.4 months). Patients had a variety of problems associated with their arrhythmia, including mitral valve prolapse, cardiomyopathy, myocarditis, prolonged QT syndrome and hypokalemia. Six patients had no clinically recognizable cardiac abnormality. The ventricular tachycardia showed a left bundle branch block contour in 10 of 17 patients, right bundle branch block in four, was multiformal in two and had an indeterminate contour in one. Sustained ventricular tachycardia was initiated and terminated reproducibly by atrial and ventricular stimulation in three of seven patients who did not have spontaneous episodes of ventricular tachycardia during the electrophysiologic study. In one other patient, short bursts of ventricular tachycardia were induced. Patients who had ventricular fibrillation, those who died, and those who are still symptomatic with poorly controlled ventricular arrhythmias had significant heart disease. In one patient, a ventricular tachyarrhythmia that had required more than 100 electrical cardioversions spontaneously disappeared after requiring 1 year of antiarrhythmic therapy.

AMBULATORY electrocardiographic recordings and invasive electrophysiologic studies have increased our knowledge about tachyarrhythmias that occur in young patients. Although ventricular tachycardia and ventricular fibrillation occur more often in adults with coronary artery disease, these ventricular arrhythmias may appear in young people, often early and late after surgery for congenital heart disease or in association with a variety of cardiac diseases, autonomic imbalance, drugs, as well as in the absence of detectable cardiac disease. When serious ventricular tachyarrhythmias occur in the young they may be misdiagnosed as aberrantly-conducting supraventricular tachycardias because of their presumed infrequency.

In this study we describe the clinical and electrophysiologic findings in 18 young patients who had ventricular tachycardia and/or ventricular fibrillation, who were extensively evaluated and followed for 4–70 months (mean 22.4 months).

Methods

Patient Evaluation

All patients underwent history and physical examination, as well as a chest x-ray and multiple 12-lead ECGs. Seventeen had 24-hour ambulatory recordings and 16 were also monitored in the hospital.

Sixteen patients had M-mode and/or two-dimensional echocardiograms and 12 had submaximal exercise stress tests. Eleven exercised on a treadmill, according to a modified Bruce protocol, and one patient (no. 9) on a bicycle ergometer. Nine patients underwent cardiac catheterization, including seven who had coronary arteriography, to rule out the presence of abnormal coronary circulation.

The diagnosis of ventricular tachycardia was made using standard scalar electrocardiographic criteria of fusion and capture QRS complexes. In patients who had spontaneous or induced ventricular tachycardia at the time of electrophysiologic study, we demonstrated that His bundle activation occurred after the onset of the QRS complex (in the absence of the Wolff-Parkinson-White syndrome), or that we could not record His bundle potentials that were previously well-recorded during sinus rhythm in the absence of obvious catheter shift. To avoid confusion, the term accelerated idioventricular rhythm has not been used.

Electrophysiologic Study

Eleven patients underwent electrophysiologic study using two to four tripolar or quadripolar electrode catheters introduced into one or both femoral veins. Cases 9 and 12 were sedated with meperidine (1 mg/kg), promethazine (0.25 mg/kg) and chlorpromazine (0.25 mg/kg). Intracavitary tracings along with scalar leads I, II, III, V1 and V6 were recorded on a multichannel oscilloscope recorder (Electronics for Medicine DR8 or VR12) at a paper speed of 100 mm/sec, using filter settings of 30 or 40–500 Hz for the intracavitary electrograms. The right atrium and apex of the right ventricle were each stimulated using a programmable stimulator (Medtronic 5325 or WP Instruments, Inc.), with rectangular stimuli of 1–2 msec duration at an intensity equal to twice the diastolic excitability threshold.

The following protocol was used to precipitate ven-
tricular tachycardia in patients who did not have it spontaneously at the time of study:

1) premature atrial stimulation during sinus rhythm and atrial pacing. The entire cardiac cycle was scanned to the point of atrial refractoriness.

