Chapter

49

Long QT Syndrome

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Case Scenario

A 16-year-old female with congenital long QT syndrome presents for endoscopic nasal surgery due to multiple sinus infections requiring antibiotics and steroids. She was diagnosed with long QT syndrome at age 13 years after having a syncopal episode while running track. An electrocardiogram showed a heart-rate corrected QT interval of >500 ms while transthoracic echocardiography revealed a structurally normal heart with normal function. Genetic testing confirmed the diagnosis of long QT2. She was started on β-blockade and despite compliance experienced additional syncopal episodes. She underwent implantable cardioverter–defibrillator placement 18 months ago and had significant postoperative nausea and vomiting after that procedure. Since then, she has had no further syncopal episodes and continues on nadolol as her only medication. Her implantable cardioverter–defibrillator was last interrogated 6 months prior and was functioning normally at that time.

Key Objectives

- Understand the major subtypes of congenital long QT syndrome.
- Discuss the anesthetic implications of long QT syndrome.
- Describe the preoperative evaluation and perioperative management of a patient with a cardiovascular implantable electronic device.
- Describe the surgical implications for a patient with a cardiovascular implantable electronic device.

Pathophysiology

What is long QT syndrome?

Congenital long QT syndrome (LQTS), with an estimated prevalence of 1 in 2,000 to 2,500, is a group of genetically transmitted disorders characterized by abnormal cardiac repolarization resulting in QT interval prolongation that predisposes patients to the acute onset of ventricular arrhythmias, most notably torsades de pointes (TdP), which may cause syncope or sudden cardiac death (SCD) [1, 2]. The ion channel dysfunction that prolongs cellular repolarization is most often caused by decreased outward potassium current I_{Ks} (LQT1, LQT5) or I_{Kr} (LQT2, LQT6), or by enhanced activity of mutant inward sodium current (LQT3). Long QT syndrome is usually transmitted in an autosomal dominant pattern. Patients who carry two abnormal LQTS genes usually demonstrate a more severe clinical phenotype, characterized by longer QT interval prolongation and a higher risk of syncope and SCD.

How is a prolonged QT interval defined?

A corrected QT interval may be determined using Bazett's formula (QT interval divided by the square root of the R–R interval). The normal range of heart-rate-corrected QT intervals (QTc) varies by age and gender. Prolonged QTc is defined as >450 ms in males and >460 ms in females. With increasingly longer QTc values, the pretest probability of a patient having an LQTS-causing mutation also increases. Genetic testing for LQTS is a Class I indication for asymptomatic, postpubertal individuals if an otherwise idiopathic QTc ≥500 ms (≥480 ms prepuberty) is detected and persists on serial ECGs [3]. Increasingly prolonged QTc values portend a higher risk of potentially lethal arrhythmic events [4].

What are the presenting signs of LQTS?

Diagnosis remains challenging as roughly 40% of patients with genotype-positive LQTS do not demonstrate QT prolongation on resting ECG [5]. Clinical manifestations are heterogenous and include presyncope, syncope, aborted cardiac arrest, cardiac arrest, and SCD. Many patients are completely asymptomatic. The first clinical manifestation of LQTS is SCD in 10%–12% of patients. Left untreated, the prognosis is poor with a 21% mortality rate within 1 year of the first syncopal episode [1, 6]. Treatment dramatically reduces the risk of cardiac events and SCD.

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Clinical Pearl

The first clinical manifestation of LQTS is sudden cardiac death in 10%–12% of patients. Increasingly prolonged QTc values portend a higher risk of potentially lethal arrhythmic events.

What are the common types of LQTS and their clinical manifestations?

The first genes implicated in congenital LQTS were discovered in the mid-1990s and since that time more than 600 mutations in 14 susceptible genes have been identified. These mutations generally involve either ***loss-of-function potassium channel*** mutations or ***gain-of-function sodium channel*** mutations. With rare exception, LQTS is a pure "channelopathy" resulting from mutations in cardiac channel α- and β-subunits. Three genotypes, LQT1, LQT2, and LQT3, account for 75%–95% of cases of congenital LQTS [2, 7].

Long QT1 (LQT1) is associated with mutations in *KCNQ1*, the slowly activating component in the delayed rectifier potassium current channel (I_{Ks}). A properly functioning channel's current is induced by sympathetic activation and is essential for QT shortening with increases in heart rate (HR). Loss-of-function mutations in the *KCNQ1* gene create a substrate in which the defective channel is unable to adapt to β-adrenergic stimulation. Therefore, these patients classically have a broad-based T wave and are ***most likely to suffer events triggered by stress, exercise, or sudden increases in sympathetic tone.*** β-Blockade is very effective therapy for patients with LQT1 and can be instrumental in mitigating the risk of TdP in these patients. Compared to other genotypes, LQT1 is associated with a shorter, frequently normal QT interval; a lower cumulative cardiac event rate; and a lower incidence of cardiac arrest or SCD. With appropriate β-blockade, the cardiac event rate may be reduced to as low as 1.2%–2% annually [1, 2, 8].

