Chapter 8

Repaired Tetralogy of Fallot

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Case Scenario

A 13-year-old boy with DiGeorge syndrome (22q11 deletion) and a history of tetralogy of Fallot repaired as an infant presents for emergency reduction of testicular torsion, orchidopexy, and possible orchiectomy. He has severe testicular pain and has been vomiting for 12 hours with minimal oral intake. He says that prior to his current illness he had occasionally become short of breath with strenuous activity and felt an occasional "funny heartbeat." His only home medication is furosemide twice daily.

He is followed yearly by his cardiologist and was last seen 11 months ago. An electrocardiogram at that time demonstrated a stable right bundle branch block.

Transthoracic echocardiography at that time showed the following:

- *A dilated right ventricle*
- *Free pulmonary insufficiency*
- *Mild to moderately diminished right ventricular function*
- *Normal left ventricular function*

Key Objectives

- Describe the anatomy of tetralogy of Fallot.
- Understand the repair of tetralogy of Fallot and its long-term implications.
- Understand the associations of DiGeorge syndrome and other genetic abnormalities.
- Describe the preoperative assessment for noncardiac surgery following repair of tetralogy of Fallot.
- Describe intraoperative management strategies.

Pathophysiology

What is tetralogy of Fallot?

Tetralogy of Fallot (TOF), more specifically tetralogy of Fallot with pulmonary stenosis (TOF/PS), describes a collection of cardiac anomalies consisting of right ventricular outflow tract obstruction (RVOTO), a ventricular septal defect (VSD), an overriding aorta, and concentric right ventricular hypertrophy (RVH). The RVOTO may involve infundibular, subvalvular, valvular, supravalvular stenosis or a combination of stenoses. These cardiac anomalies often result in cyanosis, and TOF is the most common cyanotic congenital heart disease. (See Chapter 7.)

What is the typical surgical repair for TOF?

Repair of TOF involves relieving RVOTO and closure of the VSD. The technique used will depend on the specific location of the stenoses. Relief of subvalvular obstruction is performed by resection of hypertrophied muscle bundles to open the outflow tract. This can sometimes be accomplished through the tricuspid valve but may require a right ventriculotomy and a pericardial patch. Depending on the size of the pulmonary valve annulus and the presence of any supravalvular stenosis, the pericardial patch may extend across the pulmonary valve and into the main pulmonary artery. This is commonly referred to as a transannular patch.

When possible a valve-sparing technique is performed. For a valve-sparing technique to be successful the RVOTO needs to be relieved without compromising the pulmonary valve. The intention of a valve-sparing technique is to minimize postoperative pulmonary valve incompetence while preserving RV function, minimizing conduction abnormalities, and improving exercise intolerance. When the pulmonary valve annulus is significantly hypoplastic this technique may not be possible.

While some centers may perform definitive repair of TOF in neonates, it is more common for TOF to be electively repaired between 3 and 6 months of age. Surgically created palliative systemic-to-pulmonary artery shunt procedures may be performed when necessary to augment pulmonary blood flow (PBF) during the neonatal period if anatomical complexities make elective definitive repair challenging. Another option for TOF palliation involves balloon dilation of the RVOT, with or without stent

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placement, or stenting of the PDA. Both of these procedures are performed in the cardiac catheterization laboratory.

What are the implications of a transannular patch in a TOF repair?

A transannular patch crosses the pulmonary valve annulus to relieve obstruction of the RVOT. However, while relieving obstruction, the patch distorts the pulmonary valve and often creates pulmonary insufficiency (PI). Over time PI can progress, ultimately compromising RV function, and leading to right ventricular dilation and failure.

Clinical Pearl

A transannular patch creates free pulmonary insufficiency that in time leads to right ventricular dilation and dysfunction or failure.

What is the implication of the ventriculotomy in TOF repair?

In neonates and small infants, a ventriculotomy is created in order to close the VSD and to aid in resection of the RVOT obstruction. This approach increases the risk of conduction system injury, causing local conduction block and predisposing patients to arrhythmias. A right bundle branch block is a common consequence of TOF repair, often seen immediately following repair. Limiting the size of the ventriculotomy can minimize the risk of arrhythmias and subsequently help to preserve RV function. Ideally, a transatrial-transpulmonary approach avoids a ventriculotomy and allows maximal preservation of the pulmonary valve, annulus, and the infundibulum. This approach is challenging and dependent on the patient's weight, and many patients may not have favorable anatomy for this repair.

