Chapter 31

Duchenne Muscular Dystrophy

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Case Scenario

A 20-year-old male with Duchenne muscular dystrophy presents to the cardiac catheterization laboratory for automatic implantable cardioverter defibrillator insertion. He was recently evaluated for palpitations and found to have intermittent runs of nonsustained ventricular tachycardia. He follows up annually with his cardiologist. He had spine surgery at age 12 years for scoliosis, is no longer ambulatory, and uses a wheelchair. He is also followed by a pulmonologist. Pulmonary function tests 2 weeks earlier were consistent with severe restrictive lung disease with forced expiratory volume 10% predicted, forced vital capacity 8% predicted, and pCO_2 100 mm Hg. He was started on bilevel positive airway pressure respiratory support at night, which he tolerates. His current vital signs are heart rate 110 beats/minute, respiratory rate 20 breaths/minute, blood pressure 102/64, and SpO_2 93% on room air.

His last echocardiogram, performed 11 months ago, demonstrated the following:

- *Moderate-to-severe left ventricular dysfunction (ejection fraction 30%)*
- *Posterolateral akinesis*

Key Objectives

- Discuss the cardiopulmonary morbidity associated with Duchenne muscular dystrophy.
- Describe the preoperative assessment of patients with Duchenne muscular dystrophy.
- Understand important considerations for the anesthetic management of these patients.
- Evaluate various strategies for intra- and postoperative cardiac and respiratory support of patients with Duchenne muscular dystrophy.

Pathophysiology

What is Duchenne muscular dystrophy and what is the underlying pathophysiology?

Duchenne muscular dystrophy (DMD) is a progressive neuromuscular disorder that is inherited in an X-linked recessive pattern, therefore predominantly affecting the male offspring of maternal carriers. However, nearly 30% of cases are due to random mutation rather than hereditary transmission. The incidence is approximately 1:5000 to 1:3500 live births. Duchenne muscular dystrophy is caused by a mutation on Xp21 that results in the absence of dystrophin, a structural protein critical to the dystroglycan complex found in skeletal and cardiac muscle cells. This complex stabilizes the muscle cell membrane during contraction. Without the dystroglycan complex, the cellular membrane is fragile, leading to cell damage and eventual necrosis over time. The subsequent scarring with fatty infiltration and fibrosis results in progressive muscular weakness and dysfunction.

What is the typical timeline for presentation and progression of symptoms in patients with DMD?

Duchenne muscular dystrophy is characterized by progressive muscle weakness and damage that impacts both skeletal and cardiac muscle tissue. Symptoms typically manifest as clumsiness, weakness, or failure to meet gross motor milestones by 3–5 years of age. Loss of ability to ambulate usually occurs around age 12 years. Cardiac muscle damage and fibrosis lead to clinically evident cardiomyopathy in the mid-to-late teen years, though evidence of cardiac damage can be seen much earlier on cardiac magnetic resonance imaging (MRI) and electrocardiogram (ECG) evaluations. Chest wall and diaphragmatic weakness eventually result in respiratory insufficiency and the need for respiratory support in the late teens to early 20s. In the past, respiratory insufficiency and infections were the leading cause of death for patients with DMD, but now, with early noninvasive respiratory support, heart failure and arrhythmias are often life limiting.

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What are the cardiac implications of DMD?

The absence of dystrophin in cardiac muscle cells causes gradual damage and destruction of cardiac muscle fibers and increasing fibrosis. Patients develop a progressive form of cardiomyopathy with a steady decline in cardiac function and ejection fraction (EF). Diagnostic studies can reveal early myocardial changes in asymptomatic children. For instance, ECG abnormalities have been noted in >50% of children <5 years of age. Early fibrosis and decrease in cardiac strain capacity can be seen on cardiac MRI within a similar age group. Echocardiographic and clinical evidence of cardiomyopathy typically present in 50% of patients by age 15, though clinical symptoms are not always readily apparent in nonambulatory patients. More than 90% of patients will have clinically significant cardiomyopathy by age 18.

