Chapter 18

Hypertrophic Cardiomyopathy

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Case Scenario

A 5-year-old boy with a history of hypertrophic cardiomyopathy is booked for an emergency post-tonsillectomy bleed after he presents to the emergency department spitting out blood. Five days ago, he underwent an elective tonsillectomy and adenoidectomy and preoperative evaluation at that time included an electrocardiogram that showed normal sinus rhythm and biventricular hypertrophy.

Following the adenotonsillectomy and an uneventful postoperative recovery, he was discharged home. This morning he woke up and started vomiting blood. He is awake and alert but anxious. His current blood pressure is 80/40 mm Hg, heart rate is 145 beats/minute, and hemoglobin is 10 g/dL. He takes metoprolol daily but did not take his medication this morning.

Transthoracic echocardiography prior to adenotonsillectomy demonstrated:

- *Severe hypertrophic cardiomyopathy with hyperdynamic left ventricular systolic function*
- *Mildly impaired left ventricular relaxation*
- *Severe asymmetric septal hypertrophy*
- *Left ventricular outflow gradient 40 mm Hg at rest and 60 mm Hg with Valsalva*

Key Objectives

- Describe the genetics and pathophysiology of hypertrophic cardiomyopathy.
- Identify the clinical symptoms and diagnosis of hypertrophic cardiomyopathy.
- Discuss an appropriate preoperative evaluation of patients with hypertrophic cardiomyopathy.
- Describe anesthetic considerations for elective or emergent procedures in patients with hypertrophic cardiomyopathy.

Pathophysiology

What is hypertrophic cardiomyopathy?

Hypertrophic cardiomyopathy (HCM) is the most common autosomal dominant cardiac disease and is characterized by asymmetric septal hypertrophy. It is a genetically heterogeneous disease characterized by myocyte disarray, intermittent left ventricular outflow obstruction (LVOTO), diastolic dysfunction, and sudden cardiac death (SCD) [1].

What are the genetics of HCM?

Hypertrophic cardiomyopathy is genetically heterogeneous due to incomplete penetrance and variable expressivity. It is estimated that as many as 1 in 200 people may carry the gene but only 1 in 500 develop the disease [2]. Along with incomplete penetrance, HCM has variable expressivity. For example, some patients with HCM can have septal hypertrophy but no left ventricular (LV) outflow gradient and normal sinus rhythm, while others have near complete outflow tract obstruction or life-threatening arrhythmias.

The genetic heterogeneity leads to a variable clinical presentation and many people remain asymptomatic and undiagnosed. It is estimated that 750,000 people in the United States have HCM but only 100,000 have been diagnosed. However, lack of symptomatology does not imply benign expression of this disease. Sudden cardiac death (SCD) has occurred when neither the patient nor the provider was aware of the diagnosis. In fact, HCM is the leading cause of SCD in young athletes and the diagnosis is typically established postmortem. A thorough understanding of the pathophysiology is needed to safely care for a patient with HCM.

Clinical Pearl

Asymptomatic and undiagnosed patients with HCM may present for an anesthetic, but lack of symptomatology does not imply benign expression of the disease.

https://doi.org/10.1017/9781108657341.019 Published online by Cambridge University Press

What genes are involved?

Mutations in at least 11 genes that encode proteins of the cardiac sarcomere are responsible for HCM [3]. The majority of mutations are missense with a single amino acid substitution, but there are also insertions, deletions, and splice mutations that result in abnormal proteins being incorporated into the cardiomyocyte contractile apparatus. Both actin and myosin filaments of the sarcomere may be involved. Most frequently, the mutation involves the β-myosin heavy chain but there are as many as 1400 variants and every part of the contractile apparatus can be abnormal. The myriad of sarcomere proteins involved contributes to the heterogeneity of clinical expression.

What are the effects on the myocardium?

Abnormal sarcomere proteins can alter actin–myosin crossbridge cycling, potentially affecting both contraction and relaxation. These altered physical and functional properties of the sarcomeres cause abnormal growth, disorganized architectural patterns, and myocyte disarray [4]. Abnormal growth leads to septal hypertrophy, the hallmark of the disease. Disturbed cardiomyocyte architecture may result in rhythm disturbances and the abnormal sarcomere function can result in decreased cardiac relaxation.

