Open Journal of
Clinical & Medical
Case Reports

Volume 3 (2017)
Issue 11

ISSN 2379-1039

First manifestation of a congenital problem in advanced old age: not all VF is ischemic!

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Abstract

We describe a patient with known congenital long QT syndrome (LQTS), who remained asymptomatic thorough adulthood and only manifested cardiac symptoms after the age of 88 years.

He presented to the emergency department with collapse with loss of consciousness, and went on to suffer two ventricular fibrillation (VF) arrests.

LQTS is generally thought of as a disease that manifests in childhood and early adulthood, and the data on patients who have survived beyond their 60s without an event is limited. This case demonstrates that older LQTS patients also need risk stratification and the challenges that comorbidities bring. It also demonstrates that congenital pathology should be included in the differential diagnosis even in elderly patients.

In this case it was thought that the LQTS was exacerbated by age related cardiac conduction system disease and bradycardia, and therefore he was treated by implantation of a permanent pacemaker.

Keywords

long QT syndrome; bradycardia; pacemaker

Abbreviations

LQTS: long QT syndrome ; VF: ventricular fibrillation ; TdP: Torsades-de-Pointes; ICD: implantable cardioverter defibrillator; QTc: corrected QT interval; ECG: 12-lead electrocardiogram

Introduction

The congenital long QT syndrome (LQTS) is a hereditary channelopathy of myocardial repolarization first described in 1975. More than 15 genetic types have been identified [1] however there are three main subtypes: long QT 1 (KCNQ1 mutation, characterized by broad based T waves and exertional symptoms), long QT 2 (KCNH2 mutation characterized by biphasic T waves and auditory triggers, and long QT 3 (SCN5A mutation with a long isoelectric segment on the ECG and resting symptoms) [2].

Long QT can also be acquired as a result of drug or electrolyte disturbance, but it is recognized that in some cases of acquired LQTS there is also an underlying genetic predisposition [3].

The prevalence of LQTS is around 1:2000 [4].

The characteristic ventricular tachyarrhythmia that underlies the cardiac events of LQTS is Torsades-de-Pointes (TdP), a generally self-limiting polymorphic ventricular tachycardia (VT) that produces transient syncope but that can also degenerate into ventricular fibrillation (VF) and cause cardiac arrest and sudden death [5-7].

It is generally thought that patients under the age of 40 are at highest risk, however the first study to report the clinical course of LQTS patients older than 40 years showed that LQTS subjects maintain a high risk for life-threatening cardiac events after this age [8].

Risk factors identified include female gender, QTc duration, and a history of syncope in the past 10 years in non-genotyped subjects, and the presence of the LQT3 genotype in genetically tested individuals. It has been suggested that patients with these risk factors should be considered for primary therapies such as beta-blockers and implantable cardioverter defibrillators (ICDs) even when identified after the age of 40 [8,9].

Case Presentation

We describe a case of a man with known congenital LQTS who remained asymptomatic thorough adulthood and only manifested cardiac symptoms after the age of 88 years. The genotype was not identified but his ECG and symptoms would be compatible with long QT 3.

This patient was known to have fainted approximately once a year between the ages of 7 and 14, but investigations done at the time had not identified any specific etiology.

His daughter, who is in her fifties, is also diagnosed with LQTS but she continues to be treated with beta-blockers alone having declined an ICD, despite two previous episodes requiring defibrillation (one related to swimming and the other with an emotional trigger, possibly exacerbated by erythromycin). His son was asymptomatic despite a borderline ECG and his parents were not known to have suffered from syncope.

The patient had previously suffered from falls, and been identified to be relatively bradycardic, however after visits to the falls clinic and to cardiology services, it was decided that his falls were not cardiac in origin and that a pacemaker was not warranted. He had a background of ischemic heart disease and of moderate Alzheimer's dementia, managed with memantine (which can cause bradycardia, but is not reported to prolong the QT interval). He had previously been on beta-blockers but these were stopped in 2011 because of bradycardia.

On the day of his presentation, he was sitting at the table eating his breakfast when he felt lightheaded and collapsed on the floor. After approximately 5 minutes he regained consciousness and his wife called the ambulance.

