Chapter

25

Truncus Arteriosus

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Case Scenario

A 3-year-old female with a history of truncus arteriosus and DiGeorge syndrome presents for elective repair of a cleft palate. She was diagnosed prenatally with truncus arteriosus and underwent uncomplicated repair with placement of a right ventricle-to-pulmonary artery conduit at 5 days of age. She has been followed since that time and over the past year has developed a slowly increasing gradient across her conduit. A report from her cardiologist in her hometown 3 months ago described her conduit stenosis as moderate to severe. Her truncal valve was also mild to moderately regurgitant at that time. She has mild developmental delay and has recently begun to have some diminished exercise tolerance compared to her preschool classmates. On exam she has normal work of breathing with SpO_2 of 96% on room air. Heart rate and blood pressure are within normal limits.

Key Objectives

- Understand the anatomy and physiology of truncus arteriosus before and after surgical repair.
- Describe potential sequelae of truncus arteriosus repair.
- Describe the preoperative assessment of children with repaired congenital heart disease.
- Identify the anesthetic implications of DiGeorge syndrome.
- Describe the implications of cardiac disease in a patient undergoing cleft palate repair.

Pathophysiology

What is truncus arteriosus?

Truncus arteriosus (TA) is a rare form of cyanotic congenital heart disease (CHD) in which the embryologic separation of the arterial trunk remains incomplete. The primary anatomic lesion is a common arterial trunk that supplies the pulmonary, coronary, and systemic circulations. (See Figure 25.1.) A single semilunar "truncal valve," often with an abnormal number of cusps, takes the place of the aortic and pulmonic valves. (See Figure 25.2.) Nearly all cases of TA are associated with a ventricular septal defect (VSD); therefore, mixing of deoxygenated and oxygenated blood occurs both in the ventricular chambers and in the common arterial trunk [1, 2]. Truncus arteriosus accounts for up to 3% of congenital cardiac defects and is estimated to occur in 60 to 140 patients per million live births [3]. It is frequently prenatally diagnosed with routine fetal echocardiography [4, 5].

Clinical Pearl

The primary anatomic lesion is a common arterial trunk that supplies the pulmonary, coronary, and systemic circulations. A single semilunar "truncal valve," often with an abnormal number of cusps, takes the place of the aortic and pulmonic valves.

How are the anatomic variations classified?

Two classification systems are currently used to categorize anatomic variations of the pulmonary arteries (PAs) in TA.

Collett and Edwards (C & E) originally described four different variations, three of which are still in use.

- ***Type 1***: The main PA originates from the common arterial trunk prior to bifurcation into the branch PAs.
- ***Type 2***: The right and left PAs arise separately from the posterior aspect of the main arterial trunk.
- ***Type 3***: The right and left PAs branch from the right- and left-lateral aspects of the main arterial trunk, respectively.

The ***modified Van Praagh classification*** [6] utilizes slightly different criteria.

- ***Type 1*** (analogous to C & E type 1): The main PA arises from the arterial trunk.
- ***Type 2*** (analogous to C & E types 2 and 3): The branch PAs arise separately from the arterial trunk.
- ***Type 3***: Only one branch PA arises from the arterial trunk, and the other PA is supplied by the ductus arteriosus or collateral vessels.

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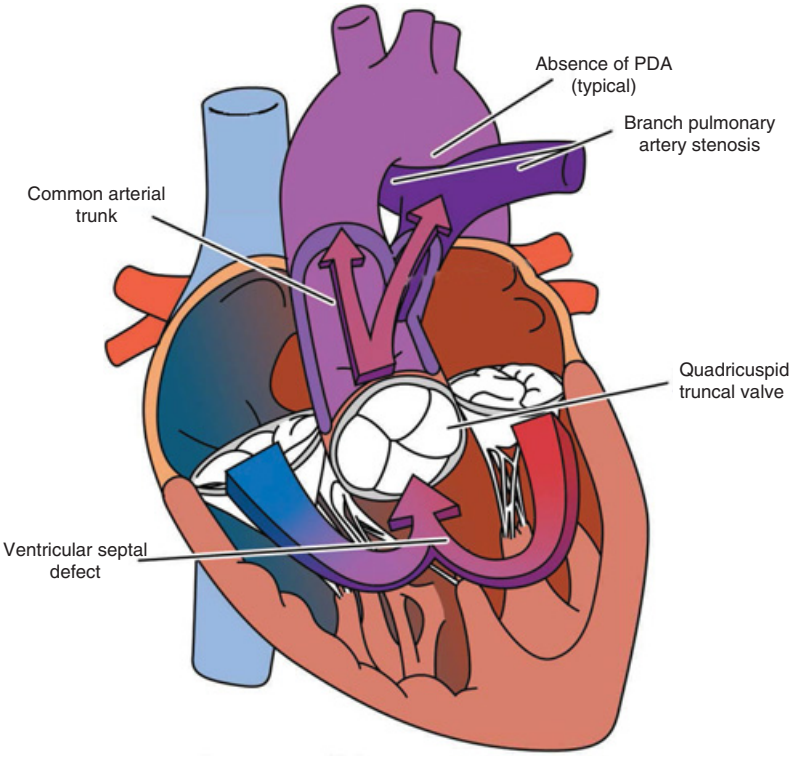

Figure 25.1 Truncus arteriosus. Drawing by Ryan Moore, MD, and Matt Nelson.

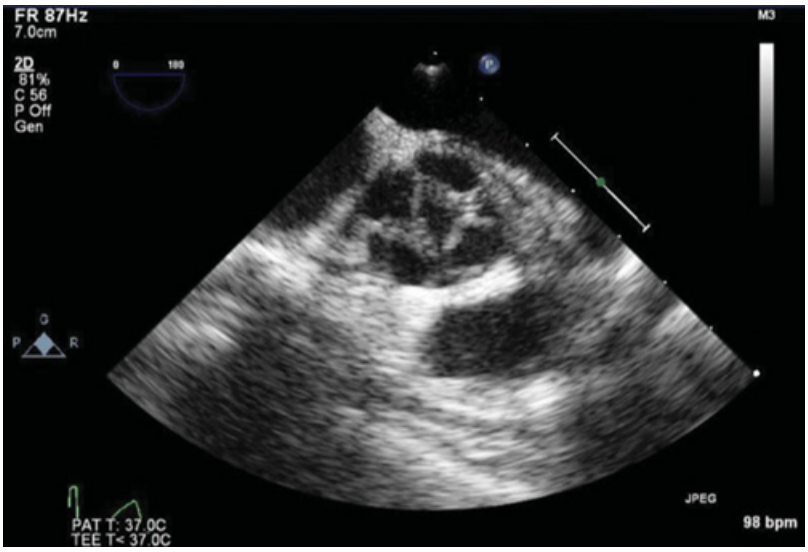

Figure 25.2 **Quadrileaflet truncal valve,** mid-esophageal short-axis image. Courtesy of Faith Ross, MD.

- A further subclassification denotes the presence (A) or absence (B) of a VSD.

In all cases of TA, a detailed understanding of the anatomy is important for surgical planning, but the physiologic principles remain largely the same.

What other congenital conditions are associated with TA?

Although TA can occur in isolation, it is commonly associated with comorbid cardiac and extracardiac congenital conditions. Patent foramen ovale (PFO) and other atrial

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septal defects (ASDs) are the most common coexistent cardiac malformations, followed by anatomic variations in the coronary arteries and the aortic arch (right-sided or interrupted) [7].

The most common extracardiac congenital malformations associated with TA are those within the heterogeneous spectrum of 22q11 deletion syndrome (22q11DS), found in 30%–40% of cases [3, 8]. While the incidence of TA has significant correlation with trisomy 21, TA remains rare in this population.

Clinical Pearl

The most common extracardiac congenital malformations associated with TA are those within the heterogeneous spectrum of 22q11DS; they are found in 30%–40% of cases.

What is the distribution of pulmonary and systemic blood flow in a neonate with unrepaired TA?

In the neonate with unrepaired TA, the relative distribution of pulmonary (Q_p) and systemic (Q_s) blood flow is dynamically related to pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR). This relationship is expressed as the ratio Q_p:Q_s. In the immediate peripartum period, PVR remains relatively high; thus, the distribution of blood flowing through the arterial trunk favors the systemic circulation (low Q_p:Q_s). With the physiologic decrease in PVR over the first hours, days, and weeks of life, the ratio of cardiac output entering the pulmonary circulation increases (increasing Q_p:Q_s).