2) atrial pacing at progressively shorter cycle lengths until atrioventricular (AV) block occurred;

3) premature ventricular stimulation during right ventricular pacing. The entire cardiac cycle was scanned to the point of ventricular refractoriness. Stimulation at two cycles lengths was tried before we concluded that the ventricular tachycardia could not be started;

4) if single premature ventricular stimulation failed to induce ventricular tachycardia, two premature ventricular stimuli were introduced. After the first premature ventricular stimulus at the shortest coupling interval that consistently produced a ventricular response, a second premature ventricular stimulus was delivered at progressively shorter coupling intervals to the first, until the point of ventricular refractoriness.

**Patient Follow-up**

Within 4 months of the preparation of this report, all survivors underwent a continuous 24-hour, two-channel, ambulatory recording (Avionics, model 445).

Tapes were scanned visually (Avionics Cardioscanner model 660A) by an experienced technician. Ventricular ectopy in the tapes of patients 1, 4, 11-15 and 18 was counted using a Honeywell 716 computer program.14

**Results**

**Patient Profile (Table 1 and 2)**

The age range was 4 days to 24 years (mean 16.6 years). There were nine females and nine males, and all patients were white. Most patients were symptomatic with their arrhythmia. Twelve patients had organic heart disease. No patient had evidence of the Wolff-Parkinson-White syndrome by surface ECG or electrophysiologic evaluation. Three patients died at 5, 34 and 37 months after symptoms began.

Patients are presented below in groups according to their cardiac diagnoses. The clinical course of some patients is elaborated in greater detail.

**Mitral Valve Prolapse**

Four patients (1-4) had mitral valve prolapse diagnosed by physical findings16 and echocardiogram.16 Three patients (2-4) are taking antiarrhythmic drugs.

**Cardiomyopathy**

Three patients (5-7) were diagnosed as having severe congestive cardiomyopathy by the presence of generalized reduced ventricular wall motion, dilated chambers and normal coronary arteries. They had no evidence of asymmetric septal hypertrophy by echocardiography.

Case 5 presented with palpitations and dyspnea on exertion. Despite aggressive management of his ventricular arrhythmias, he suffered sudden death 37 months later while taking quinidine and propranolol.

Case 7 was noted to have premature ventricular complexes 5 years before admission but received no therapy and had no problems. Heart size was normal by echocardiogram and chest x-ray. She was 5 months pregnant with her first pregnancy when she had a syncope spell and presented to her local hospital in ventricular fibrillation. After resuscitation, she continued to have episodes of ventricular tachycardia and was referred for therapy. An echocardiogram showed biventricular enlargement and generalized poor wall motion consistent with a cardiomyopathy. Because she was pregnant, cardiac catheterization was not performed. During her hospitalization she spontaneously delivered stillborn. Despite therapy with conventional and investigational drugs that appeared to limit the ventricular arrhythmia to multiformal ventricular complexes and three or four beat episodes of ventricular tachycardia, she died suddenly 5 months after her initial presentation with syncope. This patient illustrates the potential latency between the beginning of a cardiomyopathy and the development of serious problems. In this patient the cardiomyopathy may have begun with the onset of premature ventricular

**Table 1. Patient Profile**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (years)/ sex</th>
<th>Diagnosis</th>
<th>Symptoms with Arrhythmia</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>22/F</td>
<td>MVP</td>
<td>Syncope</td>
</tr>
<tr>
<td>2</td>
<td>15/M</td>
<td>MVP</td>
<td>Syncope</td>
</tr>
<tr>
<td>3</td>
<td>22/F</td>
<td>MVP</td>
<td>Palpitations with extremes of exercise</td>
</tr>
<tr>
<td>4</td>
<td>18/F</td>
<td>MVP</td>
<td>Presyncope, precordial pain</td>
</tr>
<tr>
<td>5</td>
<td>21/M</td>
<td>Cardiomyopathy</td>
<td>Palpitations, dyspnea on exertion</td>
</tr>
<tr>
<td>6</td>
<td>22/M</td>
<td>Cardiomyopathy</td>
<td>Syncope</td>
</tr>
<tr>
<td>7</td>
<td>24/F</td>
<td>Cardiomyopathy</td>
<td>Syncope</td>
</tr>
<tr>
<td>8</td>
<td>15/M</td>
<td>Myocarditis</td>
<td>Syncope</td>
</tr>
<tr>
<td>9</td>
<td>11/F</td>
<td>Aortitis, carditis</td>
<td>Syncope, dyspnea</td>
</tr>
<tr>
<td>10</td>
<td>17/F</td>
<td>Long QT</td>
<td>Syncope</td>
</tr>
<tr>
<td>11</td>
<td>4 days/M</td>
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<td>Uncompromised</td>
</tr>
<tr>
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<td>17/F</td>
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<td>22/M</td>
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</tr>
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</tr>
<tr>
<td>18</td>
<td>21/M</td>
<td>None</td>
<td>Palpitations</td>
</tr>
</tbody>
</table>