What is the pathophysiology of HCM?

Hypertrophic cardiomyopathy is predominantly an obstructive disease. Obstruction occurs when there is mechanical impedance to outflow of blood from the LV, produced by the hypertrophied septum and systolic anterior motion (SAM) of the mitral valve. The mechanism of SAM is multifactorial and includes decreased LV diameter, Venturi effect, and twisting mechanism of the ventricle during systole. The enlarged septum and dysfunctional mitral valve result in distortion of the LV cavitary anatomy, which impedes blood flow from the ventricle. In addition, the mitral leaflets may fail to coapt in a normal fashion, causing regurgitation. (See Figure 18.1.)

What are the symptoms of HCM?

Symptoms of the disease may include dyspnea, angina, syncope, and palpitations. Children may present with asthma-like symptoms and fail to keep up with their peers. Infants may present with failure to thrive, tachypnea, and diaphoresis with feeding.

Why do these patients have dyspnea?

In patients with HCM, diastolic dysfunction and/or mitral regurgitation may lead to increased left atrial pressure (LAP). Dyspnea occurs when the elevated LAP impedes pulmonary venous outflow leading to congestion of the lungs. Higher pulmonary capillary pressure drives fluid into the interstitium, impeding diffusion of oxygen and resulting in symptomatic shortness of breath.

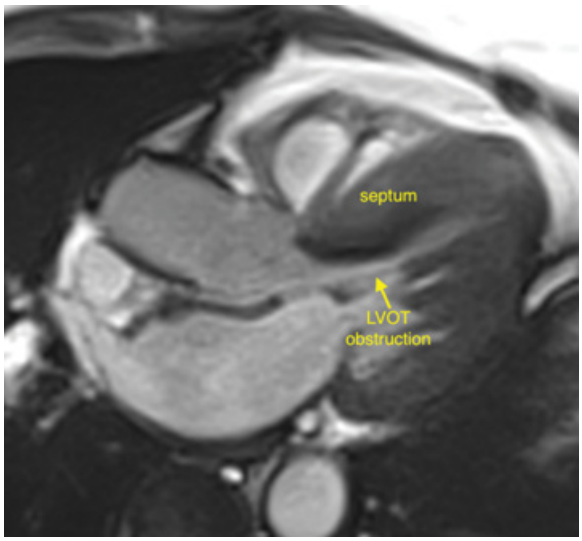

Figure 18.1 Hypertrophic cardiomyopathy. Magnetic resonance imaging of severe hypertrophic cardiomyopathy resulting in left ventricular outflow tract obstruction. Courtesy of Michael Taylor, MD.

Why do patients with HCM experience angina?

The etiology of angina in HCM is subendocardial ischemia from an imbalance of myocardial oxygen supply and demand. Oxygen demand is increased due to the myocardial hypertrophy; more muscle mass requires additional oxygen for actin–myosin crossbridge cycling. Even though myocardial oxygen demand is higher, capillary density is 33% lower compared to the normal heart, and this too results in lower oxygen delivery [5]. In addition, when LV end-diastolic pressure is elevated due to diastolic dysfunction, the pressure gradient driving coronary flow is diminished, and flow through the coronary arteries decreases, thereby further reducing oxygen supply. The result of the supply/demand imbalance is subendocardial ischemia, experienced subjectively as angina.

Why do HCM patients have syncope?

Syncope is due to inadequate cerebral perfusion. With HCM, decreased cardiac output (CO) and insufficient cerebral perfusion can occur from either increased LVOTO or dysrhythmias.

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What is the etiology of palpitations in HCM patients?

Rhythm disturbances are frequently observed with HCM, including sinus pauses, intermittent atrioventricular (AV) block, premature atrial and ventricular contractions, atrial fibrillation and flutter, and supraventricular and ventricular tachycardia. Sudden cardiac death is predominantly caused by ventricular fibrillation during intense physical exertion or stress despite a normal sinus rhythm at baseline.

How is HCM diagnosed?

