Hypertrophic cardiomyopathy surgery: Perioperative anesthetic management with two different and combined techniques

ABSTRACT

Hypertrophic cardiomyopathy (HOCM) is the most common genetic heart disorder and the most common cause of sudden cardiac death among young population and a major cause of disability for patients of any age. An extended transaortic septal myectomy is the definitive treatment. It is very important to have a good knowledge of the characteristic pathophysiology of the disease in order to optimize intraoperative treatment of these patients. We present a case of a 68-year old woman who underwent hypertrophic elective cardiomyopathy surgery. Anesthetic management is crucial to guarantee maximum safety, since HOCM has the capacity to produce hemodynamic events of such severity that put patient’s life at risk. The use and combination of intraoperative transesophageal echocardiography (TEE) and direct measurement of the left ventricular outflow tract gradient provides vital information to ensure successful surgical outcome in patients with HOCM.

Key words: Cardiomyopathy hypertrophic; cardiac surgery; hemodynamics; transesophageal echocardiography

Introduction

Hypertrophic cardiomyopathy (HOCM) is the most common cardiovascular genetic disorder. HOCM has a prevalence of 0.2% and is the most common cause of sudden cardiac death among young people and provokes substantial disability in patients of all ages.

The symptoms of HOCM include dyspnea, palpitations, arrhythmia, syncope, heart failure, stroke, and sudden death. Electrocardiographic (ECG) findings in these patients reveal abnormalities such as left axis deviation, increased QRS voltage, left ventricular hypertrophy (LVH), and ST-segment alterations. Transthoracic echocardiography (TTE) is the most widely used tool to screen for the disease and diagnose patients, while magnetic resonance imaging (MRI) scans are reserved for cases of uncertain TTE findings. Beta blockers are the first-line drug therapy for these patients due to their negative inotropic and chronotropic effects; placement of a pacemaker and implantable cardioverter-defibrillator use are reserved for cases that are refractory to medical treatment. Surgery is recommended for patients presenting symptoms of advanced heart failure in spite of maximum drug therapy and with a peak left ventricular outflow tract (LVOT) gradient...
on TTE of >50 mmHg.\textsuperscript{[6]} Transaortic septal myectomy is the surgical approach of choice for most patients, as it is associated with an improvement in symptoms and greater long-term survival\textsuperscript{[7,8]} and the rate of procedural mortality in care facilities experienced in the technique is <1%.\textsuperscript{[9]}

Attending anesthesiologists must be familiar with the characteristic physiopathology of this disease in order to optimize perioperative treatment, as HOCM has the potential to cause severe and life-threatening hemodynamic instability. Additionally, as anesthesiologists may come into contact with cases of undiagnosed HOCM, they must be prepared to anticipate the hemodynamic changes and cardiovascular instability sometimes associated with the disorder, especially sudden hypotension that fails to improve or even worsens with inotropic drug treatment. In such cases, HOCM should not be ruled out until a transesophageal echocardiography (TEE) has been conducted.\textsuperscript{[1]}

**Case Report**

A 68-year-old woman (165 cm; 64 kg) with no known allergies to medication and a past history significant for arterial hypertension, chronic bronchitis, ischemic heart disease (stent in the circumflex artery in 2017) and obstructive HOCM was scheduled to undergo an elective extended transaortic septal myectomy. Her usual treatment consisted of bisoprolol, torasemide, disopyramide, pantoprazol, acetylsalicylic acid and bronchodilators. She was referred to our hospital from another center following a diagnosis of HOCM manifesting clinically as intense dyspnea was referred to our hospital from another center following a diagnosis of HOCM manifesting clinically as intense dyspnea was referred to our hospital from another center following a diagnosis of HOCM manifesting clinically as intense dyspnea. She was then given intravenous midazolam (2 mg), propofol (60 mg), fentanyl (150 µg) and rocuronium (50 mg). The trachea was intubated with 7.5 cuffed tube. Intraoperative TEE revealed severe hypertrophy (20 mm) of the intermuscular septum and LVOT obstruction with SAM associated with the mitral valve [Figure 1] as well as a peak gradient between the left ventricle (LV) and the aorta (Ao) of 100 mmHg. Following the sternotomy, another measurement was taken from the surgical field using two catheters placed directly in the LV and the Ao respectively, revealing a gradient of 116 mmHg. Anesthesia was maintained with sevoflurane, fentanyl and rocuronium. As the patient displayed a tendency toward hypotension, she required single-dose boluses of phenylephrine and continuous perfusion of noradrenaline at a rate of 0.04 mcg/kg/min to achieve an average arterial pressure of >60 mmHg until cardiopulmonary bypass was performed. An extended transaortic septal myectomy was conducted without incident. When CPB was discontinued, the patient presented ventricular fibrillation, which resolved with amiodarone (300 mg) and 3 automated internal defibrillator shocks of 10 J. At this time, the patient entered sinus rhythm at 40 bpm and atrial and ventricular pacemaker electrodes were placed at a rate of 80 bpm. Postoperative TEE evidenced resolution of hypertrophy in the LVOT [Figure 2], an LV-Ao gradient of 8 mmHg, mild MI, absence of iatrogenic ventricular septal defects and normal biventricular function. LV-Ao gradient measurement from the surgical field was 7 mmHg [Figure 3], which closely resembled the value obtained using TEE and thereby confirmed that the intervention had been done successfully. Given the absence of complications associated with the surgical procedure CPB was terminated and the patient was shifted to the intensive care unit with stable hemodynamics. The trachea was extubated after 6 h and shifted to hospitalization ward after 24 h.