What variations exist in TOF and how do they impact surgical repair?

A wide disease spectrum exists for TOF. In addition to TOF/PS, TOF can occur with pulmonary atresia (TOF/PA), with an absent pulmonary valve (TOF/APV), with major aortopulmonary collateral arteries (TOF/MAPCAs), or with an atrioventricular septal defect (TOF/AVSD). These complexities may alter the timing of repair and/or require a staged repair to establish adequate PBF and to separate pulmonary and systemic circulations. (See Chapters 7, 10, and 47).

What chronic postoperative sequelae are associated with repair of TOF?

Increased morbidity is seen as patients who have undergone TOF repair approach adulthood. Common long-term problems in patients following TOF repair are related to PI, RV dysfunction and an increasing incidence of arrhythmias over time.

Pulmonary insufficiency is initially well tolerated but places an increased volume load on the RV. Over time, long-standing severe PI leads to RV enlargement and dysfunction, heart failure, and tachyarrhythmias. Long-standing RV dysfunction can also cause LV dysfunction due to interventricular dependence. As PI is a long-standing, slowly progressive pathology in most of these patients, there is adaption over time and thus even patients with severe PI may not present with symptoms of right heart failure such as exercise intolerance and dyspnea. (See Figure 8.1.)

Atrial reentrant tachycardias and ventricular tachycardias are both possible chronic sequelae of TOF repair. These arrhythmias may result from abnormal structural conduction pathways, surgical scar tissue, and/or consequences from the chronic effects of RV dilation stretching the conduction system. These arrhythmias place patients at risk for sudden cardiac death (SCD), necessitating placement of an implantable cardioverter-defibrillator (ICD) in some patients. Monitoring for increased QRS duration will aid in identifying patients at increased risk of SCD.

Additionally, in patients who require right ventricular–pulmonary artery (RV–PA) conduits as part of their repair, the need for conduit revision due to conduit stenosis or patient growth is common. Aortic root dilation or aortic valve insufficiency may also occur as a result of the initial surgical repair. Aortic valve insufficiency may result from tension on the valve leaflets from the initial VSD repair. While it is unclear how aortic root dilation develops in patients with repaired TOF, its presence indicates a multifactorial disorder that is likely to impact long term prognosis [1].

Clinical Pearl

Chronic postoperative sequelae of TOF repair include RV enlargement and dysfunction, heart failure, and tachyarrhythmias such as atrial reentrant tachycardia and ventricular tachycardia.

When should placement of an ICD be considered?

There is an increased incidence of SCD in patients with repaired TOF, with the majority of events due to sustained

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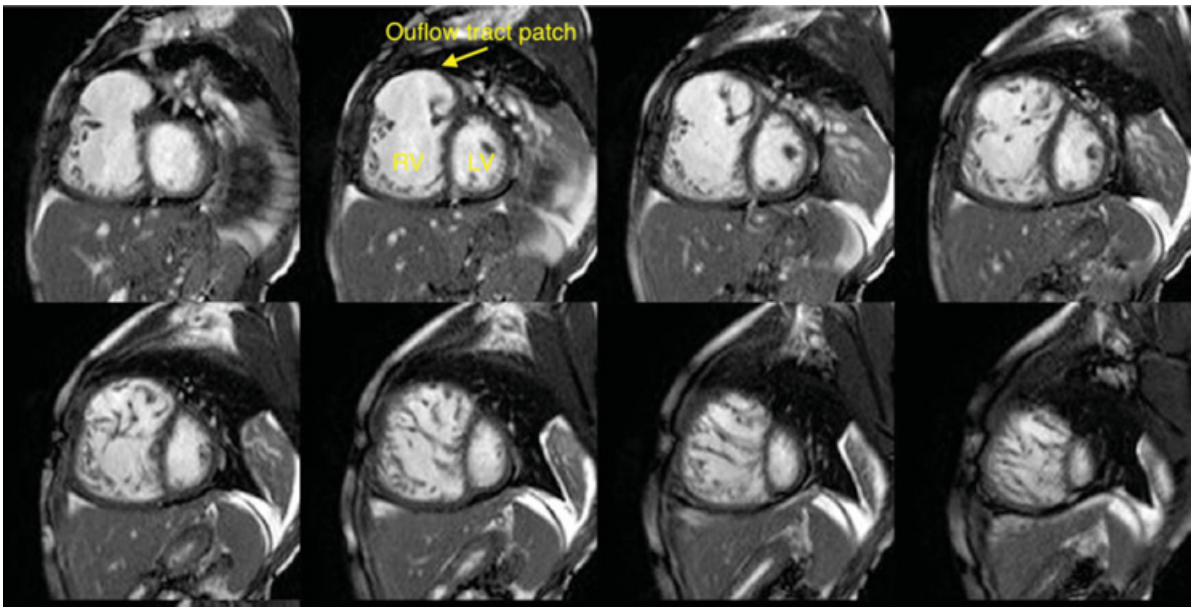