The dilated cardiomyopathy seen in patients with DMD is characterized by systolic and diastolic dysfunction. Diastolic dysfunction usually precedes systolic dysfunction and may be present even in young children. Over time, EF decreases and cardiac output (CO) falls. The ventricles are often dilated with elevated left atrial and ventricular end-diastolic pressures, potentially resulting in valvular dysfunction (mitral or tricuspid regurgitation). The myocardium may also become increasingly arrhythmogenic; this can manifest as simple ectopy, atrial or ventricular dysrhythmias, and heart block.

What is the current treatment for patients with DMD? Are there special considerations for the perioperative period?

The cardiomyopathy associated with DMD is often treated with a variety of medications. These can include cardioselective ß-blockers to prevent excessive sympathetic activation, angiotensin converting enzyme inhibitors (ACEi) or angiotensin receptor blockers (ARBs) for afterload reduction, diuretics to reduce volume overload, antiarrhythmics, and occasionally inotropic support.

- ***Corticosteroids*** are used to preserve muscle strength and prolong functional abilities such as walking. Depending on the dose, duration, and dosing regimen, they can be associated with numerous side effects, including iatrogenic adrenal insufficiency and fracture risk. For more invasive procedures, use of a perioperative stress dose of steroids should be considered.
- ***ß-blocker***: ß-blocker therapy should be continued in the perioperative period to avoid reflex tachycardia, unless discontinued by the electrophysiologist in preparation for the procedure.
- ***Afterload reduction***: ACEi or ARBs should be withheld for 24 hours prior to anesthesia or sedation to prevent hypotension.
- ***Diuretics***: Chronic diuretic therapy can lead to significant electrolyte disturbances, especially hypokalemia, and should prompt preoperative electrolyte checks. Vasodilation during anesthesia or sedation often unmasks latent hypovolemia and can result in significant hypotension, requiring careful fluid resuscitation and temporary vasoconstrictor therapy.
- ***Antiarrhythmics***: Generally, antiarrhythmics should be continued in the perioperative period, but occasionally electrophysiologists prefer to withhold longer acting medications to facilitate testing during the procedure. Close communication with the cardiologist is important.
- ***Inotropic support***: Some patients are on long-term home inotropic support with milrinone, which can cause platelet dysfunction. If possible, central access dedicated to this medication should not be disconnected or used for anesthesia induction, as an inadvertent milrinone bolus can result in significant hypotension that is difficult to treat.
- ***Destination therapy with left ventricular (LV) assist devices***: While DMD patients are not candidates for heart transplantation, under special circumstances ventricular assist devices may be implanted to improve the remaining quality of life.

What are the pulmonary implications of DMD?

Duchenne muscular dystrophy causes progressive destruction and weakness of the respiratory muscles, a decline in total lung capacity, and eventually the inability to maintain adequate tidal volumes. As the disease advances, patients typically require noninvasive respiratory support only at night initially, but later it may also be necessary during the day. The need for continuous respiratory support may prompt placement of a tracheostomy for ongoing mechanical ventilatory support. It is important to clarify the patient's respiratory status at the time of surgery and discuss their plans for future respiratory support.

Cough and airway clearance ability are frequently impaired. Some patients use cough assist devices to prevent atelectasis and mucous plugs. Weakness of the oropharyngeal muscles results in decreased ability to manage secretions and increased risk of aspiration. Patients with DMD frequently suffer from recurrent pneumonias and respiratory tract infections.

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Anesthetic Implications

How should this patient's cardiac function be evaluated preoperatively? What abnormalities might be expected?

Preoperative evaluation of cardiac function should include a baseline ECG to screen for arrhythmias or conduction abnormalities and a recent echocardiogram or cardiac MRI to assess ventricular and valvular function.

Left ventricular hypertrophy is the most common finding in young DMD patients, with the ECG demonstrating Q-waves in lead III or V6. Older patients typically have deep Q waves in leads I, aVL, V5, and V6 with right ventricular (RV) hypertrophy and a shortened PR interval. A pseudo-infarction pattern consistent with a posterior or posterolateral infarct is common. Many patients develop persistent sinus tachycardia, defined as a heart rate >100 beats/minute after 12 years of age. This tachycardia is termed "disordered automaticity" and is presumably due to a sympathetic imbalance or impaired autonomic function. Atrial fibrillation, atrial flutter, and ventricular tachyarrhythmias are also common.

