

Case report

Pregnancy in long QT syndrome with implantable cardioverter-defibrillator. A case report and review of the literature

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Summary

Implantable cardioverters-defibrillators have decreased morbidity and mortality as well as improved quality of life in patients with life-threatening cardiac arrhythmias and allowed an increasing number of young women to reach their reproductive years. New questions and tasks arise for medical professionals as to organize appropriate management of these patients, because little is known regarding the risk and outcomes of such pregnancies. The aim of this report is to describe our centre's first experience of pregnancy and delivery management in patient with an implantable cardioverter-defibrillator as primary prevention of ventricular arrhythmias in congenital long QT syndrome.

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Introduction

Thirty years passed since the first implantable cardioverter-defibrillator (ICD) was used for the treatment and prevention of life-threatening arrhythmias [1]. Subsequently, the number of device implants has increased each year all over the world [1]. Although, the majority of devices are implanted to patients with acquired heart disease, the indications for ICD implantation expanded to include younger age groups, in particular those with inherited and congenital heart disorders [2]. For these younger patient cohorts, survival to reproductive maturity and beyond is now the usual process [3], leading to a new and unique group of young women with an ICD in situ who wish to become pregnant.

The effects of various pharmacological agents to treat arrhythmias during pregnancy have been evaluated [4]; however, little is known regarding the outcome of pregnancy in women with ICDs.

Pregnancy outcome results for such patients are mostly limited to case reports [4–6]. The objective of this article is to describe our experience in management of pregnancy and delivery in patient with long QT syndrome and ICD, and also to review publications on this topic.

Case report

A 19-year-old female in third trimester of her first pregnancy was referred to our cardiology out-patient department for the risk evaluation and recommendations for follow-up and delivery. Two years ago she had three syncopal episodes in 2 months. Before fainting she felt pain around the neck and then she collapsed. All episodes of fainting were similar, but only once one her mother heard a thump and found her on the floor disoriented and with no recollection of what had happened. Congenital long QT syndrome (LQTS) was diagnosed from her ECG changes and positive family history. LQTS was diagnosed for her mother after an episode of syncope during pregnancy, but she had never used antiarrhythmic medications and remained asymptomatic.

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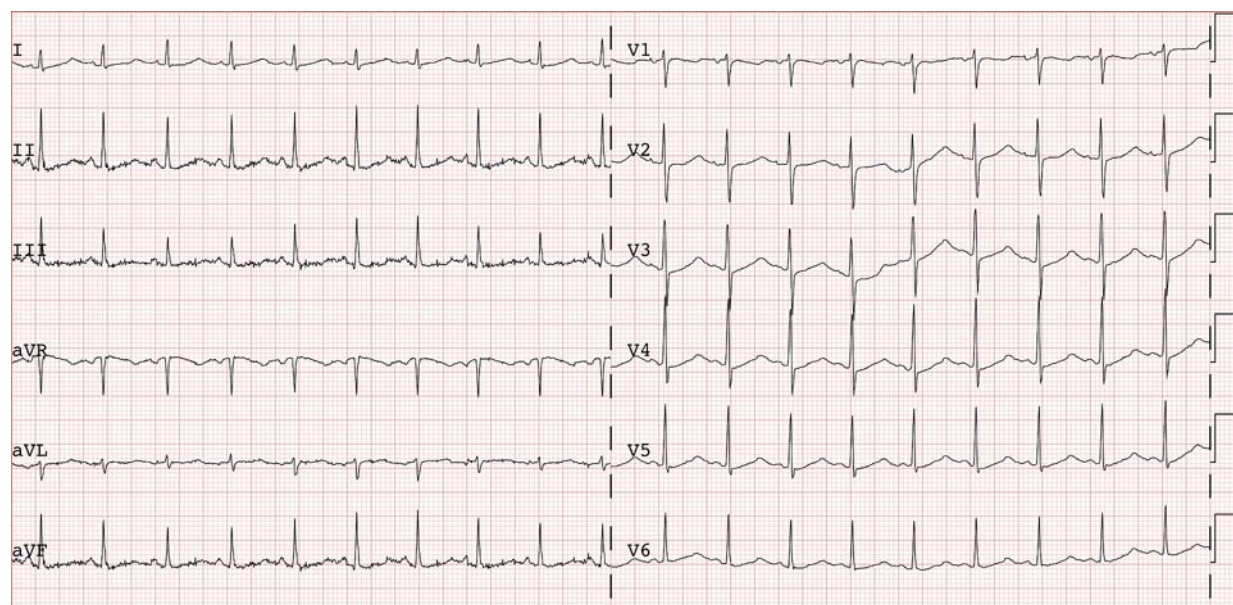


