Chapter 23

l-Transposition of the Great Arteries ("Corrected" Transposition)

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Case Scenario

A 12-year-old female presents with acute appendicitis, vomiting for more than 18 hours with minimal oral intake during this time. She has a history of levo-transposition of the great arteries diagnosed in utero and developed complete heart block requiring permanent pacemaker insertion at age 7 years. Her last outpatient cardiology appointment was 8 months ago. Her mother reports that since then she has been doing well and participating in normal activities. Cardiology was consulted, as outpatient records could not be obtained. Electrocardiogram shows a heart rate of 110 beats/minute with ventricular pacing spikes. Pacemaker interrogation revealed the following settings: DDD, intrinsic atrial rate 110 beats/minute, 0% atrial pacing, 100% ventricular pacing, and 3 years of pacemaker longevity remaining. Her current vital signs are heart rate 110 beats/minute, respiratory rate 20 breaths/minute, SpO_2 98% on room air, and blood pressure 95/55 mm Hg. Her abdomen is tender, but not distended. Her hemoglobin is 14 and hematocrit 42. Abdominal ultrasound is indicative of acute appendicitis. All other laboratory work is within normal limits.

Echocardiography reveals:

- *Mild right (systemic) ventricular dysfunction with ejection fraction 50%*
- *Moderate systemic atrioventricular valve regurgitation*
- *Dilation of the tricuspid valve annulus*

Key Objectives

- Describe the anatomy of levo (l-looped) transposition of the great arteries.
- Understand associated cardiac abnormalities and their clinical presentation.
- Identify long-term sequelae associated with uncorrected l-transposition.
- Describe the options for surgical repair to "correct" l-transposition.
- Discuss the preoperative assessment of patients with pacemakers.
- Describe intraoperative management and surgical considerations for this patient.
- Discuss whether a laparoscopic surgical approach is appropriate.
- Describe appropriate discharge criteria and disposition planning.

Pathophysiology

What are the anatomic characteristics of l-transposition of the great arteries?

Levo-transposition of the great arteries (l-TGA) is a rare anomaly, comprising less than 1% of all forms of congenital heart disease (CHD), and is characterized by both atrioventricular (AV) and ventriculoarterial (VA) discordance. In this anomaly, the right atrium (RA) connects via the mitral valve (MV) to the morphological left ventricle (LV), which supplies the pulmonary artery (PA). The left atrium (LA) connects via the tricuspid valve (TV) to the morphological right ventricle (RV), which supplies the aorta. (See Figure 23.1.) Because the TV and RV are connected to the aorta in l-TGA, they are also commonly referred as the systemic AV valve and systemic ventricle, respectively.

How does embryologic development affect the anatomy of l-TGA?

One of the crucial embryologic processes for correct anatomic alignment of the four chambers of the heart occurs during the third week of gestation. During this period the straight primitive heart tube "loops" to the right (dextro or d-loop), resulting in the normal morphologic position of the RV. However, looping to the left (levo or l-loop) leads to abnormal positioning of the ventricles and to abnormal

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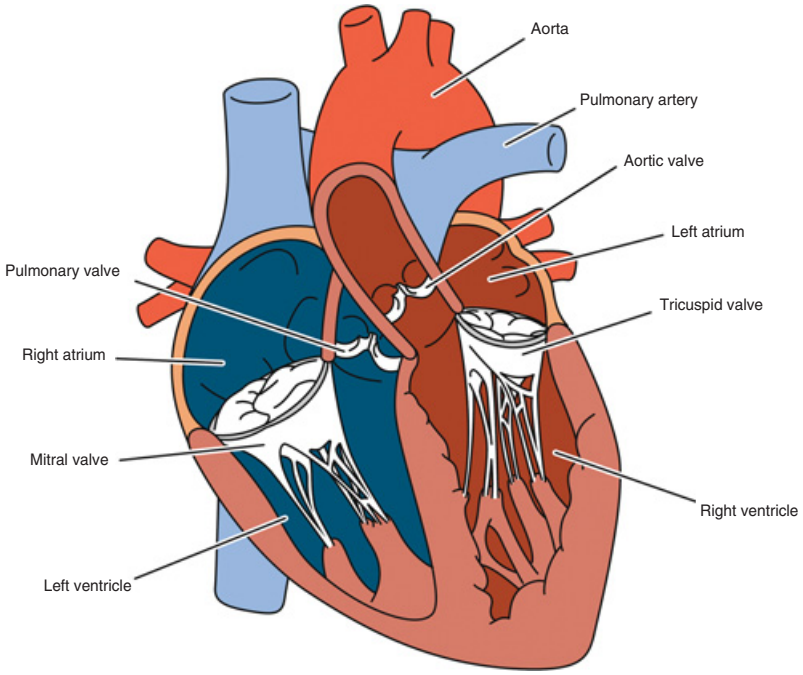

Figure 23.1 L-transposition of the great arteries. Drawing by Ryan Moore, MD, and Matt Nelson.

connections among the atrial, ventricular, and arterial segments of the heart.

What is the flow of deoxygenated and oxygenated blood in the heart of the l-TGA patient? Why is the term "congenitally corrected TGA" used in describing this lesion?

Deoxygenated systemic venous blood returns to the correctly positioned RA via the superior (SVC) and inferior vena cava (IVC). From the RA it flows to the discordant LV via the MV and into the lungs through the PA. Oxygenated blood from the lungs returns to the correctly positioned LA via the pulmonary veins. From the LA it flows to the discordant RV via the TV and returns to the systemic circulation through the aorta.

Based on the preceding description of blood flow, one can appreciate that the term "congenitally corrected TGA" is appropriate; despite ventriculoarterial discordance, oxygenated and deoxygenated blood flows in the correct physiologic direction resulting in normal oxygenation and perfusion. L-transposition of the great arteries is also commonly referred to as "double discordance" or "ventricular inversion." (See Figure 23.2.)

What are the anatomic variations of l-TGA and the nomenclature for describing positions of the heart, atria, ventricles, and great arteries?

The most common anatomic arrangement of l-TGA is situs solitus with l-looping of the ventricles and with the aorta abnormally positioned anterior and leftward of the PA (S, L, L). Situs solitus refers to normal positioning of the thoracic and abdominal organs. Anatomically this means the heart is located in the left chest with a leftward pointing apex, with the morphologic RA located on the right and the morphologic LA located on the left. In a patient with normal cardiac anatomy, including situs solitus, d-looped ventricles, and normal great artery connections, the aorta is located rightward and posterior to the PA (S, D, D).

In a less common variation of l-TGA with situs solitus, the aorta is located anterior and rightward (S, L, D). An even less common variant of l-TGA includes dextrocardia, wherein the heart is positioned and oriented in the right chest, and situs inversus, where there is inversion of the abdominal viscera, including the liver, spleen, and stomach. It is an anatomic mirror image of l-TGA. Although the anatomy is different, the pathophysiology is the same, and thus it is also classified as l-TGA. Its designation is "I" (i.e., I, D, D) which describes situs inversus with d-looping

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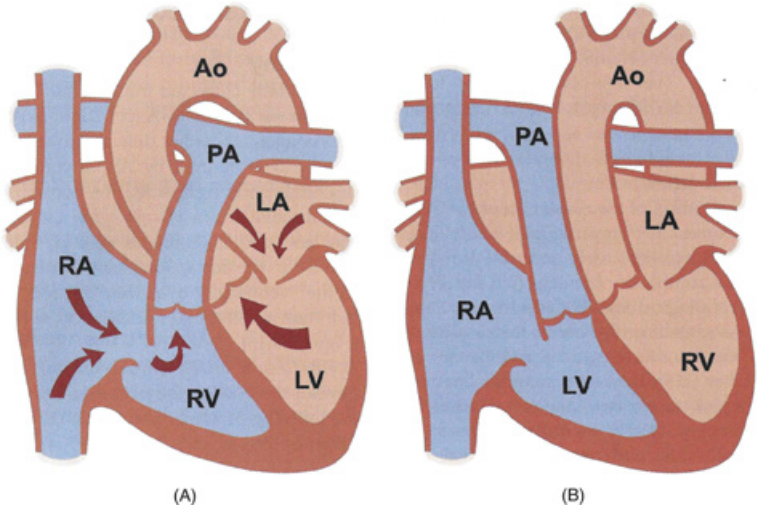

Figure 23.2 Congenitally corrected transposition of the great arteries (ccTGA). (A) Normal anatomy. (B) ccTGA – blood flow in corrected transposition. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle. From McEwan A. and Manolis M. Anesthesia for transposition of the great arteries. In Andropoulos D. B., Stayer S., Mossad E. B., et al., eds. *Anesthesia for Congenital Heart Surgery*, 3rd ed. John Wiley & Sons, 2015: 542–66. With permission.

of the ventricles, and the aorta positioned anterior and rightward of the PA.

Clinical Pearl

Situs solitus refers to normal positioning of the thoracic and abdominal organs. Anatomically this means the heart is located in the left chest with a leftward pointing apex, with the morphologic RA located on the right and the morphologic LA located on the left. In a patient with normal cardiac anatomy, including situs solitus, d-looped ventricles, and normal great artery connections, the aorta is located rightward and posterior to the PA (S, D, D).

How does coronary artery anatomy differ in l-TGA?

Coronary artery anatomy in patients with l-TGA is variable. The coronary arteries in l-TGA are inverted. Thus, the morphologic left coronary artery (LCA) arises from the patient's right-sided sinus and the morphologic right coronary artery (RCA) from the left sided sinus. The LCA (vessel originating from right-sided sinus) has a circumflex (left circumflex or LCX) and anterior descending (LAD) branch, while the RCA (vessel originating from the left-sided sinus) has an epicardial distribution. Although the above described coronary distributions are most common, many other variations have been described, including both RCA and LCA having a common single sinus origin, while other descriptions note hypoplasia of the left anterior descending branch. Ultimately, careful preoperative delineation of the coronary anatomy is important before the double switch operation (DS), which is described later in this chapter.

Clinical Pearl

The coronary arteries in l-TGA are inverted. Thus, the morphologic LCA arises from the patient's right-sided sinus and the morphologic RCA from the left-sided sinus.

What is the typical clinical presentation of a patient with *isolated* l-TGA?

L-transposition occurs as an isolated defect in fewer than 10% of affected patients. Prenatal diagnosis is challenging and therefore often missed. Patients without other structural cardiac defects are asymptomatic at birth and during early childhood and usually present later in life, with some studies reporting lack of symptoms for up to 70–80 years. Older children and adults may present with signs and symptoms related to either cardiac conduction defects (bradycardia, fatigue, poor exercise tolerance) or systemic right heart failure (dyspnea, fatigue, fluid retention, decreased exercise tolerance).

Because the morphologic RV is not well suited to perform the workload of the systemic ventricle, progressive tricuspid regurgitation (TR) ensues. A controversial topic is whether poor RV function leads to TR or whether dysfunction is a result of long-standing TR. Either way, RV failure is a frequent sequel to systemic AV valve regurgitation, and surgical repair of the TV should be considered early, prior to onset of irreversible ventricular dysfunction. Tricuspid valve regurgitation increases volume overload

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and worsens ventricular failure. Additionally, the morphologic RV is perfused by a single coronary artery. Because the RV has higher systemic workload, it is hypothesized that its limited coronary arterial blood supply in relation to its demand places it at higher ischemic risk.

In l-TGA, the cardiac conduction system is often abnormal and unstable. The AV node and His–Purkinje conduction system (His bundle) have an unusual position and course, and many patients have dual AV nodes. The second anomalous AV node and bundle are usually located anteriorly, and the elongated bundle is vulnerable to fibrosis with advancing age. Thus, these patients are prone to developing complete heart block (CHB) and reentrant tachyarrhythmias (i.e., Wolff–Parkinson–White syndrome). The risk of developing CHB increases linearly with age; the progressive incidence is 2% per year.

Clinical Pearl

l-TGA occurs as an isolated defect in fewer than 10% of affected patients. They are asymptomatic at birth and during early childhood, and typically present later with either arrhythmias or systemic RV failure. Because their cardiac conduction system is often abnormal and unstable, they are prone to developing CHB and reentrant tachyarrhythmias.

What associated anatomic cardiac abnormalities exist for patients with l-TGA and how do these impact the physiology?

More than 90% of patients with l-TGA have additional cardiac anomalies. The most frequent are ventricular septal defects (VSD), pulmonary outflow tract obstruction, and abnormalities of the systemic (tricuspid) AV valve. The clinical presentation and course in this cohort of patients with l-TGA depends on the presence and nature of the associated defects.

- ***Ventricular septal defects*** occur in up to 80% of patients with l-TGA. Most commonly the VSD is a large, perimembranous outflow (subpulmonary) defect, although the defect can be located in any region of the ventricular septum. If the defect is small, or restrictive, a systolic murmur may be present, becoming evident once the pulmonary vascular resistance (PVR) sufficiently falls after birth. This results in left-to-right shunting, specifically shunting of blood from the morphologic RV (left-sided) to the morphologic LV (right-sided). Patients with a large or unrestrictive VSD usually present in infancy or childhood with congestive heart failure (CHF).
- ***Pulmonary outflow tract obstruction*** (used synonymously with LV outflow tract obstruction or LVOTO) occurs in approximately 30%–60% of patients with l-TGA and often accompanies a large VSD. The obstruction is commonly subpulmonary (subvalvular) and less frequently secondary to valvular pulmonary stenosis or atresia. Subpulmonary stenosis may be due to a fibromuscular membrane, fibrous tissue tags, or aneurysmal tissue of the interventricular septum. Neonates will often present with cyanosis if pulmonary outflow tract obstruction and concomitant VSD are present, as there is limited pulmonary blood flow (PBF) with significant pulmonary-to-systemic ventricular level shunting.
- ***Tricuspid (systemic AV) valve abnormalities*** have been reported in up to 90% of patients at autopsy, but not all clinically manifest during a patient's lifetime. Regurgitation is common and generally progressive. There is a wide range of variability in severity of the abnormality, with severe "Ebstein" anomaly being the worst, accounting for up to 55% of TV abnormalities. In these patients the systemic AV valve is inferiorly displaced, located closer to the cardiac apex. Additionally, these delicate valves are inherently prone to developing regurgitation, which is an additional burden for the systemic morphologic RV. If left uncorrected, systemic valve abnormalities lead to development of ventricular dysfunction and heart failure.

Clinical Pearl

The most commonly associated anatomic cardiac abnormalities in l-TGA are VSDs, pulmonary outflow tract obstruction, and TV abnormalities.

What are the surgical options to "correct" l-TGA?

There are several surgical options. Some focus on repairing the associated anatomic lesion (i.e., VSD, TV abnormality, pulmonary outflow tract obstruction) and others focus on definitive surgical intervention, with anatomic correction of the ventricular relationships. The presence of associated lesions remains the major determinant regarding the timing and surgical repair of l-TGA.

1. ***Double switch***: The definitive option for anatomic repair offered to patients with l-TGA devoid of significant pulmonary outflow tract obstruction is the double switch operation (DS), which involves both atrial and arterial switch components. (See Figure 23.3.) An intraatrial baffle (Mustard or Senning

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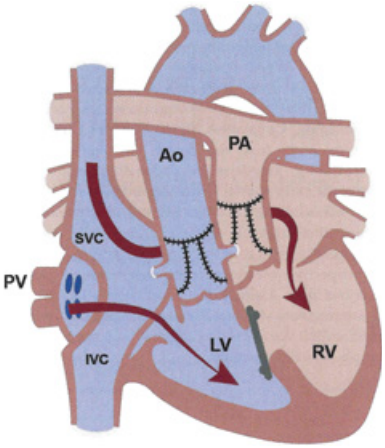