The patient was admitted to the Acute Frailty Unit where he suffered a VF followed by a Torsades-de-Pointes arrest each responsive to a DC shock (Figures 1 and 2).

The 12-lead electrocardiogram (ECG) post cardiac arrest showed atrial fibrillation with a rapid ventricular response of 127 bpm, left axis deviation, a QRS duration of 80 ms and a corrected QT interval of 482 ms (Figure 3). This was temporarily rate controlled with bisoprolol. The blood tests found no significant electrolyte or other abnormalities (K^+ 5.3). Chest radiograph showed a possible right lower

lobe consolidation, which was treated with co-amoxiclav.

After being transferred overnight to the Coronary Care Unit the patient was in sinus rhythm with a heart rate of 40 – 58 bpm and a corrected QT interval of 605 ms (Figure 4). The LQTS was initially unrecognized and the patient had a further VF arrest, successfully resuscitated with a single 150 J DC shock and treated with intravenous amiodarone and magnesium sulphate as per Advanced Life Support guidelines [10]. The subsequent ECG showed sinus bradycardia with a corrected QT interval of 720 ms (Figure 5). The beta-blocker was stopped as it was thought that the bradycardia was worsening the QT interval prolongation and therefore contributing to the VF arrests. The amiodarone was also stopped. The patient received a dual chamber PPM (Medtronic Sensia device with St Jude Medical Tendril leads) (measurements at the time of pacemaker implant show a corrected QT interval of more than 700 ms with macrovolt T wave alternans, Figure 6) and was afterwards treated with a beta-blocker. His pacemaker was set at a base rate of 80 beats per minute. He had no further arrests during this admission and was doing very well at discharge.

Discussion/Conclusions

The first pacing checks post discharge was satisfactory with no further arrhythmias documented.

Unfortunately after a short recovery the patient started to gradually deteriorate and lost 2 stone in weight over a 3-month period. He was subsequently readmitted to hospital for blood stained diarrhoea and vomiting. He suffered a further ventricular fibrillation cardiac arrest despite normal electrolytes and other biochemical parameters, and reported compliance with his beta-blockers, and it was decided that further resuscitation attempts would not be in his best interests. He passed away three months after his first congenital LQTS cardiac event.

Long QT syndrome is generally thought of as a cause of cardiac arrest in young patients. As in this case, cardiac arrests in the elderly are usually initially assumed to be due to ischaemic heart disease or heart failure and this can lead to inappropriate treatment. LQTS is exacerbated by bradycardia, however, conduction system disease related bradycardia is common in the elderly. It is therefore entirely plausible that after a peak of symptomatic LQTS in younger patients, there is another peak in the elderly. We are not aware of similar published cases but given the 1:2000 prevalence of LQTS and tendency to bradyarrhythmia in older age, it seems that there is a potential for under recognition of this entity.

In this gentleman there was extensive debate as to whether he should have a pacemaker or an implantable cardiac defibrillator. This was discussed by pacing and electrophysiology specialists, the medical and nursing team, and with the patient's family. The patient was also involved in the discussion but was not thought to have capacity to make an informed decision regarding treatment. Theoretically it made sense that his latent LQTS had become manifest because of age related conduction disease and bradycardia, and therefore that treating the bradycardia by means of a pacemaker should be sufficient to restore him to his previously asymptomatic state. In a younger, fitter patient, however, one would probably have implanted a defibrillator as in the presence of known LQTS it is hard to be certain that there will never be another non bradycardia related trigger for arrhythmia.

In hindsight, given that this gentleman went on to suffer a further long QT related arrest, it is right to revisit the question of a defibrillator. This gentleman became even more frail over the intervening

3-month period and was felt not to be suitable for resuscitation. An ICD would have made decisions at readmission more complicated and any prolongation of life would have been associated with discomfort and disability.

