ANAESTHESIA FOR A PATIENT WITH LONG QT SYNDROME UNDERGOING RENAL TRANSPLANTATION

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Summary

Anaesthetizing patients with Long QT Syndrome is a major challenge, as the potential for sudden catastrophic cardiovascular collapse is well known. We present a 15-year-old boy with Long QT Syndrome who presented for an elective renal transplant. All electrolyte concentration abnormalities were corrected preoperatively and adequate β blockade was maintained. The patient was given a taper controlled infusion of propranolol together with opioids and anaesthesia was uneventful and the patient was extubated at the end of the surgical procedure.

Key Words: Long QT Syndrome, Arrhythmias, Torsade de Pointes, Renal transplantation

Introduction

Long QT Syndrome, although rare, can be associated with catastrophic complications especially during the perioperative period. Many anaesthetic drugs have been implicated in precipitating malignant arrhythmias in susceptible individuals. The management of these patients is a delicate matter requiring a multidisciplinary approach, often involving the cardiologist, surgeon and anaesthetist. We present a patient with symptomatic Long QT syndrome, presenting for renal transplantation.

Case report

A 15-year-old boy was electively scheduled for renal transplantation. Two years prior to surgery, he was diagnosed with end-stage renal failure and has been receiving Continuous Ambulatory Peritoneal Dialysis.

The patient gave a history of recurrent syncopal attacks during the past eight months. These attacks were preceded by emotional and physical stress, usually lasted less than one minute, and was not associated with residual neurological deficits. Following further investigations, he was diagnosed to have Long QT Syndrome, and was started on propranolol 80 mg tds. The frequency of syncopal attacks decreased and he was symptom-free in the three months prior to surgery.

Preoperative assessment revealed a thin boy with a sallow appearance. He had a blood pressure of 130/80 mmHg and a regular pulse rate of 72 per min. Apart from having kyphoscoliosis due to tertiary hyperparathyroidism, examination of the various systems was essentially unremarkable.

Although preliminary investigations showed a prolonged QT interval (QTc interval of 480 ms), the latest ECG done prior to operation did not show any evidence of a prolonged QT interval.

Preoperative investigations revealed that serum electrolytes (potassium, calcium, magnesium and sodium) were within normal limits. Serum creatinine
was 1340mg dl⁻¹. The patient had a haemoglobin concentration of 12.9 g dl⁻¹, a normal platelet count and normal coagulation profile. The chest radiograph showed a normal cardiac silhouette and no signs of fluid overload. Echocardiography revealed an ejection fraction of 52% with normal valves and heart chambers. Spirometry revealed a mild restrictive pattern.

On the day of surgery, the patient was given oral midazolam 3.75mg as premedication. External pacing apparatus, a defibrillator and drugs necessary for management of cardiac arrhythmias were made available before induction of anaesthesia.

General anaesthesia was induced with intravenous fentanyl 100 μg, followed by a target controlled infusion of propofol at 6 μg ml⁻¹. Muscle relaxation was achieved and maintained with atracurium. Invasive blood pressure monitoring was established immediately after induction of anaesthesia. The trachea was intubated after achieving an adequate depth of anaesthesia to blunt the sympathetic effects. A central venous catheter was then inserted into the right internal jugular vein.

Shortly after intubation of the trachea, the mean arterial pressure (MAP) dropped transiently to a minimum of 50 mmHg. Intravenous fluids and 3 mg of Ephedrine were given. The entire episode, with the MAP being below 60 mmHg, lasted less than 10 minutes. No arrhythmia followed the injection of ephedrine. Anaesthesia was maintained with a target concentration of propofol between 2 and 4 μg ml⁻¹. Analgesia was provided with judicious boluses of fentanyl and morphine. Oxygen saturation, end-tidal carbon dioxide concentration, blood glucose and electrolyte levels were maintained within optimal operative limits. Mean arterial pressure was maintained at 60 –100 mmHg, and central venous pressure was kept between 10 –15 mmHg. ECG monitoring revealed a stable sinus rhythm throughout the whole operation. Total blood loss was 400 mls, and was replaced with a combination of crystalloids and colloids. Hypothermia was kept to a minimum.

The operation proceeded uneventfully, and lasted about four hours. At the end of the procedure, muscle paralysis was reversed with neostigmine 1.25 mg together with atropine 0.5 mg. The drugs were given in 4 divided doses over 10 minutes. Extubation of the trachea was uneventful. The patient was subsequently transferred to the Intensive Care Unit. Monitoring of the ECG continued for 24 hours postoperatively. Adequate analgesia was provided using intravenous morphine given via a Patient Controlled Analgesia (PCA) device. Beta-blockade was given through the intravenous route until resumption of oral maintenance therapy. Postoperatively, the patient produced a satisfactory urine output, and the serum creatinine returned to normal limits. The patient was transferred to the general ward after three days.

Discussion

QT Syndrome (LQTS) is a disorder arising from cardiac ion channelopathies, affecting myocardial repolarization and results in a predisposition to ventricular tachydysrhythmia, in particular torsade de pointes. β-adrenergic blocking agents has been the mainstay of treatment of congenital LQTS since 1975. However, β-blockade is not completely protective and treated patients remain at risk of life-threatening episodes of torsade de pointes in the perioperative period.

The implications LQTS have on anaesthesia has been recently reviewed. Practical considerations for patients with LQTS should include immediate management of torsade de pointes, and avoidance of factors that increase the risk of precipitating this condition. The effects of various anaesthetic agents on the QT interval have been extensively studied. The choice of the specific anaesthetic agent for a patient with LQTS almost always requires individualization to the particular case confronted.

Several aspects in the management of this patient deserve attention, notably the use of pharmacological agents during the perioperative period. Propofol was chosen as the primary anaesthetic agent as recent review articles are unequivocal in supporting its use in patients with LQTS. Atracurium was chosen as the muscle relaxant as its metabolism is renal function independent, and is the most appropriate for patients with end stage renal failure.

We took precautions to ensure our patient did not receive excessive ventilating pressures. High peak airway pressures and long inspiratory to expiratory ratios should be avoided, as the Valsalva manoeuvre may increase the QT interval. Our patient had mild restrictive lung disease, but did not require high ventilating pressures to maintain adequate oxygenation.
Using sympathomimetic and anticholinergic agents

After induction of anaesthesia, our patient developed hypotension transiently before the surgical incision. This was probably because of the combined effects of β-blockade and cardiovascular depression by the anaesthetic agents. The use of α adrenergic agonists is discouraged in patients undergoing renal transplantation for fear of causing renal vasoconstriction. However, drugs with β-agonist activity may induce ventricular arrhythmias in patients with LQTS. As β-blockade was likely to be contributory to the hypotension, a small dose of ephedrine was given to reverse this effect. We continued to challenge the patient with intravenous fluids while keeping a close watch on the central venous pressure. If the blood pressure had remained low, an infusion of dopamine would have been started.

Anti-muscarinic drugs have been reported to prolong the QT interval in healthy subjects. As neostigmine is never given without atropine or glycopyrrolate, reversal of neuromuscular block in known LQTS patients is probably best avoided. After careful consideration, we decided that the risk of developing ventricular dysrhythmias would be minimal if reversal was carried out with a reduced dose of atropine and neostigmine. Our patient was well β-blocked, and surgical stimuli had not raised the heart rate beyond 70 beats per minute at any time during the operation. The neostigmine-atropine mixture was given in small doses under continuous ECG monitoring so that any cardiac rhythm abnormality could be immediately detected and appropriate intervention commenced.

In summary, patients with LQTS are at risk of developing malignant arrhythmias under anaesthesia. Such patients require careful preoperative assessment, meticulous intraoperative care and postoperative ICU management. Correction of any preoperative electrolyte abnormality and adequate β-blockade will help ensure an uneventful operative course.

References