Long QT2 (LQT2) is caused by a mutation in the *KCNH2* gene, which is responsible for the rapidly activating component of the delayed rectifier potassium current. These patients tend to have a notched or low-amplitude T wave and are ***classically triggered by startle, fright, or emotion***. Sex hormones are also known to affect the risk of cardiac events in patients with LQT2, with females experiencing a significant increase in risk associated with puberty. β-Blockade is the first-line therapy in these patients but is not as effective as in patients with LQT1. Long QT2 patients have a 6%–7% risk of cardiac arrest while on β-blocker therapy. These patients are also exquisitely sensitive to potassium levels, often requiring perioperative repletion, chronic potassium supplementation, or spironolactone therapy [1, 2].

Long QT3 (LQT3) differs from the previous two types in several ways. Although both LQT1 and LQT2 are associated with mutations in the same potassium channel, LQT3 is associated with mutations in *SCN5A*, a gene responsible for encoding an inward sodium current channel. Long QT3 is often associated with a gain-of-function mutation, whereas both LQT1 and LQT2 are associated with loss-of-function mutations. Patients with LQT3 have a long, flat ST segment, a tendency toward abnormal bradycardia, and are ***most susceptible to cardiac events while sleeping or at rest,*** as the QT interval prolongs excessively with slowing of the HR. Treatment in this subgroup can be very challenging, particularly in those who present at an early age [7]. β-Blockade remains a first-line therapy despite reduced efficacy and a 10%–15% rate of major cardiac events even with medical compliance. Sodium channel blockers, such as mexiletine and flecainide, have been shown to shorten the duration of QT interval in patients with LQT3; however, studies suggest that their clinical effectiveness may be mutation specific [2].

Clinical Pearl

Patients with LQT1 classically have a broad-based T wave and are most likely to suffer events triggered by stress, exercise, or sudden increases in sympathetic tone. LQT2 patients tend to have a notched or low-amplitude T wave and are classically triggered by startle, fright, or emotion. Patients with LQT3 have a long, flat ST segment, a tendency toward abnormal bradycardia and are most susceptible to cardiac events while sleeping or at rest.

What are the genetics of LQTS and what is the role of genetic testing in LQTS?

Congenital LQTS is inherited in both an autosomal dominant and recessive pattern with variable penetrance. Whereas some gene carriers demonstrate QT prolongation, syncope, and even SCD, other gene carriers with the same mutation may show no prolongation and no symptoms. Therefore, while a family history of SCD may lead to a diagnosis of LQTS in a surviving family member, the survivor's individual risk is best predicted by his or her own history of symptoms and QT interval length [9].

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Genetic testing should be performed if congenital LQTS is suspected after considering clinical features, family history, and ECG characteristics described earlier. Exercise or pharmacologic stress testing and Holter monitoring may increase the diagnostic yield in selective cases. Asymptomatic patients should be evaluated if they have an otherwise unexpected and persistently prolonged QT interval (QTc >480 ms for prepubertal patients and >500 ms for adults). In the presence of a clinical diagnosis of LQTS, the yield for genetic testing is about 75% [3].

Clinical Pearl

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What is acquired LQTS?

Acquired QT prolongation is defined as QT prolongation and subsequent TdP caused by medications, electrolyte disturbances, or disease. Although most cases of acquired QT prolongation are not genetically based, the first manifestation of congenital LQTS may occur in the setting of acquired QT prolongation. Reversible causes of LQTS include medication, myocardial ischemia, hypothermia, and electrolyte abnormalities.

Are any clinical syndromes associated with congenital LQTS?

Timothy syndrome is a rare, often lethal form of LQTS. In addition to cardiac conduction abnormalities, including prolonged QT interval, atrioventricular block of varying degrees, and bradycardia, these patients may have syndactyly, facial dysmorphisms, and neurodevelopmental abnormalities such as autism. The most common cause of death is ventricular tachyarrhythmia at a mean age of 2.5 years. ***Jervell and Lange–Nielson syndrome*** is a rare, highly malignant autosomal recessive form of LQTS associated with bilateral sensorineural deafness [5]. ***Anderson–Tawil syndrome*** is a rare autosomal dominant disorder associated with periodic paralysis and developmental abnormalities including the face, skeleton, and limbs [10].