Case report learning points are as follows:

- Congenital conditions can first produce symptoms in advanced old age and therefore we should remain vigilant for these diagnoses
- Even in asymptomatic LQTS it is important to record the diagnosis and avoid exacerbating drugs
- Family history does not cease to be important in the elderly
- Despite being prominent in cardiac arrest guidelines, amiodarone is not always the most appropriate drug for recurrent VT and VF.

Figures

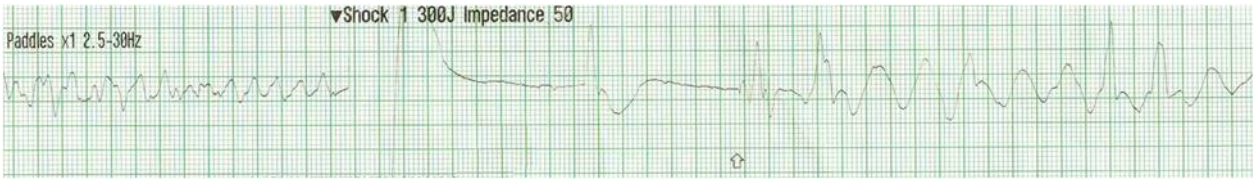

Figure 1: Continuous telemetry rhythm strip recorded during the initial cardiac arrest showing ventricular fibrillation (VF).

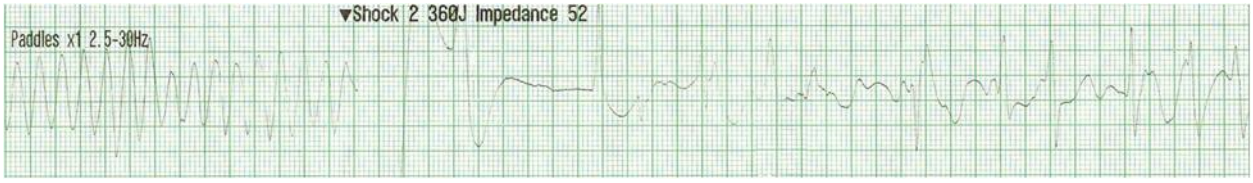

Figure 2: Continuous telemetry rhythm strip recorded during the initial cardiac arrest showing Torsades-de-Pointes (TdP).

