Prognosis in Primary Ventricular Tachycardia in the Pediatric Patient

DAVID M. BERGDahl, M.D., J. GEOFFREY STEVENSON, M.D., ISAMU KAWABORI, M.D., AND WARREN G. GUNThEROTH, M.D.

SUMMARY Five male pediatric patients with primary ventricular tachycardia are described. Although three were initially in congestive heart failure due to the tachycardia and were extremely difficult to manage, all have completely recovered, are not taking medication, and are free of arrhythmia. Three of the patients required long-term management with quinidine, with a therapeutic goal of controlling the heart rate rather than abolishing the arrhythmia. No growth disturbances were found in those three patients. A review of reported cases revealed 71 infants and children with ventricular tachycardia not associated with heart disease or systemic disorders; only four deaths were reported (5.6%). In the primary form of ventricular tachycardia in children, complete pharmacologic suppression may not be achieved without seriously endangering the normal electrophysiologic functions. Controlling the rate to an asymptomatic level with pharmacologic means is safer for a problem that may be self-limited.

PRIMARY ventricular tachycardia (VT) is an uncommon cardiac arrhythmia in childhood, and the choice of treatment may be influenced by experience with VT in the adult. In adults, this arrhythmia is life-threatening because of rapid deterioration to ventricular fibrillation, and emergency steps to convert to sinus rhythm are justified. Similarly, VT in children with profound hypoxia, or severe congenital heart disease may be a terminal event and cardioversion is urgent, although not necessarily effective. Primary or idiopathic VT in children, by contrast, is less grave; the dangers of an unplanned assault with the goal of complete suppression of the ectopic mechanism may prove more dangerous than the arrhythmia. In this report we present our experience with primary VT in infants and children to clarify the prognosis of this arrhythmia when managed conservatively.

Case Reports (Table 1)

Case 1

This case has been reported in the psychiatric literature as psychophysiological induced VT.1 This previously healthy 11-year-old male was hospitalized elsewhere in September 1963 with VT (rate 170 beats/min) and signs of congestive heart failure, but with normal blood pressure. The VT persisted despite treatment with digitalis and diuretics. DC conversion was successful, but the VT recurred rapidly, and quinidine was started. Sinus rhythm was attained initially, but over the next 6 months he was in and out of VT. A psychiatrist felt that these episodes were caused by emotional stress, although a cardiologist thought that the emotional stress was secondary to the episodes of VT. In October 1965 the episodes of VT became more frequent and the patient complained of fatigue and pallor. He was admitted to the University of Washington Hospital for cardiac catheterization and spontaneously converted to sinus rhythm during the procedure. The catheterization results showed no evidence of heart disease except for the arrhythmia. The next day he reverted to VT and was again treated with quinidine. A 6-hour Holter ECG was done, and was interpreted by the psychiatrist as VT due to emotional stress. One of us reviewed the tape and concluded that the VT was primarily exercise-induced. The patient received psychotherapy, and was maintained on a dose of quinidine sufficient to keep his overall heart rate under 130 beats/min. He decided on his own to stop taking quinidine at the age of 17 years, and for the past 10 years has been asymptomatic, doing moderate manual labor, and reports being free of VT. When examined in August 1979, he was completely normal to auscultation and had normal sinus rhythm on ECG.

Case 2

This previously healthy 11-month-old male was admitted to another hospital in March 1968 because of vomiting and prostration. He had a tachycardia of 250 beats/min, initially diagnosed as paroxysmal atrial tachycardia with Wolff-Parkinson-White syndrome. Because he was in heart failure, he was given digitalis. Within 24 hours the patient had two episodes of ventricular fibrillation (VF), which required cardioversion, drug therapy and resuscitation. Lidocaine, quinidine and dilantin were tried, and he was discharged in sinus rhythm. Over the next 4 months he was hospitalized three times with recurrences. During the second hospitalization, only quinidine and dilantin were needed to convert his VT to sinus rhythm. Two weeks later, in the hospital, propranolol and quinidine with dilantin were tried, with only transient results. A consulting cardiologist from outside the area made a telephone interpretation of supraventricular tachycardia, and recommended digitalization. Again, the
patient went into VF and required resuscitation. The patient was then transferred to our care. The arrhythmia was accepted, without further attempts to abolish it, because control of the overall rate allowed the patient to be asymptomatic. He was discharged on quinidine with an overall rate of approximately 150 beats/min. During a fourth hospitalization at the other hospital, his hospital chart could not be located, and the resident physician gave him digitalis, which again resulted in VF. After resuscitation, he was put back on quinidine and he improved. Over the next 3 years, he was maintained carefully on quinidine with only intermittent VT of 150 beats/min. He was free of any arrhythmia for the last 6 months on quinidine, and the medication was discontinued in 1971. He has been asymptomatic and free of tachyarrhythmias since that time, enjoying robust health. He was last seen in May 1979.

