Chapter

33

Mixed Cardiomyopathy

Adam C. Adler

Case Scenario

A 3-year-old male weighing 11 kg presents for semielective inguinal hernia repair following recurrent bowel incarceration. The past medical history includes arthrogryposis and severe kyphoscoliosis in addition to complex heart disease. Home medications include metoprolol. The patient is asymptomatic but is wheelchair bound due to limb deformities. The parents are requesting information regarding the safety of anesthesia for their child and the anesthetic options.

Recent transthoracic echocardiogram revealed the following:

- *Severe mixed cardiomyopathy with hypertrophic and restrictive physiology*
- *Severe biventricular hypertrophic cardiomyopathy with near complete cavitary obliteration*
- *Left ventricular diastolic septal thickness consistent with a z-score of +19*
- *Right and left ventricular outflow tract obstruction (see Figure 33.1)*
- *Ejection fraction 74%*
- *Dynamic left ventricular outflow tract obstruction from systolic anterior motion of the mitral valve*
- *Prominent reversal of flow in the hepatic veins and inferior vena cava*

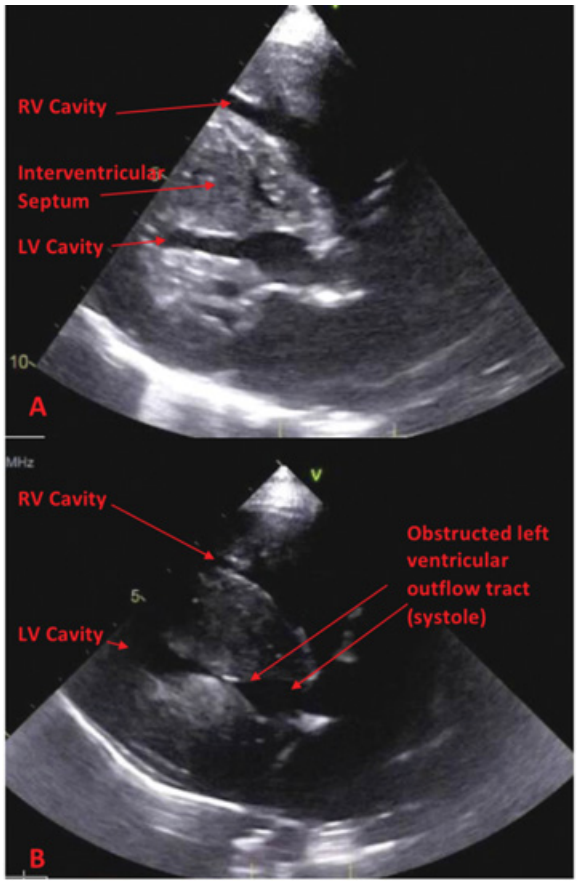

Figure 33.1 **Preoperative transthoracic echocardiography in parasternal long axis view demonstrating (A) severe right and left ventricular cavity obliteration from thickened ventricular free wall and interventricular septum and (B) dynamic obstruction of the left ventricular outflow track.** Courtesy of Adam C. Adler, MD.

Key Objectives

- Identify the subtypes of pediatric cardiomyopathies.
- Review the pathophysiology and clinical presentation for pediatric cardiomyopathies.
- Formulate a safe anesthetic plan for children with severe cardiomyopathy.

Pathophysiology

What are the subtypes of pediatric cardiomyopathy?

Pediatric cardiomyopathy subtypes include:

- Dilated cardiomyopathy (DCM)
- Hypertrophic cardiomyopathy (HCM)
- Restrictive cardiomyopathy (RCM)
- Noncompaction cardiomyopathy (NCM)
- Arrhythmogenic right ventricular dysplasia (ARVD)

While distinct classifications exist, in practice, the variants occur in combination. However, there is generally a predominant phenotype diagnosed by echocardiography

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that allows multicenter registries to track epidemiology, management, and outcomes related to pediatric cardiomyopathy. This chapter will focus on mixed hypertrophic and restrictive forms of cardiomyopathy. (See Chapter 32.)