Figure 23.3 Double switch operation. Senning operation with the arterial switch procedure. Ao, aorta; IVC, inferior vena cava; LV, left ventricle; PA, pulmonary artery; PV, pulmonary veins; RV, right ventricle; SVC, superior vena cava. From McEwan A. and Manolis M. Anesthesia for transposition of the great arteries. In Andropoulos D. B., Stayer S., Mossad E. B., et al., eds. *Anesthesia for Congenital Heart Surgery*, 3rd ed. John Wiley & Sons, 2015: 542–66. With permission.

procedure) is created, which diverts deoxygenated systemic venous return into the morphologic RV and oxygenated pulmonary venous return into the morphologic LV. The arterial switch operation (ASO) involves the transection of both the PA and aorta, and translocation of the vessels to the opposite root, resulting in anastomosis of the PA to the morphologic RV, and aortic anastomosis to the morphologic LV. The DS operation establishes both AV and VA concordance. In other words, systemic deoxygenated blood returning to the RA is baffled across the atrial septum to the TV and into the morphologic RV, which is now connected to the PA, and into the pulmonary circulation. Additionally, oxygenated pulmonary venous blood returning to the LA is baffled across the atrial septum to the MV, into the morphologic LV, and then into the newly transposed aorta to the systemic circulation. The ASO also requires coronary artery transfer as part of the great artery transfer. (See Chapters 21 and 22.)

Clinical Pearl

The double switch operation offers anatomic repair of l-TGA, comprising both atrial and arterial switch components. Normal AV and VA concordance is established, which results in systemic deoxygenated blood returning to the RA to be rerouted to the RV and pulmonary circulation and pulmonary venous oxygenated blood returning to the LA to be rerouted to the LV and systemic circulation.

2. ***Senning-Rastelli procedure***: For l-TGA patients with a large VSD and pulmonary outflow tract obstruction, the Senning–Rastelli procedure is typically performed; it has two components. An intraatrial baffle (Senning tunnel) is created, diverting deoxygenated systemic venous return from the RA into the morphologic RV and oxygenated pulmonary venous return from the LA into the morphologic LV. Additionally, the Rastelli procedure is performed. This involves closure of the VSD with an intraventricular baffling technique so that blood from the right-sided morphologic LV courses across the ventricular septum into the aorta. Because the Rastelli baffle obliterates a possible pulmonary outflow tract, the main PA is transected from the morphologic LV, and an extracardiac conduit is placed between the morphologic RV and PA to restore PBF.

Clinical Pearl

The double switch operation is performed in patients without significant pulmonary outflow tract obstruction. The Senning–Rastelli operation is performed in patients with a large VSD and pulmonary outflow tract obstruction.

3. ***Physiologic repair***, also called "conventional" or "classical" repair, addresses only coexisting defects and does not lead to anatomic correction of l-TGA. The surgical techniques are dictated by the type and severity of associated lesions. Patients with a VSD may undergo only a VSD closure, while those with pulmonary outflow tract obstruction may undergo LV to PA conduit placement. Those with TR may undergo TV repair or replacement, which is ideally performed prior to onset of RV failure (defined as RV ejection fraction <40%).
4. ***Single ventricle palliation*** may be considered in the presence of complex straddling of AV valves across the ventricular septum or ventricular hypoplasia.

What long-term sequelae are associated with uncorrected l-TGA?

The long-term sequelae of patients with uncorrected l-TGA include RV dysfunction, systemic atrioventricular valve regurgitation, CHF, arrhythmias, and CHB. The natural history of l-TGA is largely determined by the presence of associated anatomic cardiac abnormalities and the progressive dysfunction of the morphologic RV as the systemic pump. Several studies have shown that a substantial number of unoperated patients with l-TGA have compromised function and a reduction in quality of life. Patients devoid of associated significant lesions may live normal lives into early adulthood, with many reports of women having successful

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pregnancies. However, by age 45 years, 67% of patients with l-TGA develop heart failure if they have associated cardiac lesions as opposed to a 25% incidence of heart failure in patients without significant lesions.

Clinical Pearl

By age 45 years, 67% of patients with l-TGA develop heart failure if they have associated cardiac lesions, as opposed to a 25% incidence of heart failure in patients without significant lesions.

What are indications for PA banding?

1. Infants with a ***large, unrestrictive VSD*** are at risk for developing heart failure secondary to excess PBF, which worsens as PVR falls during the first few weeks of life. These infants may require PA banding if unresponsive to medical management. The PA band, a surgically placed circumferential restriction of the PA, reduces PBF by mechanically increasing PVR, thereby improving heart failure symptoms.
2. Pulmonary artery banding is also used to ***"train" the underdeveloped morphologic LV*** in preparation for definitive surgical repair. Placing a PA band increases the PVR, placing a higher afterload on the LV. This exposure to near-systemic pressures results in gradually increasing LV posterior wall thickness or hypertrophy, thus preparing it for future anatomic repair or the DS operation. Pulmonary artery banding is usually done within the first few months to years of life, with timing of the subsequent DS operation variable. Studies report the median time from PA banding to the DS operation ranges from 2 to 14.5 months.
3. Pulmonary artery banding may also be indicated for patients with ***severe or progressive regurgitation of the systemic AV valve (TV)***, as it has been shown to stabilize systemic AV valve regurgitation and thus improve the systemic ventricular function.

What are the long-term outcomes for physiologic versus anatomic repair?

Early mortality for physiologic repair is low; however, long-term outcomes are poor due to progressive RV dysfunction and heart failure, with 84%, 75%, and 61% survival reported at 1, 5, and 15 years, respectively. Risk factors associated with poor outcome include TV replacement, preoperative RV dysfunction, postoperative CHB, subvalvular pulmonary stenosis, and Ebstein-like malformation of the TV.