Importantly, critical disruptions in the balance between Q_p and Q_s can precipitate rapid clinical deterioration, as in the setting of anesthetic induction or significant sympathetic stimulation. A dramatic rise in PVR or fall in SVR (very low Q_p:Q_s) will worsen hypoxemia, even to the point of critical systemic ischemia in the setting of right-to-left shunt. Alternatively, a substantial decrease in PVR (very high Q_p:Q_s) or rise in SVR is associated with pulmonary overcirculation and low systemic cardiac output that may be insufficient to meet systemic metabolic demands. The myocardium is at particular risk in this state, as low mixed venous oxygen saturation combined with low coronary perfusion pressure can severely impair the supply of oxygen to the myocardium.

What is the expected oxygen saturation in a neonate with TA? Why is this important?

Peripheral oxygen saturation in neonates with TA depends on the dynamic relationship between Q_p and Q_s. Oxygen saturations are often in the mid-80s shortly after birth, at which time PVR and SVR are relatively balanced. Saturations increase as PVR declines and may even reach the mid-to-high 90s if pulmonary overcirculation is severe. It is important to note that in this scenario, a saturation in the high 90s is not a reassuring sign, as it indicates severe pulmonary overcirculation and is thus concerning for accelerated decompensation toward congestive heart failure (CHF) and pulmonary vascular disease.

Clinical Pearl

Near-normal arterial saturation in a patient with unrepaired TA can be an indication of severe pulmonary overcirculation and is thus concerning for risk of inadequate systemic cardiac output and cardiac arrest, with intermediate to long-term risks of accelerated decompensation toward CHF and pulmonary vascular disease.

What is the natural history of unrepaired TA?

Patients with TA become symptomatic early in infancy because PVR falls over the first days to weeks of life, resulting in pulmonary overcirculation (high Q_p:Q_s). Ultimately, most patients with unrepaired TA succumb to high-output heart failure in the setting of pulmonary overcirculation, a condition that is accelerated by an incompetent truncal valve, which further loads the volume of the left ventricle and increases ventricular end-diastolic pressures. (See Figure 25.3.) The small number of patients who survive infancy develop severe pulmonary vascular disease, ultimately culminating in pulmonary hypertension. Pulmonary hypertension further contributes to critical changes in Q_p:Q_s that underlie the near-universal childhood mortality associated with unrepaired TA.

How are coronary blood flow and myocardial perfusion compromised in unrepaired TA?

Truncus arteriosus may be associated with abnormalities in the coronary arteries, such as single coronary artery, abnormally positioned coronary origins, coronary ostial stenosis, or intramural coronary arteries. In all variations of coronary anatomy, myocardial perfusion may be further compromised by diastolic steal into the pulmonary arteries or by low truncal root diastolic pressures in the setting of truncal valve insufficiency. Increased ventricular end-diastolic pressure in the setting of valve insufficiency and CHF further impairs myocardial perfusion. Finally, the risk is compounded by the baseline hypoxemia in the setting of a mixing lesion, and hypoxemia may be transiently worsened in the setting of an acute rise in right-to-left shunt.

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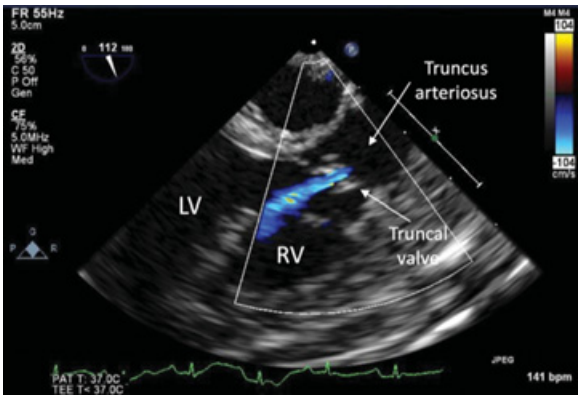

Figure 25.3 Truncal valve insufficiency and large ventricular septal defect, mid-esophageal long-axis image. Courtesy of Faith Ross, MD.

Clinical Pearl

In all variations of coronary anatomy, myocardial perfusion may be further compromised by diastolic runoff into the pulmonary arteries or by low truncal root diastolic pressures in the setting of truncal valve insufficiency.

At what age is TA typically repaired? Why is early repair important?