Abbreviations: MVP = mitral valve prolapse; VSD = ventricular septal defect.
complexes, but the patient was asymptomatic until she presented with ventricular fibrillation 5 years later.

Myocarditis

Two patients (8 and 9) were diagnosed as having myocarditis, one presumed viral and one without known etiology. Case 8 had ventricular tachycardia that exhibited slow rates. Cardiac catheterization showed generalized hypokinesis with increased end-diastolic volume and end-systolic volume. No treatment for the ventricular tachycardia was given. During 70 months of follow-up, the patient has remained asymptomatic. He exercises without restriction and his recent 24-hour ECG recording demonstrated no ventricular tachycardia. His ventricular arrhythmia has apparently spontaneously disappeared with resolution of the viral myocarditis. His echocardiogram is now normal.

Case 9 had numerous episodes of ventricular tachycardia and ventricular fibrillation that were associated with syncope and presyncope. Cardiac catheterization demonstrated supravalvular aortic stenosis (systolic gradient of 12 mm Hg), mild aortic insufficiency and luminal aortic defects consistent with an arteritis. No evidence for an infectious or collagen disease was found. She had progressive cardiac decompensation and more frequent episodes of ventricular tachycardia and died 34 months after her initial presentation.

Long QT

Case 10 had spontaneous ventricular fibrillation from which she was resuscitated. Several ECGs demonstrated the QTc interval to be 480 msec. Family history revealed that her sister died suddenly at age 21 years. Two other siblings and her mother have prolonged QTc with intervals of 490, 500 and 460 msec, respectively.

Congenital Heart Disease

A 4-day-old infant (patient 11) had ventricular tachycardia and cardiac findings consistent with a small ventricular septal defect.

Unknown Heart Disease

Case 12 was admitted with continuous, multiform ventricular tachycardia interrupted by widely spaced sinus beats (fig. 1). She presented with syncope at age 20 months and required more than 100 electrical cardioversions in the 6 months before her referral. Cardiac catheterization was normal and echocardiography revealed an enlarged left atrium. Conventional antiarrhythmic therapy, a trial of overdrive pacing, tocanide, and left and right stellate ganglion block did not control the ventricular tachycardia. The combination of aprindine and quinidine slowed the rate of the ventricular tachycardia to approximately 100–120 beats/min, but did not suppress it. She continued to have “controlled” ventricular tachycardia on this treatment program for 12 months, at which time she was noted to have more sinus beats on routine ECGs. Finally, only sinus rhythm was recorded by ECG and ambulatory 24-hour recording. Since it appeared that the ventricular tachycardia had now disappeared, she was slowly weaned from antiarrhythmic therapy without recurrence of ventricular tachycardia and ventricular fibrillation. Presently she is a healthy 3½-year-old, free of symptoms 6 months after the discontinuation of all drugs. She is discussed in greater detail because she appears to show the inexplicable, spontaneous remission of a life-threatening ventricular arrhythmia.

No Cardiac Disease Demonstrable

On the basis of normal physical examination, 12-lead ECGs, cardiac enzymes, chest x-rays and echocardiograms, six patients (cases 13–18) were felt not to have any clinically recognizable cardiac abnor-
mality except for the rhythm disturbance. Since only case 14 underwent cardiac catheterization, we cannot exclude the possibility that catheterization might have revealed the presence of occult cardiac disease as a cause of the ventricular tachycardia in these patients.

Case 13 was admitted after a syncopal spell and was found to have ventricular tachycardia consistent with torsade de pointes\(^9\) (fig. 2). She was the only patient in whom ventricular tachycardia was not proved at the time of electrophysiologic study or by the presence of fusion or capture beats. Serum potassium was 2.2 mEq/l. Although intensively evaluated, no etiology for the hypokalemia was determined and no cardiovascular disease was found.

