

# The Prolonged Q-T Syndrome Presenting as a Focal Neurological Lesion

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**A patient with a prolonged electrocardiographic Q-T interval suddenly developed left hemiplegia after an episode of ventricular fibrillation. A CT scan showed a cerebral infarction in the right internal capsule. The prolonged Q-T interval, by virtue of its association with cardiac arrhythmias, may cause cerebral hypoperfusion that commonly results in generalized neurological deficits. The lack of evidence for embolization in this patient suggests that decreased cerebral perfusion was responsible for the genesis of her neurological signs and symptoms. The prolonged Q-T interval is an easily overlooked cardiac abnormality that must be considered in a patient who exhibits a focal neurological deficit.**

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The prolonged Q-T interval of the electrocardiogram has been reviewed in numerous publications. This condition is characterized by ventricular arrhythmias, syncope, and sudden death, usually as an aftermath of physical exertion or emotional stress [3, 8, 9]. Three types of prolonged Q-T interval have been defined: the autosomal recessive Jervell-Lange-Nielsen syndrome associated with congenital deafness, the autosomal dominant Romano-Ward syndrome, and the idiopathic state [1, 2, 5, 6]. In addition, a prolonged Q-T interval has been noted as a consequence of drug therapies such as quinidine (Quinidex), procainamide (Pronestyl), and phenothiazines (chlorpromazine, Stelazine, Vesprin), as well as homeostatic alterations of hypocalcemia, hypokalemia, hypomagnesemia, hypothermia, cerebrovascular disease, arterioventricular block, and various operations involving the neck.

Although the etiology of the arrhythmias found in these syndromes is under debate, most authors agree that the neurological symptoms are the results of reduced cerebral

perfusion due to cardiac malfunction [4, 7, 8]. The neurological signs and symptoms found in these cases are usually generalized.

We are reporting a case of a young woman who was stuporous and developed focal neurological signs and symptoms after an episode of ventricular fibrillation. Evaluation revealed a prolonged Q-T interval on her electrocardiogram and an infarction in the right basal ganglia on computerized tomographic (CT) scan.

## Case Report

A 21-year-old white woman entered the hospital with a history of seizures and ventricular fibrillation. For two years she had experienced monthly syncopal episodes lasting a few seconds. These episodes occurred when she was fatigued or ill, never after alcohol consumption, intercourse, rigorous physical activity, or childbirth. Headaches, frequently associated with these syncopal episodes, occurred in the neck and radiated to the left frontal area.

The patient's family history included a high frequency of myocardial infarctions and strokes, plus the sudden death of a 19-year-old maternal aunt that was attributed to cardiac failure. The patient denied alcohol or drug abuse. She reported no edema, palpitations, paroxysmal nocturnal dyspnea, orthopnea, dyspnea on exertion, polyuria, nocturia, scotoma, nausea, viral illness, eclampsia, Raynaud's phenomenon, chronic infection, incontinence, loss of hearing, use of oral contraceptives, or recent trauma.

Two days prior to her admission, the patient had been feeling quite tired. She experienced lightheadedness, followed by loss of consciousness, symmetrical tonic-clonic movements of the limbs, eye rolling, and tongue biting; she was sleepy for an hour thereafter. The patient was taken to a local emergency room, where her physical examination was reported to be normal. Subsequently, several short syncopal episodes occurred on the day before admission.

On the day of admission, the patient awoke suddenly in the early morning and had several spells of lightheadedness while supine in bed. Later in the morning, she suddenly lost consciousness and had a cardiac arrest. The life squad reported that the patient's electrocardiogram showed ventricular fibrillation, which responded to the application of direct current countershock. At another hospital, the pa-