Case 3

This 19-month-old male, with a poor social situation and unremarkable medical history, presented in July 1969 with increased "nervousness," and a decrease in activity, appetite and weight gain over the previous 4 months. He was admitted with VT (rate 200 beats/min). He converted to sinus rhythm after quinidine was started. A noninvasive cardiovascular work-up showed no other abnormalities. He was discharged on a quinidine suspension. Two months later he was admitted with uncontrolled VT due to gastrointestinal intolerance of the quinidine suspension that was due to the vehicle (sorbitol) and not the quinidine. Cardiac catheterization with angiography revealed only mild, generalized chamber enlargement. Over the next 3 months, increasing dosages of quinidine were required to suppress the VT rate. He was then hospitalized for a trial of propranolol, but he spontaneously converted to sinus rhythm before the medication was given. He was discharged with no medication, and for 3 months he had only occasional, brief runs of VT. In April 1970, he was readmitted with VT and "pneumonia." The respiratory problem cleared on quinidine, ampicillin and a diuretic, and the VT converted to sinus rhythm. He was maintained on quinidine for the next 8 months. With no recent evidence of premature ventricular complexes (PVCs), quinidine was discontinued. He has subsequently been asymptomatic and free of VT, and was last seen in July 1979.

Case 4

This male infant was admitted to the neonatal intensive care unit in October 1977. He was premature (weight less than 2000 g) and in respiratory distress. On the sixth day of life he was noted to have dysrhythmia, three ectopic ventricular beats followed by three normal sinus beats. A noninvasive cardiac workup revealed no evidence of structural abnormality, and the child was clinically asymptomatic. There was a family history of asymptomatic dysrhythmias in an uncle and the uncle's children. Because this infant was asymptomatic, no treatment was started, and the PVCs diminished. However, he had one episode of VT at 3 weeks (rate 180 beats/min) for approximately 15 seconds. A rhythm strip taken before discharge at 6 weeks revealed no PVCs. After 20 months of follow-up and no medication this infant has been asymptomatic and free of VT.

Case 5

This full-term male infant was born on December 21, 1978 with an irregular heart beat; feeding problems were soon noted. The ECG revealed occasional PVCs, but there was no evidence of intrinsic cardiac disease. We elected to observe the infant without treatment. On the third day of life the PVCs abruptly worsened with an episode of VT (rate 194 beats/min) that lasted 13 minutes. With the administration of i.v. lidocaine, the rhythm improved to bigeminy and the PVC frequency gradually decreased. The child was then started on oral quinidine, and the lidocaine was gradually discontinued. By discharge on the seventh day of life he had no PVCs. The infant was sent home on quinidine, which was stopped by the infant's mother 1 week later. On well-child examination, no PVCs have been observed, and the child has thrived. He was last seen in May 1979.

Discussion

The diagnoses in our five cases of ventricular tachycardia were based on the criteria of Katz and Pick: atrioventricular dissociation, bizarre QRS complexes with divergent T-wave vector, and fusion beats at the onset and termination of the arrhythmia. In the first two patients, intracardiac recordings were made. However, all five patients went in and out of the arrhythmia, providing numerous opportunities to document the ventricular origin of the tachycardia (fig. 1).

Although three of our patients were in congestive heart failure due to a high rate early in their course, they were subsequently shown to be free of heart disease. On this basis, we diagnosed primary VT, a distinction critical to the prognosis of an individual case. The good long-term prognosis should condition the acute and long-term management of the patient toward minimal risk. DC cardioversion was used in the second case with ventricular fibrillation, and electrocally in the first case, but the rapid recurrence of VT in each instance suggests that this unpleasant treatment has little benefit when applied to a patient who is not hypotensive.

After observing the stable, compensated state in the first two patients with chronic VT, our therapeutic goal became control of the overall heart rate to levels compatible with a compensated state. In general, this was a heart rate of less than 150 beats/min in infants and 130 beats/min in the older children. This goal permits the use of a moderate, noncumulative schedule of quinidine. We use a dosage range of 5-15 mg/kg at 6-
hour intervals. This management was successful in all patients. Of particular importance in the pediatric population, growth and development were completely normal with maintenance of the quinidine for as long as 6 years. Because there are no commercial syrups of quinidine, the suspension for the younger pediatric patients must be made up by the pharmacist. We found that cherry syrup was acceptable to infants and children, with pulverized quinidine sulfate tablets. Although we found no proof of deterioration, we prepared only a month’s supply at a time.