Transthoracic echocardiography (TTE) is the gold standard for diagnosis of HCM. Asymmetric septal hypertrophy is the hallmark and LV function is typically preserved.

- ***For adults***, septal wall thickness of >15 mm in the absence of abnormal loading conditions is a diagnostic criterion [1].
- ***For children***, the criterion is a maximal LV wall thickness with a *Z* score >2 (greater than 2 standard deviations above average, corrected for body surface area) [6].

Pertinent features of the history include the patient's symptoms and any family history of HCM. Additionally, patients suspected to have HCM should undergo an electrocardiogram (ECG) to rule out dysrhythmias.

Cardiac magnetic resonance imaging (MRI) has been utilized to aid in the diagnosis of patients where TTE has yielded equivocal results due to poor acoustic windows or hypertrophy localized to regions not well visualized by TTE. Late gadolinium enhancement on MRI is indicative of myocardial fibrosis, which likely represents areas subjected to poor perfusion and subendocardial ischemia [7].

Genetic testing is an option, but the heterogeneity of the disease and variable penetrance make interpretation of the results difficult. In addition, recent studies indicate that identifying a particular sarcomere abnormality does not predict prognosis and cannot be used to guide treatment [8].

Clinical Pearl

Transthoracic echocardiography is the gold standard for diagnosis of HCM. Asymmetric septal hypertrophy is the hallmark and LV function is typically preserved.

What is the difference between HCM and HOCM?

Hypertrophic cardiomyopathy and hypertrophic obstructive cardiomyopathy (HOCM) are different expressions of the same disease. In a resting state, a ventricle may have asymmetric hypertrophy without obstruction defined as HCM; but obstruction can be provoked by different physiologic states and hence become HOCM. Tachycardia, hypovolemia, and hypotension may all cause outlet obstruction in these patients. With outlet obstruction, CO is impaired, leading to a downward spiral of hypotension, inadequate perfusion of the cerebral and cardiac vessels, syncope, ischemia, dysrhythmias, and death.

Clinical Pearl

In a resting state, a ventricle may have asymmetric hypertrophy without obstruction defined as HCM; obstruction can be provoked by different physiologic states and hence become HOCM. Tachycardia, hypovolemia, and hypotension may all cause LVOT obstruction in these patients.

What happens when a patient with HCM becomes tachycardic?

Increased heart rate is deleterious in a patient with HCM for a number of reasons.

- Diastolic time decreases, allowing less time for isovolumetric LV filling. Ventricular diameter decreases and LVOTO is provoked.
- Shortened ventricular relaxation time impairs oxygen delivery because coronary perfusion occurs during diastole.
- Finally, tachycardia increases workload because the myocardium develops tension more frequently each minute, thereby increasing oxygen demand. The net result is a supply/demand imbalance that increases the risk for subendocardial ischemia.

What happens when these patients become hypovolemic?

If a patient has a decrease in intravascular volume, preload diminishes, and consequently both LV filling and diameter decrease, causing LVOTO. Impaired venous return will elicit reflex mechanisms to maintain CO, resulting in tachycardia that further exacerbates the situation.

How does a HCM patient decompensate when he or she becomes hypotensive?

The decrease in both systolic and diastolic blood pressures is problematic. Elevated afterload mechanically stents open the outflow tract, helping to maintain ventricular geometry. Lower systolic blood pressures can result in outflow

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tract collapse, provoking or worsening the obstruction. Lower diastolic pressure decreases coronary perfusion, limiting oxygen delivery to the myocardium.

How is HCM medically treated?

The main goal of treatment is to decrease or prevent outlet obstruction. Medical aims are as follows:

- ***Maintain low-normal heart rate*** to allow for a longer ventricular filling time and hence improved coronary perfusion.
- ***Avoid increases in sympathetic stimulation*** to decrease and/or prevent obstruction during ejection.
- ***Prevent dysrhythmias.***
- ***Decrease diastolic dysfunction.***

β-Adrenergic receptor blockade for negative inotropy and chronotropy has been the mainstay of pharmacologic treatment. Virtually all patients diagnosed with HCM will take ß-blockers unless there is a contraindication. Calcium channel blockers are used with caution. Their negative inotropic and lusitropic properties are helpful, but they both vasodilate and venodilate, decreasing both preload and afterload. Disopyramide is sometimes used as an antiarrhythmic and to decrease contractility. Amiodarone may also be used to suppress dysrhythmias.

