Progress in Cardiovascular Diseases 54 (2012) 503 – 511
www.onlinepcd.com

Anesthesia Management of Patients with Hypertrophic Obstructive Cardiomyopathy

Michal Gajewski, Zak Hillel*

St Luke's–Roosevelt Hospital, Columbia University, College of Physicians and Surgeons, New York, NY

Abstract Hypertrophic obstructive cardiomyopathy presents a challenge to the anesthesiologist. Because the condition is relatively prevalent, it is important for anesthesiologist to be aware of the pathophysiology. In this review, we draw upon case reports and studies of the anesthesia management of patients with hypertrophic obstructive cardiomyopathy to enhance medical decision making. The scope of this article ranges from the preoperative period, when the severity of the obstruction needs to be assessed; the intraoperative period, with monitoring, as well as general management guidelines; and finally, the postoperative period, when it is important to minimize the sympathetic response. Furthermore, we address the management of the obstetric patient, with particular focus on neuraxial anesthesia, and extrapolate how this type of anesthesia may be applied to the management of patients undergoing nonlaboring, noncardiac surgery. (Prog Cardiovasc Dis 2012;54:503-511)

Keywords: Obstructive hypertrophic cardiomyopathy; Hypertrophic cardiomyopathy; Anesthetic management

Since the first documented case of subaortic stenosis and asymmetric myocardial hypertrophy by Brock[1] in 1957 and Teare[2] in 1958, there have been significant advances in the understanding of the pathophysiology and in the management of hypertrophic obstructive cardiomyopathy (HOCM). It is not uncommon for these patients to now have normal lifespans.[3] This means that sooner or later, most anesthesiologists will encounter HOCM, not only during a therapeutic surgical septal myectomy but also for many routine, noncardiac cases. Because two-thirds 3 of patients have left ventricular (LV) obstruction either at rest or after physiologic provocation,[4] the anesthesiologist must be capable of managing this unique condition. The purposes of this article are to review the current literature and to provide evidence-based recommendations for patients with HOCM.

Definition

As established by Maron and Epstein[5] in 1979, hypertrophic cardiomyopathy (HCM), or HOCM when obstruction is present, is the preferred nomenclature for this disease. Part of the confusion with HOCM pertains to the differentiation of obstructive vs nonobstructive pathophysiology because HCM, in the resting patient, is mostly nonobstructive[6] and that physiologic changes in preload, afterload, and contractility may alter or even produce de novo LV outflow tract (LVOT) obstruction.[7]

Prevalence, pathophysiology, and prognosis

Initially believed to be relatively rare, it is now known that HCM occurs in about 2 of 1000 patients, potentially affecting close to 600 000 people in the United States.[8-10]

Statement of Conflict of Interest: see page 509.

* Address reprint requests to Zak Hillel, PhD, MD, Anesthesiology, St Luke's–Roosevelt Hospital Center, 1111 Amsterdam Ave, New York, NY 10025.

E-mail address: zh2@columbia.edu (Z. Hillel).

0033-0620/$ – see front matter © 2012 Elsevier Inc. All rights reserved.
doi:10.1016/j.pcad.2012.04.002

Abbreviations and Acronyms

CVP = central venous pressure

HCM = hypertrophic cardiomyopathy

HOCM = hypertrophic obstructive cardiomyopathy

LVOT = left ventricular outflow tract

PCPW = Pulmonary capillary wedge pressure

SAM = systolic anterior motion

TEE = transesophageal echocardiography

Many cases manifest during adolescence[11]; however, late-onset cardiomyopathy may develop at any age, and long-term survival may be affected by the time of presentation, with those identified in children showing a decreased survival as com pared with those identified in adulthood.[3]