Comparing the benign outcome of our cases with those reported in the literature is complicated by the varied therapy in some of the early reports and by the failure to deal with primary VT as a separate entity from VT occurring with heart disease, as well as with hypoxia, acidosis and electrolyte imbalance. Some of the reported cases were not autopsied and some had persistent marked cardiomegaly, suggesting primary myocardial disease as an underlying cause of death, if not of the arrhythmia. Table 2 lists patients who appear to have reasonably well-documented primary forms of VT and any associated mortality. Seventy-one cases have been reported in the literature, most of them as single cases, with an overall mortality rate of 5.6%. We cannot agree with the assertion that “because of this high incidence of morbidity and death in children with paroxysmal ventricular tachycardia” that “immediate termination of the acute arrhythmia in the symptomatic patient and long-term prevention of recurrences are vital.”

Rate reduction is a safer goal, and is more humane than repeated DC cardioversions. That is not to deny that the situation in a given patient may be extremely grave and may require aggressive treatment, including electrical cardioversion, if the child is hypotensive. Assuredly, all VT should be carefully monitored until the condition stabilizes. Scrupulous attention to oxygenation is mandatory, and to the treatment of acidosis, if present. Intravenous lidocaine is indicated for rates.

### Table 1. Summary of Case Reports

<table>
<thead>
<tr>
<th>Pt</th>
<th>Sex</th>
<th>Age at onset</th>
<th>Age resolved</th>
<th>Therapy tried</th>
<th>Therapy successful</th>
<th>Recurrences</th>
<th>Follow-up</th>
<th>Comments</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>11 years</td>
<td>17 years?</td>
<td>Laxin</td>
<td>Quinidine</td>
<td>None after 6 years</td>
<td>15 years</td>
<td>Date patient was free from arrhythmias unknown</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Quinidine</td>
<td>Psycotherapy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>11 months</td>
<td>4 years</td>
<td>Digoxin</td>
<td>Quinidine</td>
<td>Intermittent VT until age 4 years</td>
<td>11 years</td>
<td>Digitalization led to VF</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Lidocaine</td>
<td>Quinidine</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Quinidine</td>
<td>and Dilantin</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Propranolol</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>19 months</td>
<td>4 years</td>
<td>Quinidine</td>
<td>Procaineamide</td>
<td>None</td>
<td>10 years</td>
<td>3 months off medications at age 2 years with only occasional PVCs</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>6 days</td>
<td>6 weeks</td>
<td>None</td>
<td></td>
<td>None</td>
<td>21 months</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>Birth</td>
<td>2 weeks</td>
<td>Lidocaine</td>
<td>Quinidine</td>
<td>None</td>
<td>6 months</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Quinidine</td>
<td></td>
<td></td>
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</table>

Abbreviation: VT = ventricular tachycardia.

**Figure 1.** Three limb leads recorded consecutively from patient 3, at 2 years of age, illustrating atrioventricular dissociation, bizarre QRS complexes with altered T-vector and fusion beats, characteristic of ventricular tachycardia.3
greater than 150 beats/min, particularly if the infant or child is in congestive failure. Oral medication should be started as soon as possible, using procainamide or quinidine (which is our choice), and a gradual reduction in the lidocaine will be possible.

When the patient stabilizes, a thorough cardiovascular examination is mandatory to rule out myocardial disease or congenital heart disease and the rare instance of a cardiac tumor. We had one patient with a fibroma at the apex of the heart who presented with VT. Before establishing the cause of the tachycardia in this patient, quinidine was successful in maintaining the patient in an asymptomatic condition by controlling the overall heart rate. He subsequently deteriorated despite the quinidine, but surgical removal of the tumor restored normal rhythm, with no recurrence. (Two similar cases were reported that same year, with identical results.) A seventh patient, when first seen at 4 years of age with a mild coarctation of the aorta, had only rare PVCs. Five years later, on routine follow-up examination, he was found to have short bursts of VT. Although he was asymptomatic, we admitted him for monitoring and cardiac catheterization. Catheterization confirmed the mildness of the coarctation. Quinidine was started, and no further episodes of VT were observed. He was maintained on quinidine at home for 4 months. The quinidine was stopped after no PVCs were observed on exercise testing with a heart rate of 160 beats/min. He has been followed for 4 years with no recurrence of either VT or PVCs. We have not included him in the primary VT category because of the presence of heart disease, although it seems unlikely that the VT was related to his heart disease. Nevertheless, quinidine was effective in this case, as in the primary cases.

The infant or child with primary VT appears to have a functional disorder as the basis of his arrhythmia, although the disorder may cause congestive failure and, if untreated, death. In these patients the complete suppression of the arrhythmia may require a drug level that will also dangerously suppress the normal pacemaker tissues. This emphasizes the need for conservative management in these patients, i.e., controlling the heart rate to an asymptomatic level. Complete abolition of the arrhythmia may produce only transient benefits at greater risk.

