Don’t stress – managing a patient with CPVT

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Introduction

Background
• Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare disorder with <100 reported cases (1-3)
• It is characterized by lethal tachyarrhythmias triggered by stress or exercise (1-3)
• Typically, it is discovered when a child presents with a syncopal episode that is further evaluated (1-3)
• Catecholamines induce bi-ventricular tachycardia that can deteriorate to cardiac arrest (1)
• Perioperative management of these patients can be very challenging

Case History

History of Present Illness
• A 15 year old male with an ICD and CPVT presented for knee arthroscopy, nasal septoplasty, and turbinate reduction
• The patient was adopted at 6 months old
• However, family history was known and concerning
  • 10 year old biological sister had sudden cardiac death
  • 12 year old brother had a “heart attack” climbing steps

Case History (continued)

Preoperative Course
• The patient received flecainide and nadolol (home medications) prior to hospital arrival; refused oral midazolam as premedication

Intraoperative Course
• In the operating room prior to induction, defibrillator pads in addition to ASA standard monitors were placed
• The “pacemaker team” was present to deactivate the patient’s ICD
• Inhalational induction with oxygen/nitrous oxide/sevoflurane was uneventful
• An intravenous line and arterial line were placed
• The patient was given a bolus of fentanyl 50mcg prior to direct laryngoscopy, and the trachea was intubated atraumatically
• General anesthesia was maintained with sevoflurane in addition to infusions of dexmedetomidine and remifentanil
• During the case, the patient’s heart rate ranged from 60 – 70 beats per minute while blood pressure ranged from 110/70 – 70/40 mmHg
• Hypotension was treated with judicious fluid boluses
• At the end of the procedure, the patient was awakened and successfully extubated
• Prior to removing the defibrillator pads, his ICD was reactivated and interrogated
• The patient was then transported to the PACU on supplemental oxygen

Discussion
• The goal for management of patients with CPVT is to minimize adrenergic stimulation that may potentially result in a fatal arrhythmia
• Therefore, it is important to continue beta blocker therapy perioperatively
• Intraoperative tachycardia should be addressed quickly
• Issues with hypotension can be treated with fluid, or an agent with pure alpha agonist activity
• Hypotension secondary to bradycardia can be treated with atropine, pacing, or glucagon (if beta blocker overdose is suspected) (1-3)

Case History (continued)

Intraoperative Course
• The patient received flecainide and nadolol (home medications) prior to hospital arrival; refused oral midazolam as premedication

Postoperative Course
• His postoperative course was uneventful
• After discussion with the cardiac, orthopedic, and otolaryngology teams, the patient was discharged home

References