Lifestyle modification is part of the treatment plan. Avoidance of dehydration, caffeine, and competitive sports is advisable. Both genetic and family counseling is helpful for patients and families with the diagnosis.

Clinical Pearl

The main goal of treatment for HCM patients is to decrease or prevent LVOT obstruction.

Are there surgical therapies to address HCM?

Surgical myectomy can be performed, in which a portion of the hypertrophied muscle is excised from the ventricular septum, enlarging the outflow tract. It is the primary therapeutic option for patients who are refractory to pharmacologic intervention and have a LVOT gradient >50 mm Hg at rest or with provocation [9]. An additional benefit with myectomy is that the mitral apparatus may function in a more normal fashion if SAM and mitral regurgitation are improved.

Catheter-based alcohol ablation performed in the cardiac catheterization laboratory is a potential alternative to surgical myectomy. Alcohol is injected into an artery supplying the septum, causing transmural myocardial infarction, reducing septal wall thickness, and enlarging the LVOT diameter. Patients can avoid cardiopulmonary bypass with this technique, but potential side effects of alcohol treatment include increased conduction abnormalities and residual LVOT gradient.

Automatic implantable cardioversion devices (AICDs) have been used prophylactically to prevent SCD. Indications for AICD placement include a family history of SCD, unexplained syncope, repetitive episodes of nonsustained ventricular tachycardia, massive left ventricular hypertrophy (LVH), and extensive late gadolinium enhancement on MRI or fibrosis. Patients with systolic or diastolic heart failure who are refractory to pharmacologic and surgical treatment may require implantation of a left ventricular assist device (LVAD) or cardiac transplantation.

Anesthetic Implications

What are the potential anesthetic complications in a patient with HCM?

Anesthesia and surgery can cause catecholamine surges resulting in tachycardia, hypotension, and hypovolemia, all of which may exacerbate LVOTO. Adverse perioperative outcomes for HCM patients can include congestive heart failure, myocardial ischemia, systemic hypotension, dysrhythmias, and death [10]. Adverse events in children are more likely to occur in the setting of LVH, LVOTO, diastolic dysfunction, and a history of ventricular arrhythmias [11]. In an adult study, patients with HCM and a LVOTO gradient >30 mm Hg at rest and intraoperative hypotension had a higher rate of adverse events [12].

What preoperative evaluation is necessary for a 5-year-old patient with HCM presenting for elective adenotonsillectomy?

Identification of high-risk patients can help guide clinical management and ensure adequate allocation of resources. Evaluation includes a thorough family history to rule out familial SCD, patient history to ascertain symptomatology, and an ECG to rule out rhythm disturbances. In addition, a chest radiograph may show pulmonary edema or an enlarged cardiac silhouette due to LVH and/or left atrial enlargement. An echocardiogram or cardiac MRI is mandatory to determine if the patient has outflow obstruction at rest or with provocation, the latter typically elicited with a Valsalva maneuver to decrease venous return. Preoperative instructions should include short nil per os (NPO) times to minimize hypovolemia and maintain preload, and continuation of pharmacologic management

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such as ß-blockers to blunt the sympathetic nervous system (SNS) response to anesthesia and surgical stress.

Clinical Pearl

An echocardiogram or cardiac MRI is mandatory to determine if the patient has outflow obstruction at rest or with provocation, the latter typically elicited with a Valsalva maneuver to decrease venous return.

The surgeon would like to perform the adenotonsillectomy at an outpatient facility. What is the most appropriate venue?

Caution should be exercised regarding location of surgery. Due to the increased risk of adverse events and death when patients with HCM are anesthetized, it is not advisable to anesthetize a patient with HCM in a free-standing ambulatory setting [12]. It is recommended to proceed in a specialized center with pediatric anesthesiologists, cardiologists and intensivists that are experienced in managing a child with HCM.

If a patient with HCM has an AICD, how should it be managed in the perioperative period?