Figure 8.1 Tetralogy of Fallot. Short axis magnetic resonance imaging stack showing markedly dilated right ventricle with a dilated outflow tract patch after repair of TOF. Chronic pulmonary regurgitation results in a dilated RV and eventually leads to RV dysfunction. The RV dilation and dysfunction can lead to tricuspid regurgitation, arrhythmias and rarely sudden death. Courtesy of Michael Taylor, MD.

ventricular tachycardia (VT). These patients are typically older, over 20 years of age, and have had multiple prior cardiac operations.

Implantable cardioverter–defibrillator implantation is recommended when the following risk factors are present [2]:

- Severe PI
- Severe RV dilation
- Left ventricular dysfunction (LV end-diastolic pressure ≥12 mm Hg)
- QRS ≥180 msec on ECG
- Nonsustained VT captured on Holter monitor
- Inducible VT during an electrophysiological study

When should pulmonary valve replacement be considered?

Residual complications of TOF repair can necessitate intervention to restore pulmonary valve competence and preserve RV function before irreversible ventricular damage develops. This can be accomplished by a pulmonary valve (PV) replacement. Pulmonary valve replacement may now be performed surgically or percutaneously in the cardiac catheterization laboratory. While patients with surgically performed PV replacement have a high survival and a low rate of reintervention, transcatheter PV replacement has a high rate of success and is a suitable, less invasive alternative for many patients that allows avoidance of surgery and cardiopulmonary bypass.

Pulmonary valve replacement is indicated when RV end-diastolic volumes are greater than 160–170 mL/m^2 or RV end-systolic volumes are greater than 80–85 mL/m^2 [3]. The presence of significant PI and/or pulmonary stenosis, RV dilation, or RV dysfunction is also considered when deciding when to proceed with PV replacement. Cardiac magnetic resonance imaging (MRI) has proven to be a valuable asset for following these patients and assessing the correct timing for valve replacement.

Following PV replacement, patients experience an improvement in symptoms, with decreased QRS time on ECG, a decrease in RV size, and decreased RV volumes. It has been noted that ejection fractions do not significantly improve following PV replacement, lending support for early intervention before severe RV dysfunction occurs [3].

Is genetic testing helpful in patients with TOF?

Genetic data can be useful to help stratify risk in patients with cardiac and noncardiac manifestations of disease. Genetic abnormalities in TOF occur equally in males and females and can be syndromic or nonsyndromic. Up to 25% of TOF patients have chromosomal abnormalities, with trisomy 21 and 22q.11.2 microdeletion being the

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most frequent. Tetralogy of Fallot can also be associated with *JAG1* (Alagille syndrome), *NKX2-5*, *ZFPM2*, and VEGF mutations.

What is DiGeorge syndrome and what is its relevance in TOF?

DiGeorge syndrome is the most severe form of the 22q.11.2 microdeletion and affects the development of the thymus and parathyroid glands. It includes palatal abnormalities, dysmorphic facies, learning disabilities, immune deficiencies, and hypocalcemia. Aortic arch and branching anomalies are also more common in patients with 22q11 deletion; however, they are usually not associated with a vascular ring in TOF.

