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CASE REPORT

Anaesthesia for left thoracoscopic sympathectomy for refractory long QT syndrome: three case reports

Madhusudan Upadya[a]*, Julie CR Misquith[a] and Rammoorthi Rao[a]

[a]*Department of Anaesthesia, Kasturba Medical College, Manipal University, Mangalore, India*
**Corresponding author, email: madhusudan.upadya@manipal.edu*

Congenital long QT syndrome (LQTS) is a rare genetic disorder that has been associated with various genetic mutations including life-threatening cardiac arrhythmias and sudden death. Left thoracoscopic sympathectomy is an effective treatment for patients who are refractory to medical therapy or who need frequent epicardial internal cardio defibrillator intervention. The authors report three cases, one adult and two children, who underwent successful left thoracoscopic sympathectomy. All three patients remained clinically stable without arrhythmias through 3 months of follow-up. It is suggested in the literature that 77% of patients experienced immediate relief of symptoms. The results of this case report suggest that left thoracoscopic sympathectomy is a safe and effective approach for treating patients with LQTS.

Keywords: congenital condition, left cardiac sympathetic denervation, left thoracic sympathectomy, prolonged QT syndrome, video-assisted thoracoscopic surgery

Introduction

Long QT syndrome (LQTS) is caused by malfunction of cardiac ion channels impairing ventricular repolarisation.[1] This can be congenital or acquired. Age of presentation can be anywhere from in utero to adulthood. The stress of anaesthesia and surgery can predispose to the development of the polymorphic ventricular tachycardia torsade de pointes and some drugs used during anaesthesia can prolong the QT interval. This may either revert spontaneously back to sinus rhythm causing syncope or degenerate to ventricular fibrillation causing sudden death. There is evidence that the left stellate ganglion is dominant, and therefore it has been the focus of attention.[2–4] About 30% of the affected population fail to respond to medical treatment and remain symptomatic. Hence thoracoscopic denervation techniques have been tried.

Therapeutic options for patients with LQTS include β-blockade calcium channel blockers, implantable cardioverter defibrillators (ICDs) and left cardiac sympathetic denervation (LCSD). For patients who continue to have cardiac events while taking β-blockers, receiving frequent shocks from ICDs or are unable to tolerate pharmacotherapy, LCSD is another therapeutic option.[3,20]

Left cardiac sympathetic denervation (LCSD), a modification of the left stellectomy, is a proven alternative for high-risk patients.[2,3,5–14] The procedure involves an inferior stellate ganglionectomy and ganglionectomy of the upper 4 or 5 thoracic ganglia as shown in Figure 1. Newer techniques include using thoracoscopic sympathectomy while leaving the upper stellate ganglion intact to minimise the risk of causing Horner syndrome. In a retrospective study, Schwartz *et al.*[3] showed a dramatic > 90% decrease in the yearly number of cardiac events per patient following LCSD. Collura *et al.* published the Mayo Clinic experience with videoscopic LCSD in 20 paediatric and adult patients. Appropriate ICD shocks have been completely eliminated so far in 8 of 11 patients who presented with a history of ICD shocks or aborted cardiac arrest. One patient, who received 15 ICD shocks in the year prior to surgery, has had only one shock in the 36 months since her procedure. The two remaining patients have had less successful results with a minimal reduction in the frequency of cardiac events/ICD shocks postoperatively. The mean follow-up time was 16.6 ± 9.5 months.[20] Hofferberth *et al.*[23] in their study found no major perioperative complications. Longer-term follow-up was available in 22 of 24 patients at a median follow-up of 28 months (range, 4–131 months). Sixteen (73%) of the 22 patients experienced a marked reduction in their arrhythmia burden, with 12 (55%) becoming completely arrhythmia free after sympathectomy. Six (27%) of the patients were nonresponsive to treatment; each had persistent symptoms at follow-up and concluded that video-assisted thoracoscopic left cardiac sympathetic denervation can be safely and effectively performed in most patients with life-threatening ventricular arrhythmias. This minimally invasive procedure is a promising adjunctive therapeutic option that achieves a beneficial response in most symptomatic patients. These results support the inclusion of thoracoscopic cardiac sympathetic denervation within the treatment armamentarium in all patients with ventricular arrhythmias refractive to conventional medical therapy.

In the present article, we report our experience and review the literature to summarise outcomes of thoracoscopic sympathectomy in two children and a young adult. Videothoracoscopic surgery has become increasingly popular for cervical sympathectomy. This procedure is performed under general anaesthesia, with the use of double lumen tube (DLT) oro-tracheal intubation or endobronchial intubation with the patient in the right lateral decubitus position.

Case report

History and presentation

Three patients, two of whom were children — a male aged 14 years and a female aged 4 years — and an adult female aged 24 years were diagnosed with LQTS. The 14-year-old male and the 24-year-old female were siblings with autosomal dominant inheritance, dental abnormalities, clinodactyly, and bidirectional VT with long QT. The 4-year-old female child was a sporadic case who was diagnosed after the child repeatedly clenched her chest

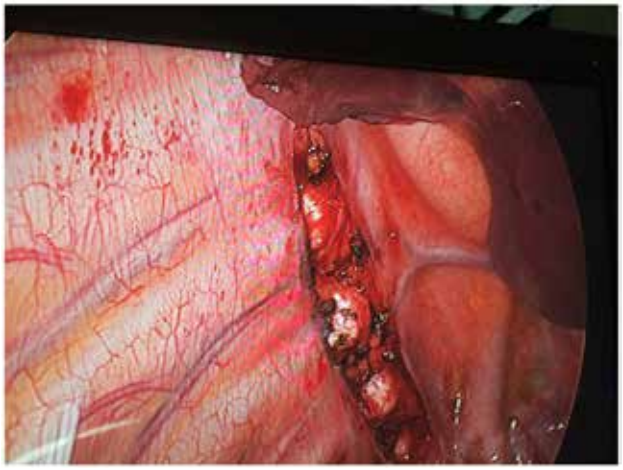

Figure 1. Video assisted thoracoscopic sympathectomy.

while crying or under stressful conditions and had history of seizures and sensorineural deafness. Several different genetic mutations have been implicated in causing LQTS, and most are inherited in an autosomal dominant pattern.[21] Rarely, patients present with a prolonged QT interval in association with congenital deafness, also known as Jervell and Lange-Nielsen syndrome (JLNS). JLNS is inherited in an autosomal recessive pattern.[22]

Table 1 gives a brief summary of the symptoms and outcome of the patients after thoracoscopic sympathectomy.

Anaesthetic management

Evaluation of patients was done, which included a detailed history, routine laboratory investigations and a complete cardiac and neurological workup. The patients were deemed fit for surgery by a combined team of specialists that included the cardiologist, cardiothoracic surgeon, paediatrician, anaesthetist and general surgeon who were involved in the management and workup of these patients. The procedure was explained to all patients and written informed consent was taken from the patient or, in the case of children, the patient's parents. Preoperative fasting guidelines were followed as per the age of the patients and medications were continued following the cardiologist's advice.

On arrival in the operating theatre, all three patients were secured with intravenous cannulas appropriate for their age. Intravenous fluid infusions were initiated. Monitors were connected which included five-lead electrocardiography (ECG), noninvasive blood pressure (NIBP) monitoring and pulse oximetry for oxygen saturation monitoring. All patients were premedicated with 2 µg/kg fentanyl and 0.02 mg/kg midazolam. They were preoxygenated with 100% oxygen for five minutes. Induction was done with propofol 2 mg/kg and neuromuscular blockade was achieved with rocuronium 1 mg/kg. After complete muscle relaxation, the 24-year-old female patient and 14-year-old male patient were intubated with 32 Fr DLT and the 4-year-old female patient was intubated with a 4.5 uncuffed tube which was initially fixed at 14 cm but once single lung ventilation was required was repositioned and pushed to the right bronchus fixing at 18 cm at the angle of the mouth.

Once intubated, end tidal carbon dioxide ($EtCO_2$), temperature and urine output monitoring was done. Invasive arterial blood pressure was monitored after securing an age-appropriate adequate size arterial cannula for each patient. Central venous cannulation was also established for all three patients. External defibrillator pads were secured in all patients. Once this was done, the patient was put in the right lateral position. Anaesthesia was maintained with N_2O, O_2, isoflurane 1% and intermittent doses of vecuronium as per requirement.

Surgery was performed via a left-sided video-assisted thoracoscopic approach as seen in Figure 2.

The lung was collapsed so that the sympathetic chain could be viewed. Gas insufflation of the thoracic cavity was not done in any of the three cases. All three surgeries were uneventful and vitals were stable throughout the procedure. Figure 3 shows the intraoperative vitals of the 14-year-old male child.

All three patients were extubated at the end of the procedure after neuromuscular blockade reversal with neostigmine 0.05 mg/kg and 0.01 mg/kg glycopyrrolate. None of the patients needed to be defibrillated or cardioverted during surgery. All patients were shifted to the high-dependency unit postoperatively where adequate monitoring was carried out.

Postoperatively, analgesia was provided with i.v. opioids. The 4-year-old female patient had one run of ventricular tachycardia three hours following surgery for which a lignocaine infusion was started and stopped after two days, following which no similar episodes recurred. The other two patients had no further arrhythmias.

All three patients had relief of symptoms and remained asymptomatic even after nine months of follow-up.

Schwartz *et al.*[13] report the largest data set available on LCSD, one of the three modalities of treatment for LQTS. The main findings were that LCSD significantly reduced the frequency and occurrence of both syncope and cardiac arrest; that among

Table 1: Summary of cases of LQTS treated with video assisted thoracoscopic sympathectomy

Age (years)/sex	Indications/ symptoms	Internal defibrillator *in situ*	Preoperative medications	Duration of symptoms	Syndrome	Complication
24/F	Palpitations, syncope, pseudo seizures, family history	No	Beta blocker	2 years	Anderson– Tawil*	No
14/M	Palpitations, breath-lessness, family history	No	Beta blocker	3 months	Anderson – Tawil*	No
4/F	Recurrent VT, pseudo seizures	No	Beta blocker, Ligno-caine infusion	1 month	Jervell and Lange-Nielsen[16, 22]**	One run of VT 3 hours after surgery

*Micrognathia, clinodactyly, ventricular arrhythmias, low-set ears.
**Prolonged QT, severe bilateral sensorineural deafness.

Figure 2. Patient position for thoracoscopic sympathectomy.

Figure 3. Intraoperative vitals of the patient during surgery.

patients with only syncope before surgery, the five-year postoperative cumulative probability of aborted cardiac arrest (ACA) or sudden death (SD) was 11% and actual survival at five years was 97%. The five-year survival for the entire group, including preoperative ACA, was 95%. In addition, LCSD was associated with significant shortening of the QTc.

Discussion

Hughes[14] reported the first endoscopic thoracic sympathectomy in 1942. In 1954, Kux[15] reported his experience of more than 1400 procedures. However, it has become more popular since the introduction of video endoscopic techniques into surgery in the 1980s.

In the most recent article by Costello *et al.*, following their retrospective review of paediatric patients with ion-channelopathies, the authors suggest that a prospective comparison needs to be done between surgical management and cardio-defibrillator use, which implies that we do not yet know enough to recommend LCSD as sole management strategy.[24]

Preoperatively, these patients may be on β-blocker therapy, may have implantable cardioverter defibrillators (ICD)[6] or have permanent pacemakers. Moss *et al.*[11] found a significant reduction in the mean rate of cardiac events with β-blocker therapy. Although β-blockade helps reduce the incidence of cardiac events, it is not entirely effective in preventing torsade de pointes and sudden death in LQTS.

Anaesthesia in patients with untreated LQTS carries a very high risk of intraoperative malignant ventricular arrhythmias,[17–19] which may prove refractory to treatment. The practical considerations of anaesthesia for patients with LQTS therefore include immediate management of torsade de pointes, and, in known cases, avoidance of factors that increase the risk of precipitating torsade de pointes. Inadequate or light plane of anaesthesia, hypertension, bradycardia, tachycardia, hypoxaemia and hypocapnia or hypercapnia must be avoided because they all potentially affect repolarisation of the cardiac myocyte and increase sympathetic tone, which can precipitate these arrhythmias. Patients should have adequate pain relief to avoid any stress response. Patients should be monitored throughout the perioperative period in a calm and quiet environment, and special attention should be paid to monitoring the QT interval.

There are various techniques of intubation for thoracoscopic sympathectomy. The commonly performed techniques are single-lumen endotracheal intubation with or without intrathoracic CO_2 insufflation. The advantage of this technique is familiarity with the technique and avoidance of the hazards of tube misplacement. One disadvantage is lung deflation, and surgical access may be less successful than with lung isolation techniques. The other is double-lumen endobronchial intubation with or without intrathoracic CO_2 insufflation. The advantage of this technique is good lung isolation if correctly positioned. The disadvantage is the risk of tube malpositioning.[20]

While anaesthetising these patients consideration must be given to avoid drugs that prolong QT interval, in order to decrease the chances of precipitating malignant arrhythmias and torsade de pointes.

A review article published in the British Journal of Anaesthesia in 2014 by Staikou *et al.*[25] discussed the effects of anaesthetic agents on the QT interval and torsadogenicity, particularly in patients with LQTS. All volatiles, especially isoflurane and desflurane, have been found to prolong QTc, while sevoflurane probably has no effect on TDR. Among the intravenous agents, propofol seems to be superior due to its minimal effects on QTc and TDR; in fact, a decrease in QTc has been demonstrated in many studies. Regarding opioids, fentanyl, alfentanil and remifentanil produced no effects on QTc, while sufentanil, at high doses, may induce QT prolongation. Succinylcholine, but not the non-depolarising neuromuscular blockers, produces QTc prolongation, which can be attenuated by opioids and beta blockers. Reversal of neuromuscular block with anticholinesterase–anticholinergic combinations has been associated with significant QTc prolongation, while such an

effect has not been demonstrated for sugammadex, even at high doses. Local anaesthetics probably have no intrinsic action on duration of repolarisation; nevertheless, an extensive subarachnoid sympathetic block may increase the duration of QTc. Conversely, thoracic epidural anaesthesia has been associated with a decrease in both QTc and TDR. Among adjuvants, midazolam seems to have no effect on QTc and TDR, while commonly used antiemetics, such as droperidol, domperidone and most 5-HT3 antagonists, produce significant QT prolongation. For patients with pre-existing repolarisation abnormalities, the effects of anaesthetic drugs and techniques on electrocardiographic conduction should be considered in the perioperative management.[25]

Conclusion

Long QT syndrome (LQTS) is being recognised with increasing frequency among the general population. It requires special attention and careful management in the perioperative period. The existing literature provides some insight into management of these patients. However, there are no definitive guidelines for anaesthetic management of LQTS. The results of this case report suggest that left thoracoscopic sympathectomy is a safe and effective approach for treating patients with LQTS.

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