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Intraoperative diagnosis of hypertrophic cardiomiopathy

I. Bekiroglu[1], R. Heng[1], J. D. Tobias[1,2]

[1]Department of Anesthesiology & Pain Medicine, Nationwide Children's Hospital, Columbus, Ohio, USA
[2]Department of Anesthesiology & Pain Medicine, The Ohio State University College of Medicine, Columbus, Ohio, USA

Corresponding author: I Bekiroglu. Department of Anesthesiology & Pain Medicine, Nationwide Children's Hospital, Columbus, Ohio, USA. Email: Ismail.Bekiroglu@Nationwidechildrens.org

Keypoints

1. Hypertrophic cardiomyopathy (HCM) is the most common genetic disease of the heart with autosomal dominant inheritance leading to mutation in one of the genes encoding for proteins in the sarcomere of the myocardium thereby leading to abnormal myocyte growth with hypertrophy.
2. Although generally asymptomatic in the early phases, the diagnosis may be made when the patient or a close relative manifests clinical signs and symptoms such as chest pain, arrhythmias or sudden cardiac death.
3. Echocardiographic analysis demonstrates left ventricle wall thickening, predominantly in the area of the anterior ventricular septum) in the majority of patients with HCM.
4. The goals of perioperative care for patients with HCM include limiting the dynamic consequences of HCM by controlling heart rate with avoidance of tachycardia, controlling arrhythmias with maintenance of a normal sinus rhythm, obtaining a normal to slightly decreased myocardial contractility, while maintaining baseline preload and afterload.

Abstract

Hypertrophic cardiomyopathy (HCM) is the most common genetic disease of the heart. Autosomal dominant inheritance leading to mutation in one of the numerous genes encoding for proteins in the myocardial sarcomere leads to abnormal myocyte growth with hypertrophy. As the process generally remains asymptomatic during the early phases, diagnosis is frequently made when the patient or a close relative manifests clinical signs and symptoms such as chest pain, arrhythmias or sudden cardiac death. We present a 16-year-old adolescent who presented for anesthetic care during repair of a fractured fibula. Intraoperatively, electrocardiographic (ECG) abnormalities were noted leading to the diagnosis of HCM. The differential diagnosis of intraoperative ECG changes is discussed and a diagnostic algorithm presented.

Keywords

hypertrophic cardiomyopathy; ST segment changes; cardiac arrest; pediatric anesthesia

Introduction

Hypertrophic cardiomyopathy (HCM) is a congenital autosomal dominant disorder of cardiac myocytes that may remain relatively asymptomatic during the initial phases of progression. An abnormal myocyte growth pattern is seen in HCM due to a mutation in one of the genes encoding proteins in the sarcomere of the myocardium.[1,2] HCM has an incidence of approximately 0.2%, representing the most common genetic disease of heart.[2] It

remains a common etiology of sudden cardiac death in young adults and athletes.[3,4] Echocardiographic analysis demonstrates left ventricle (LV) wall thickening (predominantly in anterior ventricular septum) in the majority of patients with HCM. As the process can remain asymptomatic, diagnosis is frequently made when the patient or a close relative manifests clinical signs and symptoms such as chest pain, arrhythmias or sudden cardiac death.[5,6] We present a 16-year-old adolescent who presented for anesthetic care during repair of a fractured fibula. Intraoperatively, electrocardiographic (ECG) abnormalities were noted leading to the diagnosis of HCM. The differential diagnosis of intraoperative ECG changes is discussed and a diagnostic algorithm presented.

Case report

Presentation of this case report was in accordance with the Institutional Review Board at Nationwide Children's Hospital (Columbus, Ohio). Written consent for anesthetic care and publication was obtained from a parent. A 16-year-old, 86.3 kg adolescent male with no remarkable past medical history, presented for surgical repair of a right ankle fracture. He had no past surgical or anesthesia history. There was no history of tobacco, alcohol, or recreational drug use. He was physically active with no restrictions, playing high school football. The preoperative evaluation and physical examination were unremarkable. The patient was scheduled for an open reduction and internal fixation of the fibula. He was held *nil per os* for 6 hours and transported to the operating room where standard American Society of Anesthesiologists' monitors were placed. Anesthesia was induced with propofol (350 mg), fentanyl (100 µg), and lidocaine (60 mg). A size 4 laryngeal mask airway was placed on the first attempt. Following induction, a single dose of ephedrine (5 mg) was administered for a transient decrease in blood pressure. Anesthesia was maintained with sevoflurane (expired concentration 2-4%) in air and oxygen. Cefazolin (2 grams) was administered for surgical antibiotic prophylaxis. Shortly after anesthetic induction, it was noted that the 3-lead intraoperative ECG demonstrated ST-T wave changes. Monitoring was changed to a 5-lead ECG which showed ST segment depression and ST-T wave changes (figure 1).