An echocardiogram should be performed to evaluate the degree of ventricular dysfunction and potential valvular involvement. Left ventricular posterior wall thinning, posterolateral akinesis, LV dilation and decreased shortening/ejection fraction are typical findings. However, many DMD patients are technically challenging to scan, as thoracic deformities result in poor acoustic windows and suboptimal image quality.

A cardiac MRI (CMR) is currently the preferred imaging modality for basic cardiac assessment in patients with DMD. It can provide more detailed information about ventricular function and myocardial strain patterns, and, with late gadolinium enhancement, allow for early detection of myocardial fibrosis, well before the onset of clinical manifestations. Furthermore, unlike echocardiography, CMR is not affected by acoustic windows or lung artifact. Unfortunately, previous spine surgery with extensive instrumentation often prevents adequate CMR imaging in many DMD patients.

Preoperative assessment should also include a discussion with the patient about any recent functional changes, cardiac symptoms, or alterations to their medication regimen. It is important to note, however, that even when significant cardiomyopathy is noted on imaging studies symptoms are often absent due to the patient's limited ambulation and physical activity.

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How should this patient's pulmonary function be evaluated?

Preoperative assessment of pulmonary function should include a thorough discussion of the patient's current respiratory status, their baseline requirement for respiratory support and oxygen therapy, and their ability to lie flat.

Baseline oxygenation and gas exchange, lung volumes, and the patient's ability to cough and clear the airway have significant implications for perioperative management. Oxygenation can be assessed using pulse oximetry. Inadequate oxygen saturations on room air may be an early sign of respiratory insufficiency and an indication for initiation of ventilatory support. Oxyhemoglobin saturations lower than 95% on room air should prompt further evaluation. Awake carbon dioxide tension measured with capnography or blood gas sampling (arterial or venous) can determine the presence of hypoventilation with hypercarbia.

In addition, spirometric pulmonary function tests (PFTs) should document forced vital capacity (FVC), forced expiratory volume (FEV_1), maximal mid-expiratory flow rate, maximum inspiratory and expiratory pressures, and peak cough flow. According to the American College of Chest Physicians consensus statement on respiratory support in DMD, a decreased FVC may be associated with an increased risk of postoperative respiratory failure. Patients with an FVC < 50% predicted have an increased risk of respiratory complications while those with an FVC < 30% predicted are at high risk of complications. For high-risk DMD patients, preoperative training in noninvasive ventilation should be considered to facilitate extubation and the transition to noninvasive positive pressure ventilation (NPPV) following surgery. A measured peak cough flow <270 L/minute identifies patients with ineffective airway clearance who could benefit from cough assistance techniques and should also be preoperatively trained with a mechanical insufflation–exsufflation device.

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Patients with DMD and an FVC <30% predicted are at high risk for postoperative complications and should receive preoperative training in noninvasive ventilation.

Are there other implications for DMD patients?

Many patients with DMD also suffer from gastrointestinal symptoms and can present with poor nutritional status. Gastric paresis and intestinal dysmotility lead to frequent vomiting, inadequate oral intake, and constipation. Adequate nutrition is important for respiratory function (increased work of breathing) and for wound healing. In bilevel positive airway pressure dependent patients, it can often be achieved only with special enteral formulations administered through a gastrostomy tube.

Significant contractures and poor mobility can result in development of pressure ulcers and increased risk for fractures. Positioning can be very difficult and requires careful attention to adequate padding of all pressure points.

What other issues should be discussed with the patient/family?

The preoperative discussion should address goals of care and resuscitation preferences. The patient's wishes regarding prolonged dependence on mechanical ventilation should be documented and respected. If not already done, this is also the appropriate time to offer assistance with advanced directives, living wills, and the identification of a medical proxy. Ideally the discussion should include both the patient and their designated medical proxy to avoid any confusion or uncertainty. Finally, prior to any procedure, there should be a clear understanding and agreement between members of the patient's care team whether advanced resuscitative techniques and hemodynamic support options such as extracorporeal membrane oxygenation (ECMO) and/or LV assist device placement are medically possible and desired by the patient and family.

