Case report

Prolonged QT syndrome: a probable cause of a drowning death in a recreational scuba diver

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Diving, scuba, death, prolonged QT syndrome, case reports

Abstract

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This case report concerns the death of a 17-year-old scuba diver that was probably due to prolonged QT syndrome (LQTS). LQTS is a cause of sudden death and may be responsible for drownings for which no obvious reason can be found from post mortem or the history of events. Some aspects of LQTS are reviewed and its potential contribution to diving fatalities considered. Consideration should be given to genetic screening of young victims for LQTS in cases of 'in water' sudden death.

Introduction

Unexplained loss of consciousness in young adolescent and adult swimmers and divers is uncommon but well documented anecdotally.[1] This case report concerns the death, which was probably associated with prolonged QT syndrome (LQTS), of a 17-year-old, male, recreational scuba diver. His death was one of a cluster of nine diving-related deaths recorded in South Australia in a twenty-month period (2001–2002).[2] Other deaths from this unfortunate series have been reported previously.[3]

Case report

The deceased, a newly qualified diver, and his father were diving for crayfish. The weather was warm, surface sea conditions calm, and underwater visibility was good with no current. The diving depth was to 7 metres for 30 minutes. The divers were separated for about a minute. The deceased's father found him lying but not wedged under a ledge; he was unresponsive to his father tugging on his legs. An absence of stirred sand causing decreased visibility indicated that there had not been a struggle or perhaps a grand mal convulsion. His rescue was uneventful. His regulator was out of his mouth when found and was not replaced during ascent. Cardiopulmonary resuscitation (CPR) was commenced once he had been retrieved to the boat (of the two rescuers only one had done a basic CPR course). He was taken to the local hospital and a medical retrieval team from the Royal Adelaide Hospital (RAH) was summoned. He was intubated and ventilated (Oxylog ventilator, 100% oxygen [F_iO_2 = 1.0] with a positive end expiratory pressure [PEEP] of 5 cm water) during transport.

On arrival at the RAH, his Glasgow Coma Scale score was noted to be three, he was tachycardiac but in sinus rhythm, and his blood pressure was normal. He had not received any medication. He was normothermic on arrival.

His initial arterial blood gases showed an increased A-a gradient (F_iO_2 = 1.0), no hypoxaemia but a mixed acidosis, the transport ventilator proving to be inadequate against non-compliant oedematous lungs. Initial serum biochemistry was abnormal and he was haemoconcentrated (Table 1). A chest X-ray showed pulmonary oedema and

Table 1
Relevant investigations on admission

Arterial blood gas analysis

F_iO_2	1.0; Oxylog Ventilator; 5 cm PEEP
pH	6.99
pO_2	95 mmHg
pCO_2	80 mmHg
HCO_3	18 mmol.l^{-1}

Biochemistry

Na^+	157 mmol.l^{-1} (135–148)
K^+	4.8 mmol.l^{-1} (3.5–5.3)
Cl^-	119 mmol.l^{-1} (98–106)
Mg^{++}	3.12 mmol.l^{-1} (0.7–0.95)
Creatinine	0.133 mmol.l^{-1} (0.05–0.12)
Glucose	8.9 mmol.l^{-1} (3.9–6.2)
Anion gap	25.1 mmol.l^{-1} (>18)
CK	9753 µg.l^{-1} (0–270)
CKMb	358 µg.l^{-1} (0–7.0)
CKMb/CK	3.7% (<2.6)