Clinical Pearl

Timothy syndrome is a rare, often lethal form of LQTS. Children with syndactyly should be evaluated for the presence of LQTS prior to anesthesia.

What is the overall mortality of LQTS? Are certain patient subsets at higher risk for SCD?

Annual mortality associated with LQTS is around 1% per year with highest risk subsets approximately 5%–8% per year.

Indicators of increased risk for SCD include:

- QTc >500 ms
- History of TdP-mediated syncope
- Specific genotypes
- Males and postpubertal females
- Cardiac events occurring during the first year of life

Sudden cardiac death risk in congenital LQTS patients varies by genotype: patients with LQT3 have the highest risk, followed by patients with LQT2 and finally LQT1. Patients with multiple mutations, including Jervell and Lange–Nielsen syndrome, have a particularly severe phenotype with markedly increased risk, even with treatment [11].

Patients who have their first event at <1 year of age belong to a particularly high-risk category, with a 2.3-fold increased risk of either aborted cardiac arrest (ACA) or SCD during the next 10 years compared with patients without events at less than 1 year of age [12]. Medication is less effective in this high-risk group, contributing to a poor prognosis. Alternative therapeutic interventions such as ICDs are also limited and are rarely implanted due to high complication rates in the very young.

What medications are typically used to treat LQTS?

The mainstay of therapy for congenital LQTS is medical management with β-blockade and avoidance of known triggers.

- ***Long QT1***: β-Blockade (nadolol or propranolol) is extremely effective at reducing syncope, ACA and SCD [5]. With compliance, the mortality rate may be as low as 0.5%, with a rate of ACA near 1%.
- ***Long QT2***: The risk of syncope despite β-blocker therapy is 40%. Cardiac arrest risk remains high at 6%–10%; however, most of these patients are able to be resuscitated.
- ***Long QT3***: Patients are most refractory to β-blockers with a 10%–15% rate of cardiac arrest. Cardiac events in LQT3 patients are also more likely to be fatal. Sodium channel blockers may be considered as standalone or concomitant therapy with propranolol, as they markedly shorten the QT interval in some patients, reducing the cardiac event rate [8].

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Do additional nonpharmacologic therapeutic options exist?

Permanent pacemaker (PM) placement may be considered in LQT1 or LQT2 patients experiencing clinically significant bradycardia while on β-blocker therapy. In addition, PM placement may be indicated in patients with LQT3 to provide HR consistency, decrease repolarization heterogeneity, and reduce the incidence of bradycardia- and pause-dependent TdP, which predisposes patients to TdP.

Left cardiac sympathetic denervation (LCSD) is an increasingly utilized surgical option for patients who either demonstrate recalcitrant symptoms or are unable to tolerate first-line medical management. This approach involves removal of the first four thoracic ganglia and may be performed either thoracoscopically or via extrapleural approach. Left cardiac sympathetic denervation may be suggested for patients with ventricular fibrillation (VF)-terminating implantable cardioverter–defibrillator (ICD) shocks, patients with cardiac events despite medical therapy, patients unable to tolerate β-blockade, and high-risk patients too small for ICD implantation who are inadequately protected with medical therapy [13].

What is CIED therapy?

Cardiovascular implantable electronic device (CIED) technology includes conventional PMs as well as ICDs with pacing capabilities. Creation of perioperative care algorithms for patients with CIEDs is complicated and perioperative advisories have been developed by the American Society of Anesthesiologists (ASA) and the Heart Rhythm Society (HRS) [14, 15].

Who should have an ICD placed?

Implantable cardioverter–defibrillator therapy should be considered in:

- History of an ACA, regardless of genotype
- Documented TdP or syncope despite adequate β-blocker therapy
- Intolerance of primary pharmacotherapy
- Prior LQTS-triggered cardiac event with excessive QT prolongation (>550 ms)
- Women with LQT2 and QT >500 ms [16]

Long QT3 patients also may benefit from ICD therapy [17]. Other high-risk subgroups include Jervell and Lange–Nielsen syndrome and patients with compound mutations. While ICDs may be lifesaving in children, inappropriate shocks and lead failures are common in this population, perhaps related to continued growth and activity placing a strain on leads.

Anesthetic Implications

What are specific preoperative considerations in LQTS patients?

Clarifying the severity of a patient's clinical presentation is essential to better assess their perioperative risk.