Ideally, TA is repaired within the first weeks of life. Patients with severe pulmonary overcirculation refractory to medical management or who require ongoing mechanical ventilation require repair in the first few days of life. Significant pulmonary overcirculation results in decreased systemic flow and low cardiac output (high Q_p:Q_s), myocardial ischemia, heart failure, and accelerated pulmonary vascular hypertensive disease, all of which can be improved with early surgical repair. Repair after 3 months of age carries increased risk for perioperative mortality.

What does the surgical repair of TA achieve?

Definitive repair of TA achieves three anatomic endpoints to restore separate pulmonary and systemic circuits:

- Removal of the pulmonary arteries from the arterial trunk such that the latter, now called the aorta, provides only systemic outflow
- Closure of the VSD
- Creation of a right ventricle to pulmonary artery (RV–PA) conduit

Truncal valve and aortic arch repair may occur during the same surgery if defects in either are likely to compromise successful recovery from the initial repair, or these may be delayed until an older age if a "wait and watch" approach is appropriate. Truncal valve replacement is occasionally performed at the time of initial repair if truncal valve dysfunction is severe; however, due to the complications associated with frequent valve replacements in a growing child, a significant degree of truncal valve dysfunction is typically accepted before resorting to valve replacement.

What postoperative complications and long-term sequelae follow surgical repair of TA?

Fortunately, >90% of patients survive initial surgical repair of TA. Perioperative mortality increases with older age at intervention, additional cardiac defects, need for postoperative tracheostomy, and comorbid 22q11DS. An estimated 40%–60% of patients who survive to discharge may require subsequent surgical repair of the truncal valve, endovascular or surgical reintervention to decrease right ventricular outflow tract obstruction in the setting of RV–PA conduit stenosis, or surgical repair of residual defects of the aortic arch within the first decade of life [9]. Those with coronary artery anomalies have an increased risk of coronary insufficiency.

Clinical Pearl

Over 90% of children with TA survive their initial repair. Perioperative mortality increases with older age at intervention, additional cardiac defects, need for postoperative tracheostomy, and comorbid 22q11DS.

Anesthetic Implications

Apart from TA, what other anomalies are associated with 22q11DS?

There is substantial phenotypic heterogeneity in patients with 22q11DS, some of which are recognized constellations of multiorgan anomalies including DiGeorge, Catch-22, conotruncal facial, and velocardiofacial syndromes. 22q11 deletion syndrome is associated with immunodeficiency; hypocalcemia in the setting of parathyroid dysfunction; craniofacial abnormalities including cleft lip/palate; and cardiac defects including tetralogy of Fallot, interrupted aortic arch, VSD, and TA. Other anomalies of the genitourinary, renal, cognitive, endocrine, neurologic, and ophthalmologic organs are not uncommon in these patients.

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How are these anomalies relevant to anesthetic management?

Even in the absence of CHD, features of 22q11DS can pose challenges to anesthesiologists. Neurodevelopmental delay may complicate the induction of anesthesia and necessitate careful preoperative anxiolysis. Dysmorphic facies, including micrognathia, retrognathia, palatal abnormalities, and laryngotracheomalacia, can impede airway management. Notably, Pierre Robin sequence is present in 10%–20% of patients with 22q11DS. In preparation for the cleft palate repair in this scenario, a thorough airway exam and review of previous anesthetic records are crucial to developing an appropriate plan for airway management in the setting of facial dysmorphology. Deficits in cell-mediated immunity necessitate irradiated blood products when transfusion is indicated. Limb hypoplasia and other malformations may make peripheral venous and arterial access challenging to obtain. Hypoparathyroidism may result in critical hypocalcemia requiring active replacement, especially in the setting of transfusion of citrated blood products [10].

Clinical Pearl

Patients with impaired cell-mediated immunity related to DiGeorge syndrome continue to require irradiated blood products throughout life, and blood products should be ordered well in advance to ensure adequate preparation time.

What clinical, laboratory, and imaging findings are critical to the preoperative evaluation of a patient with repaired TA prior to elective noncardiac surgery?

Assuming the child has fully recovered from the original cardiac surgical repair, the anesthesiologist should solicit a careful history of the patient's recent health, focusing on new or worsening symptoms of heart failure; valve insufficiency; or pulmonary vascular disease, including declines in exercise tolerance, diaphoresis, failure to thrive, syncopal episodes, unexpected weight gain or loss, or unusual shortness of breath. A physical exam complements the patient's history; increased work of breathing, new cyanosis, peripheral edema, a congested or tender liver, or changes in cardiac murmur are all concerning for complications of TA and its repair that may require intervention prior to elective noncardiac surgery.