Electrocardiographic Characteristics (Table 3)

The resting ECG during normal AV conduction demonstrated normal sinus rhythm in 17, and one patient had atrial fibrillation. The QRS complex was normal in 14 and demonstrated incomplete or complete right bundle branch block in four. The QTc was normal except for case 10. Thirteen patients had ventricular tachycardia only, one had ventricular fibrillation only and four had both ventricular tachycardia and ventricular fibrillation. During ventricular tachycardia, the QRS morphology exhibited a left bundle branch block in 10 patients, right bundle branch block in four, was indeterminant in one and multiform in two. The ventricular rate ranged from 66–250 beats/min. Sixteen patients manifested AV dissociation. In 11 patients the AV dissociation was incomplete owing to supraventricular captures and/or fusion beats. Patient 17 had episodes of ventricular tachycardia at rates of 188 beats/min and 66 beats/min (fig. 3), both of which produced incomplete AV dissociation.

Electrical Cardioversion

Nine patients required one or more electrical cardioversions, including the five with ventricular fibrillation.

Exercise Stress Test Results

Twelve patients had one or more submaximal exercise stress tests. In three patients, exercise increased the degree of ventricular ectopy by precipitating ventricular tachycardia (patients 3 and 14) (fig. 4) and by precipitating pairs of consecutive premature ventricular complexes in a patient who had
 Только один предсердный комплексы в покое (пациент 1).

Двое пациентов увеличивают частоту предсердных комплексов в период после тренировки. Пациент 6, у которого не было предсердных экстрасистол перед тренировкой, развил предсердные экстрасистолы в период после тренировки. Пациент 18 имел пары предсердных экстрасистол после тренировки, но только один предсердный комплекс перед и во время тренировки.

Вентрикулярная тахикардия, присутствующая в покое, подавлялась тренировкой в пациентах 8 и 16. Тренировка подавила более медленную вентрикулярную тахикардию у пациента 17; более быстрая вентрикулярная тахикардия не проявлялась.

<table>
<thead>
<tr>
<th>Case</th>
<th>VT/VF</th>
<th>PVC QRS morphology</th>
<th>QRS morphology</th>
<th>Rate (beats/min)</th>
<th>AV dissociation</th>
<th>Fusions/captures</th>
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<td>1</td>
<td>Yes/no</td>
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<td>RBBB and R axis</td>
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<td>4</td>
<td>Yes/no</td>
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<td>RBBB and L axis</td>
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<td>Complete</td>
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<tr>
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<td>LBBB</td>
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<tr>
<td>6</td>
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<td>9</td>
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<td>10</td>
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<td>RBBB</td>
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<td></td>
<td></td>
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<tr>
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<tr>
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<tr>
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<tr>
<td>18</td>
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<td>LBBB</td>
<td>LBBB</td>
<td>188</td>
<td>Incomplete</td>
<td>Yes/yes</td>
</tr>
</tbody>
</table>

*Вентрикулярная фибрилляция только.
† Торс ade de pointes.

ОБозначения: VT = вентрикулярная тахикардия; VF = вентрикулярная фибрилляция; PVC = вентрикулярный комплекс; LBBB = левый пучок ветвей; RBBB = правый пучок ветвей; L axis = левая ось (−30°); R axis = правая ось (+120°).
during exercise in this patient. Ventricular tachycardia returned in all three patients after exercise. Exercise did not elicit ventricular ectopy in patients 2, 9, 10 and 13.

Electrophysiologic Studies (Table 4)

Eleven patients underwent an electrophysiologic study. Three patients (cases 9, 12 and 15) had frequent, spontaneous bursts of ventricular tachycardia that precluded pacing attempts to initiate or terminate the ventricular tachycardia in any sort of a routine fashion. Case 8 had 3–5-beat episodes of spontaneous ventricular tachycardia unrelated to pacing, but that could be captured at faster pacing rates. Programmed stimulation of the atrium and ventricle failed to initiate ventricular tachycardia in cases 1, 8, 10 and 13.
Phenylephrine, norepinephrine and isoproterenol infusions, right and left stellate ganglion block, plus premature ventricular stimulation after each stellate block also failed to precipitate ventricular tachycardia or ventricular fibrillation in patient 10. Single spontaneous ventricular responses occurred after premature ventricular stimulation during ventricular pacing in cases 1, 8, 10 and 13.