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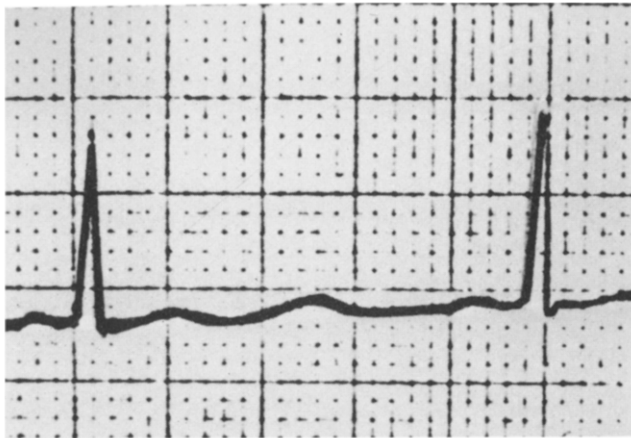


Fig. 1. Admission electrocardiogram, with a Q-T interval of 0.58 sec.

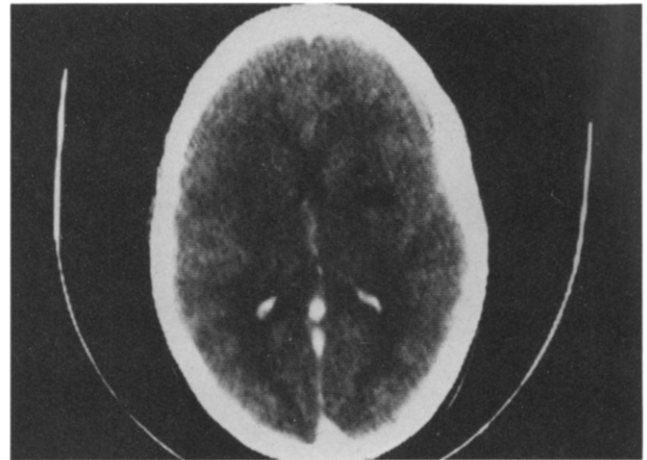


Fig. 2. CT scan demonstrating focal low density in the right basal ganglia.

tient's blood pressure was recorded as 40/0, and infusion of dopamine was begun. Altering levels of consciousness, variable pupillary reactions, limb flaccidity, and a left Babinski sign were found at that time.

On arrival at The Christ Hospital, the patient was stuporous. Her blood pressure was 100/62; heart rate, 100 and irregular; respirations, 16; and temperature, 38.2°C rectally. Examinations of the head, neck, chest, abdomen, and limbs were normal. Peripheral pulses were full. There was an S<sub>4</sub> gallop. The patient was oriented only to person; she could not follow commands, exhibited slow, dysarthric speech, and ignored her left side. There was left central-facial weakness, left hemianopia, decreased tonus of the left leg, left Babinski sign, and choreoathetoid movements of both arms. Deep tendon reflexes were normal, and there were no carotid bruits.

The hematocrit was 37 and the white blood count was found to be 22,500 with 82 polymorphonuclear leukocytes, 5 band forms, 10 lymphocytes, and 3 mononuclear cells. The blood glucose was 277 mg/dl, and the sedimentation rate was 5 mm. Cerebrospinal fluid pressure was 100 mm H<sub>2</sub>O; glucose, 153 mg/dl; protein, 21 mg/dl; and 5 white cells per cubic millimeter. Cerebrospinal fluid cultures were negative. The electrocardiogram showed premature ventricular contractions, supraventricular tachycardia, a rate of 155 per min., a QRS interval of 0.12 sec.; and a Q-T interval of 0.58 sec. (Fig. 1). A CT scan demonstrated low-density areas in the right internal capsule (Fig. 2).

The patient was placed in the intensive care unit, where her arrhythmias were vigorously treated with lidocaine and Pronestyl. Over a period of three days, her mental state and neurological examination returned to normal. A repeat electrocardiogram showed a continued prolongation of the Q-T interval. A cardiac stress test and an echocardiogram were normal. Therapy with propranolol was initiated, and by the time of discharge, the patient's Q-T interval was 0.39 sec. (Fig. 3).