Hypertrophic cardiomyopathy is described as a heterogenous autosomal dominant disease of the myocardial sarcomere that may involve 1000 different mutations in 18 different proteins.[3,4] Microscopically, HCM is characterized by myocardial disarray.[12] Sarcomeric protein mutations may cause abnormal activation of myocyte growth patterns, which then lead to uncoordinated muscle function and eventual hypertrophy.[12,13] Obstructive HCM may be subaortic, or midventricular, or both.[14,15] These characteristics will determine the degree of obstruction, subsequent symptoms, alterations in hemodynamics, and the incidence of arrhythmias—all of which have implications for anesthetic management. Subaortic obstruction is caused by systolic anterior motion (SAM) of the anterior mitral leaflet. There is now consensus that drag, the pushing force of flow, is the dominant hydrodynamic force on the mitral valve that causes SAM.[4,15,16] This causes the mitral leaflet to make contact with the ventricular septum causing obstruction, with concomitant mitral regurgitation due to deformation of leaflet position.[4] The mitral valve may undergo distortion and thickening due to hemodynamic or in situ abnormalities, predisposing it to endocarditis and, very rarely, rupture of the chordae tendinae.[14] With midventricular pathology, the obstruction occurs at the papillary muscle level[10,15] due to systolic apposition LV wall or due to a hypertrophied anomalous papillary muscle.[17,18] Unlike subaortic obstruction, with midventricular obstruction, mitral regurgitation is often absent.[10]

As described elsewhere, the peak instantaneous LV outflow gradient, rather than the mean gradient, guides treatment, and therefore, all therapeutic suggestions will focus on the peak gradient. There are 3 categories of dynamic LV outflow tract obstruction. There is resting obstruction with gradients of 30 mm Hg or greater at rest; latent obstruction without gradients at rest, but gradients greater than 30 mm Hg with physiologic provocation; and provocable obstruction with mild, less than 30 mm Hg gradients at rest, but high gradients after provocative maneuvers.[4] Obstructed patients either at rest or with physiologic provocation (exercise) are two-thirds of patients.[4,19] It is important to differentiate the obstructive and nonobstructive forms of the disease because this distinction guides anesthetic considerations. Patients with obstructive cardiomyopathy generally tend to be more symptomatic and prone to perioperative decompensation.

In addition to LVOT obstruction and mitral regurgitation, the pathophysiology of HCM also may include diastolic dysfunction, myocardial ischemia, and arrhythmias, all of which can have adverse implications during anesthesia and surgery. Indeed, diastolic dysfunction oftentimes may be the reason for decompensation when nonobstructive HCM is present. This is due to slow relaxation and elevated end-diastolic pressures, which can mimic a picture of left and (infrequently) right-sided heart failure with dyspnea and chest pain.[9,15] Myocardial ischemia may occur at any time in patients with HCM because there is a mismatch of oxygen supply and demand due to the hypertrophied muscle.[20] There is also a degree of microvascular dysfunction in patients with HCM, which may, on its own, be a predictor of clinical decompensation and death.[21] Lastly, although many different arrhythmias may occur in patients with HCM, atrial fibrillation is the most common sustained arrhythmia, occurring in 20% to 25% of patients.[10] Atrial fibrillation is also associated with mortality due to heart failure and stroke.[3,10,22] Thus, patients with HCM and atrial fibrillation should be anticoagulated with warfarin or dabigatran, unless there is a contraindication.[4,22]

Left ventricular outflow tract obstruction not only defines the hemodynamics of HCM, but is also an independent determinant of heart failure and cardiovascular death.[23] In a 2003 study, Maron et al[24] found that patients with *LVOT obstruction* (defined as a basal gradient of at least 30 mm Hg) had an increased risk of death from HCM or progression to congestive heart failure, which was more than 4 times that of patients without obstruction. In the same study, it was found that patients with obstruction, particularly if older than 40 years, have a higher probability of progression to New York Heart Association class III or IV heart failure and death.[24] Patients with HOCM have an annual mortality rate of 1.3% (of which 0.7% is attributable to sudden cardiac death) and a higher rate of morbidity than their healthy peers.

Diagnosis

Because surgery and anesthesia have the potential to increase the myocardial workload and decrease preload, patients with HCM are at risk for increased LVOT obstruction and arrhythmic complications. It is, thus, prudent to routinely follow guidelines set forth in Assessing and Reducing the Cardiac Risk of Noncardiac Surgery and the American College of Cardiology/American Heart Association 2007 guidelines on perioperative