References


20. Pedersen DH, Zipes DP, Foster PR, Troup RJ: Ventricular

<table>
<thead>
<tr>
<th>Year of report</th>
<th>Number of cases of primary VT</th>
<th>Deaths</th>
</tr>
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<tbody>
<tr>
<td>1942</td>
<td>8</td>
<td>1</td>
</tr>
<tr>
<td>1947</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>1949</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>1958</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>1960</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>1962</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>1964</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>1965</td>
<td>8</td>
<td>3</td>
</tr>
<tr>
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<td>1</td>
<td>0</td>
</tr>
<tr>
<td>1970</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>1971</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>1972</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>1973</td>
<td>17</td>
<td>0</td>
</tr>
<tr>
<td>1974</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>1975</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>1976</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>1980 (present study)</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>71</td>
<td>4 (5.6%)</td>
</tr>
</tbody>
</table>

Table 2. List of Reported Cases of Primary Ventricular Tachycardia and Associated Mortality
tachycardia and ventricular fibrillation in a young population. Circulation 60: 988, 1979

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Paroxysmal Hypotension Associated with Sympathetic Withdrawal

A New Disorder of Autonomic Vasomotor Regulation

R. Sanders Williams, M.D., and Thomas M. Bashore, M.D.

SUMMARY We evaluated a patient who had transient episodes of hypotension with clinical and laboratory features apparently distinct from previously recognized disorders of vasomotor regulation. In between his abrupt attacks of hypotension, the patient is asymptomatic and demonstrates normal autonomic modulation of heart rate and blood pressure in response to changes in body position, Valsalva maneuver, cold, and exercise. During periods of hypotension, his plasma norepinephrine falls markedly and he has blunted or absent responses to stimuli that normally have a pressor effect due to sympathetic efferent discharge. Mechanical or known hormonal disorders that produce episodic hypotension have been excluded by extensive testing. We suggest two possible causes for our patient's paroxysmal sympathetic withdrawal: first, a centrally mediated inhibition of sympathetic discharge to peripheral resistance and capacitance vessels, but with no afferent stimulus reflexly producing sympathetic withdrawal readily evident; or second, an episodic release of an unknown endogenous compound with inhibitory effects upon central or preganglionic sympathetic neurons or upon postganglionic sympathetic neurons by a presynaptic inhibition of norepinephrine release.

HYPOTENSION associated with abnormal autonomic modulation of vasomotor function is a prominent feature of several neurologic disorders1-11 (table 1A). Other disorders may also lead to paroxysmal hypotension by one of three major mechanisms: episodic or variable vascular obstruction,18-22 abnormal activation of vasodepressor reflexes,23-26 or abnormal episodic release of endogenous vasoactive substances27-32 (table 1B).

We recently evaluated a patient with paroxysmal hypotension in whom extensive laboratory investigation revealed features that appear to be distinct from any recognized disorders of vasomotor regulation.

Case Report

A 50-year-old Caucasian man was referred to our Cardiovascular Laboratory for the evaluation of frequent episodes of presyncope associated with hypotension. His spells of lightheadedness occurred one to three times daily and lasted 1 minute to 4 hours. He had to remain supine to avoid syncope. The onset of the spells was not related to body position. There were no premonitory symptoms, no apparent periodicity, and no evident precipitating events (such as exertion, meals or emotional stress) associated with the episodes. He occasionally had bitemporal headaches during or after a period of hypotension, but had no other associated symptoms. He had no flushing of the skin. In between the episodes he was asymptomatic, with no orthostatic lightheadedness or abnormalities of sweating or micturition.

He first noted similar spells in 1971, though from that time until late 1978 they had been less frequent (one or two per month) and were shorter (5-60 minutes) than the more recent spells. Previous therapeutic trials of ephedrine, atropine and fludrocortisone acetate had been ineffective in preventing or moderating his symptoms.

His medical history included numerous episodes of superficial and deep thrombophlebitis of the lower extremities, dating from a deep-vein thrombosis during hospitalization for an appendiceal abscess at age 12 years, and including several vein-stripping procedures. In 1971 he suffered an angiographically documented pulmonary embolus despite systemic anticoagulation, and underwent percutaneous insertion of an umbrella filter device into his inferior vena cava. He also had a history of abuse of alcohol and minor tranquilizers and reported several hospital admissions for psy-

From the Division of Cardiology, Duke University Medical Center, and the Veterans Administration Medical Center, Durham, North Carolina.

Address for correspondence: R. Sanders Williams, M.D., P.O. Box 3945, Duke University Medical Center, Durham, North Carolina 27710.

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