CASE REPORT

Ventricular fibrillation after elective surgery in an adolescent with long QT syndrome

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SUMMARY

Congenital long QT syndrome (LQTS) is a potentially lethal but highly treatable channelopathy. Along with multiple risk reduction measures, a recommendation for left sympathetic cardiac denervation therapy and/or implantable cardioverter defibrillator is made for higher risk patients. Despite its relatively common incidence in paediatric patients, there are no formal recommendations regarding perioperative management and discharge criteria for LQTS patients undergoing ambulatory surgery. This report describes a 17-year-old girl, diagnosed with congenital LQTS at 9 years of age, who had an episode of ventricular fibrillation the day after elective ear, nose and throat surgery. Despite several risk factors, she had a same-day dismissal, was not adequately monitored postoperatively and her cardiologists were not notified of her procedure. For the high-risk LQTS patient, we recommend monitoring of perioperative electrolytes and rhythm, postoperative ECG, adequate β -blockade therapy, avoidance of particular pharmacological agents, consideration of overnight observation and communication with the patient's cardiologist prior to procedure, and at discharge.

BACKGROUND

Congenital long QT syndrome (LQTS) is a potentially lethal but highly treatable cardiac channelopathy characterised by its trademark dysrhythmia of torsade de pointes, which can manifest as syncope, seizures or sudden death.¹ A comprehensive risk reduction programme for these patients often includes various measures such as avoidance of QT prolonging medications, electrolyte replenishment, fever reduction, pharmacologic therapy and, potentially, left sympathetic cardiac denervation (LCSD) therapy and/or an implantable cardioverter defibrillator (ICD), when appropriate, based on a patient's long QT risk stratification.^{2–5} This consideration is especially important in the perioperative setting, where several case reports and case series have demonstrated the QT prolonging effect of electrolyte abnormalities and pharmacological agents on patients, specifically in children with clinical LQTS.^{4 6–9}

In 2006, in the USA, the average number of children undergoing ambulatory surgeries was about 3.2 million.¹⁰ Despite an estimated incidence of 1 in 2000 for LQTS¹¹ and the large number of children undergoing ambulatory surgeries annually, there are no formal recommendations regarding the perioperative management, discharge criteria and follow-up recommendations for children with LQTS who need non-cardiac surgery. We present a

case of a 17-year-old girl, diagnosed with clinical LQTS at 9 years of age, who had an episode of ventricular fibrillation (VF) the day after elective ear, nose and throat (ENT) surgery.

CASE PRESENTATION

A previously healthy 9-year-old girl was diagnosed with Hodgkin's lymphoma in December 2005, when she presented with a chest mass. In 2006, while undergoing chemotherapy for Hodgkin's lymphoma, she developed a PICC line infection and had an echocardiogram performed. A screening ECG performed at that time demonstrated QT prolongation, which was thought originally to be chemotherapy (Adriamycin)-related. β -Blocker therapy with atenolol at 25 mg twice daily was initiated, however, the patient's QT interval did not normalise on follow-up evaluations. Subsequently, genetic testing was initiated, which elucidated a pathogenic LQT2 mutation, p.Asn629Ser-KCNH2, and she was continued on atenolol β -blocker therapy. An extended family history was negative for known LQTS and also for multiple unexplained sudden deaths, other than a paternal great grandfather, who died suddenly at 42 years of age.

The patient remained asymptomatic from the standpoint of her LQTS until the age of 13 years, when she experienced a syncopal episode while walking to school. This episode was preceded by a 15 min prodrome of feeling lightheaded. She was incontinent of urine during the episode and was observed to have leg convulsions, but she recovered spontaneously after the episode. β -Blocker therapy was then changed to nadolol 40 mg daily, which was dosed appropriately for a weight of 52.6 kg, and Mexiletine 150 mg twice daily was added with subsequent attenuation in the girl's QT interval.

The patient was deemed to be high risk given her female gender, postpubertal status, possible arrhythmia-mediated syncopal episode, persistent QTc >500 ms and LQT2 genotype, with her LQT2 mutation localising to the pore of the KCNH2-encoded Kv11.1 potassium channel. Therefore, in 2010, she underwent endocardial single chamber ICD placement and LCSD, which were performed without any postoperative complications. The patient was then weaned off the Mexiletine and continued on nadolol 40 mg daily. She then did well with the ICD over the next few months with the exception of an episode of inappropriate ICD shocks for sinus tachycardia while she was horseback riding in June 2013. This resolved after the VF zone limit on the ICD was increased (previous VF zone at 200 bpm).