In case 5, rapid atrial and ventricular pacing precipitated short bursts of ventricular tachycardia that had a contour similar to those that the patient experienced spontaneously.

In the other three patients (cases 2, 4 and 14) rapid atrial pacing at cycle lengths of 300–350 msec, for 10–20 seconds, initiated sustained ventricular tachycardia with a QRS contour identical to the patient’s spontaneously occurring ventricular tachycardia (fig. 5). Premature atrial or rapid ventricular stimulation (case 14) and premature ventricular stimulation (case 4) also precipitated ventricular tachycardia, while rapid atrial stimulation (case 14), rapid ventricular stimulation (cases 2 and 14) and premature ventricular stimulation (case 4) terminated the ventricular tachycardia in these three patients.

Case 14 was restudied while taking oral propranolol, 40 mg four times daily. Propranolol prevented atrial induction (or termination) of the ventricular tachycardia, because the shortest interval between consecutive ventricular complexes that conduct from the atrium was only 410 msec. However, rapid ventricular pacing still induced (and terminated) the ventricular tachycardia (table 4, fig. 5).

In all patients, the measured electrophysiologic parameters were normal.

**Autopsy Results**

Patients 5, 7 and 9 died and underwent autopsy examinations.

The heart of patient 5 weighed 650 g, and had uniform enlargement and myocardial thickening. The left ventricular myocardium was 2 cm thick. The valves and coronary arteries were normal. The aorta was narrow, 6.5 cm in circumference, compared with the pulmonary artery, which was 9.5 cm in circumference.

The heart of patient 7 demonstrated four-chamber dilation and weighed 400 g. All four valves and the coronary arteries were normal. Fibrosis was present basally in the ventricular septum, near the AV node.

The heart of patient 9 weighed 385 g, and was covered with a thick, adherent fibrous pericardium. Sections of the myocardium showed foci of fibrosis, and dense fibrous tissue with chronic inflammation along the epicardial surface. The right ventricular wall was 5 mm thick and the left ventricular wall was 9–11 mm thick. All four valves were normal. The ascending aorta showed minimal scarring and wrinkling. The aorta, great vessels and coronary arteries had foci of reduplication of the internal elastic membrane associated with uneven fibrous thickening of the media. At the level of L4 there was almost total occlusion of the aorta. An exhaustive search for organisms with multiple sections, stains and cultures was negative. She was felt to have carditis and arteritis of unknown etiology.

**Discussion**

**Nature and Natural History**

This study provides information on the nature and natural history of ventricular tachyarrhythmias in a
group of young patients. From our data, as well as from reports in the literature,\textsuperscript{5, 10-12} it is clear that ventricular tachycardia may occur in young patients without clinical evidence of organic heart disease. Its presence does not always imply increased risk of syncope or sudden death. In our patients, the important factor influencing the natural history appeared to be the nature and severity of the underlying heart disease.\textsuperscript{22, 23} Those patients who had ventricular fibrillation, those who died, and those who continue to be symptomatic with poorly controlled ventricular arrhythmias all have had significant organic heart disease. Only patient 10 developed ventricular fibrillation without clinically recognizable organic heart disease. However, she has the prolonged QT syndrome. Although sudden death may occur in young people who have ventricular arrhythmias with apparently normal hearts,\textsuperscript{11, 24} patients without clinically recognized organic heart disease in our study in general have done well, with and without therapy.
Other parameters did not appear to be useful as predictors of outcome. For example, response during the electrophysiologic study in our small group of patients did not define a high-risk population. The three patients in whom we initiated sustained ventricular tachycardia are all doing well on drug therapy. Three of four patients who had a repetitive ventricular response had no ventricular ectopy during the latest 24-hour recording, while the fourth patient had 27 premature ventricular complexes. Nor were the results of stress testing helpful prognostically. Exercise precipitated the ventricular arrhythmia or increased the frequency of premature ventricular complexes in five of 12 patients, suppressed the ventricular tachycardia in three, and did not elicit ventricular ectopy in four patients. The response of the arrhythmia to stress testing did not appear to be related to the nature of the heart disease, the severity of the ventricular tachyarrhythmia or the symptoms it produced.