- ***Mutation type*: *LQT1 and 2*** patients are frequently triggered by increased sympathetic stimulation and will benefit from adequate premedication and a calm induction. ***LQT2*** patients are susceptible to sudden loud noises and should be kept in a quiet room. ***LQT3*** patients experience events at rest or during sleep and should be monitored closely during premedication and recovery from anesthesia.
- ***Medications***: Continuation of β-blocker therapy may be the most important strategy for mitigating the risk of perioperative TdP in patients with LQTS. It is important to determine what medications patients are taking and to continue these medications perioperatively.
- ***Longer QTc intervals***: These predict a higher likelihood of cardiac events. Patients should have an ECG performed prior to their procedure.
- ***Electrolytes***: Patients with LQTS are exquisitely sensitive to metabolic derangements; hypokalemia, hypomagnesemia, and hypocalcemia may predispose patients to the development of ventricular arrhythmias [18]. Patients with LQT2 are sensitive to changes in potassium and should be monitored closely for hypokalemia.
- ***Cardiovascular implantable electronic device***: For patients who have required CIED technology, understanding and planning management of their CIED is an important part of their preoperative assessment and will be discussed in the text that follows.

While medication interactions are the biggest consideration in LQTS patients, what other considerations exist?

Many of the medications administered during an anesthetic affect the QT interval; a list of medications and their effect on the QT interval may be found at www.crediblemeds.org.

Electrolyte derangements are poorly tolerated and should be monitored, particularly in procedures with significant volume shifts. It is reasonable to consider treatment with magnesium (30 mg/kg) given the low toxicity risk and stabilizing effect on the myocardium. Temperature should be monitored and modulated to keep the patient normothermic,

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as hypothermia has also been shown to prolong the QT interval and conversely, fever >39°C has been shown to develop TdP. Finally, because these patients are often receiving β-blockers, they may tolerate hypovolemia and fluid shifts poorly.

Clinical Pearl

A list of medications and their effect on the QT interval may be found at www.crediblemeds.org.

Are there special monitoring considerations in a patient with LQTS?

Ideally patients with LQTS should be fully monitored before induction of anesthesia, including a multiple-lead ECG tracing. The level of monitoring should be based on the severity of the patient's clinical condition and the procedure to be performed, with a low threshold for invasive monitoring. Central access may be helpful in the event that rapid institution of transvenous pacing is needed; however, the decision to place a central venous catheter remains largely based on procedural indications.

Are certain premedication agents safer than others in patients with LQTS?

Midazolam has not been implicated in QT prolongation. Ketamine has not been shown to prolong the QT interval; however, it is a sympathomimetic and should be avoided if possible in these patients [19]. Dexmedetomidine is being used with increasing frequency both for premedication and for sedation in the pediatric population. The risk profile of dexmedetomidine for LQTS patients is clouded by conflicting studies. One study performed in children demonstrated a prolongation of the QT interval [20], while other studies have reported a reversal of acquired and iatrogenic prolonged QT in adults and a protective effect in laboratory animals. The QT interval prolonging effect appears to be related to bradycardia, a known side effect that may be mitigated with dose adjustment and slow titration. Dexmedetomidine is likely safe when used cautiously in patients with LQTS; however, particular attention must be shown to those with LQT3 who are predisposed to cardiac events while at rest or with a slower HR.

Clinical Pearl

Dexmedetomidine is likely safe when used cautiously in patients with LQTS; however, particular attention must be shown to those with LQT3 who are predisposed to cardiac events while at rest or with a slower HR.

Are intravenous induction and maintenance agents safer than volatile anesthetic agents?

For induction, one must balance the potential stress associated with the placement of an intravenous (IV) line in an awake child against the potential risk of QT prolongation with a volatile anesthetic agent. Laryngoscopy and intubation can provoke profound sympathetic stimulation and prolong the QT if not appropriately blunted by adequate anesthetic depth. Propofol does not prolong the QT interval, does not accentuate the transmural dispersion of repolarization, and has been shown to reverse sevoflurane induced QTc prolongation in healthy patients. Etomidate has little effect on the QT interval [21].

Nitrous oxide has been used safely in case reports; however, it does possess sympathomimetic properties and should be used cautiously. When compared with commonly used IV agents, volatile agents have demonstrated the ability to prolong the QT interval when studied in healthy children. Whether this prolongation is enough to trigger an episode of TdP in LQTS patients is unclear [17]. QTc changes produced by sevoflurane are concentration dependent and can occur at clinically relevant concentrations. It has been suggested that perioperative continuation of β-blocker therapy may be sufficiently protective against volatile anesthetic agent induced QT prolongation and the development of TdP. However, LQT2 patients may be more susceptible to volatile agent induced arrhythmias than other genotypes, and volatile agents either should be avoided or used with great caution in these patients [22].

Clinical Pearl

Long QT2 patients may be more susceptible to volatile agent induced arrhythmias than other genotypes, and volatile agents either should be avoided or used with great caution in these patients.

Are nondepolarizing muscle relaxants safer than succinylcholine?