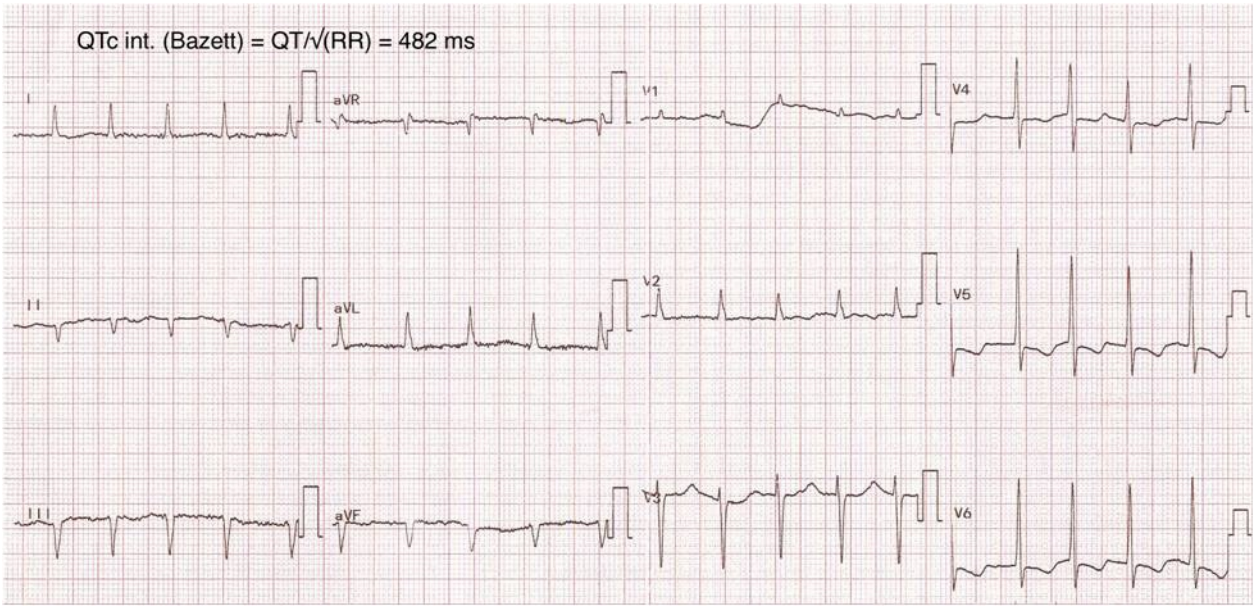

Figure 3: Post initial cardiac arrest 12-lead ECG showing atrial fibrillation (AF) with a rapid ventricular response of 127 bpm, left axis deviation, a QRS duration of 80 ms and a corrected QT interval of 482 ms.

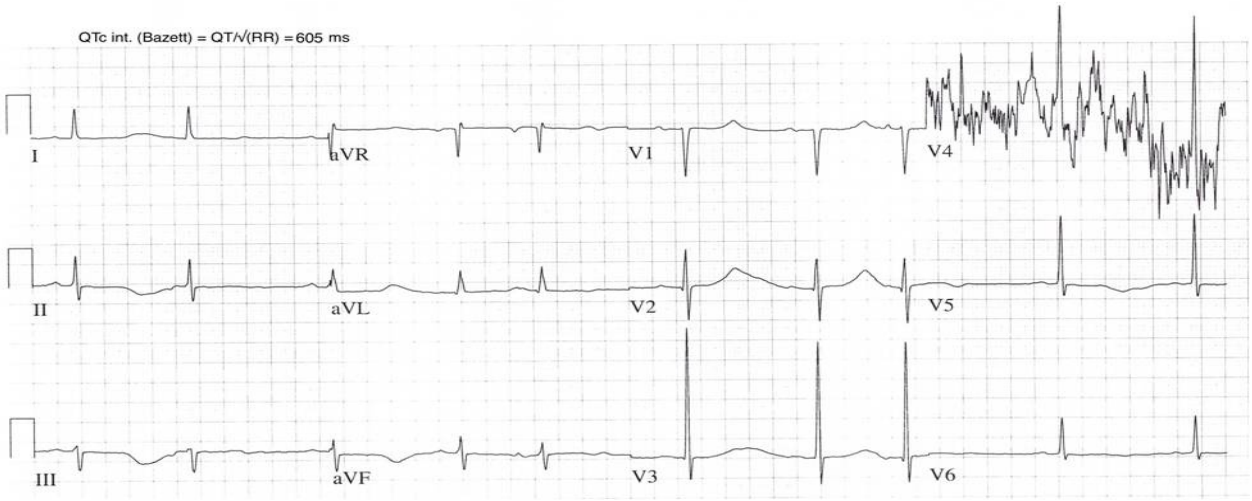

Figure 4: 12-lead ECG demonstrating sinus rhythm with a heart rate of 40 – 58 bpm and a lengthening of the corrected QT interval of 605 ms.