Liver enzymes elevated; normal bilirubin

Haemoglobin	186 gl^{-1}
Haematocrit	55

Platelet and white cell counts normal

Figure 1
Twelve-lead electrocardiogram from 17-year-old diver. Also shown is a separate lead II rhythm strip in which the long QT interval is more apparent.
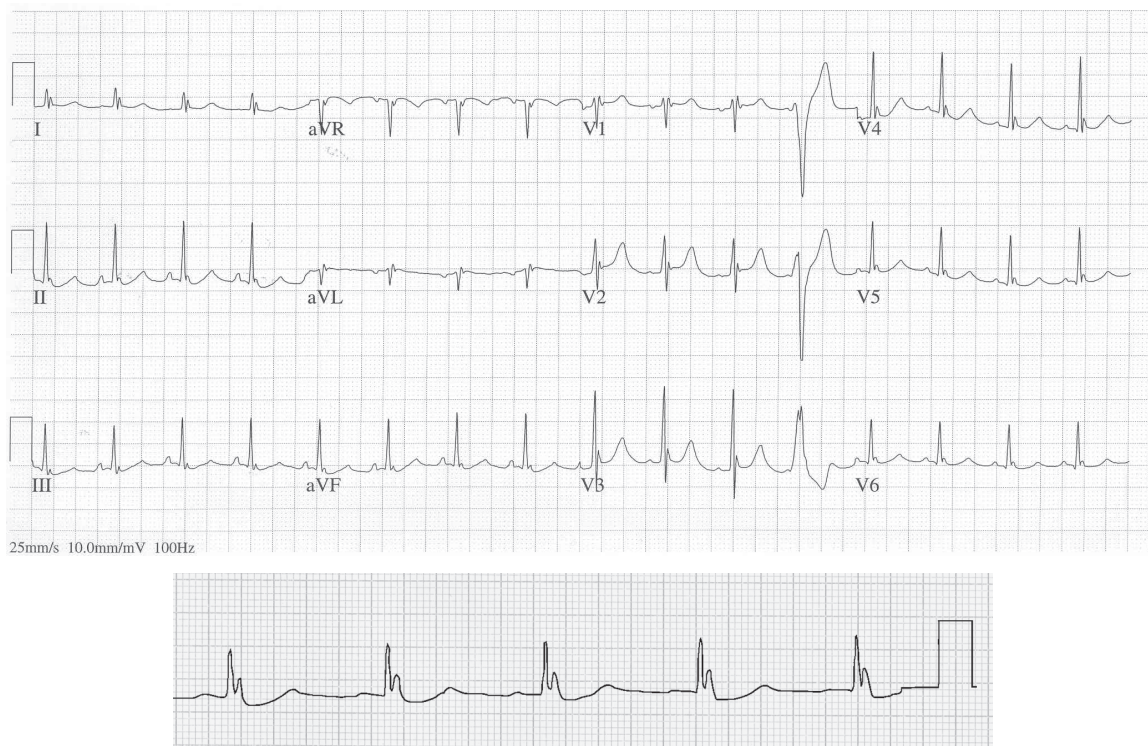
his initial ECG a prolonged QT interval at 483 msec at a pulse rate of 62 min^{-1} (despite the elevated serum Mg^{++}) with an intraventricular conduction defect (Figure 1). During his admission he had several episodes of spontaneous ventricular tachycardia, which spontaneously reverted. These episodes may have been a result of electrolyte abnormalities, cerebral oedema or LQTS. He did not require inotropic support during his resuscitation.

Within 12 hours of admission to the RAH he developed bloody osmotically induced diarrhoea. This is the result of gut ischaemia and is a poor prognostic sign. His pulmonary function improved; the F$_i$O$_2$ was decreased because the risk of decompression illness was thought to be negligible from his dive profile.[4] However, his cerebral oedema failed to improve and his intracranial hypertension resulted in 'coning', brain death occurring within 24 hours of admission. At post mortem no cause for his drowning was found. Genetic screening for LQTS was not performed. The cause of death was reported to be due to salt-water drowning, presumably due to an arrhythmia caused by LQTS.

Specific questioning of his family revealed a history of 'drop attacks' witnessed by his uncle but not by his parents. These were not reported to his general practitioner and were not investigated, and, therefore, it is unknown if they were syncopal attacks. He had had a diving medical prior to commencement of his diving course by his local general practitioner. His family have agreed to undergo genetic investigation performed by their local general practitioner; however, these results are not available to the author.