Succinylcholine should be avoided because of known QT prolongation and its propensity to cause abrupt potassium shifts. Vecuronium and rocuronium have been shown to have little effect on serum histamine concentration and do not prolong the QTc, making them the most suitable agents [23]. Anticholinergics such as atropine and glycopyrrolate prolong the QT interval in healthy patients and have been implicated in the development of TdP in patients with LQTS [17]. Bradycardia associated with neostigmine also can lead to QT prolongation and should be

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used with caution. With the availability of sugammadex, which does not prolong the QT interval, rocuronium now has the added advantage of easy reversibility with the avoidance of traditional reversal agents [24].

Clinical Pearl

Succinylcholine should be avoided because of known QT prolongation and its propensity to cause abrupt potassium shifts.

What is the appropriate management of TdP?

While TdP is the most common arrhythmia requiring treatment, LQTS patients may present with either brady- or tachyarrhythmias. Although most episodes of TdP are self-limited, prolonged episodes may be associated with hemodynamic instability, VF, and cardiac arrest. Rapid and short-acting β-blockers should be readily available to prevent and treat tachycardia in patients with LQT1 and 2. Cardiac pacing capabilities should be available to treat bradycardia in LQT3 patients. Magnesium sulfate is the first-line treatment for TdP; a 30–50 mg/kg bolus may be repeated after 15 minutes as needed, followed by an infusion. If TdP persists, temporary pacing may be attempted to terminate the arrhythmia. Defibrillation and cardiopulmonary resuscitation should be carried out swiftly should the patient deteriorate to VF. Amiodarone should not be used for VF, as it can prolong the QT interval. Lidocaine may be effective for refractory ventricular arrhythmias. While patients with acquired QT prolongation and TdP may benefit from isoproterenol or dobutamine to prevent bradycardia, these medications may worsen QT prolongation and increase the risk of arrhythmias in patients with congenital LQTS.

Clinical Pearl

Amiodarone should not be used for treatment of ventricular fibrillation, as it can prolong the QT interval.

What is the best approach to pain management in patients with LQTS?

Fentanyl, alfentanil, remifentanil, and morphine have been used safely in patients with LQTS; however, methadone and sufentanil prolong the QTc. Commonly used local anesthetics including bupivacaine, ropivacaine, and lidocaine do not significantly affect QT interval. Lidocaine actually may shorten repolarization time [21] and may prevent QTc prolongation during intubation if given at induction. Epinephrine should be avoided as an adjunct to local anesthetics because it has been shown to prolong the QT interval in patients with LQTS. Spinal, epidural, and caudal anesthesia has been used successfully in patients with LQTS. Spinal anesthesia is associated with QT prolongation in healthy patients; however, it may be advantageous because it reduces the stress response and provides dense analgesia. Epidural anesthesia may be preferred because the onset of the block is more gradual, and the risk of abrupt hypotension is lower.

What antiemetics are safe to use in patients with LQTS?

Stress associated with postoperative nausea and vomiting can be severe enough to trigger an arrhythmia in patients with LQTS, but unfortunately many commonly prescribed antiemetics have been linked to prolonged QT. A black box warning for droperidol was issued by the US Food and Drug Administration (FDA) in 2001 because of the potential for cardiac arrest precipitated by QT prolongation. The 5-hydroxytryptamine type 3-receptor antagonists (i.e., ondansetron) are highly effective antiemetics, but they have been shown to prolong the QT interval. The clinical significance of this QT prolongation is unclear. Data are lacking on the specific safety profile of ondansetron in LQTS; however, current recommendations include cautious administration at the lowest effective dose with continuous cardiac monitoring [17]. The latest US FDA recommendations state that the QT prolongation associated with ondansetron is dose dependent and that no single IV dose of ondansetron should exceed 16 mg. Metoclopramide and dexamethasone can be safely administered as part of an antiemetic regimen.

Are there specific postoperative considerations for the patient with LQTS?

Long QTS patients should have continued ECG monitoring postoperatively. Whether they are monitored in the postanesthesia care unit or intensive care unit must be determined on an individual basis, taking into account the severity of the patient's disease, the complexity of the surgical procedure, and the patient's intraoperative ECG. While there are no specific guidelines, it is reasonable to propose that these patients be monitored for at least 24 hours postoperatively. Patients need to understand the importance of resuming their home regime of medications following discharge.

Clinical Pearl

Long QTS patients need to resume their oral medication regime on discharge and let their physician know if they are not able to do so.

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Management of Implanted Devices

What are the indications for PM placement and what are the common lead configurations?

