



# Anaesthesia Application for Cardiac Denervation in a Patient with Long QT Syndrome and Cardiomyopathy

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Long QT syndrome is a congenital disorder that is characterized by a prolongation of the QT interval on electrocardiograms and a propensity to ventricular tachyarrhythmias, which may lead to syncope, cardiac arrest or sudden death. Cardiomyopathy and pulmonary hypertension diseases have additional risks in anaesthesia management. In this study, we emphasize on one lung ventilation, pacemaker-implantable cardioverter-defibrillator and the anaesthesia management process in a patient with long QT syndrome, cardiomyopathy and pulmonary hypertension who underwent thoracic sympathectomy.

**Keywords:** Long QT syndrome, ketamine, cardiomyopathy, implantable cardioverter-defibrillator, one lung ventilation, ICD

## Introduction

Long QT syndrome is a dysfunction of the ion channels affecting the repolarisation phase of the myocardial action potential. With characteristic findings of a prolonged QT interval ( $QT_c > 0.44$  s) on electrocardiography (ECG) and T wave changes, a torsade de pointes (TdP)-type ventricular tachycardia that emerges because of exercise or emotion can result in syncope or sudden death. Dysfunction of the ion channels may be hereditary or acquired. The acquired form emerges with the drugs used that prolong the QT interval or the disorder of electrolyte balance. It is known that the inherited type is caused by mutations in several genes encoding the ion channels in the myocyte membrane (1, 2).

Because of the development of life-threatening arrhythmias, these patients are advised to stay away from drugs that prolong the QT interval. High doses of beta-blockers need to be applied, and an implantable cardioverter defibrillator (ICD) needs to be used in treatment. Ganglionic sympathetic denervation is performed in selected cases. The fact that the QT interval can be affected by the anaesthetics and the interaction of ICD with electrocautery and other electrical appliances makes maintenance difficult in the intraoperative period.

A number of cases have been reported in the literature about the anaesthetic management in long QT syndrome. However, a cardiac denervation case in which simultaneous cardiomyopathy and pulmonary hypertension existed, automated external defibrillation was used instead of ICD and an anaesthetic management was provided with a single-lung ventilation was not encountered in the literature.

The processes of one lung ventilation, pacemaker-ICD and anaesthetic management in a thoracic denervation surgery planned for a patient who has hereditary QT syndrome, cardiomyopathy (CMP) and pulmonary hypertension (PHT) and whose arrhythmias did not decrease despite medical treatment is highlighted in this presentation.

## Case Presentation

A 19-year-old young woman (weight, 55 kg) applied to our hospital with complaints of quickly getting tired, weakness and shortness of breath that she had experienced for about 10 years. In the family history of the patient, who was diagnosed in examinations with non-compaction dilated cardiomyopathy, 2<sup>nd</sup> degree mitral insufficiency and pulmonary hypertension, it was found that her two siblings also had dilated cardiomyopathy (CMP) and her two uncles had died suddenly at a young age. Complaints of sudden fainting and loss of consciousness had begun in 2008 in the patient, in whom heart drugs and follow-up were started and, in the advanced tests performed afterwards, a long QT syndrome diagnosis was made and ICD was

Table 1. Intraoperative haemodynamic and laboratory data of the patient

	Before induction	After induction	30 min	60 min	90 min	120 min
SAP (mmHg)	110	100	95	98	88	92
DAP (mmHg)	70	65	60	62	55	59
HR (beats min <sup>-1</sup> )	90	88	75	78	68	70
CO	4	3.4	3.1	3.6	4.1	4
SVV	7	9	3	4	3	5
SpO <sub>2</sub> (%)	95	99	99	99	99	99
CVP (mmHg)	-	12	13	13	13	13
Sodium (mmol/l <sup>-1</sup> )	137	137	139	141	140	138
Potassium (mmol/l <sup>-1</sup> )	4.1	4	4.2	4.1	4.1	4
Calcium (mmol/l <sup>-1</sup> )	1.11	1.1	1	0.95	0.98	1
Glucose (mg dL <sup>-1</sup> )	112	110	115	118	116	120

