Anesthetic Management of Hypertrophic Cardiomyopathy and Challenges in the MRI Suite

Moderators: Nina A. Guzzetta, M.D.; Elizabeth C. Wilson, M.D.

Institution: Emory University School of Medicine, Children’s Healthcare of Atlanta – Egleston

Objectives:

1. Understand the underlying anatomy and pathophysiology of hypertrophic cardiomyopathy (HCM).
2. Review the morbidity and mortality associated with HCM when combined with anesthesia.
3. Assess the medical workup of children with HCM in order to evaluate severity of disease.
4. Understand the effects of anesthetic drugs on the hemodynamic management of children with HCM.
5. Discuss considerations for providing off-site anesthesia specifically as it relates to care of patients in the magnetic resonance imaging (MRI) suite.

Case History:

A seven-year-old, 28 kg male with HCM and Attention Deficit Hyperactivity Disorder presents for cardiac MRI under general anesthesia after having failed sedation provided by a non-anesthesia sedation service. The patient failed sedation on a separate occasion secondary to significant hypotension with the administration of propofol.

Questions:

1. What is the anatomy of HCM?
2. What type of patients present with HCM?
3. What type of signs and symptoms do these patients present with?
4. How does the physiology of HCM explain these symptoms?
5. What additional information would be helpful to you prior to administering anesthesia?

Case continued:

On physical exam, the patient had a normal airway with good mouth opening and a normal thyromental distance. Upon chest examination a pansystolic high-frequency murmur was heard throughout the chest but best appreciated at the apex. The lungs fields were clear. Patient medications included risperidone and methylphenidate. Echocardiogram showed moderate asymmetric left ventricular hypertrophy with a left ventricular outflow tract (LVOT) peak gradient of 28 mmHg and systolic anterior motion of the mitral valve at rest. A stress test was adequate and demonstrated a decrease in blood pressure with exercise and an increase in the LVOT peak gradient to 60 mmHg. There were no ST segment changes or arrhythmias.

Questions:
6. What is the typical medical management of HCM and how does it apply to this patient?  
7. Why do you think this patient failed propofol sedation?  
8. What type of NPO instructions would you give this patient?  
9. What type of anesthetic would you choose for this patient and why?  
10. What risks would you discuss with the parents or guardians prior to the anesthetic?  
11. How would you premedicate this patient?  
12. Assuming a general anesthetic, how would you approach induction?  
13. How would you secure the patient’s airway?  
14. Would you want any additional monitoring for this case?  

Case continued:  

After premedication with oral midazolam, the patient underwent an inhalational induction with subsequent placement of a peripheral intravenous line. Hypotension and ST segment changes were noted on induction.  

15. What would your intervention be?  

Blood pressure and ST segments improved with administration of intravenous volume and phenylephrine and the blood pressure that correlated with normal ST segments was noted. A laryngeal mask airway was used to secure the airway. Anesthesia was maintained with sevoflurane, nitrous oxide and oxygen and the patient was kept spontaneously ventilating. Further ST segment evaluation during the MRI was limited. During the procedure the patient experienced two episodes of non-sustained ventricular tachycardia.  

16. Why is ST segment evaluation limited during a cardiac MRI?  
17. How would you respond to the non-sustained ventricular tachycardia? Would you abort the MRI?  
18. What additional concerns can you think of that are unique to working in the MRI suite?  
19. Where would you recover this patient?  
20. Would you obtain any post-procedure tests or consults?  

On completion of the MRI the patient woke without complication and was recovered in the post-MRI suite.  

Discussion:  

Hypertrophic cardiomyopathy (HCM) is one of several cardiomyopathies, a heterogeneous group of disorders of cardiac muscle that are classified according to ventricular morphology and pathophysiology. Today there are five major classifications of cardiomyopathies: hypertrophic cardiomyopathy, dilated cardiomyopathy, restrictive cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy and unclassified. HCM is defined as hypertrophy of the left ventricle in the absence of abnormal loading conditions such as valvular disease, hypertension or other congenital malformation sufficient to explain the degree of hypertrophy. The frequency of disease in children is 0.3 to 0.5 cases per 100,000 births. HCM occurs more frequently in males and in the first year of life with another peak occurring in adolescence. The primary concern of HCM is sudden death which is currently the leading cause of death in young athletes. Recent population-
based reports quote an overall annual rate of sudden death of 1% to 1.5% per year beyond infancy. Other causes of death from HCM include thromboembolism, progressive cardiac failure and infective endocarditis.