In 2008, the American College of Cardiology, American Heart Association, and HRS updated guidelines for PM implantation, including recommendations for pediatrics and congenital heart disease (CHD) [25]. Permanent PMs can be attached to the endocardium via a transvenous approach or to the epicardium via a surgical approach. The decision for a surgical approach is dependent on several factors including size of the patient and the cardiac anatomy. Regardless of the system used, it is generally believed that the greatest risk factor for damage to implanted leads is age at time of implant and the presence of CHD [26].

Pacemakers are either single chamber, dual chamber, or trichamber (cardiac resynchronization PMs) depending on the patient's size and device indication. Leads are typically placed in the right atrium (RA) and/or right ventricle (RV). In resynchronization systems, leads are usually placed in both the right and left ventricle. For endocardial systems, the LV lead is usually placed within the coronary sinus, but atypical configurations have been described in patients with CHD. In patients with single ventricle physiology, a resynchronization system may signify two epicardial leads on a single ventricle.

- ***Single chamber devices*** will sense intrinsic electrical activity from the corresponding chamber within a preset time limit, either inhibiting or triggering pacing of that chamber depending on device programming
- ***Dual chamber devices*** can sense and pace both in the atrium and ventricle, maintaining AV synchrony in patients without intact AV node function

How are the location and function of PMs described?

An internationally recognized code developed by the North American Society of Pacing and Electrophysiology (NAPSE) and the British Pacing and Electrophysiology Group (BPEG) [27] is used to describe pacemakers and is known as the NASPE/BPEG Generic (NBG) Pacemaker Code. (See Table 49.1.) The five-position code shown in Table 49.1 is often shortened to the first three positions. The dual-chamber mode is the most sophisticated and commonly used mode.

What is rate modulation?

Rate modulation, or rate adaptation, denoted by "R" in the fourth position of the NBG code, describes a pacemaker's ability to automatically change the pacing rate in response to certain monitored parameters in patients with chronotropic incompetence. This function is commonly used in patients who do not have intact sinus node function, and it enables the PM to automatically increase the HR to meet metabolic demands. Most patients with a permanent PM are programmed to the DDDR mode [28].

What is multisite pacing?

The fifth position of the NBG code conveys information regarding the performance and location of multisite pacing: pacing both atria, pacing both ventricles, or multiple pacing sites in a single chamber [28]. Biventricular pacing is a technique of using simultaneous or near simultaneous pace activation of one or both ventricles to improve ventricular dysynchrony and cardiac function.

What are the indications for ICD placement?

The first implantable cardioverter–defibrillator was implanted in 1980. Current indications include:

- Hemodynamically significant ventricular tachycardia
- Ventricular fibrillation
- Conditions associated with SCD (e.g., long QT syndrome, Brugada syndrome, arrhythmogenic RV dysplasia, and infiltrative cardiomyopathies)

Implantable cardioverter–defibrillators are also useful for primary prevention of SCD in patients with hypertrophic cardiomyopathy, post–myocardial infarction with an ejection fraction (EF) of <30%, or cardiomyopathy with an EF of <35%.

Table 49.1 Generic Pacemaker Code

Position I	Position II	Position III	Position IV	Position V
Chamber(s) paced	**Chamber(s) sensed**	**Response(s) to sensing**	**Programmability**	**Multisite pacing**
O = None A = Atrium V = Ventricle D = Dual (A + V)	O = None A = Atrium V = Ventricle D = Dual (A + V)	O = None T = Triggered I = Inhibited D = Dual (T + I)	O = None R = Rate modulation	O = None A = Atrium V = Ventricle D = Dual (A + V)

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How are ICDs coded?

ICDs also have an international generic code. (See Table 49.2.) In addition to tachyarrhythmia therapies, including defibrillation of tachyarrhythmias and pace termination of tachyarrhythmias, all transvenous ICDs are equipped with pacing capabilities. For complete identification, position IV is expanded to include all five pieces of information conveyed by its full pacemaker NBG code. For example, most devices with a rate responsive PM and ICD will be identified as VVE-DDDR.

What is important to determine during the preoperative evaluation in a patient with a CIED?

Preoperative assessment should include:

- Type of device
- Indication for placement and other coexisting cardiovascular pathology
- Determination of "pacemaker dependence"
- Determination of device function

The device identification card for the patient will include the make, model, and serial number of the device. Important information can also be obtained by referring to consult notes from the cardiologist or PM clinic.

The only reliable method of assessing battery status, lead placement, current settings, adequacy of PM/ICD function, and magnet mode is direct interrogation with a programmer.

What if the manufacturer's card or clinic notes are not available?

If no other data are available, a chest radiograph can often be utilized to identify an x-ray code that can help identify the manufacturer of the device. The chest radiograph can also help in determining the type of device (ICD vs. PM) and whether it is single, dual, or biventricular. It can also be used to determine the number, position, and integrity of leads as well as any unusual configurations of lead/generator placement and tunneling that may impact the surgical approach.