In contrast, early mortality following anatomic repair remains high, up to 10%. However, long-term outcomes seem to be comparable or superior to those observed in physiologic repair patients. Since the DS and the Senning–Rastelli procedures were only introduced in the 1990s, long-term data are limited but growing. In the series of patients followed longest, the 20-year survival rates for patients following DS and Senning–Rastelli were similar (83% vs. 76%, respectively). The sequelae following anatomic correction are primarily due to conduction abnormalities (CHB and tachyarrhythmias), left ventricular dysfunction, and neoaortic valve regurgitation. In addition, some patients may experience baffle-associated complications, such as obstructions or leaks. Narrowing of the baffles created during the atrial switch procedure (i.e., Senning or Mustard) may present as chylothorax, SVC syndrome, or pulmonary venous congestion, depending on the site of the obstruction. Baffle leaks cause atrial level shunting, which may present as a simple cyanosis or in more severe cases as a cerebrovascular accident.

Up to 25% of patients following anatomic repair will require surgical reintervention. In the Senning–Rastelli group, RV–PA conduit replacements or interventional conduit procedures will be required; in the DS group, aortic valve replacements and intraatrial baffle interventions are more prevalent.

How can functional status be assessed in the pediatric patient with heart failure?

Heart failure (HF) in children is a progressive clinical and pathophysiologic syndrome with characteristic signs and symptoms that include edema, fatigue, respiratory distress, and growth failure. It is also accompanied by circulatory, neurohormonal, and molecular derangements. Heart failure is a risk factor for major adverse cardiac events after surgery. Even with minor procedures such as an appendectomy, patients with chronic stable HF have been shown to have increased morbidity and mortality. As the well-established New York Heart Association Heart Failure Classification is not applicable to most of the pediatric population, the Modified Ross Heart Failure Classification has been used in children. (See Table 23.1.)

It may be difficult to assess functional class in older children, especially teenagers who are excellent at masking their symptoms and limitations. Often questions like "What do you like to do for fun?", "What is your favorite activity in physical education?", or "Do you walk your dog?" provide more insight into functional capacity.

Heart failure in patients with unrepaired l-TGA is likely multifactorial but primarily attributed to the morphologic RV being structurally ill-equipped to perform the workload

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Table 23.1 Modified Ross Classification for Pediatric Heart Failure

Class I	Asymptomatic
Class II	Mild tachypnea or diaphoresis with feeding in infants Dyspnea on exertion in older children
Class III	Marked tachypnea or diaphoresis with feeding in infants Prolonged feeding times with growth failure Marked dyspnea on exertion in older children
Class IV	Tachypnea, retractions, grunting, or diaphoresis at rest

Adapted from Ross R. D. *Pediatr Cardiol* 1992; 13: 72–5 and Ross R. D. *Pediatr Cardiol* 2012; **33**: 1295–300. With permission.

of the systemic ventricle. This can often be accentuated by volume overload from TR secondary to an abnormal TV. Additionally, the RV is supplied by a single coronary, whereas the LV has dual supply (LAD and LCX). This mismatch places the RV at higher ischemic risk, as it is functioning as the systemic ventricle. Finally, the TV support system has small, numerous papillary muscles originating from the septum as well as the free wall, making it poorly designed to handle RV dilation, leading to worsening TR.

Clinical Pearl

Heart failure in patients with unrepaired l-TGA is likely multifactorial but is primarily attributed to the morphologic RV being structurally ill equipped to perform the workload of the systemic ventricle. This can often be accentuated by volume overload from TR secondary to an abnormal TV.

Pacemaker/Implanted Device Management

What are the potential risks of surgery in a patient who is pacemaker-dependent or has an implantable cardioverter-defibrillator?

The most significant threat to the pacemaker (PM) and the patient is from electromagnetic interference (EMI), which may disrupt the operation of the device implanted in the patient if it is located in the area of the electromagnetic field. Electromagnetic interference is capable of precipitating pacing inhibition by oversensing. Electromagnetic waves are erroneously interpreted as the patient's intrinsic heart activity. This may lead to bradycardia or even asystole. In patients with an implantable cardioverter-defibrillator (ICD), EMI may be misinterpreted as tachyarrhythmias, resulting in either antitachycardia pacing or inappropriate shock therapy. Additionally, EMI may stimulate the sensor for rate-responsive pacing, precipitating inappropriate tachycardia. It can activate "power on reset" of the device, reversing it to the factory program parameters such as VVI pacing at the lower rate. Finally, it can induce damage at the lead–tissue interface, worsening pacing thresholds or even causing direct damage to the device. Essential elements of preoperative evaluation of cardiovascular implantable electronic devices (CIEDs) are given in Table 23.2.

How can it be determined if a patient is PM dependent? What special precautions should be taken?

Determination of PM dependence can be difficult, especially if the interrogation report or reprogramming device is not available. It is prudent to obtain a 12-lead ECG or rhythm strip before surgery. If there are PM spikes in front of all or nearly all P waves and/or QRS complexes, assume dependency. Patients with a history of syncope or AV node ablation should be considered reliant on their PM. It is essential to have a backup pacing plan for these high-risk patients. At minimum, transcutaneous pacing and defibrillation pads should be placed in an anterior–posterior position, away from the generator, prior to draping. Standard American Society of Anesthesiologists (ASA) monitors and invasive monitoring (i.e., arterial line) aid in recognizing pacing inhibition. Plethysmography confirms a perfusing rhythm, while 5-lead ECG with appropriately adjusted gain allows for detection of pacing spikes.

Clinical Pearl

It is prudent to obtain a 12-lead ECG or rhythm strip before surgery. If there are PM spikes in front of all or almost all P waves and/or QRS complexes, assume dependency.

What are sources of EMI? How is the electrosurgical risk of cardiovascular implantable electronic devices interference minimized?

The most common source of EMI in the operating room is electrocautery. Other sources capable of interfering with CIEDs include external defibrillation, radiofrequency ablation, electroconvulsive therapy, and scanning systems for detecting retained surgical instruments, among others.

The ASA and the Heart Rhythm Association published a joint consensus statement on the perioperative management of patients with CIEDs, recommending programming the PM in asynchronous mode only if significant inhibition

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Table 23.2 Essential Elements of Preoperative Evaluation of Cardiovascular Implantable Electronic Devices

Device type	Pacemaker (PM), implantable cardioverter defibrillator (ICD), chronic resynchronization therapy (CRT)
Date of last interrogation	<12 months for PM <6 months for ICD <3 months for CRT
Manufacturer and model	Chest radiograph may aid in identification if information not readily available
Device indication	PM: Sick sinus syndrome, atrioventricular block, syncope ICD: Primary or secondary prevention CRT: Heart failure
Device safety parameters	**Battery longevity** >3 months preferable **Capture threshold** <3 mV (the least amount of electrical stimulus required to consistently depolarize the myocardium) **Lead impedance** between 300 and 1200 Ohms **Sensitivity** >1.5 mV for P wave and > 4 mV for R wave detection (ability of the device to detect the patient's intrinsic rhythm; the greater the amplitude of the sensed complex, the higher likelihood of patient's own activity being sensed)
Leads	If implanted >3 months they are less susceptible to displacement
Programming essentials	**PM**: Mode, lower, and upper tracking rates **ICD**: Lowest heart rate for shock delivery, lowest heart rate for anti-tachycardia pacing **Rate adaptation**: Type of sensor (*mechanical* sensor detecting acceleration or *physiologic* sensor tracking minute ventilation)
Underlying rhythm/PM dependency	Determine underlying rhythm and information regarding ability to sustain adequate cardiac output
Magnet response	Will differ with type of device, manufacturer, and battery life Low battery life will cause pacing at lower rates What is magnet pacing rate? Does the device allow for magnet application function to be disabled to "no response"?

is observed, even in PM dependent patients. Certain steps can be taken to minimize EMI, such as keeping the presumed electric current path at least 6 inches away from the generator and/or the leads by properly positioning the electrocautery return pad. If not feasible, then bipolar cautery or short bursts of unipolar cautery at a lower power setting should be used (<5 seconds long with at least 2-second pauses between the bursts). In all cases, a magnet needs to be immediately available if preoperative reprogramming was not chosen or available.

Is magnet use always safe and feasible?

A magnet applied to the PM will prevent generator inhibition and avert inappropriate tracking or rate response by initiating asynchronous pacing.

Each pacemaker manufacturer has a slightly different magnet response, as follows:

- A ***Medtronic*** pacemaker will pace DOO or VOO at 85 bpm. However, if it is at the end of life or at elective replacement indicator, pacing drops to 65 bpm.
- A ***St. Jude*** device will pace at 100 bpm or 85 bpm depending on how much battery life is left. A response to the magnet should be confirmed preoperatively, because the response could be disabled in pacemakers manufactured by St. Jude Medical, Boston Scientific, or Biotronic.
- In the case of ***ICDs***, a magnet will suspend tachyarrhythmia detection and therapy but will not affect the pacing. Therefore, EMI may cause oversensing and undesired inhibition of the ICD's PM function leading to inconsistent pacing. This may be catastrophic in patients with an ICD who are also PM dependent.

Generally, asynchronous pacing is well tolerated. However, in some patients magnet use may lead to deleterious hemodynamic effects. Pediatric patients, particularly those in HF, depend on AV synchrony and higher HR to maintain their cardiac output. Those with biventricular pacing for chronic resynchronization therapy should always be considered pacer dependent and not capable of tolerating asynchrony. For these high-risk patients preoperative reprogramming of their PM or ICD is preferred over using a magnet.

Clinical Pearl

A magnet applied to a pacemaker will prevent generator inhibition by initiating asynchronous pacing, but each pacemaker manufacturer has a slightly different magnet response. Magnet response should be confirmed preoperatively or the pacemaker should be reprogrammed.

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Anesthetic Implications

What are the challenges of using a magnet during surgical procedures?

An essential element of safe and efficient management of patients with CIEDs is open communication between the surgical team, anesthesiologist, and CIED team (i.e., cardiologist, cardiac electrophysiologist). Surgical input should include procedure details such as site, positioning, possible sources of EMI, risk of fluid and electrolyte shifts, and need for blood transfusion. The magnet should be placed over the generator to elicit the desired response. If the surgical site is near the generator pocket and not readily accessible, a sterile magnet should be available on the surgical field, where it can be quickly placed if needed.

In pediatric patients with complex CHD, an epicardial pacing system is typically preferred to a transvenous system. Historically, the PM generator pocket has been formed in the abdominal wall within the rectus abdominus muscle sheath, often at the level of umbilicus. Other locations include subxiphoid and retrocostal approaches. Therefore, it is crucial to know the exact location of the generator pocket. Obtaining chest and abdominal radiographs is often indicated to confirm the PM location.

Which surgical approach, open or laparoscopic, is safer and more beneficial for this patient?

Benefits of laparoscopic surgery versus an open surgical procedure in children without CHD have been well established. Laparoscopic approaches have been associated with decreased morbidity and transfusion rates, as well as shorter length of stay. A recent retrospective cohort study from 2013–2014 National Surgical Quality Improvement Project-Pediatrics (NSQIP-P) data compared morbidity and mortality outcomes in children with minor, major, and severe CHD undergoing laparoscopic versus open procedures. Within the minor CHD group, laparoscopic procedures were associated with lower 30-day mortality, in-hospital mortality, and 30-day morbidity in comparison to open procedures. These benefits declined as the severity of CHD increased. Overall, analyzed outcomes were comparable between laparoscopic and open techniques. The risks of mortality and 30-day morbidity are significantly higher for patients with severe CHD (OR 12.31, 95% CI) and major CHD (OR 3.46, 95% CI). Patients with minor CHD have similar mortality as patients without CHD. However, the risks of increased morbidity (including reintubation, infection, and readmission) remained significantly higher even in the patients with minor CHD. Given that the patient in the aforementioned scenario has only mildly decreased RV function with good functional status, utilizing a laparoscopic approach is a reasonable surgical option for management of acute appendicitis. (See Table 23.3.)

What are the possible pitfalls of a laparoscopic approach?

Laparoscopy requires peritoneal insufflation of carbon dioxide (CO_2) resulting in increased intraabdominal pressure and $PaCO_2$. Pneumoperitoneum elevates the diaphragm, which worsens lung compliance, functional residual capacity, and atelectasis. Neonates and infants are particularly vulnerable to changes in respiratory mechanics. Hemodynamic changes such as diminished preload, rise in systemic vascular resistance (SVR), and a rise in PVR should be anticipated and treated accordingly. Patients with severe TR or significantly depressed RV function may not tolerate laparoscopy. Persistent hypoxia or hypotension may prompt conversion to an open procedure.

What are the perioperative considerations in anesthetizing a PM-dependent patient with unrepaired l-TGA presenting for emergent laparoscopic surgery?

A focused history and physical provides crucial information on a patient's functional status and severity of CHD, which

Table 23.3 NSQIP-P Definition and Classification of CHD Severity

Classifications	Definitions and Criteria
Minor CHD	• Cardiac condition with or without medication and maintenance, unrepaired (asymptomatic) • Repaired CHD with normal cardiovascular function and no medication
Major CHD	• Repaired or palliated CHD with residual hemodynamic abnormality with or without medications
Severe CHD	• Uncorrected cyanotic heart disease • Documented pulmonary hypertension • Ventricular dysfunction • Listed for heart transplant

CHD, congenital heart disease; NSQIP, National Surgical Quality Improvement Program.
Adapted from Faraoni D., et al. *J Am Coll Cardiol* 2016; **67**: 793–801. With permission.

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aids in determining anesthetic, surgical, and disposition goals. Open communication between the surgeon, cardiologist, and anesthesiologist is essential in creating a safe and feasible plan.

To decide on magnet use versus reprogramming, assess the EMI risk with the surgeon and consult the cardiologist if the last PM interrogation was >12 months ago. If it is determined that the patient will tolerate either brief pacing inhibition, or asynchronous pacing, a sterile magnet should be available on the surgical field. The PM pocket location should be known to the surgical team and included in the prepped field. Reprogramming the PM prior to the procedure is not indicated in this patient scenario.

Another point of discussion with the surgeon should address laparoscopy. Despite being a safe and better option for this particular patient, laparoscopy may not be tolerated in every patient with major CHD. Criteria for conversion to open procedure need to be established a priori.

Clinical Pearl

To decide on magnet use versus reprogramming, assess the EMI risk with the surgeon and consult the cardiologist if the last PM interrogation was >12 months ago. If it is determined that the patient will tolerate either brief pacing inhibition, or asynchronous pacing, a sterile magnet should be available on the surgical field.

What methods are appropriate for induction and maintenance of anesthesia?

The anesthetic plan should address the issue of suboptimal preload due to prolonged fasting time and ongoing vomiting. Intravenous (IV) access should be established before induction to allow for sufficient rehydration, administration of IV premedication, and rapid sequence induction (RSI). Although our patient is in stable HF, the potential for hemodynamic instability should be expected and addressed. Ketamine or etomidate are both acceptable induction options in those with significant systemic AV valve regurgitation and compromised heart function. Inotropes (i.e., epinephrine, ephedrine, norepinephrine, and/or dopamine) should be readily available. Avoidance of both increases in SVR and decreases in heart rate is advisable, as these conditions worsen systemic AV valve regurgitation and lower cardiac output. Thus, inotropes with primarily α-adrenergic effects (i.e., phenylephrine and vasopressin) are discouraged as they will exacerbate systemic AV valve regurgitation, causing cardiac output to worsen. A backup pacing plan should be in place if the asynchronous rhythm becomes intolerable; place transcutaneous pacing pads away from the generator (anteroposterior position) prior to draping. For this case standard ASA monitors, specifically plethysmography waveform (pulse oximetry), are sufficient for hemodynamic monitoring and detection of a nonperfusing rhythm. Arterial line placement is useful in assessing pacing effectiveness and pulsatility but is not required.

Is prophylaxis for prevention of bacterial endocarditis indicated for this patient?

According to the 2017 American Heart Association/American College of Cardiology Focused Update on antimicrobial prophylaxis for the prevention of bacterial endocarditis, antibiotic prophylaxis is recommended only in high-risk cardiac conditions, if ongoing gastrointestinal infection is present. The child in our scenario is in the low-risk category and therefore does not need prophylaxis. However, antibiotics should be administered if a surgical indication exists, for example, bowel perforation. (See Table 23.4.)

Should the patient's PM be reprogrammed or interrogated postoperatively?

Although reprogramming prior to the procedure is not necessary, postoperative interrogation may be needed if the patient experiences significant pacemaker-related intraoperative events (i.e., significant EMI, hemodynamic instability, cardiac arrest, need for defibrillation or transcutaneous pacing). If elected, postoperative interrogation must be completed prior to patient's discharge from telemetry. Follow-up with the patient's cardiologist within one month of discharge to complete PM interrogation is also recommended.

Clinical Pearl

Any pacemaker or ICD that has been reprogrammed preoperatively should be interrogated and reprogrammed to its preoperative or most optimal settings PRIOR to the patient's discharge from telemetry.

When is it safe to discharge a patient with uncorrected l-TGA following appendectomy?

Following surgery in this scenario, the unrepaired l-TGA patient with pacemaker should remain hospitalized overnight for observation. Close monitoring on telemetry would be warranted if any unusual perioperative events occurred or if the pacemaker was reprogrammed or affected in any way. For a scheduled or nonemergent procedure, same day discharge could be considered after an uneventful routine procedure assuming the patient is

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Table 23.4 Indications for Antimicrobial Prophylaxis for the Prevention of Bacterial Endocarditis

Highest Risk Patients	Highest Risk Procedures
• Prosthetic heart valves, including mechanical, bioprosthetic, and homograft valves (transcatheter-implanted as well as surgically implanted valves are included)	• Dental procedures that involve manipulation of either gingival tissue or the periapical region of teeth or perforation of the oral mucosa; this includes routine dental cleaning
• Prosthetic material used for cardiac valve repair, such as annuloplasty rings and chords	• Procedures of the respiratory tract that involve incision or biopsy of the respiratory mucosa
• A prior history of infective endocarditis	• Gastrointestinal (GI) or genitourinary (GU) procedures in patients with ongoing GI or GU tract infection
• Unrepaired cyanotic congenital heart disease	• Procedures on infected skin, skin structure, or musculoskeletal tissue
• Repaired congenital heart disease with residual shunts or valvular regurgitation at the site or adjacent to the site of the prosthetic patch or prosthetic device	• Surgery to place prosthetic heart valves or prosthetic intravascular or intracardiac materials
• Repaired congenital heart defects with catheter-based intervention involving an occlusion device or stent during the first 6 months after the procedure	
• Valve regurgitation due to a structurally abnormal valve in a transplanted heart	

afebrile, tolerating a regular diet, and either free of pain or with pain well controlled on nonnarcotic agents.

Suggested Reading

Atallah J., Rutledge J. M., and Dyck J. D. Congenitally corrected transposition of the great arteries (atrioventricular and ventriculoarterial discordance). In Allen H., Driscoll D. J., Shaddy R. E., et al., eds. *Moss and Adams' Heart Disease in Infants, Children and Adolescents*, 8th ed. Philadelphia: Lippincott Williams & Wilkins; 2013, 1147–60.

Beauchesne L. M., Warnes C. A., Connolly H. M., et al. Outcome of the unoperated adult who presents with congenitally corrected transposition of the great arteries. *J Amer Coll Cardiol* 2002; **40**: 285–90.

Chu D. I., Tan J. M., Mattei P., et al. Mortality and morbidity after laparoscopic surgery in children with and without congenital heart disease. *J Pediatr* 2017; **185**: 88–93.

Crossley G. H., Poole J. E., Rozner M. A., et al. The Heart Rhythm Society (HRS)/American Society of Anesthesiologists (ASA) expert consensus statement on the perioperative management of patients with implantable defibrillators, pacemakers and arrhythmia monitors: facilities and patient management: this document was developed as a joint project with the American Society of Anesthesiologists (ASA), and in collaboration with the American Heart Association (AHA), and the Society of Thoracic Surgeons (STS). *Heart Rhythm* 2011; **8**: 1114–54.

Graham T. P., Bernard Y. D., Mellen B. G., et al. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. *J Am Coll Cardiol* 2000; **36**: 255–61.

Hornung T. S. and Calder L. Congenitally corrected transposition of the great arteries. *Heart* 2010; **96**: 1154–61.

McEwan A. and Manolis M. Anesthesia for transposition of the great arteries. In Andropoulos D. B., Stayer S., Mossad E. B. et al., eds. *Anesthesia for Congenital Heart Disease*, 3rd ed. Hoboken, NJ: John Wiley & Sons; 2015; 542–66.

Nishimura R. A., Otto C. M., Bonow R. O., et al. 2017 AHA/ACC focused update of the 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *J Am Coll Cardiol* 2017; **70**: 252–89.

Ross R. D. The Ross classification for heart failure in children after 25 years: a review and an age-stratified revision. *Pediatr Cardiol* 2012; **33**: 1295–300.

Warnes C. A. Transposition of the great arteries. *Circulation* 2006; **114**: 2699–709.

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