Discussion

Many of the laboratory findings reported in Table 1 are the combined result of salt-water aspiration and severe hypoxaemia. His elevated serum magnesium (Mg^{++}) level, haematocrit and sodium concentration reflect the fluid shifts that occur when large amounts of salt water are aspirated.[5] The decreased bicarbonate concentration and increased anion gap were due to metabolic acidosis, whilst the elevated creatine phosphokinase indicated muscle and myocardial ischaemia. Presumably his elevated liver enzymes indicated a degree of hepatic ischaemia (although at post mortem he was noted to have Wilson's disease). The cause of the victim's pulmonary oedema is obvious, but there may also have been a degree of myocardial failure due to ischaemia and an elevated Mg^{++}-induced myocardial depression.[4] However, inotropic support was not required during resuscitation.

LQTS has been implicated in cases of 'sudden death' in young adults and children and has been overlooked as a cause of drowning.[6–8] A study in Finland by Lunetta et al reported that LQTS was responsible for one out of 165 drownings and suggested that this may be one cause of 'dry drownings'.[8] Other cardiac causes for sudden death are listed in Table 2.

LQTS is an arrhythmogenic cardiovascular disorder resulting from mutations in the cardiac ion channels. Persons with LQTS have a disorder of cardiac electrical activity causing a delay of repolarisation. The clinical

Table 2
Some causes of sudden death in adults, age <35 years (modified from Hockings[7])

Cardiac
- Hypertrophic cardiomyopathy
- Dilated cardiomyopathy
- Ventricular pre-excitation syndromes (Wolff-Parkinson-White syndrome)
- Coronary artery anomalies (particularly the left coronary artery from the non-coronary sinus of the aortic root)
- Arrhythmogenic right ventricular dysplasia
- Long QT syndrome
- Myocarditis
- Aortic valve stenosis (associated with congenital bicuspid valve)
- Aortic dissection (Marfan's syndrome)
- Left ventricular hypertrophy
- Coronary artery disease – myocardial infarction
- Concussion of the heart (commotio cordis) due to an accidental precordial thump
- Brugada syndrome (idiopathic ST-segment elevation) – electrical familial disease caused by a genetic ion-channel defect

Other
- Drug-induced
- Intracranial events
- Asthma

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features of LQTS result from an episodic ventricular tachyarrhythmia called 'torsades de pointes' (twisting of the points – the QRS axis). Traditionally it has been classified as either inherited – the Romano Ward syndrome – or acquired.[6,7] The inherited disorder is autosomal dominant; hence, if either parent has LQTS then the child has a 50% chance of inheriting the disorder. Synchronised ion movement into and out of cells (potassium, calcium and sodium) produces cardiac electrical activity. These ion channels are genetically encoded with at least six genotypes, and three genes specific for LQTS have now been isolated.[7,9] In addition, it is thought that the acquired variety may have a silent mutation of one of these six genotypes and will remain asymptomatic until exposed to certain conditions, for example hypokalaemia or drugs.[7]

LQTS occurs rarely, 1 in 20,000, and is characterised by sudden death, recurrent syncope and seizures. Indeed, some patients have been diagnosed as epileptic.[10] It can present at any age. Often the first manifestation is syncope or cardiac arrest precipitated by emotional or physical stress or loud noises.[6,7,10] Overall, the world data suggest that 40% of patients are asymptomatic at the time of diagnosis. Weintraub et al published a series of 23 Australian children with congenital LQTS.[11] The median age at time of referral was 10 years (range 4 days to 19 years); 14 (61%) had a family history of the syndrome, and 19 (82%) were symptomatic at the time of diagnosis. Syncope was the main

Table 3
Diagnostic criteria in LTQS (from Booker et al[6]). A score >4 is considered diagnostic

ECG	Points
QTc:	
>480 ms	3
460–470 ms	2
450 ms (males)	1
Torsades de pointes	2
T-wave alternans	1
Notched T wave in 3 leads	1
Low heart rate for age	0.5
Clinical history	
Syncope with stress	2
Syncope without stress	1
Congenital deafness	0.5
Family history	
Family member:	
with LQTS	1
unexplained sudden death <30 yrs	0.5

presenting symptom (69%), then aborted sudden death (26%) and near drowning (5%).[11]

The QT interval varies with heart rate, and because of this variation the QT interval is corrected for rate and presented as QTc. A QTc interval greater than 440 msec is considered prolonged. Lead II is the best lead in which to calculate it. Diagnosing LQTS in patients is difficult – up to 12% of affected individuals may have a normal ECG.[7] However, a clinical scoring system is used (Table 3) and a score >4 is diagnostic of LQTS.[6] Using this scoring system the deceased had a score of 3–5 (2–3 for ECG findings and either 1 or 2 for syncopal episodes – it is unknown whether these were related to stress, see later).

