

Anesthesia management in left cardiac sympathetic denervation with catecholaminergic polymorphic ventricular tachycardia

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ABSTRACT

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare familial cardiac arrhythmia characterized by RYR2 or CASQ2 gene mutation. This arrhythmia occurs in patients with a structurally normal heart. The syndrome may cause sudden cardiac arrest due to exercise or emotional stress related ventricular fibrillation. Most frequent clinical symptom is syncope. Currently, no definite anesthetic approach is available for the management of these patients. In this article, we report a case of CPVT and discuss anesthetic management.

Keywords: Anesthesia; catecholaminergic polymorphic ventricular tachycardia; left cardiac sympathetic denervation.

The catecholaminergic polymorphic ventricular tachycardia (CPVT) is potentially lethal, cardiac channelopathy. Treatment strategies include pharmacotherapy with β -blockers, implantable cardioverter defibrillators, and left cardiac sympathetic denervation (LCSD).

CASE REPORT

A 19-year-old female patient with a prior history of exercise-related syncope for two times was admitted to our clinic. She was five- and eight-year-old during her first and second syncopal episode, respectively. Further investigation demonstrated premature heartbeat, bidirectional ventricular tachycardia on 24-h Holter monitoring. The patient was diagnosed with CPVT accompanied by typical electrocardiogram (ECG) findings (Figure 1). Because of recurrent episodes of exercise-related syncope, she was treated with bisoprolol and verapamil. Although she was on medical treatment, she had cardiac arrest. Her mother applied basic life support and she was brought to the hospital. An implantable cardiac defibrillator (ICD) implantation was recommended with beta-blocker therapy. The first ICD was implanted; however, it was discharged in five months. Then, the second ICD was implanted. Nevertheless, it was discharged one month later. As a result, she was treated with a daily dose of bisoprolol 10 mg three times a day and flecainide 100 mg bid. The patient remained asymptomatic for five years. At five years, she had a repeated syncopal

episode as assessed by regular examinations. Following this episode, LCSD was recommended.

After written informed consent was obtained, she was pre-medicated with midazolam 3 mg (intramuscular) before the entrance to the operation room. Upon arrival to the operating room, an external defibrillator pad was placed and monitored through electrocardiogram (recording leads D2 and V5), non-invasive blood pressure and pulse oximetry.

Induction was performed with propofol 2 mg/kg, fentanyl 2 μ g/kg and rocuronium 0.5 mg/kg. Single-lung ventilation was achieved with a 35 F Portex endobronchial double lumen tube (Smiths Medical International Ltd., Keene, NH, USA). The patient was positioned on the operating table with her right side down. An epidural catheter was inserted into the thoracic epidural space 5-6th and then 3 mL 0.5% bupivacaine, 50 μ g fentanyl and 3 mL normal saline were injected through the catheter. Total volume was 7 mL. For the maintenance of anesthesia, propofol 2 mg/kg/h was applied.

The sympathetic ganglia were identified through the pleura, which were dissected to expose

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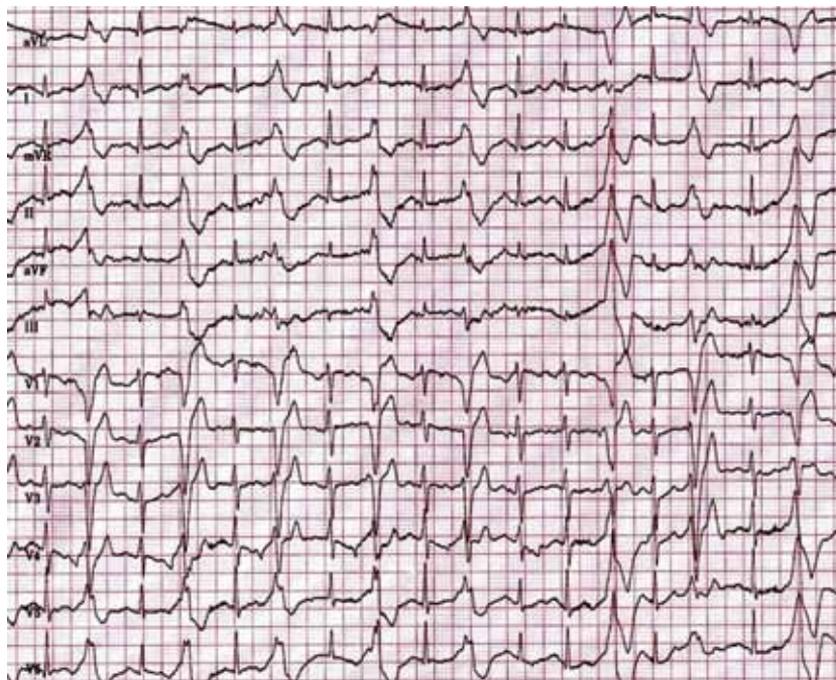


Figure 1. Electrocardiogram finding; catecholaminergic polymorphic ventricular tachycardia.

the left-sided sympathetic chain from T₄ to T₁ via videoscopic transthoracic approach. The patient received bupivacaine 15 mg and fentanyl 50 µg for intraoperative pain management through a thoracic epidural catheter. For nausea prophylaxis, the patient was given ondansetron 4 mg perioperatively and the patient received glycopyrrolate and neostigmine for the reversal of paralysis. The patient was, then, extubated in the operation room and monitored during 24 hours postoperatively.