SAP: systolic arterial pressure; DAP: diastolic blood pressure; HR: heart rate; CO: cardiac output; SVV: Stroke volume variation; SpO<sub>2</sub>: arterial oxygen saturation obtained by pulse oximetry

placed. Radiofrequency ablation procedures were performed on three different ectopic focuses with an electrophysiological study (EPS) in 2012 in the patient with continuing palpitation. Despite medical treatment (including the highest dose of beta-blockers that can be tolerated) and EPS, because ICD was continually active due to frequently ongoing ventricular extra systoles and a TdP rhythm, the patient's daily life was impaired. The patient was put on the list of transplantation-ventricular assist device because of non-compaction dilated CMP and inherited long QT syndrome in cardiac transplant council. Meanwhile, it was decided to perform a left cardiac sympathectomy on the patient. 12-lead electrocardiogram, pulse oximetry, invasive radial artery pressure and cardiac output monitoring with a flotrac device were performed on the patient, who was taken to the operating room without premedication, after being evaluated in the outpatient clinic and after the necessary preparations were completed. Meanwhile, only the required personel were allowed in the operating room in case of arrhythmias might occur with the patient's emotional stress. To reduce potential stress, the preparation of materials by the nurses was postponed and a quiet and relaxing environment was created in the room as far as possible. In the beginning of the operation, properties of the ventricular tachycardia and fibrillation treatment of the ICD were switched off and the pacemaker function was switched to the mode of a lower rate of 50 beats min<sup>-1</sup> VVI. In case of dangerous arrhythmias that might occur during the pre-operative period, application of external defibrillator at a supine position with the head 45 degrees up and under sterile drapes might have been ineffective and therefore, automated external defibrillators (AED) pads were attached as a more practical and effective method. Instead of inhalation anaesthetics, which are thought to prolong the QT interval, total intravenous anaesthesia was preferred; 300 mcg of fentanyl

in induction, 3 mg midazolam, 50 mg of rocuronium and 0.5 mg kg<sup>-1</sup> of ketamine in titrated doses were administered. The maintenance was continued with 3 mcg kg<sup>-1</sup> of fentanyl, midazolam 0.03 mg kg<sup>-1</sup>, 0.3 mg kg<sup>-1</sup> rocuronium and a 50% O<sub>2</sub>-air mixture in 40-min intervals. On the patient intubated with a left double-lumen tube inserted at once without causing sympathetic stimulation, right internal jugular vein and bladder catheterization were performed later, since there were pacemaker pads on the left. Then, the left arm of the patient, who was put in a supine position with the head 45 degrees up, was suspended. After the left lung was deflated, the camera and process ports were placed through the left anterior axillary line intercostal 4<sup>th</sup> space. During one lung ventilation (OLV), 7 mL kg<sup>-1</sup> tidal volume and 5 cm H<sub>2</sub>O positive end expiratory pressure were applied to the right lung. The left thoracic sympathetic chain was followed at the level of T1-4 and it was dissected from the surrounding tissues and excised. After possible accessory nerves were cauterized, a drain was inserted and the incision was closed. Bupivacaine injection was applied to the intercostal nerves in the incision area by surgeons. Following the completion of the surgery, the patient was put in double-lung ventilation, the position was restored and the patient was extubated comfortably using Sugammadex. The carbon dioxide and oxygen partial pressures remained at normal levels within the period of implementation of one lung ventilation. Electrolytes and blood sugar levels were periodically monitored through blood gas analysis, and normothermia and normovolemia were provided. ECG changes, defibrillation needs and haemodynamic changes were not observed during the attempt (Table 1). At the end of surgery, analgesia was achieved with 25 mcg intravenous fentanyl and in later stages, 25 mcg of fentanyl was applied only once to the patient who did not complain much about pain as long as she stayed in the intensive care unit. After

the extubation in the operating room, ICD was reactivated, the pacemaker settings were made and the patient was taken to the intensive care unit. The patient, while still being monitored seamlessly in the cardiology clinic, was informed, and written informed consent was taken in order to make this presentation.