**Discussion**

HOCM is an autosomal dominant hereditary cardiac disorder characterized by hypertrophy of the LV. The clinical presentation of HOCM varies depending on the location and extent of the disease. Associated cardiac anomalies include LVOT obstruction, MI, diastolic dysfunction, and cardiac ischemia. Some of these abnormalities are dynamic in nature

**Figure 1**

Near-infrared spectroscopy (INVOS™ 5100C Cerebral/Somatic Oximeter, Covidien\textsuperscript{®}), body temperature using an esophageal thermometer, central venous pressure and hourly urine output. When the patient arrived in the operating room, her arterial pressure was 145/69 mmHg and she had a heart rate of 72 bpm and oxygen saturation of 96%.

Upon entry to the operating room, the patient was monitored for the following: arterial blood pressure by invasive monitoring through the left radial artery, ECG with ST-segment analysis, oxygen saturation through pulse oximetry, capnography, Bispectral Index (BIS™) monitoring to measure the hypnotic effects of anesthesia,
and exacerbate with decreases in preload and postload, with tachycardia and with an increase in contractility.\[^{10}\]

There are no published guidelines for intraoperative anesthetic management of patients with HOCM and they must be in optimal condition prior to surgery, particularly in terms of blood volume before sedation as this reduces the risk of vasodilatation-associated LVOT obstruction triggered by anesthetic drugs. Preoperative drug therapy must be maintained throughout the perioperative period, especially Beta-blockers, and proper anxiolysis is also beneficial in reducing catecholamine levels in the bloodstream.\[^{11}\]

Invasive monitoring of arterial pressure before induction of anesthesia is essential in detecting hypotensive events, to which these patients respond poorly. Propofol or thiopental sodium may be used, though dose levels of these drugs must be measured carefully due to the myocardial depressant effects and decreased postload they cause. Etomidate is also an attractive option due to the low impact it has on the cardiovascular system.\[^{4}\]

Any decrease in arterial pressure must be treated with an appropriate increase in intravascular volume and in postload; \(\alpha\)-adrenergic agonists such as phenylephrine and noradrenaline are the agents of choice for this due to their ability to increase systemic vascular resistance and diminish LVOT obstruction.\[^{14}\]

It is also important to avoid the use of drugs that have \(\beta\)-adrenergic effects (i.e., dobutamine, dopamine, adrenaline, isoproterenol), as these worsen LVOT obstruction due to their positive inotropic and chronotropic effects.\[^{4}\]

It is also of utmost importance to maintain sinus rhythm, as HOCM reduces LV compliance, thus increasing reliance on atrial contraction to maintain cardiac output. This makes immediate cardioversion necessary in cases of sudden atrial fibrillation that destabilize the patient.\[^{2}\] Sevoflurane is a mild myocardial depressant that produces a more modest decrease in systemic vascular resistance and blood pressure than isoflurane or desflurane and only a slight increase in heart rate or none at all. Mechanical ventilation with high tidal volume and high positive end-expiratory pressure is harmful, as it reduces preload and causes greater LVOT obstruction.\[^{11}\]

When performed by experienced professional TEE is an essential tool for intraoperative monitoring of these patients as it determines the cause of hemodynamic instability (e.g., hypovolemia, LVOT obstruction, SAM or systolic/diastolic dysfunction) and thus makes optimal management possible.\[^{12}\] It is also crucial to confirm that the surgery has been carried out successfully by verifying reduction of the left ventricular outflow tract gradient, improvement of SAM and absence of iatrogenic ventricular septal defects. Unfortunately, in some cases it may be technically difficult to measure LVOT gradient by TEE because
the Doppler beam must be aligned parallel to the maximal velocity vector, so in our institution we also perform direct intraoperative measurement of pressures in the LV and the Ao because it provides important hemodynamic data in addition to the intraoperative TEE findings and ensure favorable surgical outcome.

To conclude, an extended transaortic septal myectomy is the definitive treatment for HOCM. It is important to have a good knowledge of the characteristic pathophysiology of the disease in order to optimize intraoperative treatment of these patients. The use and combination of intraoperative TEE and direct measurement of pressures in the LV and the Ao are crucial to ensure the adequacy of HOCM surgery.

Declaration of patient consent
The authors certify that they have obtained all appropriate consent forms. The patient have given her consent for the images and other clinical information to be reported in the journal. The patient understand that her name and initial will not be published and due efforts will be made to conceal identity.

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Conflicts of interest
There are no conflicts of interest.

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