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Do patients with DMD carry an increased risk for developing malignant hyperthermia?

There is no association between DMD and risk of malignant hyperthermia. Patients with DMD have the same risk of malignant hyperthermia as the general population. However, DMD is associated with a significant risk of rhabdomyolysis and hyperkalemia during exposure to volatile anesthetics and succinylcholine. Use of volatile anesthetics in this patient population remains controversial. While numerous case reports of rhabdomyolysis with even low-level volatile anesthetics have been published, many volatile anesthetics have been performed in patients with DMD without any evidence of rhabdomyolysis. The exact mechanism is unknown, but it is possible that volatile anesthetics can induce additional destabilization of the cellular membrane in patients with DMD. The risk of rhabdomyolysis seems to decrease with increasing age, most likely due to loss of muscle mass and increasing fibrosis.

Given the risk of rhabdomyolysis and hyperkalemia, volatile anesthetics are generally avoided in this population. Succinylcholine should never be administered to a patient with DMD because of the likelihood of a hyperkalemic cardiac arrest. Total intravenous anesthesia is the preferred technique. If intravenous (IV) access is unattainable, even with oral or intramuscular premedication and nitrous oxide (which is safe), a short exposure to a volatile anesthetic for induction and venous access may be acceptable after careful risk/benefit evaluation. Prolonged exposure, however, is not recommended.

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Duchenne muscular dystrophy is NOT associated with an increased risk for malignant hyperthermia, but exposure to volatile anesthetics and succinylcholine can cause significant rhabdomyolysis and hyperkalemia.

What are some of the considerations for using monitored anesthesia care for this procedure?

Automatic implantable cardioverter–defibrillator (AICD) insertion is a minimally invasive procedure that typically requires transvenous access to the heart via the subclavian or cephalic vein as well as the creation of a subcutaneous pocket for the generator. This can often be performed under local anesthesia with IV sedation as needed. The duration of the procedure can vary significantly, depending on the time it takes to find acceptable myocardial sites for the intracardiac electrodes and the testing of the system. Occasionally,

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the presence of extensive myocardial fibrosis, an "irritable" myocardium, and poor cardiac function necessitate multiple attempts. Furthermore, rescue defibrillations can have a negative impact on the already borderline cardiac function.

Depending on the patient's baseline functional status and whether she or he is amenable, the administration of minimal sedation with midazolam, ketamine, or dexmedetomidine, and local anesthetic infiltration by the proceduralist may allow for device placement without significant negative impact on cardiopulmonary function. However, this patient is 20 years old and has experienced significant progression of his neuromuscular disease, currently requiring noninvasive respiratory support overnight. His baseline SpO_2 on room air is only 93%, a sign of borderline respiratory function. His preoperative PFT results clearly put him in a high-risk category for respiratory complications. Any level of sedation can further impair his respiratory status by decreasing his upper airway muscle tone and leading to airway obstruction. During a procedure in the supine position requiring sedation for device testing he will benefit from the use of NPPV. The potential need for more invasive ventilation strategies (i.e., intubation) in response to complications should be discussed during the preoperative evaluation and consent process.

From a cardiac perspective, patients with heart failure often require elevated levels of circulating catecholamines to maintain cardiac output. Even minimal sedation can decrease sympathetic activation and result in inadequate perfusion. The circulation time is often prolonged, and the onset of medication effects can be delayed. Sedatives should be given in small increments and carefully titrated to effect. Some patients may require low-dose inotropic support (e.g., epinephrine 0.02 mcg/kg/minute) to allow for adequate sedation. On the other hand, untreated anxiety and pain can cause increases in afterload and/or ectopy that would be poorly tolerated by a failing ventricle.

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Patients with cardiomyopathy can have a significantly prolonged circulation time and thus the onset of medication effects is often delayed. Sedatives should be given in small increments and carefully titrated to effect.

What kind of vascular access and monitoring is indicated?

Severe muscle atrophy and contractures can complicate peripheral vascular access. Anesthesia providers should allow for additional time and be prepared to use ultrasound guided techniques. The placement of additional vascular access for inotropic support should always be considered, especially in the catheterization laboratory, where access to the patient is extremely limited once the procedure has begun. Ultrasound guidance can also be useful to identify possible central venous or ECMO cannulation sites in the event of complications.