Clinical Pearl

If no other data are available, a chest radiograph can often be utilized to identify an x-ray code that can help identify the manufacturer of the device.

What is meant by the term "pacemaker-dependent"?

Pacemaker dependence can be determined by one or more of the following:

- A verbal history or medical record indication that the patient has experienced a bradyarrhythmia that resulted in syncope or other symptoms requiring CIED implantation
- A history of successful AV node ablation resulting in CIED placement
- No evidence of spontaneous ventricular activity when the pacemaker function of the CIED is programmed to VVI pacing mode at the lowest programmable rate

Who should make recommendations regarding the management of a CIED during a procedure?

Current CIED recommendations from the ASA and HRS focus on an individualized, multidisciplinary approach driven by the primary CIED management team. The best perioperative care of a patient with a CIED will result from the patient's own CIED team (or another CIED team) providing a specific prescription for CIED management to the procedural team.

How close to surgery should the CIED be interrogated?

Pacemakers should be interrogated at least every 12 months. If the PM involves CRT therapy, then interrogation should be every 3–6 months. Implantable cardioverter–defibrillators

Table 49.2 Generic Defibrillator Code

Position I	Position II	Position III	Position IV
Shock chamber(s)	**Antitachycardia Pacing chamber(s)**	**Tachycardia detection**	**Antibradycardia pacing chamber(s)**
O = None A = Atrium V = Ventricle D = Dual (A + V)	O = None A = Atrium V = Ventricle D = Dual (A + V)	E = Electrogram R = Rate modulation	O = None A = Atrium V = Ventricle D = Dual (A + V)

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should be interrogated every 6 months. Children and infants have higher resting and peak heart rates than adults, which will increase battery utilization and significantly impact the longevity of pulse generators.

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Pacemakers should be interrogated at least every 12 months. If the PM involves CRT therapy, then interrogation should be every 3–6 months. Implantable cardioverter–defibrillators should be interrogated every 6 months.

Once the device has been interrogated, what additional preoperative information will assist in management of the patient?

Preparation for patient safety and proper maintenance of the device during a procedure includes answering the following questions:

1. Is electromagnetic interference (EMI) likely to occur during the procedure?
2. Is preoperative reprogramming to asynchronous mode or disabling special algorithms (including rate adaptive functions) needed?
3. Do antitachyarrhythmia functions need to be suspended?
4. Can use of a bipolar electrocautery system or ultrasonic scalpel be considered to minimize EMI effects?
5. Are temporary pacing and defibrillation equipment available?

What is electromagnetic interference?

Electromagnetic interference can result from any device that emits radiofrequency waves between 0 and 10^9 HZ. The perioperative period is particularly problematic as patients are exposed to a number of energy sources and machinery that may generate EMI including, but not limited to, electrocautery, external defibrillation, electroconvulsive therapy, transcutaneous electrical nerve stimulation, and radiofrequency waves used in ablation procedures.

What are the potential problems associated with EMI and CIEDs?

The most common source of EMI is electrosurgical energy. Electrical current can be delivered in bipolar or monopolar configurations, and with a variety of power waveforms to produce these tissue effects. For bipolar electrosurgery, there appears to be minimal chance for an adverse CIED interaction. For monopolar electrosurgery, electrical current is applied via a small active electrode to the operative site and then flows through the patient's body to a large surface area return electrode. Monopolar electrosurgery is the most common source of EMI and CIED interaction in the operating room. Possible interactions include pacing inhibition; triggering of unneeded tachyarrhythmia therapy; damage at the lead–myocardial tissue interface causing an increase in pacing threshold; pulse generator damage; and the induction of electrical reset mode.

The most frequent CIED interaction with EMI is oversensing, which results in inappropriate inhibition of pacing output. Oversensing by an ICD has the additional problem of false detection of the tachyarrhythmia, possibly leading to inappropriate CIED therapy. The significance of oversensing is determined by a number of patient- and device-related factors. For example, ICDs require a certain duration of continuous high-rate sensing to fulfill arrhythmia detection criteria. For a patient with a robust underlying rhythm, pacing inhibition may be inconsequential, while a PM-dependent patient may experience a hemodynamically unstable underlying rhythm with prolonged pacing inhibition.

How can EMI risks be decreased?

The anatomic site of electrosurgery application, the duration of electrosurgery application, and the position of the return electrode determine the risk of oversensing. Management of potential sources of EMI associated with electrocautery includes:

- Assuring positioning of the cautery and current return pad to avoid the current pathway passing through or near CIED pulse generator and leads
- Avoiding proximity of the cautery's electrical field to the pulse generator or leads
- Use of short, intermittent, and irregular cautery bursts at the lowest feasible energy levels (less than 4 seconds, separated by at least 2 seconds)
- Use of a bipolar electrocautery system or an ultrasonic scalpel if possible

The risk is greatest if the current path crosses the CIED and/or leads and decreases when the presumed current path is kept at least 6 inches away from the CIED.