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In July 2013, when the patient was 17 years old, she underwent an elective ENT surgery for a deviated nasal septum with same-day dismissal. Neither the patient's local cardiology team nor her LQTS specialty team was notified of this procedure. After the patient was discharged, she had nausea and vomiting during the night and experienced four episodes of ventricular tachycardia between 2:35 and 4:25. Initially, she had an episode of ventricular tachycardia that degenerated into VF at a rate of >400 lasting about 40 s, which was effectively terminated with a 35 J shock from her ICD. Subsequently, she had three more episodes of non-sustained ventricular tachycardia (rates 250–429) that did not require defibrillation. She then experienced a seizure and a syncopal episode while being transported to the ED. She was given magnesium and intravenous fluids and admitted to the paediatric intensive care unit for antiarrhythmic therapy and observation.

OUTCOME AND FOLLOW-UP

In September 2014, the patient had her ICD upgraded with the addition of an atrial lead to enable a rapid pacing strategy, and has been doing well with no subsequent ICD shocks. Since then, the patient's family has undergone genetic testing for LQTS. Her four older brothers and both parents had negative results for genetic testing as well as normal ECG without QT prolongation, suggesting a sporadic de novo LQT2 mutation in our patient.

DISCUSSION

Establishing appropriate guidelines for perioperative monitoring and follow-up after ambulatory surgery is extremely crucial in patients with clinical LQTS, especially for those whose disease stratification puts them at a high risk for arrhythmias. Factors that categorise an adolescent patient as high risk for an adverse event include: (1) postpubertal female, (2) compound or digenic heterozygosity, (3) QTc $\geq$ 500 ms, (4) syncopal episode, especially while on β -blockade therapy, (5) $\geq$ 2 cardiac events before 18 years of age.^{12–13} Our patient had several risk factors, which placed her at increased threat for an adverse event after her surgery: (1) postpubertal female, (2) higher risk LQT2 mutation, (3) QTc >500 ms and (4) previous episode of syncope despite being on β -blocker therapy eventually requiring LCSD and ICD placement.^{12–16} Despite these risk factors, there was no adequate perioperative monitoring or follow-up plan outlined for this patient.

Until formal guideline-based recommendations are assembled, vetted and published, we recommend the following simple measures in the recovery room, to minimise the risk of an adverse postoperative event in patients with high-risk LQTS: (1) appropriate monitoring and correction of perioperative electrolyte abnormalities (specifically maintaining potassium at a plasma level > 4 mEq, as well as assessing calcium and magnesium levels), (2) perioperative rhythm monitoring and postoperative ECG with surveillance for T wave abnormalities and/or PVCs prior to discharge, and (3) ensuring adequate β -blockade therapy and avoiding exposure to medications with known torsadogenic potential (tier I drugs on <http://www.crediblemeds.org>), as outlined in previous studies.^{3–5}

Safe alternatives for the treatment of postoperative nausea and vomiting are metoclopramide or promethazine.⁷ In addition to these measures, the patient's primary cardiologist should be notified prior to the procedure and at the time of discharge, especially if there are any concerns. It would be reasonable to consider overnight observation for those LQTS patients with any one of the high-risk factors listed previously, ideally with

continuous ECG monitoring if the patient does not have an ICD. This would allow for early and adequate monitoring during recovery from anaesthesia, correction of electrolyte abnormalities as appropriate, and ensuring adequate management of pain and nausea, which might in fact include use of effective anti-nausea medications that reside on the QT drugs-to-avoid list. Continuous ECG monitoring during overnight observation may be considered if there are documented rhythm abnormalities during the intraoperative or immediate postoperative period.

For the low risk LQTS patient, same day dismissal for elective surgery is appropriate if the postoperative electrocardiogram and electrolytes show no abnormalities. Additionally, the patient and their family should be provided with clear instructions regarding reasons such as profound nausea, repeated episodes of emesis, inability to keep down oral fluids, palpitations and syncope, to seek medical attention. This would provide a cost effective but safe management alternative for lower risk LQTS patients.

Learning points

- Risk stratification for the patient with clinical long QT syndrome (LQTS) is essential in establishing appropriate guidelines for perioperative monitoring and follow-up after ambulatory surgery.
- For high-risk LQTS patients, we recommend monitoring of perioperative electrolytes and rhythm with postoperative ECG, adequate β -blockade therapy, avoidance of particular pharmacological agents, consideration of overnight observation and communication with the patient's cardiologist prior to procedure and at time of discharge.
- For the low risk LQTS patient, same day dismissal after elective surgery is appropriate if there are no abnormalities on postoperative ECG or electrolytes, and if the patient's cardiologist is also in agreement.

Contributors MJA conceived the study and provided critical review; SJK reviewed the case and drafted the manuscript; KNP and JMB provided input to the manuscript, and critically reviewed it.

Competing interests None declared.

Patient consent Obtained.

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