The need for invasive monitoring depends on the patient's respiratory and cardiac status. An arterial line provides accurate blood pressure readings during prolonged periods of ectopy or low cardiac output states and facilitates respiratory monitoring.

What are the considerations for induction and maintenance of a general anesthetic if this patient is unable to tolerate the procedure under sedation?

The need for general anesthesia in case of failed sedation and/or procedural complications should always be anticipated. Some patients, especially if untrained, are unable to tolerate the facemask for NPPV. Syringe pumps with the IV anesthetic(s) of choice, vasoactive infusions, and intubation equipment should be readily accessible. Given the limited access to the patient in the catheterization lab, it is best to make the decision to induce general anesthesia before the patient is fully draped and positioned, otherwise the procedure should be stopped and access to the patient optimized. Preoxygenation with 100% FiO_2 either via facemask or the noninvasive ventilation device is very important. The primary goal during anesthetic induction is maintenance of cardiac output by avoiding ventricular depression, hypotension, and tachycardia. Depending on the patient's cardiac status, it may be beneficial to start inotropic support prior to induction. Owing to their relatively minimal hemodynamic effects, ketamine and etomidate are commonly used induction agents. It is important to note that ketamine, while typically well tolerated, does have direct myocardial depressant effects that can manifest in patients with significant cardiomyopathy and chronic sympathetic activation. Propofol is an alternative but should be titrated slowly in small doses to effect to minimize its vasodilating and cardiodepressant properties. Anesthetic maintenance can be accomplished with a total IV anesthetic technique using a carefully titrated propofol infusion. Adjuvant dexmedetomidine or ketamine infusions can be helpful to decrease propofol requirements. If local anesthesia is not providing adequate analgesia, a short-acting opioid infusion such as remifentanil can be added.

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What options exist for respiratory management of this patient intraoperatively under sedation or general anesthesia?

If the patient is not at increased risk for aspiration, tolerates the facemask, and has adequate ventilation and gas exchange with acceptable pressure support, a noninvasive ventilation device can be used. Patients who use NPPV at home should always be instructed to bring their system to the hospital to use in the perioperative period when allowed.

In case of airway obstruction due to poor upper airway muscle tone or inadequate mask seal, a laryngeal mask airway (LMA) could be an alternative. For prolonged and more invasive procedures, and/or patients who are at risk for aspiration or have restrictive lung disease, a secure airway with an endotracheal tube is the best option, as it also allows clearance of secretions and mucus.

Are there any concerns about intubation in patients with DMD?

Compared to the general population, patients with DMD are at increased risk for difficult laryngoscopy. Progressive fibrotic infiltration of oropharyngeal and cervical muscles can lead to tongue hypertrophy, restricted mouth opening, and limited neck movement. Difficult airway equipment such as a video laryngoscope or a flexible fiberoptic bronchoscope should be readily available.

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Patients with DMD are at increased risk for difficult laryngoscopy due to tongue hypertrophy, restricted mouth opening, and limited neck mobility.

What are the implications of utilizing neuromuscular blocking drugs in patients with DMD?

Depolarizing neuromuscular blocking drugs such as succinylcholine are absolutely contraindicated in patients with DMD due to the risk of hyperkalemic cardiac arrest secondary to profound rhabdomyolysis. Nondepolarizing neuromuscular blocking drugs may be used with caution if muscle relaxation is necessary. The onset, duration, and time to recovery is significantly prolonged in patients with DMD. Given the increased risk of respiratory insufficiency after anesthesia, maintenance and full recovery of the remaining muscle function are critical. Therefore, neuromuscular blockade is often avoided. Adequate intubating conditions can be achieved with propofol and remifentanil. Alternatively, for patients with severe cardiomyopathy who are unable to tolerate intubating doses of propofol, rocuronium can be used, but the onset of full relaxation will be delayed. Several case reports have described the successful use of sugammadex for reversal of rocuronium-induced neuromuscular blockade in patients with DMD.