## Discussion

Anaesthesia in patients with long QT syndrome carries a high risk (3-5). Similarly, the application of anaesthesia is also risky and complicated in cases of dilated cardiomyopathy (6). In this case, the use of drugs such as ketamine, ephedrine and adrenaline that enhance sympathetic activation can create dangerous arrhythmias. On the other hand, in order to prevent hypotension-bradycardia, which may develop in the induction of anaesthesia, these drugs are important in patients with borderline ventricular function due to dilated cardiomyopathy. Therefore, although the pair of midazolam-fentanyl, which is considered to be safe for long QT syndrome in induction, was applied slowly with increasing doses with ketamine  $0.5 \text{ mg kg}^{-1}$ , a stable arterial pressure was provided (7). Even though it is recommended in the literature to avoid the use of ketamine in long QT syndrome, sympathetic activation was not seen in this patient because it was used alone and not in high doses (8). In the patient with heart failure, the use was haemodynamically positive in terms of balancing the depressing effects of the anaesthetics on myocardium.

Induction of the airway during the insertion of a double-lumen tube for one lung ventilation (OLV) may cause haemodynamic reactions. Although sympathetic stimulation can cause dangerous arrhythmias in long QT syndrome, parasympathetic stimulation may fairly depress myocardial insufficiency and may impair oxygen delivery. In addition, problems such as hypoxia, hypercapnia, acidosis development and high airway pressures that could cause excessive sympathetic stimulation and disrupt the myocardial performance may be experienced in later stages of the surgery with one lung ventilation. Therefore, ventilation parameters and blood gas analysis were closely followed in the patient during the settlement of the double-lumen tube and afterwards in the application of OLV, and no problem was experienced.

In this young patient, who needed continuous ICD shock, in addition to her worries of undergoing a surgery, many processes such as anaesthetic drugs, airway instrumentation, OLV, surgical stress, the activation of inflammatory pathways and electrolyte imbalance can easily start malignant arrhythmias. Therefore, it may cause catastrophic consequences to stop the ICD. On the other hand, electromechanical interference may occur if the ICD is left activated. In case of the development of polymorphic ventricular tachycardia, opening of surgical drapes, removal of the sterile transparent surgical cover, removal of trocars and the deployment of the paddles at sitting position can be time-consuming for defibrillation. In patients with torsades de pointes and a risk of ventricular fibrillation, the AED approach performed by placing external pads in the

intraoperative period is a recommended application (9). For all these reasons, it was decided that the most appropriate approach for such a patient was automated external defibrillator. After the operating status of the AED device brought to the operating room was tested, the pads were placed. Thus, effective defibrillation could be applied to the patient in a short time without contaminating the surgical site. After proper position and preparations, surgery was started and no malignant arrhythmias developed during the intraoperative period. At the end of the intervention, the pacemaker and ICD of the patient were reactivated.

In long QT patients who will undergo denervation with videothoracoscopy, the application of thoracic epidural anaesthesia (TEA) is also an option (9, 10). With thoracic epidural anaesthesia, minimal effects were observed on the haemodynamic parameters, such as heart rate, mean arterial pressure, central venous pressure and cardiac index, and it was said to be well tolerated by patients (11). We did not consider this option because of the inconsistency of our patient, not being able to provide a comfortable position and the demand for a surgery with a double-lumen tube.

## Conclusion

The anaesthesia of patients with long QT syndrome and dilated cardiomyopathy requires very delicate management. When these two severe clinical situations come together, a much more attentive intraoperative approach is needed. In the presented case, an uneventful operation was performed with a known history, multidisciplinary preoperative preparation and a careful anaesthetic plan.

**Informed Consent:** Written informed consent was obtained from patient who participated in this case.

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