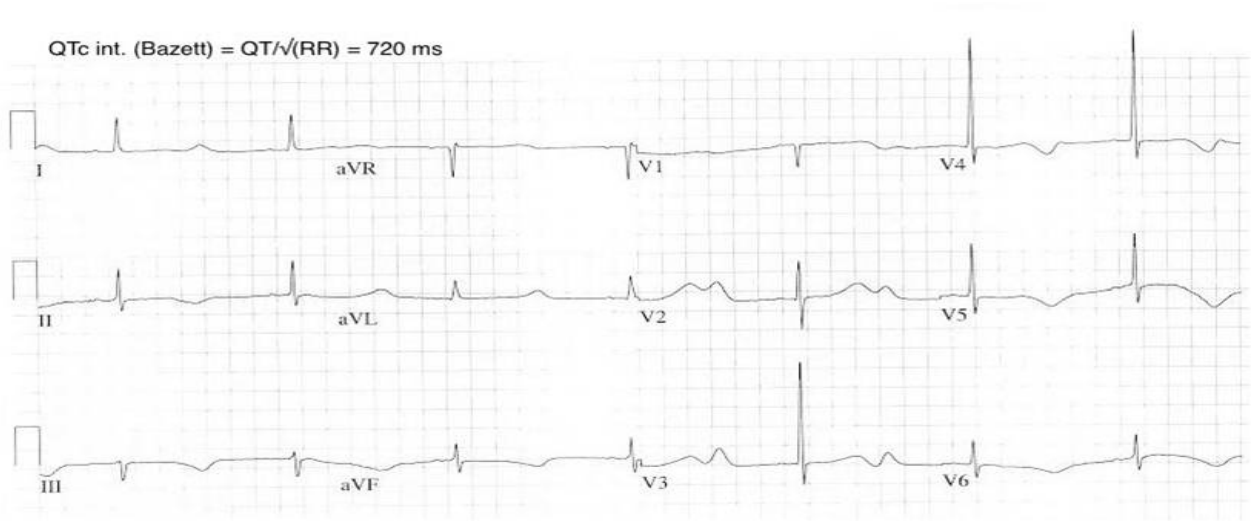

Figure 5: 12-lead ECG post a further cardiac arrest episode underlying worsening of bradycardia and corrected QT interval prolongation (720 ms).

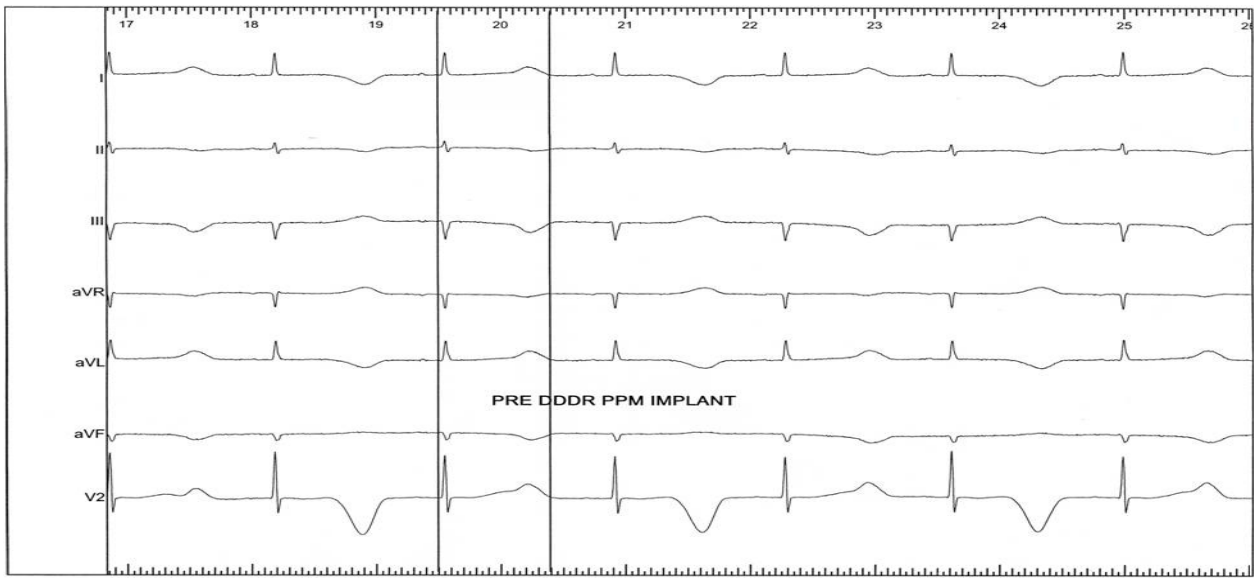

Figure 6: Pre-DDDR PPM implant highlighting a QT interval of more than 700 ms and macrovolt T-wave alternans best seen in lead V2.

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Manuscript Information: Received: March 13, 2017; Accepted: June 07, 2017; Published: June 09, 2017

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Citation: Stanley F, Wood A, Nicolson W. First manifestation of a congenital problem in advanced old age: not all VF is ischemic!. Open J Clin Med Case Rep. 2017; 1270

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