The specific anatomy of the surgical repair and any reinterventions or complications in the interim must be appreciated. Recent imaging, such as transthoracic echocardiography, cardiac magnetic resonance imaging (CMRI), computed tomography, or cardiac catheterization will aid in the understanding of each individual's cardiac anatomy and physiology. A recent ECG may reveal inherent or acquired conduction abnormalities. Laboratory investigations should focus primarily on the appropriate type and screen of blood products if transfusion is likely, as these patients have often been exposed to blood products early in life and may have acquired sensitivity to uncommon red cell antigens.

Clinical Pearl

Patients with repaired TA should undergo routine cardiology follow-up throughout their lives. Patients should have a recent cardiology evaluation prior to any elective surgery. Evaluation should focus on identifying new or worsening signs or symptoms of heart failure, RV-PA conduit stenosis, valve incompetence, or pulmonary vascular disease.

When is preoperative consultation with cardiology indicated?

Consultation with the child's cardiologist may be warranted prior to an elective surgical procedure in several settings. Cardiology consultation is appropriate to determine the need for updated or clarified diagnostic imaging, including transthoracic echocardiography, CMRI, or heart catheterization, especially if relevant results are inaccessible or if there are new or worsening signs or symptoms of heart failure, valve incompetence, or pulmonary vascular disease. In all cases of concern, elective surgery should be delayed until complete cardiology evaluation and optimization.

The patient described in this scenario has some features that are concerning for increased anesthetic risk. Her RV–PA conduit gradient has progressed to the point where conduit intervention may be indicated prior to elective surgery, particularly as she is showing signs of decreased exercise tolerance. Moderate to even severe pulmonary stenosis can be reasonably well tolerated in children when the stenosis has developed gradually enough for the RV to compensate. However, it would be wise to determine that this is the case for this child. It is important to note the progression of the RV-PA conduit gradient over time and assess whether the RV function has remained normal on echocardiographic and/or CMRI examinations. Although her truncal valve has some insufficiency, this is often well tolerated under anesthesia. Overall, the child's cardiologist should at the very least be aware of the upcoming elective surgery and deem the timing acceptable.

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Does this patient require infective endocarditis prophylaxis?

Yes. Current guidelines recommend endocarditis prophylaxis for patients with repaired CHD with bioprosthetic or homograft valves or with residual shunting or valvular incompetency at or near the site of bioprosthetic or homograft (e.g., conduit). Furthermore, although cleft palate repair is not a dental procedure per se, there is manipulation of the gingival mucosa that creates a risk for hematogenous seeding of oral bacteria. For these reasons, this patient should receive targeted antibiotic prophylaxis. Institutional guidelines often specify which antibiotic regimen is indicated for each patient, determined by the age, weight, and ability to take oral medications, but they often involve amoxicillin, cephalexin, clindamycin, or vancomycin. Note that when cefazolin is given for dual endocarditis and surgical prophylaxis, the recommended dose may be higher than for standard surgical prophylaxis [11].

Clinical Pearl

When cefazolin is given for dual endocarditis and surgical prophylaxis, the recommended dose may be higher than for standard surgical prophylaxis.

What are important features of a safe anesthetic plan for this patient?

In a well-compensated patient with repaired TA presenting for cleft palate repair, standard recommended American Society of Anesthesiologists monitors and routine peripheral venous access are sufficient. The details of the anesthetic plan should be tailored to the physiology of the individual patient, with special attention to optimizing PVR and SVR in patients with significant truncal valve insufficiency or RV–PA conduit stenosis. Effective preoperative anxiolysis can safely be employed to avoid unnecessary emotional upset, at the discretion of the anesthesia provider.

Induction of anesthesia and tracheal intubation should be swift and smooth, as hypercarbia, hypoxemia, and painful stimulation all increase PVR and can precipitate right heart failure in the setting of significant conduit stenosis, particularly if the PVR is reactive. Furthermore, increases in heart rate secondary to sympathetic stimulation impair filling of the hypertrophic and noncompliant RV. As craniofacial abnormalities can make rapid intubation a challenge at times, the anesthetic plan should include a skilled airway operator with all needed tools and personnel immediately available to them.