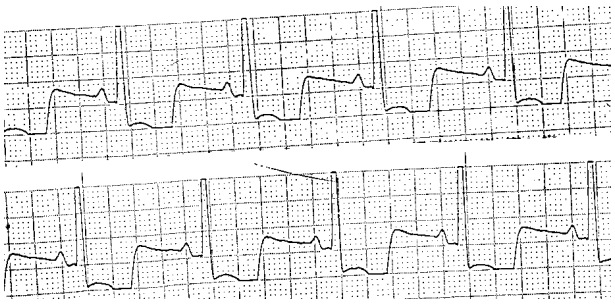

Figure 1. Intraoperative 5-lead continuous ECG monitor showing ST segment depression with ST-T wave changes and high voltage.

Repeat cardiac auscultation and examination revealed a normal sinus rhythm with no ectopy, a normal S1 and S2, and no murmur. Maintenance anesthesia was continued with inhaled sevoflurane in 100% oxygen. The surgical procedure lasted approximately 75-80 minutes and was completed without complications. Intraoperative fluids included lactated Ringer's (1000 mL). Postoperative analgesia was provide by hydromorphone (0.5 mg) and ketorolac (30 mg). Prophylaxis against postoperative nausea and vomiting included dexamethasone (8 mg) and ondansetron (4 mg). The patient was awake and conversive in the post-anesthesia care unit (PACU). He denied chest pain or other cardiac symptoms. Postoperative cardiology consultation was obtained. A 12-lead ECG and echocardiogram were obtained in the PACU. The ECG demonstrated normal sinus rhythm, QT prolongation with T-wave inversion in inferior, lateral, and anterior leads (figure 2).

The echocardiogram showed mild concentric left ventricular hypertrophy, compatible with HCM, long-standing hypertension, or athlete's heart. The patient was discharged home with a Holter monitor and scheduled for follow-up with cardiology. At his follow-up cardiology visit, an ECG demonstrated sinus rhythm with persistent diffuse T-wave inversion (II, III, aVF, V3-V6) and accompanying ST segment depression, and prolonged QTc at 465 ms.

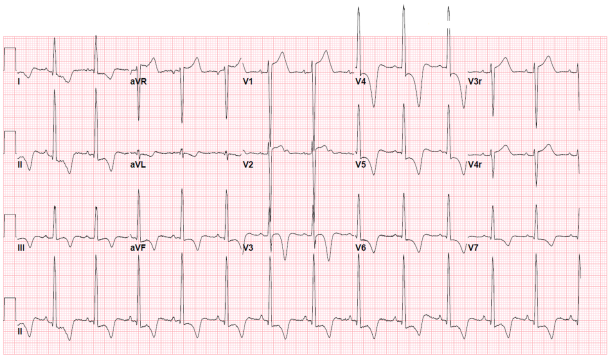

Figure 2. Postoperative ECG showing normal sinus rhythm, QT prolongation with T-wave inversion in inferior, lateral, and anterior leads.

The differential diagnosis per pediatric cardiology includes athlete's heart, LVH secondary to hypertension, and HCM. Features that would argue against the hypertrophy being secondary to conditioning (athlete's heart) include the small ventricular cavity and ECG abnormalities. However, the plan is to continue monitoring the progression or resolution of the hypertrophy with cessation of exercise to evaluate the effect of deconditioning on his heart. Follow-up 2 months after surgery with a cardiac MRI demonstrated concentric LV hypertrophy with no myocardial fibrosis. Ambulatory blood pressure monitoring was normal without evidence of hypertension.

Discussion

The current standard of care for intraoperative patient monitoring includes pulse oximetry, non-invasive blood pressure, temperature, continuous ECG, and capnography.[7] Additional equipment monitoring includes a low pressure circuit disconnection monitor and an inspired oxygen analyzer. Such monitoring has significantly decreased the incidence of perioperative morbidity and mortality. The Australian Incident Monitoring Study (AIMS) demonstrated that there is a preventable factor that can be identified in more than half of intraoperative cardiac arrests.[8] Of 1256 critical intraoperative events, which occurred in association with general anesthesia, 52% were "monitor detected". The majority were detected by either pulse oximetry (27%) or capnography (24%).[8,9] The remaining events were detected by ECG (19%), a blood pressure monitoring device (12%), a low pressure circuit disconnection alarm (8%), or an oxygen analyzer (4%). In the pediatric population, the primary causes of intraoperative cardiac arrest relate to the cardiovascular system including either hypotension from hemorrhage during major surgical procedures or hyperkalemia related to the rapid administration of blood and blood products.[10]