DISCUSSION

Catecholaminergic polymorphic ventricular tachycardia is a rare malignant inherited arrhythmia syndrome.^[1] It usually presents in childhood or young adulthood with a history of physical or emotional stress-induced syncope or cardiac arrest.^[1] Arrhythmia is characterized by bidirectional or polymorphic ventricular tachycardia in patients with a structurally normal heart.^[2-4]

Catecholaminergic polymorphic ventricular tachycardia is associated with RYR2 and CASQ2 gene mutation. The inheritance pattern is autosomal-dominant and autosomal-recessive, respectively.^[4,5] These genes are related to calcium channels.^[2] A family

history of syncope or cardiac death is positive in approximately 35% of patients with CPVT.^[3]

Catecholaminergic polymorphic ventricular tachycardia was first described in 1975 by Reid et al.^[3,4,6] Diagnosis can be challenging. As ECG usually produces normal results in asymptomatic patients, ECG is not specific.^[2,4] The diagnosis is based on ECG findings during exercise. During exercise, typical ECG finding is bidirectional tachycardia.^[1,4,7,8]

The medical treatment of CPVT is based on beta-blockers.^[1] Syncopal attacks can be controlled by beta-adrenergic blockers. Nadolol is the preferred agent thanks to its prolonged half-life. Therefore, beta-blockers are effective for the acute phase and they are the first-line of treatment.^[1,3,7,9,10] Other therapeutic options for patient with CPVT include calcium channel blockers and/or flecainide. However, an ICD should be planted in patients with recurrent arrhythmias or cardiac arrest episodes.^[1,3,4]

In this case, our patient presented due to exercise-related syncope with a prior history of cardiac arrest. She was diagnosed as CPVT based on her medical history and typical ECG findings. In the beginning, she was treated with a single beta-blocker agent, however, the patient presented again with cardiac arrest

while she was on medical treatment. An automatic ICD was implanted. The first ICD depleted in five months. As a result, the second ICD was implanted; however, it depleted one month later. The depletion was caused by inappropriate multiple ICD shocks. During a five-year follow-up, the patient remained asymptomatic. At five-years, she had a repeated syncopal episode as assessed by regular examinations. Therefore, LCSD was recommended. An ICD was recommended for CPVT patients who have cardiac arrest or syncopal attack despite receiving maximum dosage of beta-blocker therapy.^[11,12] However, when the symptoms persist despite beta blockade, calcium channel blockers, ICD, and LCSD are effective alternative treatments.^[1,11,13]

Left cardiac sympathetic denervation is described in 1971.^[14,15] Surgically LCSD involves resection of the lower half of the left stellate ganglion and the left sided sympathetic chain. Left cardiac sympathetic denervation has been used as an effective option for patient with CPVT.^[14,16] However, clinical experience of LCSD in CPVT patients is limited. In addition, there are few reports on the anesthetic procedure of patients with CPVT. The goal of the treatment is to manage the adrenergic stimulation, since adrenergic stimulation may provoke arrhythmias.^[13]

Operative management of these patients is often challenging. There is a high risk of life-threatening arrhythmia and sudden cardiac arrest. Anesthetic management should be planned carefully.^[13,17,18] Careful planning and monitorization are critical to ensure a safe operation. The operation room should be appropriate for the induction. Hyper-hypotension, bradycardia, tachycardia, hypothermia, hyper-hypocapnia, hypoxemia should be also controlled, as these conditions can effect cardiac activity.^[11,19]

Serum electrolytes should be measured. An external defibrillator and blood pressure monitoring should also be present in the operating room. Premedication is necessary to avoid increased sympathetic activity.^[19-21]

For premedication, midazolom is used in our patient. Propofol was used in our patient for the induction. Propofol has been used as induction and maintenance agent in patients with CPVT to avoid complications.^[1] Rocuronium was administered to our patient for muscle relaxation. Rocuronium can be safely administered to this group of patients.^[1] In this present case, video-assisted thoracoscopic cardiac denervation, a minimally invasive procedure, was made.

Bupivacaine and fentanyl were used for the pain management and to maintain medical sympathetic denervation. Then, a thoracal epidural catheter was placed. For the management of postoperative nausea and vomiting, ondansetron was initiated. No significant complication and adverse effects were noted during the operation.

In conclusion, anesthetic management of the patient with CPVT requires careful monitoring of cardiac parameter, understanding of risks, and good management of postoperative pain control.

Declaration of conflicting interests

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