Asymptomatic Idiopathic Syndrome of Prolonged Q-T Interval in a 45-Year-Old Woman*

Recurrent ventricular tachyarrhythmias required multiple resuscitative efforts with electrical countershock in a 45-year-old woman with previously undiagnosed asymptomatic congenital prolongation of the Q-T interval. This patient represents the oldest person with symptoms relating to idiopathic prolongation of the Q-T interval found in the literature. Exacerbating factors, including diuretic-induced hypokalemia and the concomitant administration of perphenazine, were present. In such cases, initial refractory to therapy with antiarrhythmic agents, insertion of a transvenous pacemaker with overdrive suppression of the ventricular tachyarrhythmias may be lifesaving, allowing for the institution of therapy with agents that can selectively shorten the Q-T interval.

Selected Reports

VENTRICULAR TACHYARRHYTHMIAS PRECIPITATED
BY HYPOKALEMIA AND THERAPY WITH AMITRIPTYLINE AND PERPHENAZINE

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CASE REPORT

A 45-year-old white woman was admitted to the Mount Sinai Hospital after a single episode of syncopal episodes and recurrent ventricular tachyarrhythmias necessitated multiple cardiac resuscitative efforts.

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mately two minutes' duration. During the week prior to admission, she had had several episodes of light-headedness, palpitations, diaphoresis, and vomiting. Current medications included tablets containing 2 mg of perphenazine and 25 mg of amitriptyline (Triavil) multivitamin preparations, and furosemide (40 mg/week). The last dose of furosemide was taken on the day prior to admission. The patient was also on a weight-reduction diet that was low in potassium-containing foods. There was no previous history of neurologic or cardiac disease, although an electrocardiogram taken five years prior to admission revealed a normal sinus rhythm at 60 beats per minute (Fig 1), with a Q-T interval of 0.48 second. At that time the patient was not taking any medications and had normal serum levels of electrolytes. The family history and past medical history of the patient were noncontributory. Electrocardiograms from other family members were unavailable.

Physical examination revealed a blood pressure of 120/90 mm Hg, pulse rate of 84 beats per minute, and regular, temperature of 37°C (98.6°F), and a respiration rate of 20/min. A grade 2/6 midystolic murmur was heard at the left sternal border in the third intercostal space, radiating to the apex and aortic area. Findings from the remainder of the physical examination were normal. On admission, laboratory data, including levels of cardiac enzymes, were normal, except for a serum potassium level of 3.2 mEq/L, a serum uric acid level of 8.0 mg/100 ml, and a hemoglobin level of 10.0 gm/100 ml. The ECG on admission (Fig 2) showed a normal sinus rhythm of 88 beats per minute, with frequent ventricular extrasystoles. The Q-T interval was prolonged and measured 0.56 second, with prominent U waves. The extrasystoles were not suppressed with either infusion of lidocaine or therapy with procaine amide hydrochloride, both accompanied by intravenous administration of potassium chloride. Despite this treatment, although no further lengthening of the Q-T interval occurred, the patient's rhythm progressed to ventricular fibrillation with syncpe, which responded to electrical defibrillation.

During the next 12 hours the patient had approximately 40 to 50 episodes of reentrant ventricular tachyarrhythmias and syncpe unresponsive to antiarrhythmic therapy, each requiring electrical countershock (Fig 3). Serum levels of electrolytes, including potassium, were within normal limits after infusion of 160 mEq of potassium chloride. Sinus bradycardia, which ensued after each cardioversion, did not respond to intravenous administration of atropine.

In an attempt to provide enough electrical stability for insertion of a pacemaker, a bolus of 300 mg of bretylium tosylate was slowly administered intravenously, this transiently suppressed the arrhythmia. A bipolar transvenous pacemaker was then inserted into the apex of the right ventricle and set at a rate of 150 impulses per minute. At this rate, all ectopic activity was suppressed. Hypotension, with a systolic pressure of 70 mm Hg, ensued, and an intravenous drip infusion of norepinephrine and phentolamine was initiated. During this entire period the patient was alert when in sinus or pacemaker rhythm, even in the presence of hypotension.