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Succinylcholine can cause hyperkalemic cardiac arrest and is absolutely contraindicated in patients with DMD. *Nondepolarizing neuromuscular blocking drugs should be used with caution as the onset, duration, and time to recovery are significantly prolonged.*

What intraoperative hemodynamic changes should be anticipated?

Even minimal sedation can lead to hemodynamic instability in a patient with significant cardiomyopathy. Endogenous catecholamine secretion is typically elevated at baseline and can be affected by sedative medications or general anesthesia resulting in decreased cardiac output, vasodilation, and hypotension. Early inotropic support may be required; consider epinephrine (0.02–0.04 mcg/kg/minute), dopamine (5–10 mcg/kg/minute), or milrinone (0.25–0.5 mcg/kg/minute). However, depending on the extent of myocardial fibrosis and cardiomyopathy, the heart may have limited ability to increase contractility.

Although beneficial from a respiratory standpoint, positive pressure ventilation can decrease LV wall tension and preload to the RV. Depending on the patient's volume status and the degree of diastolic dysfunction, slow and careful fluid resuscitation may be necessary.

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What complications may occur during pacemaker or AICD placement?

Possible complications during the procedure may be divided into access- and device-related problems:

- ***Access-related complications*** can include pneumo- or hemothorax, air embolism, and later, thrombosis. The risk of intraoperative pneumothorax varies from 0.6% to 5% (average 2%) depending on the procedural approach. The highest risk of pneumothorax occurs with a blind subclavian approach. Use of fluoroscopy during subclavian access somewhat decreases this risk. Vascular access via the cephalic vein, axillary vein, internal jugular vein, or femoral vein carries a lower risk. Given the impaired respiratory status of patients with DMD, pneumothorax is poorly tolerated and must be recognized and treated expeditiously.
- ***Device-related complications*** can be caused by the leads or the generator. Arrhythmias during lead placement are common but generally self-limited. Recurrent arrhythmias without adequate recovery periods (e.g., during the testing phase) can lead to significant hemodynamic compromise and may require treatment. In rare occasions, perforation of the atrial or ventricular wall by the pacing wires cause pericardial effusion and possible tamponade. The risk is increased during the placement of atrial (thin wall) or biventricular leads for cardiac resynchronization therapy.

What are the postoperative respiratory risks for patients with DMD? How can these be mitigated?

Patients with DMD are at high risk of postoperative respiratory failure. The lowest level of sedation necessary should be used during the procedure. Whenever possible, pain control should be achieved with local anesthesia and regional techniques to limit the use of opioids and the risk of respiratory depression. Noninvasive respiratory support can be extended into the postoperative period until the patient returns to his baseline level of respiratory function. If intubation or LMA placement was necessary, transition to NPPV should occur immediately after removal of the airway device for patients who use NPPV chronically. For all other patients, transition to NPPV after extubation or LMA removal should be strongly considered, particularly for those with FVC <50% predicted. Supplemental oxygen should be carefully titrated to the lowest level necessary to maintain adequate saturations; otherwise, hypoventilation, poor clearance of secretions, and atelectasis are easily masked.

Does this patient require an intensive care bed postoperatively?

The level of care during the postoperative recovery strongly depends on the degree of residual sedation, analgesic requirements, and need for respiratory or hemodynamic support. A back-up intensive care unit (ICU) bed should be available. If the patient was able to tolerate the procedure with minimal sedation and returns to his baseline status, extended recovery and inpatient overnight observation for late cardiac or respiratory complications is reasonable. Many hospitals will allow patients on stable chronic noninvasive respiratory support to return to the floor. Patients who required intubation or LMA placement and were extubated to NPPV support should be admitted to an ICU or stepdown unit where therapeutic respiratory modalities are more readily available. For some patients, the surgical procedure may unmask the need for a higher level of respiratory support with mechanical ventilation or prolonged NPPV.

How should postoperative pain be treated?

The use of opioids should be minimized whenever possible to reduce respiratory depression and cough suppression. A multimodal analgesic approach to pain control is optimal, using acetaminophen, nonsteroidal antiinflammatory agents, and local anesthesia. Pectus or intercostal nerve blocks can be useful regional anesthesia techniques to provide long-lasting analgesia after pacemaker placement.

Suggested Reading

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