HCM follows an autosomal dominant pattern of inheritance and is caused by mutations in the genes that encode proteins of the myocardial sarcomere. Depending on the specific mutation, the abnormal protein leads to ineffective contraction of the sarcomere and the development of myocyte hypertrophy. One specific mutation found on the β-myosin heavy chain has been found to be responsible for approximately one-third of all cases of HCM. Nevertheless, over 50 mutations causing HCM have been identified thus making HCM the most widespread genetic disease of the myocardium. HCM can also be associated with inborn errors of metabolism (e.g., mitochondrial diseases and Pompe’s disease), neuromuscular diseases and certain syndromes (e.g., Noonan’s syndrome).

The histological hallmarks of HCM are myocyte hypertrophy, myocyte disarray and interstitial fibrosis. Additionally, small intramural coronary arteries are often dysplastic and narrowed due to mural thickening by hyperplasia of smooth muscle cells. During infancy most children with HCM have few, if any, symptoms. The initial diagnosis is commonly made following the incidental finding of a heart murmur, an abnormal electrocardiogram (ECG) or as a result of family screening when another family member has been diagnosed with HCM. Older children will often experience chest and/or dyspnea with exertion. Syncope is a concern and may be the result of left ventricular outflow tract obstruction, atrial or ventricular dysrrhythmias or abnormal vascular responses. Exertional syncope is associated with an increased risk of sudden death. ECG changes include signs of septal and left ventricular hypertrophy, as well as ST-segment and T-wave changes. Echocardiography is a mainstay in determining the extent, location and severity of disease. The typical echocardiographic finding of HCM is ventricular hypertrophy with severely thickened ventricular walls (>17 mm). LVOT obstruction can be secondary to dynamic muscular obstruction, early systolic closure of the aortic valve or systolic anterior motion of the mitral valve which may also cause mitral regurgitation. Hyperdynamic left ventricular function is often seen and almost all patients demonstrate diastolic dysfunction with abnormal myocardial relaxation and diastolic Doppler patterns. Exercise testing is also a routine part of the work-up to determine the severity of HCM. An abnormal response to exercise is associated with an increased risk of sudden death in young children and adolescents. Cardiac MRI is particularly useful in assessing the distribution and severity of left ventricular hypertrophy and providing functional measurements of systolic and diastolic function. Additionally, cardiac MRI with gadolinium-based contrast is used to assess myocardial tissue characteristics. Extensive enhancement of the septum with gadolinium has been shown to closely correlate with sudden death and progressive left ventricular remodeling.

The mainstay of medical therapy for HCM consists of beta-blockers or calcium-channel blockers and is guided by patient symptoms and exercise testing. Atrioventricular pacing has also been used as an alternative to surgery though studies showing long-term improvement in outcomes are lacking. For patients at high risk for sudden death an implantable cardioverter-defibrillator (ICD) is indicated. Septal myectomy is reserved for patients with an LVOT gradient greater than 50 mmHg and symptoms that are unresponsive to medical therapy.
Patients with HCM are at significant risk for cardiac complications during surgery and anesthesia. Although studies looking specifically at the pediatric population are lacking, one study of adults with HCM undergoing surgery and anesthesia found a 40% risk of experiencing at least one adverse cardiac event. Anesthetic management focuses on maintaining preload and afterload, avoiding increases in contractility and avoiding tachycardia. Ideally patients with HCM should be well hydrated prior to the induction of anesthesia. Agents that maintain afterload without a significant increase in contractility or heart rate are preferable for both induction and maintenance of anesthesia. Appropriate intra-operative monitoring and a postoperative care plan are dependent on the procedure and the severity of HCM.

The MRI suite presents significant technical challenges to the anesthesiologist. The remote location means working at a distance from the operating room and with personnel who may be unfamiliar with the needs of anesthesia providers. The generated magnetic field creates multiple additional considerations. MRI-compatible equipment must be used. “Projectile injury” to the patient or other care providers can occur from ferromagnetic objects that are pulled into the scanner at rapid velocity. Battery powered devices may suddenly fail to operate and pacemakers and ICDs, even if MRI-compatible, may not operate as planned. ECG monitoring is especially difficult within the magnetic field. Spike artifacts that mimic R-waves are produced secondary to the changing magnetic fields of the imaging gradients and make it impossible to reliably detect ischemia or arrhythmias.

References:


