Long QT Syndrome in Pregnancy: Are Vaginal Delivery and Use of Oxytocin Permitted? A Case Report

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Abstract

Background: Patients with congenital long QT syndrome (LQTS) are at increased risk of ventricular arrhythmia, particularly during labour and the puerperium.

Case: A 28-year-old primigravida with known LQTS underwent induction of labour at 41 weeks' gestation using a Foley catheter balloon and IV oxytocin. Vaginal delivery with passive second stage and outlet forceps was undertaken with early epidural analgesia to prevent tachycardia and psychological stress. The patient gave birth to a healthy female, and had an uncomplicated postpartum period under continuous electrocardiogram monitoring.

Conclusion: Vaginal delivery with use of oxytocin for the induction of labour can be safely undertaken in patients with LQTS.

Résumé

Contexte : Les patientes qui présentent le syndrome du QT long congénital (syndrome du QTL) sont exposées à un risque accru d'arythmie ventriculaire, particulièrement pendant le travail et la puerpéralité.

Cas : Une primigravide de 28 ans présentant un syndrome du QTL connu a subi un déclenchement du travail à 41 semaines de gestation effectué au moyen d'une sonde de Foley à ballonnet et d'une perfusion d'oxytocine. Un accouchement vaginal (dans le cadre duquel le 2e stade a été passif et où l'on a procédé à une extraction par application de forceps à la partie basse) a été mis en œuvre et une analgésie péridurale a été pratiquée de façon précoce pour prévenir la tachycardie et le stress psychologique. La patiente a donné naissance à une fille en santé; elle a par la suite connu une période postpartum exempte de complications sous surveillance électrocardiographique continue.

Conclusion : Un accouchement vaginal s'accompagnant d'un déclenchement du travail au moyen d'oxytocine peut être mis en œuvre en toute sécurité chez les patientes qui présentent un syndrome du QTL.

Key Words: Long QT syndrome, labour, oxytocin, vaginal delivery

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INTRODUCTION

Pregnancy is associated with a variety of cardiovascular changes that healthy women can tolerate physiologically, but these may be responsible for clinical decompensation in patients with structural heart defects. The effect of pregnancy on patients with cardiac rhythm disorders is not completely characterized, and different options have to be considered for their management during pregnancy, delivery, and the postpartum period.[1]

Long QT syndrome is a cardiac disorder characterized by prolongation of the QT interval in the electrocardiogram and a propensity to develop torsades de pointes (a polymorphic ventricular tachyarrhythmia) or ventricular tachycardia, frequently leading to syncope, cardiac arrest, or sudden death, often in young, otherwise healthy individuals. It can be congenital, acquired, or a combination of these. Congenital forms are divided into types 1 to 10 according to the causal gene and the mode of transmission. Types 1, 2, and 3 are the most common and together account for 95% of cases of congenital LQTS. They are well characterized, and gene-specific ECG patterns as well as trigger factors for the occurrence of ventricular tachycardia have been established. In LQTS type 1, emotional or physical stress is a triggering factor for arrhythmia.[2]

We report here a case of hereditary LQTS type 1 in pregnancy and discuss the antenatal, intrapartum, and postpartum management of this patient.

THE CASE

The patient, a 28-year-old woman, had been found to have LQTS at the age of 11 after recurrent syncope. Genetic testing showed a truncating mutation in KCNQ1 and established the diagnosis of LQTS type 1. This hereditary form of LQTS, with a dominant pattern of inheritance and not combined with deafness, is known as the Romano-Ward syndrome.

The patient had inherited the genetic alteration from her father, and her condition had been quite stable on β-blockers for many years. At the age of 24, she underwent insertion of an implantable cardioverter defibrillator following cardiac arrest. Subsequently the ICD had functioned appropriately and the patient had been asymptomatic.

The patient was initially referred to our tertiary centre for pre-conception counselling with a maternal–fetal medicine specialist. Once she became pregnant, she attended the high-risk pregnancy clinic monthly until 36 weeks, and then weekly until delivery. A morphologic ultrasound scan was performed in the second trimester. Fetal growth was monitored clinically and assessed with ultrasound at 32 weeks' gestation. The patient was thought to have a low risk of intrauterine growth retardation because of the use of a β-blocker throughout pregnancy.

The course of pregnancy was uneventful for both mother and fetus. Treatment with a β-blocker (metoprolol 100 mg twice daily) was continued during pregnancy without any change in dose because of the clinical stability of the patient following the installation of the ICD. At 38 weeks, a multidisciplinary team discussion was organized to plan further management and the timing and mode of delivery.

Expectant management with anticipation of spontaneous labour was followed until 41 weeks, at which time induction of labour was planned. The patient had labour induced at 41 weeks' gestation using a Foley catheter balloon and IV oxytocin. Oxytocin was cautiously administered using a low concentration and without giving bolus doses. Early epidural analgesia without epinephrine was used to prevent tachycardia associated with pain. The duration of the first stage of labour was 11 hours and 41 minutes; the second stage was passive and lasted 56 minutes. Delivery was assisted by outlet forceps with avoidance of pushing in the second stage. During labour, continuous ECG monitoring was performed, and no arrhythmia was documented.

The patient gave birth to a healthy female weighing 3300 g, with an Apgar score of 9 at one minute and 10 at five minutes.

Immediately postpartum, the patient developed uterine atony, but this was controlled using IV carbetocin 100 μg. Blood loss was estimated at 1000 mL. Continuous ECG monitoring was maintained for 48 hours after delivery, and the patient had an uncomplicated recovery.