Clinical Pearl

While distinct types of cardiomyopathy exist, patients often present clinically with mixed types and physiology.

What is the incidence of pediatric cardiomyopathy?

The overall incidence of pediatric cardiomyopathy is estimated to be around 1–1.5 cases per 100,000 patients. Dilated cardiomyopathy accounts for more than 50% of all cases. Hypertrophic cardiomyopathy is the second leading cause, with restrictive cardiomyopathy being the least common.

Hypertrophic Cardiomyopathy

What is hypertrophic cardiomyopathy?

Hypertrophic cardiomyopathy (HCM) is a disease of the myocardial sarcomere characterized by thickening of the myocardium. It occurs either sporadically or as part of a familial (genetic) disorder or metabolic syndrome. On echocardiography, HCM appears as increased muscle mass without ventricular dilation. Histologically, HCM appears as myocyte disarray and fibrosis, which is best thought of as an increase in poorly or nonfunctioning myocardium.

What are primary and secondary hypertrophic cardiomyopathy and how do they differ?

- ***Primary HCM*** (most common) reflects an autosomal dominant pattern of inheritance and is characterized by left ventricular hypertrophy (LVH) with rare RV involvement. Primary HCM results from a genetic mutation affecting different elements of the muscle sarcomere. More than 1000 genetic mutations have been identified for primary HCM. The primary form of HCM typically develops in childhood or adolescence.
- ***Secondary HCM*** is generally "secondary" to a variety of metabolic and mitochondrial disorders and syndromes and is generally not isolated to the cardiac muscle. Manifestation of secondary HCM occurs in infancy and commonly affects both right and left ventricles. Some disorders associated with secondary HCM include mitochondrial diseases such as Friedreich's ataxia and metabolic disorders such as Fabry and Pompe disease.

What are the anatomic manifestations of HCM and the resultant pathophysiology?

While the phenotypic expression of HCM varies, the pattern of LVH typically involves the basal and mid-interventricular septum. Generally, the ratio of septal wall to inferolateral wall thickness is >60%. Severe septal hypertrophy often displaces the mitral valve apparatus and obstructs the left ventricular outflow tract (LVOT) in the subaortic region. This septal hypertrophy results in dynamic obstruction to the LVOT during mid-systole when the septal wall moves toward the mitral apparatus. (See Figure 33.2.) In addition to LVOT obstruction, significant diastolic dysfunction occurs with

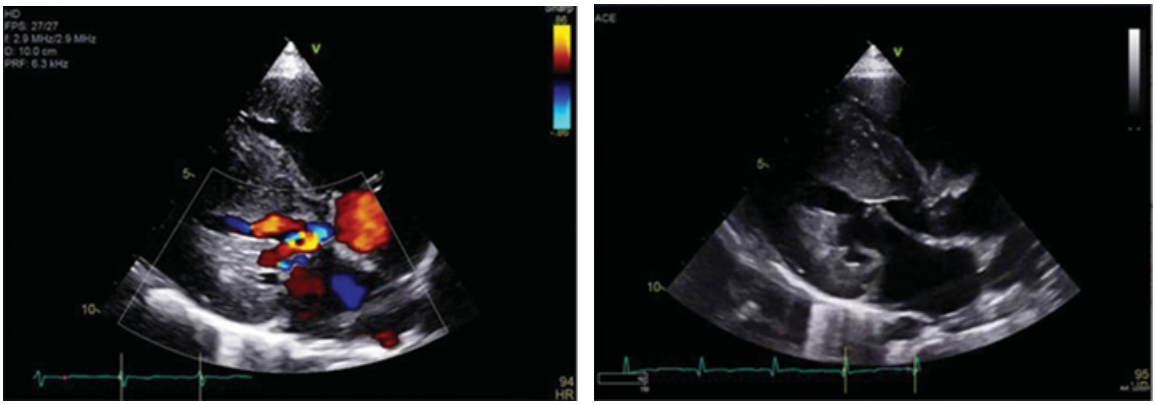

Figure 33.2 Transthoracic long axis view showing significant flow acceleration at the LV outflow tract (left) and dynamic LV outflow obstruction due to systolic anterior motion of the mitral valve (right). Courtesy of Adam C. Adler, MD.