During the next two days the pacemaker rate was unable to be decreased without the appearance of recurrent ventricular arrhythmias. By the third day of hospitalization, all intravenous medications had been discontinued, and normal sinus rhythm could be maintained. The Q-T interval remained prolonged, and oral therapy with diphenhydantoin (200 mg three times a day) was initiated. After 30 hours the Q-T interval had decreased to 0.42 second. An echocardiogram dis-
FIGURE 1. Electrocardiogram obtained on April 15, 1969, five years prior to admission. Note regular sinus rhythm of 60 beats per minute, with Q-T interval of 0.48 second.

FIGURE 2. Electrocardiogram taken on admission (Oct 29, 1974). Note sinus rhythm of 88 beats per minute, with frequent ventricular extrasystoles, Q-T interval of 0.56 second, and prominent U wave in leads 2 and V5 to V6. Lead V2 is missing.

closed no abnormality. An audiogram was normal.

The subsequent hospital course was complicated by the appearance of a pericardial friction rub on the fifth day of hospitalization, marked elevations in the serum level of creatine phosphokinase, and T-wave inversion noted in electrocardiographic leads V4 to V6. There were no subsequent episodes of premature ventricular systoles or syncope. While receiving therapy with diphenylhydantoin (Dilantin) sodium, the Q-T interval varied from 0.36 second to 0.42 second. The patient was discharged on the 16th day on a regimen of diphenylhydantoin sodium (200 mg twice a day). No other medications were prescribed. An ECG obtained 15 months after discharge revealed a persistently prolonged Q-T interval of 0.45 second. Simultaneously determined serum concentrations of potassium, magnesium, and calcium were normal. The serum level of diphenylhydantoin sodium was 12 mg/ml.

DISCUSSION

The etiology of the syndrome of a prolonged Q-T interval, associated with syncope, due to ventricular arrhythmias has never been adequately explained. These symptoms, when associated with deafness, are considered to be inherited as an autosomal recessive trait, while the genetic transmission in patients with normal hearing is considered to be autosomal dominant.

In adults a distinction between congenital prolongation of the Q-T interval and the acquired syndrome of a prolonged Q-T interval described by Motte et al8 must be made. This latter condition is characterized by spontaneous attacks of syncope and ventricular fibrillation in the absence of emotional or physical stress, associated with a specific cause of prolongation of the Q-T interval, such
as therapy with drugs, electrolyte disturbances, or defects in conduction. Although the patient described was in her fifth decade, the presence of a prolonged Q-T interval on a routine ECG taken five years prior to the current admission in the absence of hypokalemia or ingestion of medication, suggests the presence of congenital prolongation of the Q-T interval. The persistence of a prolonged Q-T interval, with documented normal levels of electrolytes in the absence of medication other than diphenylhydantoin, confirms the presence of congenital prolongation.

Although pathologic anatomic lesions of the conduction system and its vascular supply have been described in postmortem examinations, the pathogenesis of this syndrome remains to be clearly defined. The most current hypothesis postulates an imbalance of sympathetic tone to the anterior and posterior myocardium, with greater sympathetic activity on the left, rather than the right, side. Treatment with β-adrenergic blocking agents or left cervicothoracic sympathetic ganglionectomy has frequently been effective, with amelioration of symptoms.

Our patient represents an instance of asymptomatic prolonged Q-T interval of at least five years’ duration that was probably exacerbated by the presence of several extrinsic factors. The ingestion of amitriptyline may have been a contributory factor. Therapy with amitriptyline has been associated with ventricular tachyarrhythmia and with an increased incidence of sudden death in patients with cardiac disease. Perphenazine, the other ingredient of the combined drug preparation (Triavil), is a derivative of phenothiazine also known to prolong the Q-T interval.

The presence of hypokalemia probably resulted from a diet low in potassium, combined with chronic intermittent therapy with diuretic drugs. Prolongation of the Q-T interval with subsequent arrhythmias due to hypokalemia is likely to have been another initial exacerbating factor. Hypokalemia has been noted in association with one episode of ventricular fibrillation in a patient with familial prolongation of the Q-T interval.

Although treatment of the syndrome has only been variably successful, nontreatment is associated with a mortality of 73 percent. In an attempt to control the ventricular tachyarrhythmia, multiple agents have been used. Propranolol, advocated as an effective agent, was not administered initially in our patient because the diagnosis of primary prolongation of the Q-T interval was not entertained until her condition was stable. Therapy with digitalis has been found to shorten the Q-T interval in several instances, without effect on the frequency of syncopal episodes.

Administration of bretylium tosylate has been found to be of variable effectiveness. In our patient, therapy with bretylium transiently suppressed ventricular arrhythmias, allowing for placement of a pacemaker. The hypotension subsequently seen may be attributed in part to this drug. The pacemaker overdose was lifesaving. Successful suppression of reentrant ventricular tachyarrhythmias by a pacemaker in the syndrome of prolonged Q-T interval has been reported, although exacerbation of the arrhythmias with insertion of a pacemaker has also been described.

Therapy with diphenylhydantoin is also able to shorten the Q-T interval, as well as decrease synaptic transmission in the stellate ganglia, and has been used successfully in the treatment of a prolonged Q-T interval. In our patient, therapy with diphenylhydantoin was begun on the third day of hospitalization, in the face of continued prolongation of the Q-T interval after all ectopic activity had ceased. With administration of diphenylhydantoin, the Q-T interval de-
creased, there were no subsequent ventricular arrhythmias, and the patient was able to be discharged in good physical condition.

We believe that this case represents the oldest reported patient with primary prolongation of the Q-T interval who was totally asymptomatic until the appearance of exacerbating factors resulted in life-threatening arrhythmias. In view of the increasing frequency of case reports of this syndrome, it is suggested that a number of asymptomatic persons with prolonged Q-T intervals may well be at risk when provoked by extrinsic factors that tend to accentuate disturbances in repolarization. Early diagnosis of this disorder is, therefore, essential to prevent the impressive mortality associated with lack of therapy.

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Transvenous Pulmonary Embolectomy for Acute Massive Pulmonary Embolism

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Transvenous pulmonary embolectomy employing a vacuum-cupped directionally controlled catheter is a relatively new technique used in the management of major pulmonary embolism. We present the findings in a patient with acute massive pulmonary embolism who underwent transvenous pulmonary embolectomy, with immediate and marked improvement in hemodynamic function and survival. Insertion of a new intracaval filter at the same time provided protection against recurrent thromboembolism.

As if the diagnosis of pulmonary embolism did not present the clinician with enough difficulty, the subsequent management of the disorder, especially mas-

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sive pulmonary embolism, adds further confusion because of therapeutic methods which range from treatment with drugs alone to open pulmonary embolectomy with the patient on cardiopulmonary bypass.

This report concerns a patient seen shortly after the occurrence of massive pulmonary embolism and hemodynamic collapse, who failed to respond to administration of heparin and vasopressor drugs. The application of a new technique using a transvenous catheter for pulmonary embolectomy, followed by the insertion of a vena caval filter, resulted in immediate hemodynamic recovery and survival.

CASE REPORT

This 51-year-old man was admitted to a community hospital following an automobile accident which resulted in a fracture of the left tibial condyle and shaft. The patient was treated with a cast and was making an uneventful recovery until the morning of the fourth day of hospitalization, at which time he became nauseated and diaphoretic, with complaints of pain in his left calf. His blood pressure, which had been 154/100 mm Hg, fell to unobtainable levels, while his apical pulse rate increased from 50 to 130 beats per minute. The respiratory rate had increased from 18/min to 40/min, and the diagnosis of acute massive pulmonary embolism was made. Heparin was given intravenously at a dose of 15,000 units, and blood pressure was supported with administration of levarterenol (Levophed) bitartrate.

The patient was transferred to the Medical College of Virginia Hospital, where physical examination revealed that the patient was diaphoretic and confused. His blood pressure was 60/0 mm Hg, and his pulse rate was 128 beats per minute. The respiratory rate had increased from 18/min to 40/min, and the diagnosis of acute massive pulmonary embolism was made. Heparin was given intravenously at a dose of 15,000 units, and blood pressure was supported with administration of levarterenol (Levophed) bitartrate.

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