Anesthesia in a Child with Newly Diagnosed Hypertrophic Cardiomyopathy for Placement of Implantable Cardioverter Defibrillator

Rajnish Kumar, Bibha Kumari
Department of Anaesthesiology, IGIMS, Patna, Bihar, India

Abstract

Hypertrophic cardiomyopathy (HCM) is a genetic myocardial disease usually characterized by asymmetric ventricular septal hypertrophy. HCM is an important cause of sudden cardiac death in adolescents and young adults. We are presenting a case report, ten years boy came in emergency with sudden loss of consciousness (witness cardiac arrest). Child was revived after cardiopulmonary resuscitation and send to coronary care unit. Echocardiography findings were suggestive of HCM. There was history of sudden death of her mother and maternal uncle. After stabilization ICD was implanted under total intravenous anesthesia. Post procedure his hospital stay was uneventful.

Keywords: Hypertrophic cardiomyopathy, implantable cardioverter defibrillator device, total intravenous anesthesia

Introduction

Hypertrophic cardiomyopathy (HCM) is a genetic disease of myocardium disease usually characterized by asymmetric ventricular septal hypertrophy. HCM is a primary cardiac disorder that results from known or suspected genetic defects in sarcomeric proteins of the cardiac myocyte. HCM is subdivided into obstructive and nonobstructive types, depending upon the presence of left ventricular outflow tract obstruction. Incidence of HCM is common in adults and is lower in children. HCM is characterized by unexplained left ventricular hypertrophy and typically involves the interventricular septum. Sudden death is the most common cause of death in children with HCM.[1] We report the initial resuscitation and subsequent management with implantable cardioverter defibrillator (ICD) implantation in a child with newly diagnosed HCM, which was presented as cardiac arrest.

Case Report

A 10-year-old boy presented with cardiac arrest in the emergency room. Return of spontaneous circulation was within one minute of CPR. Within 2 min, he started responding and his heart rate was 68 beats/min, blood pressure was 108/70 mm Hg, and SpO₂ was 100%. He was shifted to coronary care unit. On echocardiography, the findings were suggestive of HCM (nonobstructive), asymmetrical septal hypertrophy, and mild mitral regurgitation with normal left ventricular functions [Figure 1]. He was started on tablet metoprolol 12.5 mg once daily. There was a history of sudden death of his mother and maternal uncle. He was planned for automated ICD implantation for secondary prevention of sudden cardiac death. In preanesthetic evaluation, no previous systemic illness and laboratory parameters were within normal limits. Informed parental consent for anesthesia was obtained.

In cardiac catheterization laboratory, all standard monitors were attached and intravenous line patency was checked. His heart rate was 70 beats/min, noninvasive blood pressure was 110/70 mm Hg, and SpO₂ was 100%. He received intravenous midazolam 1 mg, fentanyl 50 µg along with oxygen by nasal prong. Infiltration of the skin and subcutaneous tissue at the implant site was done with 15 ml of 1% lignocaine. The patient complains of pain during pocket formation for which he also received intravenous ketamine 0.5 mg/kg twice. Hemodynamic

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parameters were normal during 1 h of ICD implantation [Figure 2]. He shifted to coronary care unit for monitoring and rest hospital stay was uneventful.

**Discussion**

HCM with witness cardiac arrest is a very rare presentation. HCM is an autosomal dominant pattern of inheritance and is caused by mutations in the genes that encode proteins of the myocardial sarcomere. HCM is a common genetic disorder in adults (1 case per 500 populations). Patients with HCM may be asymptomatic. Symptoms if present can include shortness of breath, chest pain, presyncope, syncope, and palpitations. The most devastating complication may present as sudden cardiac arrest. Symptoms in children with HCM include dyspnea, chest pain, fainting attack, and arrhythmias. Anesthesia for HCM is challenging because of the risk of perioperative arrhythmia and myocardial ischemia in the presence of tachycardia and low diastolic pressure.[3] The goals of perioperative care for patients with HCM are to limit the dynamic consequences of HCM with avoidance of tachycardia, maintenance of normal sinus rhythm, normal to slightly decreased myocardial contractility, while maintaining baseline preload and afterload. The volatile anesthetic agents such as isoflurane may increase the heart rate; however, sevoflurane is well tolerated in all but the most severe forms of HCM.[1] Propofol is also not a good choice because it decreases preload and systemic vascular resistance. In this case, we have used low dose ketamine during the procedure because it has some advantage of preserving spontaneous breathing, diastolic arterial pressure and providing profound analgesia.[3] Ketamine, on the other hand, may increase salivation which can be troublesome and may produce laryngospasm. Kansara *et al.* have successfully given general anesthesia for the placement of an implantable cardioverter-defibrillator in an infant with congenital long QT syndrome: Anesthetic considerations. *Ann Card Anaesth* 2011;14:122-6.

**Conclusion**

Patients with HCM are at significant risk of cardiac complications during anesthesia. Anesthetic management should be based on the maintenance of preload, afterload, adequate analgesia, and avoidance any sympathetic activity during procedure.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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