Who should have their CIED reprogrammed prior to their procedure?

Perioperative management largely relies on determining the patient's CIED dependence and EMI potential. The ASA task force recommends reprogramming to an asynchronous paced mode in patients who are PM dependent when there is a significant risk of EMI. Reprogramming to

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an asynchronous mode at a rate higher than the patients' intrinsic rate will overcome potential oversensing, or undersensing, from EMI.

Clinical Pearl

The ASA task force recommends reprogramming to an asynchronous paced mode in patients who are PM dependent when there is a significant risk of EMI.

Should rate adaptive functions be disabled?

Special algorithms such as rate adaptive function should be disabled. Pacemaker rate–response algorithms may cause unwanted HR elevation during a procedure. These algorithms are specific to the particular CIED model and manufacturer. For example, transvenous PMs that correlate an increase in respiratory rate and tidal volume with exercise and a need for increased cardiac output pose a challenge for anesthesiologists. Because of the monitored parameter (i.e., respiratory rate and tidal volume via thoracic impedance) the paced rate in these devices may increase inappropriately in response to mechanical hyperventilation.

What monitors need to be in place for a patient with CIED?

Primary activities associated with intraoperative management of CIED include the following:

- Monitoring the operation of the device
- Preventing potential CIED dysfunction
- Performing emergency defibrillation, cardioversion, or heart rate support if necessary

It is essential that ECG monitoring of the patient include the ability to continuously detect PM activity. A perfused peripheral pulse should be monitored with a waveform display such as pulse oximetry or invasive pressure monitoring. Temporary pacing and defibrillation equipment should be immediately available before, during, and after the procedure. Patients with disabled ICDs should have transthoracic defibrillator pads in place.

Clinical Pearl

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How should a CIED be managed postoperatively?

Any PM that was reprogrammed for a procedure should be interrogated and the CIED function should be restored. This should occur while the patient is still being monitored in the post-anesthesia care unit or in the intensive care unit. Most manufacturers also advise reinterrogation of all devices postoperatively, especially if monopolar diathermy, significant fluid or blood component administration, or external defibrillation have occurred.

Can a magnet be used instead of reprogramming the CIED?

The appropriate role of magnets in the perioperative period remains controversial. Advisories and expert opinion statements caution against routine magnet use as a substitute for appropriate preoperative consultation and preparation. In emergency situations, where time may preclude reprogramming by qualified personnel, a magnet may be placed over the device.

How does a magnet affect a PM? Does a magnet affect an ICD differently than a PM?

Only an interrogation with a programmer can reveal current magnet response settings. However, the expected response for most PMs is the initiation of asynchronous pacing at a fixed, preset "magnet rate," as well as a fixed AV delay, which varies by manufacturer.

Although most ICDs will suspend antitachycardia therapy when a magnet is placed over the device, magnet response varies according to the manufacture and device program. While the ICD function will be suspended, the device will not be forced to revert to an asynchronous pacing mode. Patients with these devices who are PM dependent need to have their device reprogrammed to an asynchronous mode preoperatively if there is significant risk of EMI.

What are the potential disadvantages of using a magnet?

It is important to appreciate that a preset magnet rate may not be sufficient to meet the patient's metabolic demands, particularly for a pediatric patient. Cardiac function in any patient may be compromised, as asynchronous pacing may result in loss of AV synchrony, loss of CRT, loss of capture, or competition between the PM and the patient's intrinsic rate. A patient with an ICD who is PM dependent will still be at risk of

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profound bradycardia or asystole if pacing is inhibited by EMI. It may be difficult to maintain the magnet in a stable position over the pulse generator of the device, particularly if the patient is lateral or prone. Although some devices emit a diagnostic tone when a magnet is applied, whether any sound is emitted, and its meaning depend on the device manufacturer.

Are there special considerations for performing defibrillation or cardioversion in a patient with a CIED?

If a life-threatening arrhythmia occurs, follow Advanced Cardiac Life Support guidelines to provide rapid cardioversion or defibrillation. If possible, attempt to minimize the current flowing through the pulse generator and lead system by the following mechanisms:

- Position defibrillator or cardioversion pads/paddles as far as possible from the pulse generator.
- Position defibrillator or cardioversion pads/paddles perpendicular to the major access of the CIED pulse generator and leads by placing them in an anterior–posterior location.

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Suggested Reading

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