The neonate was investigated thoroughly (by ECG and cardiac ultrasound) and showed no evidence of cardiac disease. Genetic testing for the baby was offered, but this was declined by the mother, and a cardiac follow-up was organized. The postpartum period was uneventful and the patient continued with β-blocker therapy.

DISCUSSION

The prevalence of congenital LQTS is estimated at approximately 1:3000.[3,4] It is believed that congenital forms account for 3000 deaths of young people per year in the United States.[2,5] The diagnosis of LQTS can be challenging to make, but is usually based on ECG patterns, clinical symptoms, and genetic findings. The clinical course of patients with LQTS depends on the extent of corrected QT interval prolongation and history of cardiac events, and is modulated by age, gender, and genotype. Beta-blockers constitute the therapy of choice for LQTS, while left cardiac sympathetic denervation and implantation of a cardioverter defibrillator should be considered in patients who remain symptomatic despite β-blocker therapy.[5,6]

Women with LQTS are at significant risk for a cardiac event during pregnancy and the postpartum period, with 10% of patients experiencing their initial cardiac event postpartum.[1] Compared with the pre-conception time period, the postpartum period presents a 2.7-fold increased risk of experiencing a cardiac event and a 4.1-fold increased risk of experiencing a life-threatening event.[6] After this transient high-risk postpartum period, the risk of cardiac events reverts to the baseline pre-pregnancy risk. The increased risk for cardiac events in the postpartum period is significantly reduced by treatment with β-blockers.[1,4] Beta-blockers are not teratogenic, and metoprolol (a β1-selective β-blocker) is considered safe to use in pregnancy. We avoided β2-receptor

ABBREVIATIONS

ECG	electrocardiogram
ICD	implantable cardioverter defibrillator
LQTS	long QT syndrome

blockade because of associated peripheral vasodilatation and uterine relaxation.[7] Most β-blockers are secreted in breast milk, but adverse effects are low in neonates with normal renal and hepatic function. The risk of arrhythmia in a pregnant woman with LQTS outweighs any risk of β-blocker therapy to the fetus or newborn.[1]

During pregnancy, the most important factors increasing the risk of arrhythmia are the increase in sympathetic activity and increased circulating levels of estrogen and progesterone that directly or indirectly influence the number and function of mutant ion channel proteins. During the puerperium, some factors promote the risk of arrhythmia, particularly the decrease in heart rate and associated increase in the QT interval. The psychological stress and altered sleep patterns associated with caring for a newborn infant could contribute to an increase in adrenergically mediated cardiac events during the postpartum interval.[1]

The disadvantage of regional anaesthesia in LQTS is the potential for a high spinal nerve block, causing hypotension and bradycardia-induced parasympathetic override, but this complication can be avoided by using a technique of slow titration of anaesthetic solution.

Women with a cardiac pacemaker or ICD should have the type and nature of the device recorded on the antenatal chart. The effect of a magnet on the ICD should also be noted, since a magnet will deactivate the defibrillator function and the ICD program will be altered once the magnet is removed.[4] Application of a magnet is usually recommended at the time of Caesarean section to deactivate the defibrillator and detection functions.[4] Limited data are available on the optimal mode of delivery in these cases. Most published case reports describe delivery by Caesarean section.[4] Only two case reports describe successful vaginal delivery,[8,9] and in one of these cases the patient had an ICD.[9]

In our case, we planned for a vaginal delivery under epidural analgesia. Early induction of epidural analgesia was planned to decrease the sympathetic stimulation and myocardial oxygen consumption associated with labour.[10]

Management in a calm and quiet environment was essential to reduce adrenergic responses during delivery. Beta-blocker treatment was maintained, as well as continuous ECG monitoring and regular assessment of electrolytes, with replacement if necessary. We ensured that resuscitation equipment was available in the vicinity. Delayed pushing and passive second stage with assisted vaginal delivery was chosen since the Valsalva manoeuvre prolongs the corrected QT interval in healthy non-pregnant individuals.[4] Some drugs may cause adrenaline-like effects and prolong the QT interval; these include some antibiotics, antidepressants, antihistamines, ephedrine, oxytocin, phenylephrine, diuretics, some anti-arrhythmic, and inotropic drugs, cholesterol-lowering drugs, diabetes medications, and some antifungal and antipsychotic drugs. These drugs are officially listed[11] and were avoided during pregnancy and labour.[4] Since no information on the effect of prostaglandins on the QT interval was available, we chose to use a Foley catheter balloon for cervical ripening.

Oxytocin is potentially dysrhythmogenic.[12] The underlying mechanism is unclear, but a direct effect on cardiac repolarization has been suggested. Oxytocin receptors have been found in human hearts.[12] In 2001, two deaths were related to a bolus (10 IU) of oxytocin.[13] Following these events, a report in 2005 from the United Kingdom recommended that for all women bolus injections of oxytocin should be given slowly and in a dose not exceeding 5 IU.[14] Our patient received an infusion of oxytocin for induction of labour using a low concentration and no bolus doses.

CONCLUSION

Congenital LQTS is a potentially life-threatening condition. Delivery and the postpartum interval is a critical period for women with LQTS, with increased risk of ventricular tachycardia. A multidisciplinary team approach is necessary to evaluate management carefully throughout pregnancy and delivery. Vaginal delivery can be safe in women with LQTS, particularly those with an ICD. Early epidural analgesia must be offered to prevent ventricular dysrhythmias. Oxytocin may be administered prudently, without giving bolus doses, at delivery. The highest risk of arrhythmia occurs in the postpartum period, and medical treatment must be continued during this period with close ECG monitoring. Parents should be offered genetic counselling and cardiac follow-up for the newborn.

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