The anesthetic implications of DiGeorge syndrome consist of the following:

- The potential for difficult airway management
- The need for irradiated blood products due to varying degrees of immunodeficiency
- Close monitoring for and treatment of hypocalcemia
- Increased incidence of anomalous systemic arterial or venous vessels that may impact the placement of invasive arterial or central lines

Clinical Pearl

DiGeorge syndrome is the most severe form of the 22q.11.2 microdeletion and can include palatal abnormalities, dysmorphic facies, learning disabilities, immune deficiencies, and hypocalcemia.

Are there additional genetic syndromes associated with TOF with extracardiac disease manifestations?

VACTERL (vertebral and cardiac defects, anal atresia, tracheoesophageal fistula, renal and limb abnormalities) and ***CHARGE*** (coloboma, heart defects, choanal atresia, genitourinary, and ear abnormalities) associations are examples of genetic syndromes in which TOF can occur along with extracardiac manifestations. Knowledge of chromosomal abnormalities and associations will guide extracardiac anesthetic management. For instance, placement of a transesophageal echocardiography probe is often contraindicated in a patient with *VACTERL* association following repair of a tracheoesophageal fistula. Patients with these syndromes and associations may more often present with challenging airway management. These patients are also more likely to present for multiple noncardiac procedures due to associated comorbidities. (See Chapter 47.)

Anesthetic Implications

What information should be gathered from these patients during a preoperative assessment?

The preoperative assessment should include a detailed history and physical exam with specific attention to the patient's functional status, cardiac anatomy and function, long-term physiologic sequelae, and a detailed review of cardiac imaging. Additional emphasis should be paid to the following:

Cardiovascular Status Evaluation of the patient's current functional status, surgical history, presence of arrhythmias, and/or pacemaker/defibrillator dependence aids in establishing the patient's baseline health. Clinical symptoms of ventricular dysfunction may include fatiguability, dyspnea, poor feeding, diaphoresis, failure to gain weight, and vomiting. The presence of palpitations, dizziness, or syncope may indicate an underlying arrhythmia.

Respiratory Status Children with previous surgery for congenital heart disease (CHD) may have a history of prolonged intubation, vocal cord paralysis, poor lung compliance, and postoperative nerve damage (i.e., phrenic and/or vagus injury). Identifying prior respiratory issues will aid in efficient intraoperative ventilation management.

Neurologic Status Children with CHD may present with a history of a neurologic insult (i.e., cerebral vascular accidents or thromboembolic events) following prior cardiothoracic surgical repairs or following low cardiac output states. Assessing patients for residual symptomatology will guide additional perioperative management.

Medications Patients with repaired TOF are frequently taking cardiac medications. While angiotensin converting enzyme inhibitors (ACEi) are not commonly used in the management of right heart failure, they are historically linked to instability on induction of anesthesia due to excessive vasodilation. The time of last preoperative administration for medications should be noted. Typically, other cardiac medications are continued without interruption prior to anesthesia; however, it is recommended to carefully examine a patient's medication list for potential interactions or complications.

Laboratory Studies Hematologic and chemical profiles as well as coagulation studies may be indicated to assess renal and hepatic function or efficacy of anticoagulation therapy.

Cardiac Imaging and Testing All pertinent imaging should be reviewed.

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- ***Transthoracic echocardiography (TTE)*** is recommended yearly in patients with repaired TOF until 10 years of age, then once every 2 years to assess qualitative RV and LV function.
- ***Yearly ECGs*** are recommended to monitor the patient's rhythm and QRS duration. ECG changes, specifically a prolonged QRS (>180 msec), place a patient at an increased risk of VT and sudden cardiac death.
- ***Cardiac MRI*** is the preferred imaging modality in an adolescent with repaired TOF as it can assess quantitative RV and LV function, the degree of PI, presence of tricuspid valve insufficiency, and can also guide timing of PVR.

Physical Exam Each patient should be examined for cardiac-specific findings. Common concerning cardiac signs include clubbing, cyanosis, mottled skin, delayed capillary refill, lethargy, failure to thrive, murmurs, hepatomegaly, tachypnea, edema, and poor peripheral pulses.

Clinical Pearl

Patients with repaired TOF may exhibit symptoms of fatigue, dyspnea on exertion, and/or diaphoresis indicating potential ventricular dysfunction. The presence of palpitations, dizziness, or syncope may indicate an underlying arrhythmia. A QRS interval >180 msec places a patient at increased risk for sustained VT and sudden cardiac death.