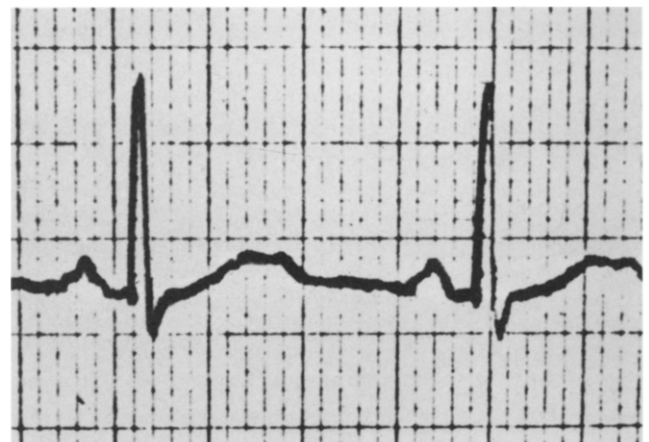


Fig. 3. Discharge electrocardiogram, with a Q-T interval of 0.39 sec.

### Discussion

The focal signs and symptoms exhibited by this patient resulted either from cerebral embolization or cerebral ischemia secondary to her cardiac arrhythmia. Vasculitis, tumor, and infection were eliminated as possible causes by the diagnostic studies performed.

The predisposition for arrhythmias found in patients with a prolonged Q-T interval suggests a cardiac origin for cerebral embolization. In this patient, the possibility of an embolus of cardiac origin is diminished by the absence of a mural thrombus or a prolapsed mitral valve on echocardiography. Embolization from an extracardiac source is doubtful, given the patient's age, absence of peripheral vascular disease, or carotid bruit. Therefore, embolization is probably not responsible for the patient's neurological deficits.

Cardiac arrhythmias may, if associated with severe, prolonged hypoperfusion, result in focal ischemic infarction.

There is ample evidence in this case for a significant period of hypotension associated with the patient's episode of ventricular fibrillation. Given the lack of evidence for other etiologies, focal infarction secondary to severe ischemia is the most plausible explanation for our patient's focal signs and symptoms.

Treatment modalities for the long Q-T syndrome include Dilantin (diphenylhydantoin), propranolol, and combined thoracic and stellate ganglion sympathectomy [5, 7]. Although none of these forms of treatment has been conclusively evaluated, propranolol is the most successful in diminishing the incidence of arrhythmias [8].

The patient has been treated with propranolol, 160 mg per day in four divided doses, and has had no further syncope episodes in three months. Headaches continue but respond to mixed analgesics. A repeat CT scan performed after three months showed continued evidence of infarction.

As this case illustrates, the prolonged Q-T interval is a cardiac abnormality that must be considered in establishing

the etiology for the sudden onset of a focal neurological deficit.

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## Letter to the Editor

### Abbreviations

May I applaud your complaint in a recent book review (*Surg Neurol* 15:273, 1981) about the use of initials as abbreviations for words. This habit is now so widespread that it makes certain types of medical literature almost unreadable, that on cerebral blood flow being a particularly conspicuous example. What presumably started as a reasonable method of shortening multiple polysyllabic chemical terms has now blossomed to a degree in which authors are too lazy to write out quite ordinary names, such as posterior communicating artery aneurysms or myasthenia gravis.

Since each author seems to regard himself or herself as having a free hand in the matter, it is necessary to familiarize oneself with the private language used before undertaking to read the article, or otherwise to refer constantly to the glossary at the beginning. The matter becomes more than merely irritating when a set of initials, e.g., *PICA*, which has gained some sort of general usage, is adopted by another author for something quite different, for instance, *proximal internal carotid artery*.

How far editors themselves may be responsible through reasons of economy in printing I do not know, but it is surely their duty to their readers to discourage an ugly, irritating, and ultimately obscurantist practice.

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### Reply

The Editors of *Surgical Neurology* make every effort to remove acronyms and other abbreviations from the papers that they publish. We agree with Mr. King and welcome his letter on this subject. The economies which may result from the use of these abbreviations is trivial. The amount of ink and paper required to spell out words instead of using abbreviations is inconsequential.

Paul C. Bucy, M.D., Editor