Figure 1. Twelve-lead ECG taken in September 2012 at 32 weeks of gestation. QTc according to Basset's formula equals 490 ms.

Patient's uncle was also affected. He died at age of 27. It was sudden unexplained death during sleep. The patient's father's ECG was documented as normal. Indeed, genetic testing was not available. Neither patient nor other family members had hearing disorders.

When she was 17 years old, she was admitted to our hospital because of syncopal episodes. 24 hours ECG recording showed normal heart rate 60–135 beats/min and QT interval prolongation to 490–630 ms, domed biphasic T waves. There were not any bradycardia episodes or pauses in 24 hours ECG. The ICD was implanted for primary prevention of sudden cardiac death in 2010. She experienced no complications, or ICD related problems after procedure. She was in a stable condition taking metoprolol succinate controlled release tablets 50 mg/d six months after procedure, with a QTc of about 490 ms at routine ECGs. Metoprolol was discontinued six months after ICD implantation because of an uneventful course. She did not have any cardiac disease or other problems that could be considered as the contraindication for pregnancy. The patient became pregnant in 2012. The time interval between ICD implantation and the pregnancy was 20 months. She was under the care of the multidisciplinary maternal cardiology team at two university hospitals (tertiary centres).

At 32 weeks the patient came to our clinic for cardiologist control. Physical examination showed blood pressure 100/60 mmHg, pulse rate 110 beats/min. ECG showed sinus tachycardia 115 beats/min and QT interval prolongation (Fig. 1). A repeated antepartum echocardiography study showed tricuspid valve regurgitation

grade 1 and the normal left ventricular size and systolic function (left ventricular ejection fraction more than 55%). 24 hours ECG showed that the heart rate throughout the day was 65–154 beats/min. There were documented 18 ventricular and 3 supraventricular premature beats.

According to the data the delivery plan was written and given to all members of the specialist team, labour ward and to the patient. She was advised to deliver her baby in specialized hospital. It was advised to try to have a vaginal delivery and that the ICD should be switched off during delivery period. During the antenatal period, device parameters remained unchanged from the non-pregnant state. The period of pregnancy was uneventful; she did not experience any arrhythmic episodes or other significant cardiac symptoms. The patient did not take any pharmacological therapeutics during pregnancy. No ICD shocks were registered before pregnancy and during pregnancy.

The patient was brought to the intensive care room at the day of planned delivery. The ICD was deactivated by cardiologist. The patient had external defibrillator pads placed and regular sinus rhythm was recorded by continuous ECG monitoring. Labour was induced with oxytocin and an epidural anesthesia was established in case to reduce emotional stress. The gestational age at delivery was 39 weeks. Our patient delivered vaginally a baby girl with Apgar's score of 9/10. The baby girl was born healthy, and ECG was normal at birth. The birth weight was 3.36 kg. The genetic testing for baby and family was recommended. The ICD antitachycardia function switched on two hours after delivery. The patient had no ar-

rhythmia episodes during delivery and in the 24- to 48-hour period after delivery too. On the third day after delivery she was discharged from the hospital without medication. Three months later the patient was contacted by phone: she feels good. She does not use any medication and feeds her baby with the breast milk. She had no events associated with ICD or arrhythmia all this period.

Discussion

Although the number of young women reaching their child-bearing age with ICD is increasing, published pregnancy outcome data for these patients are minimal [5,6].