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HCM, even in patients with mild disease. Although the LV cavity size is normal, myocardial dysfunction results in impaired LV relaxation and distensibility. Additionally, patients with HCM are prone to developing coronary ischemia. In addition to the increased myocardial oxygen demand (from increased myocardial mass), intramural coronary vasculature is often affected by the myocardial wall stress. Lastly, elevated LV end-diastolic pressure results in subendocardial ischemia.

What is the typical clinical presentation for patients with HCM?

The range of symptoms seen in patients with primary HCM varies from asymptomatic to severe, including sudden death. Most often patients have dyspnea due to LV diastolic dysfunction. Additionally, many patients experience syncope, particularly with exertion. This occurs as a result of LVOT obstruction with impedance of coronary and systemic perfusion, in addition to the multifactorial effects on the distal coronaries as noted earlier.

What management options exist for patients with HCM?

Treatment strategies for HCM include medical therapy in addition to interventional and surgical approaches. For those with mild HCM, medical management with ß-blockers and/or calcium channel blockers is initiated and guided by patient symptoms or performance on stress testing. For patients with advanced disease as evidenced by symptomatology, poor ventricular function, or a history of arrhythmias, automated implantable cardioverter-defibrillators are placed to prevent sudden death. Patients with advanced disease may be suitable for surgical septal myectomy. Surgical candidacy is guided by symptomatology (arrhythmias, dyspnea, angina, syncope) and echocardiographic findings (LVOT gradient >50 mm Hg). Patients with end-stage heart failure are often evaluated for orthotopic heart transplantation.

Restrictive Cardiomyopathy

What is restrictive cardiomyopathy?

Isolated RCM is rare, encompassing approximately 5% of pediatric cardiomyopathies. More often, more common forms of cardiomyopathy (i.e., HCM) appear in conjunction with restrictive physiology as opposed to isolated RCM.

Clinical Pearl

Restrictive cardiomyopathy is a disease process with isolated diastolic heart failure. It is distinct from other forms of cardiomyopathy that may also result in diastolic failure and restrictive physiology.

What are the common presenting signs and symptoms in patients with RCM?

Most of the presenting symptomatology results from prolonged elevation of ventricular filling pressures. Elevated end-diastolic pressure results in pulmonary edema and pulmonary hypertension, peripheral edema, hepatomegaly, and weight gain. Not uncommonly, patients present after undergoing an unrevealing workup for reactive airway disease. Restrictive cardiomyopathy may also be diagnosed during evaluation for a variety of diseases (see later). Syncope is an ominous presenting sign and is associated with a higher risk of sudden death.

What is the etiology and pathophysiology of RCM?

Restrictive cardiomyopathy is characterized by abnormal diastolic compliance or filling with normal or decreased diastolic volume in the setting of normal systolic function. Increased myocardial stiffness results in impaired myocardial relaxation and ventricular noncompliance. The resulting noncompliance causes elevated diastolic pressure in both ventricles and atria. Over time, atrial dilation occurs and may lead to arrhythmias. (See Figure 33.3.) In late stages, systolic function may also fail. Echocardiography and/or cardiac magnetic resonance imaging (MRI) can be used to differentiate RCM from constrictive pericarditis that can occur in patients with RCM, specifically those with deposition-related disorders (see later).

Although the exact etiology is unknown, RCM can be secondary to medication or chest radiation, or associated with the following diseases:

Metabolic syndromes/disorders

- Carcinoid syndrome
- Fabry disease
- Gaucher disease
- Glycogen storage disease
- Hurler syndrome

Deposition-based disorders

- Amyloidosis
- Hemosiderosis

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- Hemochromatosis
- Loeffler syndrome – hypereosinophilia

What are the management options and prognosis for patients with RCM?

Unfortunately, there are no medical management strategies that directly target diastolic dysfunction. Treatment is aimed at decreasing volume overload with diuretics, reducing systemic and pulmonary venous congestion. Antiarrhythmics are used, as abnormal rhythms frequently occur with progressive atrial enlargement. Many patients require anticoagulation due to the risk of thromboembolism, particularly in the atria.

Outcomes for patients with pediatric RCM remain poor, with a transplant-free survival rate of approximately 50% at 1 year post-diagnosis. Thus, patients are referred early for cardiac transplantation. Patients with mixed RCM/HCM have a better prognosis for transplant-free survival compared to those with isolated RCM.

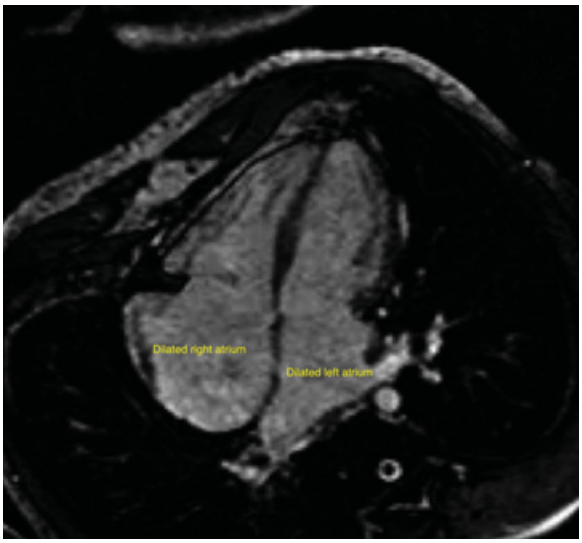

Figure 33.3 Restrictive cardiomyopathy. Four-chamber magnetic resonance imaging showing markedly dilated atria with normal ventricular sizes. The systolic function is usually normal, but diastolic relaxation is severely impaired resulting in restrictive filling of both ventricles. Courtesy of Michael Taylor, MD

Left Ventricular Noncompaction Cardiomyopathy

What is LV noncompaction cardiomyopathy?

Noncompaction cardiomyopathy (NCM) is the result of a failed process in ventricular development. Typically, the fetal LV trabeculations fill in during the course of development to form complete and circumferential ventricular cardiac smooth muscle, a process known as compaction. When compaction does not occur, the LV has deep trabeculations that are unsuited to the needs of a systemic ventricle. (See Figure 33.4.) These trabeculations result in poor subendocardial perfusion and ischemia.

The NCM subtype is associated with Barth syndrome and a variety of other genetic anomalies and syndromes (trisomy 13, 18, 21; Turner, Soto, and Marfan syndromes) along with neuromuscular dystrophies (Duchenne, Becker, multiminicore, and limb-girdle).

Noncompaction cardiomyopathy is associated with a progressive decline in systolic and/or diastolic function, multifactorial arrhythmias, and elevated risk of thromboembolism. In many cases, NCM occurs in association with other forms of cardiomyopathy, especially hypertrophic or dilated phenotypes.

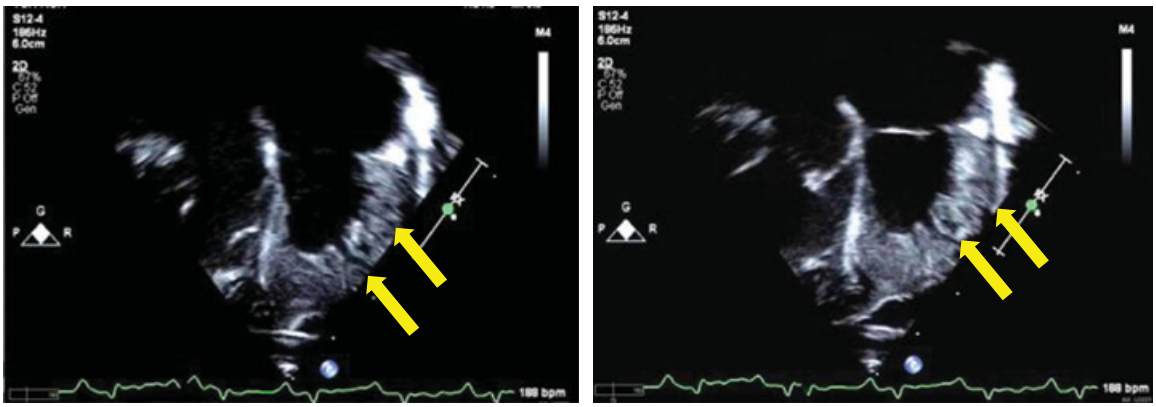

Figure 33.4 Transthoracic four-chamber view identifying deep trabeculations (yellow arrows) that are classic for LV noncompaction cardiomyopathy. Courtesy of Adam C. Adler, MD.

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Anesthetic Implications

What are the preoperative anesthetic considerations for patients with cardiomyopathy?

In addition to the perioperative considerations related to any underlying illness, preoperative evaluation in this patient population should focus on signs and symptoms associated with cardiomyopathy.

- ***Physical examination*:** Specifically, the focus should be on determining the presence and severity of dyspnea, angina, exercise tolerance, weight gain, and syncopal episodes (including frequency).
- ***Echocardiography***: A recent echocardiogram should be reviewed. Echocardiographic findings differ in patients with HCM versus RCM, but there can be overlap (e.g., HCM with restrictive physiology).
- ***Systolic function***: Many patients with HCM have hyperdynamic ventricular function. While the ejection fraction (EF) may be normal or even supranormal, this should be considered in association with the other findings (a small obliterated cavity, as seen in Figure 33.5, may completely empty, providing a normal EF despite a minuscule stroke volume). Assessment of function should also be considered given the high risk of coronary ischemia.

Clinical Pearl

In cases with ventricular cavity obliteration from myocardial hypertrophy, the EF may appear normal despite minuscule stroke volumes.

- ***Diastolic dysfunction***: Diastolic function should be reviewed in cases of HCM and RCM. The atria should be evaluated for size and volume (i.e., degree of dilation), which can reflect chronically elevated ventricular diastolic pressure and failure. Right ventricular pressure should be estimated in addition to flow patterns in the inferior vena cava and hepatic veins, as this can help differentiate RCM from constrictive pericarditis.
- ***Atrioventricular valves*** can be affected by both HCM and RCM. Septal hypertrophy in HCM can disrupt the mitral valve apparatus. In cases of RCM or late-stage HCM, ventricular enlargement can affect valve coaptation, resulting in atrioventricular valve regurgitation. Similarly, valve function can be affected as a result of ischemia.
- ***Left ventricular outflow tract***: In patients with HCM, the LV outflow tract should be examined for dynamic obstruction, including the mechanism and severity. Evaluation for systolic anterior motion of the mitral apparatus should be performed.
- ***Rhythm disorders***: A baseline electrocardiogram should be obtained along with any history of arrhythmias, as patients with prolonged diastolic failure and atrial enlargement are prone to atrial arrhythmias. Similarly, severe septal hypertrophy may disrupt the conduction pathway either mechanically or as a result of ischemia. As noted earlier, patients with cardiomyopathy are at risk for sudden cardiac death. Patients with implantable defibrillators/pacemakers should be assessed for their underlying native rhythm. (See Chapter 23, Table 23.2 for details of preoperative assessment of implanted devices.) For those requiring temporary deactivation or those at high risk of malignant arrhythmias, defibrillation pads should be placed prior to induction of anesthesia.
- ***Catheterization data***: If available, this helps inform the anesthesia provider regarding the degree of diastolic failure, degree of pulmonary hypertension, global ventricular function, coronary ischemia, and degree of LVOT obstruction.
- ***Vascular access***: Cardiomyopathy patients should have intravenous (IV) access prior to induction of anesthesia. In cases of difficult IV access or lack of patient compliance premedication may be helpful, particularly in young children. Oral midazolam or intramuscular ketamine may be used.
- ***Collaborative discussion***: A discussion should take place with the patient's primary cardiologist prior to anesthesia. If the patient is a candidate for cardiac surgical intervention, it should be performed prior to other elective interventions if possible. Additionally, if performing general anesthesia on patients with end-

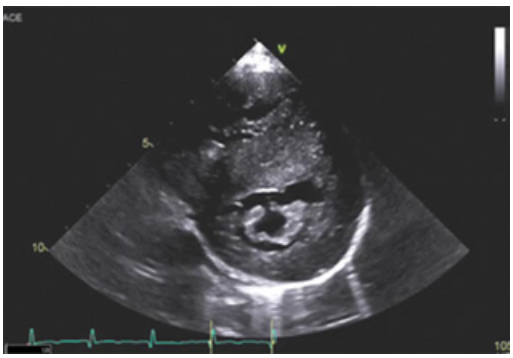

Figure 33.5 Transthoracic short axis view showing circumferential LV hypertrophy, especially in the septum, and resulting cavitary obliteration. Courtesy of Adam C. Adler, MD.

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stage cardiomyopathy, a discussion regarding extracorporeal support candidacy should occur preoperatively and in conjunction with the patient's family and primary cardiologist.

Clinical Pearl

In patients with end-stage cardiac disease, it is best to discuss management options and extracorporeal support candidacy prior to surgery for nonemergent procedures with the patient's family and primary cardiology team.

What constitutes a reasonable intraoperative plan for patients with end-stage cardiomyopathy?

The anesthetic plan should be based on the severity of the patient's cardiac illness as well as the procedural needs. Generally, most pediatric cases require general anesthesia, and it is preferable to place an airway device to ensure adequate ventilation (either a laryngeal mask airway or endotracheal tube). For procedures of shorter duration with minimal expected physiologic changes (e.g., line placement), sedation may be possible. However, it is critical to ensure the absence of airway obstruction, which can result in increased ventricular afterload. In a small subset of surgical situations, including this case, a regional technique can be proposed.

What are intraoperative anesthetic goals for patients with HCM and/or RCM?

Anesthetic goals include maintenance of normovolemia, preload and afterload. For patients with severe HCM, while the EF may remain normal, their beat-to-beat stroke volume is often minimal and dependent on adequate preload; however, ***care must be taken to avoid volume overload.***

With regard to heart rate, ***tachycardia is generally poorly tolerated***. In patients with HCM and restrictive physiology (i.e., diastolic failure), diastolic time is vital to maintain ventricular filling. Similarly, heart rate elevations are poorly tolerated with respect to the degree of LVOT obstruction. Negative inotropy is the primary medical management, usually in the form of calcium channel and ß-blockers, and should be continued perioperatively. Negative inotropy helps decrease the gradient across the LVOT, thereby reducing obstruction and promoting forward flow.

Afterload should be maintained in patients with HCM to reduce the pressure gradient across the LVOT obstruction, as well as to maintain coronary perfusion.

Anesthetic goals are similar for patients with isolated RCM and diastolic failure. While systolic function is preserved, extreme elevation in diastolic pressure from ventricular noncompliance/nonrelaxation means that large volume loads are not well tolerated. Similarly, tachycardia reduces diastolic time and ventricular filling. In the setting of significant ventricular dilation leading to mitral annular dilation and valvular regurgitation, bradycardia can also be detrimental. Ideally, the patients should be maintained at their baseline heart rate.

As mentioned earlier, dysrhythmias in cardiomyopathy patients are common and not well tolerated; for patients with a history of arrhythmia, defibrillation/pacing pads should be applied prior to induction of anesthesia.

Hypotension in HCM should be treated by augmenting preload and/or administering medication to increase afterload, while maintaining a normal heart rate. Generally, patients should undergo IV induction of anesthesia. Propofol and inhalation agents are often poorly tolerated due to the reduction in preload and afterload and compensatory tachycardia. Ketamine is a suitable induction agent, especially in the presence of opioids to reduce sympathetic activity. Similarly, an opioid-based anesthetic is generally well tolerated while preserving systemic vascular resistance (SVR).

Dilute phenylephrine should be available as a first-line therapy for hypotension. Response to inotropic drugs (epinephrine, norepinephrine) and calcium may be poorly tolerated for a variety of reasons, including increased LVOT obstruction and tachycardia. If hypotension persists, continuous infusions of phenylephrine or vasopressin should be used.

Insufflation of body cavities (thoracic or abdominal) or extremes of patient positioning (steep reverse Trendelenburg) should be performed judiciously, as they are poorly tolerated. When required, insufflation pressure should be increased gradually to allow time to assess the hemodynamic response.

When appropriate, regional or neuraxial anesthesia should be considered to blunt hemodynamic responses and potentially reduce anesthetic requirements. The use of an epidural catheter is preferable to spinal anesthesia to avoid drops in both SVR and preload. However, when using an epidural, initial bolus dosing of local anesthetic should be avoided. (See Table 33.1.)

Clinical Pearl

Drops in SVR, as may be experienced with spinal and epidural anesthesia, are often poorly tolerated in patients with dynamic outflow tract obstruction. In children, the drop in SVR with caudal/epidural anesthesia is minimal.

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Table 33.1 Pathophysiology Characteristics of Cardiomyopathies

	Dilated Cardiomyopathy (Ischemic and Nonischemic)	Hypertrophic Cardiomyopathy (Obstructive and Nonobstructive)	Restrictive Cardiomyopathy
Diastolic function and atrial transport function	• Biventricular diastolic dysfunction • Reduced biventricular compliance • Biatrial enlargement • Increased atrial pressures	• LV diastolic dysfunction • Reduced LV compliance • Left atrial enlargement • Increased left atrial pressure	• Severe biventricular diastolic dysfunction • Severely reduced biventricular compliance • Biatrial enlargement • Increased atrial pressures
Ventricular systolic function	• Decreased biventricular contractility; usually limited to LV in ischemic form • Exhausted preload reserve • Afterload mismatch	• Preserved or hyperdynamic LV contractility • Normal RV contractility	• Preserved biventricular contractility
Pulmonary vasculature	• Elevated LAP initially leads to passive elevation of PAP • Chronic elevation of LAP leads to reversible then irreversible elevation of PVR	• Elevated LAP initially leads to passive elevation of PAP • Chronic elevation of LAP leads to reversible then irreversible elevation of PVR	• Elevated LAP initially leads to passive elevation of PAP • Chronic elevation of LAP leads to reversible, then irreversible elevation of PVR
Effect of preload alterations on hemodynamics	• Exhausted preload reserve; little or no ability to recruit SV with preload augmentation	• Retained preload reserve but reduced LV compliance results in elevated LAP with preload augmentation • In obstructive form reduced LV volumes will worsen obstruction	• Baseline LV and RV EDV are low • Severely reduced biventricular compliance results in markedly elevated atrial pressures with minimal preload augmentation
Effect of heart rate alterations on hemodynamics	• Avoid bradycardia; fixed SV due to exhausted preload reserve • Tachycardia unlikely to reduce LV filling but subendocardial perfusion may be compromised in ischemic form	• Avoid tachycardia; with retained preload reserve, LV filling will be compromised, subendocardial perfusion may be compromised • Bradycardia is well tolerated	• Avoid bradycardia; fixed SV due to severely reduced LV compliance • Tachycardia may reduce LV filling resulting in reduced SV
Effect of afterload alterations on hemodynamics	• Avoid increases in SVR; large reduction in SV will occur due to reduced contractility • SVR reduction is mainstay of medical management	• Avoid decreases in SVR particularly in obstructive form; reduced LVESV will worsen obstruction • Increases in SVR well tolerated	• Avoid decreases in SVR; with fixed SV, hypotension will result • Increases in SVR well tolerated

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EDV, end-diastolic volume; LAP, left atrial pressure; LV, left ventricle; LVESV, left ventricular end-systolic volume; PAP, pulmonary artery pressure; PVR, pulmonary vascular resistance; SV, stroke volume; SVR, systemic vascular resistance; RV, right ventricle.