Continuous ECG monitoring is generally used intraoperatively for the identification and diagnosis of arrhythmias or coronary ischemia. For the pediatric population, a 3-lead ECG is generally used with a right arm electrode (white lead) placed just below the clavicle near the right shoulder, the left arm electrode (black lead) placed similarly below the left clavicle), and the left leg (red electrode) placed on the left lower chest. Using these 3 electrodes, leads I, II, and III can be displayed which can identify P -wave morphology and rhythm. Unlike the adult population, intraoperative cardiac arrest related to coronary ischemia is rare and so precordial leads or a 5-lead ECG is rarely used to evaluate V1-6 and identify ST-T wave changes suggestive of ischemia.[11,12]

In our patient, the diagnosis of HCM was first suggested by ECG changes that were noted intraoperatively. As these changes were suggestive of ischemia including ST segment depression, a 5-lead ECG was placed intraoperatively to further identify the ECG morphology. Non-pathologic ST-T wave changes may be seen with J-point depression, early repolarization, or due to sympathetic stimulation related to surgical stimulation or an inadequate depth of anesthesia. However, they may be a sign of significant cardiovascular disease including myocardial ischemia or infarction, left or right ventricular hypertrophy with strain, medication effects, inflammatory processes such as myocarditis and pericarditis, or electrolyte and acid-base disturbances (hypokalemia, hypernatremia, alkalosis).[13] ECG changes suggestive of HCM include abnormal Q waves, negative T waves, or tall R waves in the left precordial leads.[14] Based on the ECG, the clinical scenario, and the patient's age, a presumptive

diagnosis of HCM was entertained. The decision made to continue anesthetic care with attention to the determinants of myocardial oxygen delivery and demand.

Perioperative management of patients with HCM patients for any type of surgical procedure carries an increased risk of morbidity and mortality related to the propensity for arrhythmias and myocardial ischemia.[15-17] Intraoperatively, myocardial function may be impacted by alterations in preload, afterload, and myocardial contractility related to surgical stimulation, anesthetic agents, and changes in intravascular volume. Although there are limited data for the pediatric-aged patient, adult HCM patients have a significant risk of having at least one adverse cardiac event during non-cardiac surgery.[18,19] Predictors of adverse perioperative cardiac events (congestive heart failure, myocardial ischemia and dysrhythmias) include the magnitude of the surgical procedure and the duration of surgery. The goals of perioperative care for patients with HCM are to limit the dynamic consequences of HCM by controlling heart rate with avoidance of tachycardia, maintaining a normal sinus rhythm, obtaining a normal to slightly decreased myocardial contractility, while maintaining baseline preload and afterload. Control of these hemodynamic factors limits the increase in the outflow tract gradient thereby maintaining cardiac output. An additional concern perioperatively with HCM is the potential for an imbalance of the myocardial oxygen delivery-demand ratio leading to coronary ischemia. Histologically, the thickened walls of HCM may result in luminal narrowing of the intramural coronary arteries.[20] As such, diastolic blood pressure should be maintained and heart rate controlled.

Despite its prevalence, there are a limited number of previous reports regarding the perioperative care of patients with HCM.[21] The reported literature has demonstrated success with the administration of various intravenous induction agents including midazolam, propofol, thiopentone, and etomidate. Maintenance anesthesia has included opioids (fentanyl, morphine, remifentanil) in combination with halothane, sevoflurane or isoflurane, and occasionally supplementation with dexmedetomidine. In an effort to avoid or limit the need for general anesthesia, regional anesthesia including peripheral and neuraxial blockade has been reported especially in the parturient. These reports demonstrate the basic tenets of caring for such patients including: 1) maintenance of sinus rhythm with control of heart rate; 2) reduction in sympathetic activity to reduce chronotropy and inotropy; 3) maintenance of left ventricular filling pressures (preload); 4) control of variables responsible for myocardial oxygen delivery and demand; 5) maintenance of normal myocardial contractility; and 6) preservation of normal pulmonary vascular resistance.

In summary, we present the intraoperative diagnosis HCM, based on changes noted in the intraoperative ECG. Although uncommon in the general population, HCM is the most common genetic condition of the myocardium. Mutation in a gene encoding for proteins in the myocardial sarcomere leads to abnormal myocyte growth with hypertrophy. The process generally remains asymptomatic during the early phases and the diagnosis may be made only when the patient or a close relative manifests clinical signs and symptoms such as chest pain, arrhythmias or sudden cardiac death. Early diagnosis may result in control of potentially lethal complications including arrhythmias or ischemia. The goals of perioperative care for patients with HCM include limiting the dynamic consequences of HCM by controlling heart rate with avoidance of tachycardia, maintaining a normal sinus rhythm, obtaining a normal to slightly decreased myocardial contractility, while maintaining baseline preload and afterload.

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