The decision to disable an AICD is made by an electrophysiologist. The Heart Rhythm Society/American Society of Anesthesiologists (ASA) Expert Consensus Statement recommends that the decision be individualized for each patient and the decision made by the physician who monitors the patient's AICD function [13]. If the device is reprogrammed or deactivated, the Class I recommendation is for continuous cardiac monitoring during the entire time of deactivation and immediate availability of an automatic external defibrillator. The AICD should be reprogrammed in the immediate postoperative period. Prophylactic cutaneous defibrillator pads for cardioversion of a dysrhythmia is advisable if the patient's AICD has been disabled or if the patient does not have an AICD.

The patient has normal LV function with an LVOTO gradient of 40 mm Hg at rest and mild diastolic dysfunction. He is asymptomatic and ECG shows normal sinus rhythm. What perioperative monitoring would be appropriate?

Decisions regarding intraoperative monitoring are made according to the severity of HCM and nature of the operative procedure. For a minor procedure in a patient with insignificant LVOTO, standard ASA monitors with 5-lead ECG would suffice.

What are anesthetic management goals for a patient with HCM?

The primary anesthetic goal is to prevent LVOTO by avoiding tachycardia, hypotension and hypovolemia. This is achieved by reducing SNS activity as well as maintaining normal sinus rhythm, left ventricular filling and systemic vascular resistance. Fluid strategy is tailored to euvolemia, which will help maintain preload and afterload. Excessive fluid administration in the setting of a stiff ventricle with diastolic dysfunction can cause pulmonary edema.

Clinical Pearl

Anesthetic management goals in patients with HCM include avoidance of tachycardia and maintenance of normal sinus rhythm, preload, and afterload.

How should anesthesia be induced in this patient for an *elective* case?

There is no single best agent for induction of anesthesia. The aim is to keep the hemodynamic goals in mind and titrate anesthetic agents accordingly. Although inhalation induction with sevoflurane might be an option in the patient with a mild form of HCM, it is important to recognize that an inhalational induction with sevoflurane can cause significant increases in heart rate and a reduction in SVR, both of which may place the patient at risk for cardiac ischemia. In addition, without intravenous (IV) access it will not be possible to rapidly treat hypotension or rhythm abnormalities. If the child has IV access, small doses of propofol slowly titrated to maintain stable hemodynamics may be an option. However, a large bolus dose of propofol is not advisable as venodilation and vasodilation can decrease preload and afterload. If SVR drops, small doses of phenylephrine (0.5–1 mcg/kg) can be used. Etomidate is a safe choice because it has a stable cardiovascular profile, but it should be administered with another anesthetic agent such as fentanyl to aid in blunting the sympathetic response to endotracheal intubation.

Prior to direct laryngoscopy, the SNS can be attenuated with a short-acting ß-blocker (e.g., esmolol) or short-acting opioid such as remifentanil. In most cases, if muscle relaxation is required, a nondepolarizing agent such as vecuronium or rocuronium can be used, but caution is advised with agents such as atracurium that

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cause histamine release and hypotension. If a rapid-acting agent is needed, succinylcholine is an option, but it may cause bradycardia or asystole. If the fall in heart rate is hemodynamically significant, glycopyrrolate or atropine can be administered. The use of glycopyrrolate may be preferable to atropine as it can accomplish the goal of treating bradycardia while minimizing the risk of incurring tachycardia. It is also important to recognize that atropine causes a greater reduction in diastolic time than other ß-adrenergic agents. Ephedrine can be used to increase heart rate without compromising diastolic time.

What agents can be used for maintenance of anesthesia?

Maintenance of anesthesia may be accomplished with volatile agents. Sevoflurane has minimal effects on the cardiovascular system and may be preferable to isoflurane which has negative inotropic and vasodilatory properties. If volatile agents are not tolerated, IV adjuncts can be used to maintain the depth of anesthesia necessary for the operative procedure. Benzodiazepines are desirable given their anxiolytic and amnestic properties. Dexmedetomidine infusion can be useful because it blocks sympathetic stimulation and decreases heart rate, but bolus dosing can cause hypotension. In longer cases maintenance of muscle relaxation may be necessary, especially if the patient is unstable.