Specific triggers for arrhythmia in LQTS vary according to the genotype involved and are exercise, emotional stress, loud noises, rest, sleep, and neurosensorial stimulation.[6,7,10]

Table 4
Factors associated with diving that may trigger the prolonged QT syndrome

Swimming
Facial immersion
Drugs (Table 5)
Valsalva manoeuvre (particularly if fully β-blocked)
Adrenergic sympathetic stimulation:
    anxiety, physical and emotional stress
    hypoxia
Hypothermia
Sinus bradycardia
Hypokalaemia
Hypocarbia
Sudden auditory stimulation

Table 5
Drugs that need to be avoided in patients with the prolonged QT syndrome, are associated with torsades de pointes (TDP), or have been associated with a prolonged QT interval but lack substantial evidence for causing torsades de pointes[6,7,15]

Type of drug	Avoid in prolonged QT interval	Associated with TDP in clinical dose	higher dose	Possibly associated with prolonged QT interval
Anti-arrhythmic				
Class Ia		Disopyramide Quinidine Procainamide		
Class Ib			Mexilente	
Class Ic				Flecainide
Class III		Sotalol Amiodarone		
Antimalarials		Chloroquine Quinine Halofantrine		Mefloquine
Antihistamines				Terfenadine Astemizole
Prokinetic agents		Cisapride		
SSRIs			Fluoxetine Paroxetine Sertraline	
Antibiotics	Azithromycin Clarithromycin	Erythromycin	Trimethoprim Sulfamethoxazole Ampicillin	
Tricyclic antidepressants			All	
Antipsychotics		Thioridazine Haloperidol Chlorpromazine Pimozide	Lithium	
Bronchodilators	Salbutamol Most bronchodilators			
Adrenergic drugs	All except Dobutamine	Dobutamine		
Miscellaneous	Cocaine	Droperidol Methadone Some appetite suppressants	Chloral hydrate	Vasopressin

In a retrospective study by Ackerman et al of 35 cases of congenital LQTS, six (17%) had a personal history or family history of near drowning or drowning.[11] These investigators also concluded that swimming was a specific arrhythmogenic trigger for genetic-specific LQTS, but offered no explanation as to why. Another study of children with LQTS by Yoshinaga et al showed that face immersion in cold water induced T-wave alternans or a notched T wave.[12] Therefore, several factors associated with recreational diving may be arrhythmogenic triggers (Table 4). Certain drugs are associated with LQTS and torsades de pointes and are listed in Table 5. Diving medical practitioners need to be aware of these.

The treatment options of LQTS include high-dose β blockers, pacemaker, automatic implantable defibrillators (ICD) and specific gene therapy.[6,7,10] However, 25–35% of patients with LQTS on high-dose β-blocker therapy are

likely to have another syncopal attack within five years.[13] A recent case report showed that, even with an ICD, syncopal attacks while swimming are not prevented. This is not surprising because an ICD requires a short time to 'diagnose' the arrhythmia and deliver a counter shock if required, so there may be a brief syncopal episode. Hence, an implantable ICD is a contraindication to scuba diving and swimming. Besides the problems associated with an ICD, swimming appears to be a specific trigger for arrhythmias in LQTS; the reason for this is unknown but it could be due to intense sympathetic stimulation due to physical activity or a high level of anxiety. Swimming or scuba diving while on therapy should, therefore, be avoided.[1,12–14]