The rate of the ventricular tachycardia influenced the nature of the patients’ symptoms, but slower rates did not necessarily assure a good prognosis. Two of the three patients who died had ventricular tachycardia with rates of only 154 and 158 beats/min. Some patients may have ventricular tachycardia at multiple rates, as exemplified by case 17, and it is possible that we did not sample all the different rates each patient may have had. Case 12 had ventricular rates of 250 beats/min (fig. 1), but recovered. The clinical course in this patient emphasizes the fact that life-threatening ventricular arrhythmias may disappear in young people, at times for inexplicable reasons. A transient affliction of the myocardium, not clinically recognized, must certainly be suspected.

Although all three deaths occurred in patients with ventricular tachycardia of left bundle branch block contour, this finding cannot be considered specific. Seven patients who also had ventricular tachycardia with a left bundle branch block contour are still alive.

Premature ventricular complexes may occur in normal children. Their presence in patient 7 5 years before her presentation with ventricular fibrillation may have dated the onset of her cardiac disease, but in the absence of any other findings at that time, the premature ventricular complexes had no prognostic value.

**Clinical Electrophysiology**

Programmed cardiac stimulation initiated sustained ventricular tachycardia in three patients, and nonsustained ventricular tachycardia in one patient. The four patients in whom programmed stimulation failed to initiate ventricular tachycardia had a variety of clinical disorders, including mitral valve prolapse, myocarditis, long QT syndrome and idiopathic hypokalemia. Patient 8 had short runs of spontaneous ventricular tachycardia that could be captured by pacing, yet had no ventricular tachycardia inducible by programmed stimulation. This success rate in precipitating ventricular tachycardia, low compared with some series in adults, may relate to differences in stimulation protocol, the age of the patients or, more likely, the etiology of the heart disease. The majority of the patients in the adult studies have had coronary artery disease, while none of our patients had this disorder. It appears easier to induce sustained ventricular tachycardia in patients with coronary artery disease, particularly if they have a left ventricular aneurysm, than in patients with other cardiac diagnoses.

Rapid and/or premature right atrial stimulation in three patients, and rapid and/or premature right ventricular stimulation in two of these three patients, induced sustained ventricular tachycardia. Patient 14 (fig. 5), restudied while taking oral therapy with propranolol, illustrated how a test of drug efficacy by an electrophysiologic study can lead to an erroneous therapeutic conclusion. Clinically in this patient, sinus tachycardia during exertion initiated the ventricular tachycardia, an occurrence duplicated by stress testing (fig. 4). Propranolol prevented the inciting event, i.e., the sinus tachycardia, from occurring clinically, and therefore prevented a spontaneous recurrence of the ventricular tachycardia. Although propranolol also prevented atrial stimulation from achieving a sufficiently rapid ventricular rate to start the ventricular tachycardia during the second study, rapid ventricular stimulation easily induced the sustained ventricular tachycardia, as in the first study. This observation separates two potentially unrelated events required to develop a sustained tachycardia spontaneously: a mechanism necessary for initiation and a mechanism necessary for maintenance of the tachycardia. In most instances, electrophysiologic studies test the capability of the drug to prevent sustainment of a tachycardia, since programmed stimulation supplies the initiating event. It is likely that some drugs, deemed therapeutic failures on the basis of an electrophysiologic study, nevertheless could be effective clinically by preventing the inciting event, such as a premature complex or sinus tachycardia, without altering the patient’s capability of sustaining the tachycardia, once initiated.

**Acknowledgment**

The authors thank Donald Girod, M.D. and Roger Hurwitz, M.D., Division of Pediatric Cardiology, and Winston Gaum, M.D. for help during this study, William C. Roberts, M.D. of the National Institutes of Health for evaluating the heart of patient 7, and Kathy Holmes for secretarial assistance.

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D H Pedersen, D P Zipes, P R Foster and P J Troup

Circulation. 1979;60:988-997
doi: 10.1161/01.CIR.60.5.988
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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