What are the goals during emergence from anesthesia?

An important goal during anesthetic emergence is to attenuate the SNS response as the child awakens. Titration of fentanyl or morphine, and administration of nonopioid analgesics such as IV acetaminophen or administration of dexmedetomidine, 0.5–1 mcg/kg, can ameliorate the catecholamine surge with emergence.

If the perioperative course is uneventful, how should postoperative disposition be determined?

Postoperative recovery of HCM patients depends on the severity of the disease, the type of procedure, and intraoperative course. Disposition to the post-anesthesia care unit is appropriate for patients with benign expressions of HCM undergoing minor surgery who have an uneventful anesthetic. For patients with more severe symptoms, major surgery and/or perioperative cardiovascular instability, the intensive care unit (ICU) is more appropriate. Many patients fall within that spectrum and the decision should be made following a multidisciplinary discussion with the anesthesiologist, surgeon, and cardiologist/intensivist if available.

Postoperatively the patient is agitated and crying: what are the considerations and how should they be addressed?

Pain will increase catecholamines, contractility and heart rate so it is important to intervene in a timely fashion to avoid increased LVOTO. Opioid analgesics, benzodiazepines, and dexmedetomidine, alone or in combination, will help lower SNS activity. If the patient remains hyperdynamic and tachycardic despite adequate analgesia and anxiolysis, then a fast-acting ß-blocker (e.g., esmolol) can be given.

What are the anesthetic considerations in a HCM patient presenting with a post-tonsillectomy hemorrhage?

A HCM patient who has been hemorrhaging is likely to be hypovolemic and may be hemodynamically unstable with outlet tract obstruction and impending cardiovascular collapse. Hence, immediate preoperative fluid resuscitation is a must. Intravenous access should be placed, and a fluid bolus given, if necessary, prior to induction of anesthesia. In addition to being hypovolemic, the patient may also be anemic with decreased oxygen carrying capacity, which increases the risk of myocardial ischemia. Obtaining a hematocrit and blood typing and cross match should be done as soon as possible. Minimizing anxiety to prevent a SNS surge is important and may be accomplished with benzodiazepines or by utilizing support from child life specialists or distractions such as video games and movies. The parents should also be asked about compliance with cardiac medications.

If the patient has an AICD but cardiologists are not immediately available to reprogram the device, how should you proceed?

There is a risk of AICD malfunction when using monopolar electrocautery. The device may be inhibited by the monopolar electrocautery and fail to defibrillate when needed or may fire inappropriately when electromagnetic interference causes false detection of arrhythmias [14]. In an emergent situation, bipolar electrocautery should then be used, since it uses less current and will not interfere with the AICD unless directly applied to it.

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Clinical Pearl

For patients with an AICD, the electrophysiology team should interrogate, disable, and reprogram the device in elective cases. In an emergent setting, bipolar electrocautery can be used.

What perioperative monitors should be used for this patient?

Standard ASA monitoring with 5-lead ECG should be placed prior to induction. Although an arterial line is useful for hemodynamic monitoring in a potentially unstable child, placement of an arterial line in an awake child can be challenging and cause further anxiety and catecholamine release. Additionally, with an emergent post-tonsillectomy bleed the priority is achieving control of bleeding. If arterial line placement is warranted, it should take place while surgical control of bleeding is occurring. Although transesophageal echocardiography is helpful to determine volume status as well as dynamic outflow obstruction, it is not feasible given the surgical site for this patient. Transthoracic echocardiography could be utilized instead if needed. Adequate peripheral venous access for volume resuscitation should be obtained as well. Decisions should be individualized based on the clinical and hemodynamic assessment of the patient.

What are the anesthetic goals?

The primary anesthetic goal is to prevent increases in LVOTO. Although the patient did not have symptomatic LVOTO before he started bleeding, the possibility that obstruction may have been provoked by hypovolemia, tachycardia, and hypotension must always be considered. Anemia exacerbates the situation by further increasing heart rate. The goal is to reduce SNS activity while maintaining normal sinus rhythm, left ventricular filling, and systemic vascular resistance. In addition, treating intravascular depletion and restoring hematocrit to normal levels should be undertaken.