In this patient, who has both RV–PA conduit stenosis and truncal valve insufficiency, hemodynamic management must consider both of these conditions. Truncal valve insufficiency can be minimized by targeting low SVR and high-normal heart rate. However, as noted earlier, lower heart rates may be required to allow adequate right ventricular filling. These competing hemodynamic goals can make management of these patients challenging. These dueling priorities leave the anesthesiologist with the overarching goal of maintaining the child's baseline hemodynamics as much as possible, while of course providing supplemental oxygen. Cardiac output can be maintained close to baseline via a balanced anesthetic as well as the use of catecholaminergic drugs when necessary. The smooth induction of anesthesia may include the use of opioid medications, but these should be administered judiciously for the remainder of the procedure, as postoperative airway obstruction is a frequent complication of palate repair. Maintenance of euvolemia for these patients is also important especially when considering a hypertrophied and noncompliant RV.

Although postsurgical coronary artery pathology typically manifests in the first several months after repair, the possibility of coronary hypoperfusion in the setting of significant truncal valve insufficiency should be considered. Providers should pay close attention to diastolic blood pressure as this is the driving force for coronary perfusion.

What are the implications of RV–PA conduit stenosis and how does this finding affect the intraoperative anesthetic plan?

A frequent sequela of TA repair is stenosis of the RV–PA conduit. Right ventricular outflow tract obstruction (RVOTO) predisposes the patient to increased risk of RV failure. However, the low-pressure RV can withstand gradually increasing RVOTO for a long period of time in children, typically years, until there are clinical symptoms and/or signs of irreversible RV damage. Acute changes are more poorly tolerated. Acute rises in PVR, as can occur under anesthesia, contribute to an acutely increased RVOTO. However, clinically relevant increases in PVR are usually only mildly consequential when downstream from moderate to severe RV–PA conduit stenosis as in our patient.

Conversely, children with pulmonary hypertensive vascular disease or pulmonary hypertension are at risk for pulmonary hypertensive crises under anesthesia with subsequent acute RV failure, morbidity, and mortality.

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As such, in addition to the features of the anesthetic plan described for all patients with repaired TA, anesthesia providers may choose to have the following immediately available:

- Agents to support right ventricular function (e.g., milrinone, epinephrine)
- Agents to provide pressor support without increasing PVR (e.g., vasopressin)
- Agents to decrease PVR (e.g., high FiO_2, inhaled nitric oxide) should there be concern for acute right heart failure in the setting of critically increased pulmonary artery pressures

Maintenance of euvolemia is also essential.

Clinical Pearl

A frequent long-term complication of TA repair is stenosis of the RV–PA conduit, which predisposes the patient to increased risk of RV failure. Right ventricular function may deteriorate under anesthesia due to increased afterload (PVR), tachycardia, or poor myocardial perfusion.

What is the appropriate level of immediate postoperative care for this child?

All children will be admitted to the hospital after palatal surgery. For a child with repaired TA and good functional capacity who underwent uncomplicated, routine cleft palate repair, the surgical floor is an appropriate disposition. However, if there is concern for right ventricular dysfunction, pulmonary vascular disease, or myocardial ischemia, the patient would likely benefit from close monitoring in an intensive care unit, as airway obstruction following oropharyngeal surgery may be so severe as to cause hypoxemia and hypercarbia, thus precipitating right heart failure in high-risk patients.

What are the options for analgesia after cleft palate repair? Why is effective analgesia important in the setting of RV–PA conduit stenosis?

Effective postoperative analgesia remains a vital component of perioperative care for all children, and especially in the setting of residual cardiopulmonary disease such as RV–PA conduit stenosis, for the reasons described earlier. Patient controlled opioid analgesia is an excellent option for patients who are old enough to effectively use this pain control strategy. Scheduled analgesia adjuvants, such as acetaminophen, are an effective opioid-sparing approach to pain control, as is true for all postoperative patients. An acute pain specialist may provide tailored service and safe, timely adaptations of the pain control plan for some patients who experience particular discomfort. At centers with trained providers, regional anesthesia is an excellent option for some patients undergoing cleft palate repair [12].

Clinical Pearl

Analgesia following cleft palate repair in the setting of repaired TA should optimize regional and multimodal options, as hypercarbia related to airway obstruction and hypoventilation may exacerbate RVOTO and risk RV decompensation and failure.

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Suggested Reading

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