In general young women with ICDs were advised against becoming pregnant because it has not been known what effects the ICD may have on the pregnancy or how pregnancy might affect the device functioning or the underlying rhythm disorder. Data presented by Schuler et al. [2] demonstrate that pregnancy outcome overall in generally could be good with no arrhythmic episodes or other significant cardiac symptoms or ICD troubles. Despite the outcome of pregnancy in women with heart disease and an ICD implant is good, but medical and/or and device complications are frequent. Arrhythmias episodes and device shocks have been reported, but there were no adverse maternal or fetal effects [2].

Therefore, a woman who is in clinically stable condition and decides to have a child should not be routinely discouraged from doing so based on the presence of the ICD [7].

Data presented by Natale et al. [7] show that in the 11 women in whom ICD therapy was delivered, no adverse fetal outcomes were evident as a result of the ICD discharges. It seems that hemodynamic changes are only transient due to the rapid termination of ventricular tachyarrhythmia by the ICD. In addition, it is unlikely that ICD discharges could cause life-threatening fetal arrhythmias. The fetal heart has a high fibrillation threshold and the amount of current reaching the uterus should be small because therapy from internal defibrillation is a very directed [7,8].

According to authors the fetal status should be checked thoroughly after ICD shock because of the possibility of fetal hypotension associated with the arrhythmia [9].

It is also mentioned that, in women with the congenital LQTS, the risk of cardiac arrest is greater during the post-partum period in comparison with before or during pregnancy [10–13]. 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 [15].

In contrast, other authors state that women are less prone to arrhythmias during pregnancy although they commonly complain of palpitations, which are sometimes related to the increase in heart rate during pregnancy [16].

As we know the patients try to avoid drugs during pregnancy as much as possible. However many anti-arrhythmic medications are FDA category B or C, and generally considered safe, with the exception of amiodarone and atenolol (category D) [8,14]. Data have shown that ICD therapy is often combined with antiarrhythmic medications in selected patients in order to minimize the frequency of episodes requiring painful shock therapy [9]. The advent of ICDs has led to a greater arsenal of therapeutic management of patients with LQTS. ICDs have an advantage over medical treatment in pregnant patients in that the fetus is not exposed to anti-arrhythmic drug toxicity or its effects [20]. In general, if pregnancy is planned, the implantation of an ICD should be considered in patients only with the high risk factors for sudden cardiac death [10].

The International LQTS Registry reported that there are some age dependent factors in patients with congenital long QT syndrome. The increased prevalence of QT prolongation and cardiac events in females after puberty may be related to the effects of sex hormones [10,21]. According to literature endogenous estrogens appear to increase QT interval and exacerbate arrhythmia susceptibility. However, emerging evidence suggests that endogenous progesterone shortens the QT interval and protects against rhythm disturbances [21]. As was mentioned, our patient had no complaints or arrhythmia-related symptoms before puberty. The significantly higher levels of estrogen observed during pregnancy could be associated with an increased risk of cardiac events, but the substantial amounts of progesterone presenting during pregnancy may oppose the potentially pro-arrhythmic effects of estrogen on cardiac repolarization [13].

According to current data, epidural anesthesia during intrapartum period may be helpful to reduce the catecholamine release associated with pain [18], however, one must be careful to avoid reflex tachycardia due to maternal hypotension [19]. The disadvantage of regional anesthesia 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 anesthetic solution [11]. Data shows that epidural and spinal

anesthesia are safe and effective forms of anesthesia for patients with ICDs [19].

Reportedly, the hormonal and autonomic changes and the strong uterine contractions during delivery did not precipitate any arrhythmias or ICD firings [4,19]. In this respect, even though the high voltage therapy status (on/off) of the ICD at the time of delivery appears to have no effect on the overall outcome, many authors had recommend leaving the device “on” during vaginal deliveries [4,19]. If an arrhythmia develops, more prompt ICD discharge can be delivered than that by external defibrillation. Certainly, in the case of the caesarean section, the device must be “off” because electrosurgical cautery is involved [4,19].

As everyone could have such patients in their practice, we must accumulate knowledge. A multidisciplinary team approach is necessary to organize the work of service and to prepare appropriate follow-up in order to have the good outcomes of pregnancy and delivery.

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