What is an anesthetic induction strategy for this patient with post-tonsillectomy hemorrhage?

A rapid sequence induction is indicated to secure an airway with ongoing pharyngeal bleeding and a full stomach from swallowed blood. Etomidate is a safe choice for an IV induction agent. Propofol in this clinical scenario would not be advisable because it may not be safely titrated in a timely fashion. Bolus dosing of propofol in the hypovolemic patient with HCM may precipitate outflow obstruction that could lead to ventricular fibrillation. For muscle relaxation, succinylcholine or rocuronium can be used. It is important to keep in mind that succinylcholine can lead to significant bradycardia. To blunt the SNS response to airway instrumentation, judicious use of opioid analgesia may be used. Esmolol could be used (standard dosing at 0.5 mg/kg bolus with infusions of 50–300 mcg/kg/minute) to attenuate the SNS response to intubation but hypotension may occur with coexisting intravascular volume depletion.

The patient becomes hypotensive after induction: What are the concerns and possible actions?

Post-induction hypotension can precipitate outlet obstruction, decrease coronary perfusion, and evoke ventricular fibrillation. Timely treatment with a vasoconstrictor is essential to maintain afterload and coronary perfusion pressure. To increase SVR, the preferred agent is the α_1-adrenergic agonist phenylephrine, with a bolus dose of 1–2 mcg/kg and/or infusion of 0.1mcg/kg/minute. The use of inotropes such as epinephrine is inadvisable in HCM patients as epinephrine will increase contractility and heart rate and may precipitate cardiovascular collapse [1]. Caution should also be used with other ß-adrenergic agonists such as dopamine, dobutamine, ephedrine, and isoproterenol. If the response to phenylephrine is inadequate, vasopressin, a potent vasoconstrictor, is another option. An added benefit with direct vasoconstrictors is the reflex bradycardia elicited by the increase in blood pressure, which is beneficial with HCM physiology.

Post-induction hypotension can also be minimized by decreasing the concentration of volatile anesthetic agent. Giving a fluid bolus of crystalloid or packed red blood cells will expand intravascular volume and increase preload and afterload.

Clinical Pearl

The use of inotropes such as epinephrine is inadvisable in HCM patients, as epinephrine will increase contractility and heart rate and may precipitate cardiovascular collapse. If the response to phenylephrine is inadequate, vasopressin, a potent vasoconstrictor, is another option. An added benefit with direct vasoconstrictors is the reflex bradycardia elicited by the increase in blood pressure, which is beneficial with HCM physiology.

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What is an appropriate ventilation strategy for this patient?

High inspiratory airway pressures during positive pressure ventilation will decrease venous return, and the effect is exaggerated with hypovolemia. This can be achieved by avoiding high positive end-expiratory pressure, maintaining physiologic ventilation and allowing resumption of spontaneous ventilation as soon as safely possible.

The patient becomes hypertensive when the mouth gag is placed: How might this be treated?

Caution should be used with direct vasodilators as they can worsen LVOTO and precipitate cardiovascular collapse. Vasodilators such as hydralazine and nitroprusside should be avoided. In addition to provoking outlet obstruction, they can also induce ischemia by lowering diastolic pressure. A safer approach to lowering blood pressure is to decrease SNS stimulation with higher volatile agent concentrations and opioid analgesics. After ensuring adequate anesthesia and analgesia, if the patient remains hyperdynamic, consider ß-blockers to lower blood pressure into the desired range.

Clinical Pearl

Caution is indicated with direct vasodilators because they can worsen LVOTO and precipitate cardiovascular collapse.

On arrival in the ICU the patient's ECG rhythm changes to atrial fibrillation with a heart rate of 140. What is the most appropriate therapeutic action?

Patients with HCM are dependent on active ejection of blood from the atria into the LV for adequate filling. Loss of coordinated atrial systole with atrial fibrillation can be catastrophic and the recommendation is immediate direct cardioversion to restore sinus rhythm [1]. Pharmacologic restoration to sinus rhythm does not have a favorable time course and is a second line of therapy.

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Suggested Reading

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