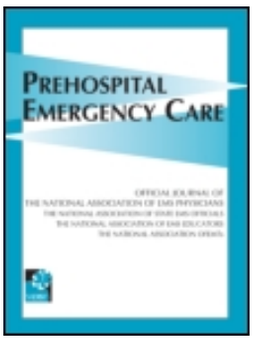

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A Near-fatal Case of Long QT Syndrome in a Teenaged Male

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Case Report

The patient was a 15-year-old male from Texas who was visiting his grandparents in Michigan for one week. The young man awoke one morning in his usual state of health, ate breakfast, spoke with his grandparents, and reportedly went back to bed. Approximately two hours later, the grandfather walked past his room and overheard abnormally loud, sonorous breathing. As he approached, he noted the patient had obviously experienced a loss of bladder and bowel function. He then witnessed a cessation of spontaneous breathing. The grandfather immediately instructed his wife to call 911. The patient's grandfather then performed two chest compressions followed by approximately 2 minutes of mouth-to-mouth ventilation without subsequent chest compressions.

According to the emergency medical services (EMS) run sheet, a call was received at 9:30 AM. The on-scene arrival time was 9:32 AM. On arrival the EMS personnel found a 15-year-old male in bed with a palpable pulse and spontaneous breathing. He was noted to be awake, but not appropriately responsive. His vital signs included a heart rate of 92 beats/min, blood pressure of 122/82 mm Hg, and respiratory rate of 16 breaths/min. Blood glucose and pulse oximetry were quickly confirmed without a need for intervention. Intravenous access was established and supplemental oxygen was provided by nasal cannula. Within 10 minutes the patient became more alert and began to respond appropriately to questions. Naloxone was not deemed necessary. He was then taken via stretcher to the ambulance. During transport the emergency medical technician–paramedic (EMT-P) monitoring the patient documented the patient to be awake, but tired. Astutely, the EMT-P also noted "an unusual appearance on the rhythm strip." The remainder of the transport was uneventful. The patient and the EMS run summary were then turned over to the emergency department physician.

In the emergency department the patient's only complaint was a mild temporal headache. He had no significant recollection of the events leading to the apparent respiratory arrest. The patient denied a previous history of seizures, syncope, or cardiac arrest. His medical problems included attention deficit disorder and intermittent asthma. He was on summer "holiday" from his Ritalin and used only his albuterol as needed. He denied any illicit drug use. The family history included a grandfather who died at 42 years of age of sudden death of unclear cause. The physical exam demonstrated a well-developed male without any significant abnormal physical findings. The lung sounds were clear and equal and the cardiac auscultation revealed a regular rate and rhythm without murmur. The neurological exam was completely intact without focal findings. Further workup included serum evaluation of electrolytes, renal function, glucose, magnesium, calcium, and comprehensive serum drug screen. Radiological evaluation included a chest x-ray, and computed tomography (CT) scan of the head. All the above were found to be normal. A 12-lead electrocardiogram (ECG) (Fig. 1) demonstrated a normal sinus rhythm, a QT interval of 528 milliseconds, and anteroseptal T-wave inversion. A repeat ECG showed a persistent prolonged QTc interval of 540 milliseconds. A cardiology consultation was obtained. With high probability it was felt the primary event was a malignant ventricular arrhythmia predisposed by a prolonged QT interval.

Discussion

Unexplained syncope is a common presentation of EMS patients. Often these patients do not have an identifiable cause of the event, especially if the patient is a child, an adolescent, or a young adult. As a result, the young often may be dismissed as having psychogenic or functional syncope. However, our understanding of syncope is changing as there is an ever-increasing body of literature on syncope and sudden cardiac death in the young.[1–4] This most likely is a reflection of improved rate of diagnosis rather than increasing incidence. Table 1 lists other etiologies of syncope and sudden cardiac death in the young.

The case of this young man is unusual in that it identifies a patient without a significant medical history who is ultimately treated as having a near-fatal dysrhythmia. His presentation is disturbing in that he was found unresponsive and incontinent, symptoms usually not found in typical low-acuity "syncopal" events. Additionally, the patient's grandfather witnessed a respiratory arrest. The patient received bystander cardiopulmonary resuscitation (CPR) by his grandfather, which may have been lifesaving or actually may have been unnecessary. There are reports of patients in the literature who are "CPR only survivors," with some debate whether these were events that indeed necessitated the provision of CPR.[5]

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Other than the history of the initial presentation and the brief period of confusion immediately following the event, there was little clinical evidence that provided a clear diagnosis.