What anesthetic risks should be discussed with these patients and their families?

Children with CHD experience higher perioperative morbidity and mortality; the Pediatric Perioperative Cardiac Arrest registry reported that the majority of cardiac arrests during noncardiac surgery occurred in patients with major CHD [4]. Repaired TOF with free PI is considered major CHD and places these patients at higher intraoperative risk. Once the baseline cardiac status has been evaluated additional concerns regarding potential intraoperative cardiac decompensation can be discussed with the patient and his family. Intraoperative decompensation is a risk in this patient given his significant dehydration secondary to his vomiting, prolonged fasting time, and use of diuretics.

Is additional testing indicated prior to this surgery?

Relief of a testicular torsion is a urological emergency as the survival of a torsed testicle is reported to be approximately 6–8 hours from symptom onset. If the patient's cardiac symptomatology has changed recently and resources are readily available, additional studies such as a TTE and ECG should be considered. In the event that the patient presents to a hospital other than the one where he is normally followed attempts should be made to obtain prior patient records. If records are not readily available, obtaining an emergent cardiology consult, ECG, and TTE should be considered. However, because of the urgency of this case it is important not to delay the procedure for additional imaging. In absence of complete data, treat the patient as if he has decompensated cardiac disease with necessary precautions.

Is bacterial endocarditis prophylaxis indicated?

The American Heart Association recommends subacute bacterial endocarditis (SBE) prophylaxis within the first 6 months following initial cardiac repair for patients with repaired TOF. Prophylaxis is also indicated in patients with prosthetic heart valves, when prosthetic material was used in the valve repair, or in patients with residual defects at a site adjacent to a prosthetic device or material. Complete guidelines for SBE prophylaxis can be referenced and individualized for each patient.

What premedication is appropriate?

Midazolam (oral, intravenous, or intranasal) is the most commonly used premedication. Intranasal dexmedetomidine has also proven helpful for anxiolysis. However, caution is advised with the use of dexmedetomidine in patients presenting with heart block from previous surgeries, those taking atrioventricular nodal blocking agents (such as digoxin), or in patients with heart failure who may not tolerate bradycardia. Given the nature of the surgery it is quite likely that this patient would benefit from premedication.

Should a peripheral intravenous catheter be placed prior to induction?

Placement of a peripheral intravenous (PIV) catheter is advantageous prior to induction given this patient's history of vomiting and presumed dehydration. Fluid replacement would also be indicated prior to induction, particularly as the RV is dilated and preload dependent. Additionally, an IV anesthetic induction offers more hemodynamic control than an inhalation induction and can limit the effects of potential myocardial depression due to volatile anesthetic agents when ventricular function is compromised. If placement of a preoperative IV is not successful, an IV placed with administration of nitrous oxide prior to induction is an option. It can at times be challenging to obtain PIV

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access in children with CHD and ultrasound guidance may be necessary.

Should additional intraoperative monitoring be considered for this procedure?

All standard American Society of Anesthesiologists (ASA) monitors should be utilized along with any additional monitoring indicated by the patient's cardiac disease and functional status. For this particular case standard ASA monitoring should suffice. A 5-lead ECG is preferred when available to monitor for cardiac arrhythmias and ischemia in all patients with CHD. Additional access and monitoring depend on the size and scope of surgery, anticipated blood loss/fluid shifts, and degree of cardiac dysfunction. An arterial line should be considered if RV function is compromised and the risk of cardiac decompensation is significant. If significant biventricular dysfunction exists, placement of a central line should be considered to infuse vasoactive drugs and monitor cardiac filling pressures. Intraoperative transesophageal echocardiography can also provide useful information in children with compromised cardiac function.

In a patient with known arrhythmias or those at high risk for arrhythmias, rapid access to a defibrillator and antiarrhythmic medications, such as adenosine, should be assured.

Clinical Pearl

Patients with repaired TOF with ventricular dysfunction or a history of arrhythmias have the potential for rapid decompensation; therefore, having safety equipment and resuscitation drugs readily available is recommended.

What type of anesthetic induction should be performed?

Patients with compromised cardiac function have the potential for rapid decompensation during induction of anesthesia due to hypotension and depressed cardiac function. A dilated, volume and pressure loaded RV with poor contractility is unlikely to be able to compensate during prolonged hypotension. In general, a slow titration of medication is the preferred means to induce anesthesia in patients with cardiac dysfunction. It is recommended for safety medications and equipment to be readily available prior to induction in the event of prolonged hypotension.