What is distraction and how can it be used?

Distraction is a technique that can replace, reduce, or augment procedural sedation for painful or anxiety-provoking procedures. Distraction works on the theory that the brain has limited ability to process sensory input and using deliberate stimuli to bombard the brain results in lesser ability for the brain to process other incoming stimuli.

Distraction can be passive or active and has been proven effective at allaying anxiety, fear, and pain in procedures ranging from small (IV placement) to more invasive (as done for this case when a neuraxial technique is offered). (See Table 33.1.) In younger children, a degree of IV sedation may also be required, while older children may tolerate procedures with distraction alone. For select procedures, adjuncts like topical or subcutaneous local anesthetic combined with distraction may be suitable and allow for avoidance of general anesthesia in challenging cardiac patients.

Clinical Pearl

Distraction techniques are designed to block the brain's input of sensory stimuli and can significantly help with painful procedures such as IV placement. Audiovisual distractors (video games, iPads, etc.) work well in children and adolescents.

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Table 33.2 Commonly Employed Forms of Procedural Distraction by Patient Age

	Cognitive Distraction		Behavioral Distraction	
	Active	**Passive**	**Active**	**Passive**
Young children	Audiovisual devices Interactive games	Music		
Adolescents	Singing Counting aloud Audiovisual devices	Music	Breathing	Reimagination of pain Emotive imagery

From A. C. Adler. With permission.

Figure 33.6 (Left) **Preoperative placement of caudal block using patient audiovisual distraction and (right) intraoperative patient positioning, proving patient comfort and continued use of audiovisual distraction during surgery.** Courtesy of Adam C. Adler, MD, MS.

What is the recovery plan for patients with cardiomyopathy following anesthesia?

Patients with end-stage cardiomyopathy have a high perioperative rate of cardiovascular complications. For minimally invasive procedures in relatively stable patients, anesthetic recovery should occur in a post-anesthesia care unit (PACU) that is familiar with pediatric and pediatric cardiovascular patients. When post-anesthesia admission is required, admission to the cardiology floor should be considered. Similarly, for more invasive surgeries and/or patients who are less stable, the ideal place for recovery is the cardiovascular intensive care unit. In situations in which the patient is admitted to the surgical floor or pediatric intensive care unit, the cardiology or heart failure team should be consulted for optimization of medical management.

Pain control in the immediate postoperative period is vital to minimize or reduce the incidence of heart failure and/or ischemia. Pain management with continuous infusion or patient-controlled analgesia pumps or via continuous regional or neuraxial techniques should be considered. Consultation with the pain management team can provide recommendations for analgesic adjuncts, as well as bridging to oral pain medications.

Case Completion

After discussion with the primary cardiologist and surgeon, it was decided that while the surgery should be done due to the near certain risk of reincarceration, it was felt that the risk of general anesthesia was unacceptably high. Unfortunately, the patient was not a cardiac surgical candidate for myectomy, transplantation, or extracorporeal support. The decision was made to proceed with a caudal block and audiovisual distraction using the patient's iPad device. The parents consented to give very light sedation if necessary but preferred to abort the procedure if this plan failed rather than convert to general

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anesthesia. Topical anesthetic was placed over the sacrum and a dense caudal block was performed using 0.25% bupivacaine. The patient was placed supine with cushions under his back in a position allowing him to watch the audiovisual device. (See Figure 33.6.) He received IV dexmedetomidine in 2-mcg aliquots for restlessness toward the end of the procedure. After PACU recovery, he was monitored by the cardiology service until discharge.

Suggested Reading

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