The patient had no previous history of seizure disorder. He was of normal body habitus, making the suggestion of valvular disease or aortic dissection due to Marfan's syndrome or other congenital structural abnormalities unlikely. He was white and not particularly muscular so that asymmetric septal hypertrophy (ASH) was less likely. This event occurred at rest, which is also not consistent with ASH. A subsequent echocardiogram performed was normal, thus virtually excluding this as an etiology. There was no historical information, physical evidence, or subsequent testing to suggest this to be a toxin or drug-induced event.

Previous medical history of attention deficit disorder and asthma was similarly not helpful in establishing a diagnosis. The patient previously took methylphenidate (Ritalin), but he had been on "holiday" for more than a month from its use at the time of the event. Patients with asthma do have a demonstrable incidence of sudden death, but there was no history that the patient was symptomatic and he denied using his inhaler at the time of the event. However, in patients with long QT syndrome, asthma has been identified as an independent predictor of an increased risk of adverse cardiac events.[6] The family history obtained later in the patient's hospitalization identified a paternal grandfather who died suddenly at the age of 42. This may or may not have relevance to this case.

TABLE 1. Etiologies of Syncope and Sudden Cardiac Death in the Young Patient[1–4]

Hypertrophic cardiomyopathy
Acute myocarditis
Anomalous coronary arteries
Single coronary artery
Tunneled coronary arteries
Conduction system abnormalities
Wolff-Parkinson-White syndrome
Long QT syndrome
Arrhythmogenic right ventricular dysplasia
Congenital valvular disease
Mitral valve prolapse
Aortic stenosis
Coarctation of the aorta
Marfan's syndrome
Idiopathic left ventricular hypertrophy
Pulmonary vascular obstruction
Commotio cordis
Toxins
Metabolic disorders

After a careful history and physical exam, the sole initial clue to diagnosis was the paramedic's cogent observation of an "unusual appearance of the rhythm strip." The patient had an abnormal and considerably prolonged QT interval on both the rhythm strip and 12-lead ECG. In this case the QTc (corrected QT interval) of 540 milliseconds is a clear, although frequently overlooked abnormality. A QTc of this duration is in

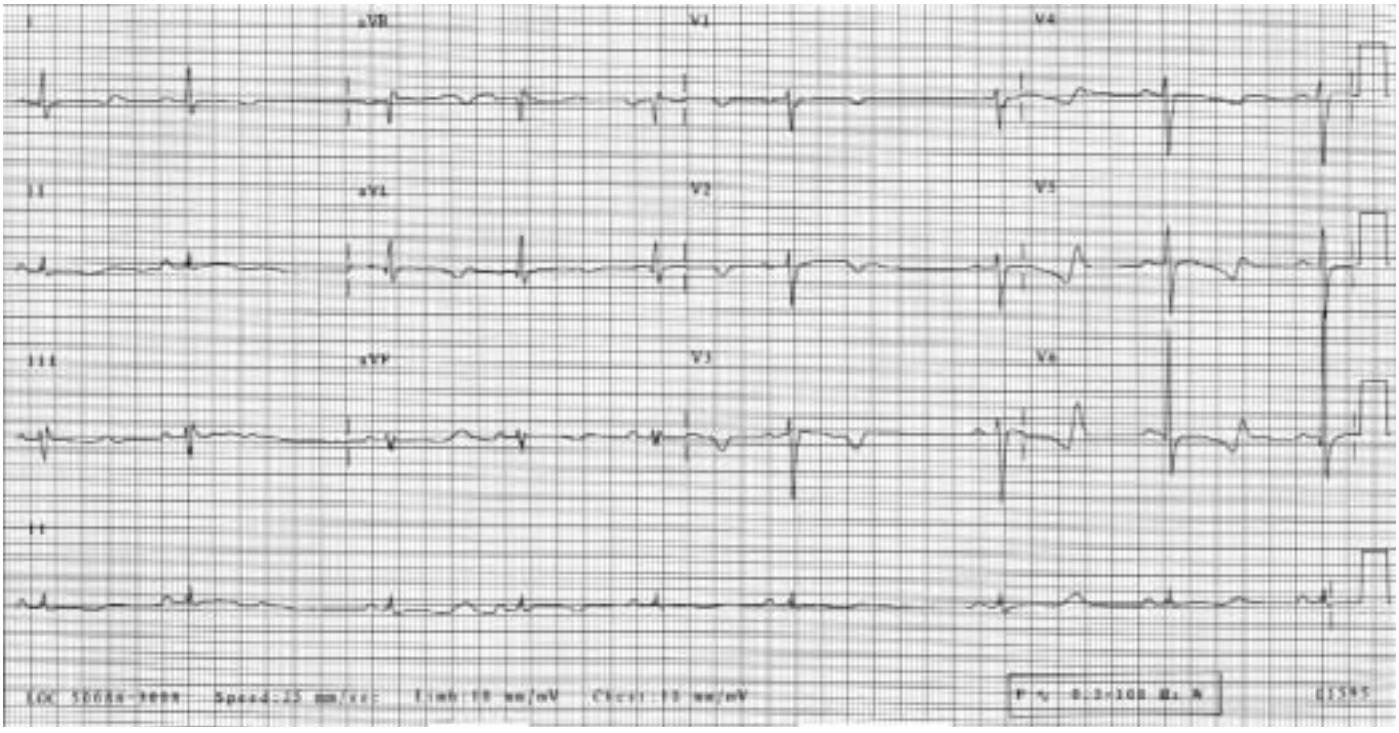

FIGURE 1. The updated ECG.

TABLE 2. Medications That Potentially Prolong the QT Interval[7,12,14]

Antibiotics
Erythromycin
Trimethoprim and sulfamethoxazole
Pentamidine
Antifungals
Fluconazole
Itraconazole
Ketoconazole
Antiarrhythmics
Quinidine
Procainamide
Disopyramide
Sotalol
Antidepressants
Amitriptyline
Phenothiazine derivatives
Rispiridone
Haloperidol
Pimozide
Asthma/anesthetics
Epinephrine
Antihistamines
Terfenedine
Astemizole
Gastrointestinal
Cisapride
Lipid-lowering
Probucol

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the 99th percentile for prolonged QT intervals. QT prolongation in the setting of a near-fatal dysrhythmia, syncopal episode, or seizure is now highly suggestive of long QT syndrome.

Abnormal repolarization of the ventricular myocardium, electrocardiographically represented by QT prolongation, predisposes the patient to torsades de pointes (polymorphic ventricular tachycardia in the setting of QT prolongation). Although torsades de pointes often terminates spontaneously, there is a significant risk of degenerating into ventricular fibrillation. Initial treatment of ventricular fibrillation consists of immediate defibrillation. Urgent treatment for torsades, or its recurrence, involves correcting underlying electrolyte abnormalities and recognizing reversible causes. Intravenous magnesium is considered first line in preventing recurrent episodes. Transvenous overdrive pacing or pharmacological intervention with isoproterenol can also be used to prevent QT interval lengthening associated with bradycardia.[7–9]

Recognition of the abnormal QT interval is the first step in preventing dysrhythmias. The general appearance of the QT interval being greater than one-half the R-R interval suggests a QT interval prolongation. Bazzet's formula (the measured QT interval divided by the square root of the R-R interval[7,10]) is used to account for the effect that heart rate has on the QT interval. In the symptomatic patient the QTc is typically greater than 440 milliseconds in males and 460 milliseconds in females. A normal QT interval, however, does not entirely exclude the diagnosis of long QT syndrome. It should also be noted that the QT interval varies with time of day, age, and sex.[10,11] Other ECG findings associated with QT prolongation include nonspecific T-wave abnormalities. T waves are typically inverted, biphasic, wide, or notched.[12] QT prolongation and nonspecific T-wave abnormalities are subtle but if missed can lead to grave medical repercussions.[13]

Long QT syndrome can be acquired or congenital. The differential diagnosis of the acquired form is broad, with the majority of cases thought to be secondary to a pharmacological source or metabolic derangement. Various antiarrhythmics, antibiotics (including antifungals), antihistamines, and antidepressants have been frequently implicated pharmacological agents associated with QT prolongation.[14] Table 2 lists medications thought to prolong the QT interval. This list is alarmingly large and ever growing. Common metabolic abnormalities leading to arrhythmia secondary to QT prolongation include hypokalemia, hypocalcemia, and hypomagnesemia. Obviously, any underlying medical condition that could precipitate a metabolic abnormality must be carefully considered. The patient in this case had a history of taking potentially cardioactive medications (methylphenidate and albuterol). These were not used, however, in temporal association with this event. The metabolic causes were proven absent with simple serum electrolyte analysis. Additionally, this patient did not have any underlying medical condition that would independently predispose him to a malignant arrhythmia. An acquired form of long QT syndrome was unlikely.

The two primary forms of inherited congenital long QT syndrome include the rare Jervell-Lange-Nielson syndrome and the much more common Romano-Ward syndrome. Jervell-Lange-Nielson syndrome is an autosomal recessive disorder characterized by neurosensory hearing loss and a pathologically prolonged QT interval.[15] The incidence is only 1 to 6 per million individuals.[12] Individuals typically present very early in life with multiple episodes of syncope, seizures, or cardiac arrest. Normal hearing excluded this diagnosis in our patient.

In contrast, the Romano-Ward syndrome is an autosomal dominant disorder with a reported incidence as high as 1 in 10,000 individuals. This roughly translates into 3,000 to 4,000 deaths per year in the United States.[16] This syndrome may be an underestimated

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cause of syncope and sudden cardiac death in the young. To date, six genotypes and multiple phenotypes have been recognized[8, 17–19] The information at the genetic and molecular level has grown dramatically during the last decade with increased understanding of the human genome and the availability of genetic testing. Fundamentally, arrhythmias occur as the result of a genetic abnormality of sodium and potassium ion channel function leading to aberrant repolarization that electrocardiographically manifests as prolongation of the QT interval. Two-thirds of gene carriers have some type of symptoms.[16] Symptoms include syncope, palpitations, seizures, or even sudden death. The most common is syncope. Patients having seizures as the initial presenting symptom of an aborted adverse cardiac event are also well described.[20–23] The typical age of initial presentation is preteen to teenage years. Events are frequently associated with emotional stress, physical stress, or noxious stimuli (such as a loud noise), although as in our case, occurrence at rest is not unusual. Screening ECGs of the parents, siblings, and offspring of patients with congenital QT prolongation are highly recommended.

Given the prior family history of sudden death and the absence of other causes for this patient's ECG abnormality, the evidence supported the diagnosis of either an inherited or a sporadic form of congenital long QT syndrome. Recognizing that there is not a correctable or reversible cause of a congenitally prolonged QT interval, treatment is mandatory as ten-year mortality rates of untreated patients may approach 50%.[9] Current long-term therapeutic options include beta-blocker therapy or cervical sympathectomy to avoid excessive sympathetic simulation, pacemaker implantation to prevent bradycardia and secondary lengthening of the QT interval, and implanted cardioverter–defibrillator (ICD) placement to abort malignant arrhythmias.[9] Individual treatment is largely based on the patient's estimated risk of future morbidity and mortality. Treatment in the asymptomatic or low-risk patient is controversial. In the high-risk patient, treatment is essential. Suppressing symptoms with beta-blocker therapy and directly aborting adverse cardiac events with an ICD are the most common treatments offered. Having what appeared to be an aborted cardiac arrest placed our patient at a high risk of repeated episodes. The patient was started on beta-blocker therapy and had an ICD implanted. At one year there had been no adverse events or recurrence of symptoms reported.

CONCLUSION

Prolonged QT syndrome has many known and likely a number of yet-to-be-discovered causes. This case exemplifies the need for a thorough evaluation of all patients with syncope. A significant event in a young child or adolescent should not be dismissed as trivial without serious consideration of all potential diagnoses. This case specifically demonstrates that a review of all prehospital information is essential. Any unusual circumstances, no matter the source or perceived insignificance, may be the only source of making a proper diagnosis. Final diagnosis in this case, and in most cases of long QT syndrome, was made based on the clinical information obtained on scene and on initial ECG.

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