While a slow induction of anesthesia is often preferable when significant cardiac dysfunction exists, in a patient presenting with significant vomiting a rapid sequence induction (RSI) may be indicated. The risk of aspiration with titrated induction versus hemodynamic instability with RSI should be weighed in context of the individual patient. Whatever induction method is chosen, a combination of medications should be chosen that prioritizes maintenance of cardiac function and perfusion pressure and emergency medications for resuscitation should be prepared.

What are the goals for maintenance of anesthesia in this child?

Anesthetic maintenance may be conducted with inhaled anesthetics and narcotics to achieve a balanced anesthetic. The goal is to maintain the patient's cardiac rhythm, blood pressure, and hemodynamic balance as close to baseline as possible. Decisions regarding anesthetic maintenance with inhaled anesthetics or additional intravenous drugs will also be guided by a patient's response to induction, intraoperative events, and ultimate postoperative plans. It is important to note that a stress response, such as a surgical stimulus, can be challenging in patients with little cardiac reserve; therefore, appropriate pain management is warranted. Fentanyl, a synthetic opioid, is known to blunt the surgical stress response with minimal effect on cardiac function, though it can lower the heart rate, especially in higher doses.

Clinical Pearl

Intraoperative hemodynamic goals include maintenance of adequate preload for a dilated RV, rapid detection and management of arrhythmias, and maintenance of low PVR.

What are the intraoperative fluid management goals for a child with this physiology?

Intraoperative fluid management can be challenging in patients with CHD. Ensuring adequate intravascular volume is important for maintaining cardiac output. In a patient with repaired TOF, although the RV may be dilated, it is also hypertrophic and often requires increased central venous pressures to optimize cardiac output. However, in a patient with RV dilation and dysfunction, excess fluid can lead to worsening function and decompensation. The goal of intraoperative fluid resuscitation in patients with repaired TOF is to replace the fluid deficit while being aware of potential right heart failure.

What rhythm disturbances might be anticipated?

The most common arrhythmias in patients with repaired TOF result from surgical scar tissue and structural

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conduction obstacles that facilitate reentrant conduction pathways. Atrial reentrant tachycardias are present in 30% of this patient population and high-grade ventricular tachycardias are observed in approximately 10% [5]. Arrhythmias may also be provoked by increasing RV strain, especially during fluid resuscitation if the RV becomes overdistended.

Clinical Pearl

Atrial reentrant tachycardias are present in 30% of this patient population and high grade ventricular tachycardias are observed in approximately 10%.

How should intraoperative arrhythmias be treated?

Pediatric Advanced Life Support algorithms should be used for management of arrhythmias, and early consultation with electrophysiology is indicated. Atrial reentrant tachycardias, typically 200 beats/minute in teenagers, consist of an accessory pathway allowing conduction through an additional circuit resulting in tachycardia. These tachycardias can be treated with vagal maneuvers (such as a Valsalva maneuver), antiarrhythmics such as adenosine, or synchronized cardioversion.

Treatment of a ventricular tachycardia will vary depending on the patient's clinical condition. If the patient is hemodynamically unstable, it is best to perform a synchronized cardioversion, with 1–2 joules/kg. If the patient is stable, intravenous lidocaine or amiodarone may be an effective alternative.

Clinical Pearl

Atrial tachycardias can be treated with vagal maneuvers, antiarrhythmics such as adenosine, or cardioversion. If the patient is hemodynamically unstable, utilize synchronized cardioversion.

Should tracheal extubation be considered in the operating room?

Tracheal extubation at the conclusion of the case is desirable and may be considered if the case is uneventful and the patient is hemodynamically stable following fluid resuscitation. Adequate reversal of neuromuscular blockade with appropriate tidal volumes should be demonstrated prior to extubation.

Where should the patient be monitored in the postoperative phase?

Following an uneventful intraoperative course, a standard post-anesthesia care unit with staff experienced in caring for children with CHD would be a reasonable location for recovery. If the case is complicated by hemodynamic instability, significant arrhythmias, or decompensated cardiac function, ongoing care in an intensive care setting is indicated.

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