The deceased had a history of 'drop attacks', which may have been syncopal episodes. These episodes were not reported to his local general practitioner and hence not investigated. It is not known if his diving medical had included an ECG, nor is it certain that the significance of a prolonged QTc interval would have been recognised and acted upon; without the history of drop attacks he would have scored 3 at best (Table 4). However, a normal QTc interval on an ECG does not exclude LQTS. Up to 12% of these individuals have been reported to have a normal ECG.[7] The author believes the importance of an ECG is understated in diving medical assessment. In his recent practice, five cases of Wolff-Parkinson-White syndrome were diagnosed in 80 consecutive diving medicals performed over a two-year period. Any history of 'fainting episodes' should be investigated with a careful medical and family history, particularly any family history regarding sudden or unexplained deaths in family members less than 30 years old. If there is any doubt, echocardiography, 24-hour Holter monitoring, exercise testing, facial water immersion with ECG monitoring, and genetic screening should be performed.

This case illustrates five points:
- An elevated serum Mg^{++} is indicative of salt-water drowning or near drowning.[5]
- The clinical observation that bloody diarrhoea is a poor prognostic sign following cardiac arrest and resuscitation is widely recognised.
- Specific questioning regarding syncopal or 'drop' attacks must be included in a diving medical and the cause for any of these attacks must be determined before the risks associated with diving can be assessed.
- 'Bystander CPR', or 'just doing something', may be effective.
- All investigations of 'in-water sudden deaths' must include a thorough family and medical history and perhaps genetic screening for genetic disorders associated with 'sudden death' if the family and medical history warrant it.

References

1 Ackerman MJ, Tester DJ, Porter CJ. Swimming, a gene-specific arrhythmogenic trigger for inherited long QT syndrome. *Mayo Clin Proc*. 1999; 74: 1088-94.

2 Courts Administration Authority, South Australia. <http://www.courts.sa.gov.au/courts/coroner/findings/index.html>

3 Acott CJ. South Australian diving-related deaths 2001–2002. A coroner's inquiry. *SPUMS J*. 2004; 34: 130-6.

4 Cohen DS, Matthay MA, Cogan MG, Murray JF. Pulmonary oedema associated with salt water near drowning: new insights. *Am Rev Respir Dis*. 1992; 146: 794-6.

5 Acott CJ, Doolette D. Simulations of near drowning and decompression sickness – a preliminary study. *SPUMS J*. 2002; 32: 35-40.

6 Booker PD, Whyte SD, Ladusans EJ. Long QT syndrome and anaesthesia. *Br J Anaesth*. 2003; 90: 349-66.

7 Hockings BEF. Sudden death in apparently normal young adults. *ADF Health*. 2001; 2: 18-22.

8 Lunetta P, Penttilia A, Levo A, Sajantila A. Molecular screening of selected long QT syndrome (LQTS) mutations in 165 consecutive bodies found in water. In: *Proceedings of International Congress on Drowning*, Amsterdam, 2003, in press.

9 Wang Q, Chen Q, Li H, Towbin JA. Molecular genetics of the long QT syndrome from genes to patients. *Curr Opin Cardiol*. 1997; 12: 310-20.

10 Moss AJ, Robinson JL. The long-QT syndrome. *Circulation*. 2002; 105: 784-6.

11 Weintraub RG, Gow RM, Wilkinson JL. The congenital long QT syndromes in childhood. *J Am Coll Cardiol*. 1990; 16: 674-80.

12 Yoshinaga M, Kamimura J, Fukushige T, Kusubae R, Shimago A, et al. Face immersion in cold water induces prolongation of the QT interval and T-wave changes in children with the nonfamilial long QT syndrome. *Am J Cardiol*. 1999; 15: 1494-7.

13 Moss AJ, Zareba W, Hall WJ. Effectiveness and limitations of β blocker therapy in congenital long-QT syndrome. *Circulation*. 2000; 101: 616-23.

14 Ott P, Marcus F, Moss AJ. Ventricular fibrillation during swimming in a patient with the Long QT Syndrome. *Circulation*. 2002; 106: 521-2.

15 Arizona Center for Education and Research in Therapeutics. <www.torsades.org>

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