Chapter 10

Tetralogy of Fallot, Pulmonary Atresia, and Aortopulmonary Collaterals

Mikel Gorbea and Lisa Wise-Faberowski

Case Scenario

A 2-year-old child weighing 11.5 kg with a history significant for tetralogy of Fallot, pulmonary atresia, multiple aortopulmonary collaterals, and DiGeorge syndrome presents for thyroglossal duct cyst excision under general anesthesia. He had a prenatal diagnosis of tetralogy of Fallot and pulmonary atresia, and at age 4 months underwent unifocalization of his left- and right-sided aortopulmonary collaterals via thoracotomies, along with placement of a 4 mm central shunt from the aorta to the unifocalized neopulmonary artery. The ventricular septal defect remains open. He has had multiple cardiac catheterizations for balloon dilation of pulmonary arteries and sees his cardiologist regularly.

Physical exam is notable for dysmorphic facies, slight clubbing, and bilateral thoracotomy incision scars. A holosystolic murmur is auscultated at the left sternal border. Lungs are clear and liver edge is 2 cm below the right costal margin. The hemoglobin count is 15 and hematocrit 44%, with all other laboratory results within normal limits. Current vital signs are heart rate 111 beats/minute, blood pressure 90/50 mm Hg, respiratory rate 25 breaths/minute, and SpO_2 82% on room air.

Recent transthoracic echocardiography shows the following:

- *Atretic pulmonic valve*
- *Large ventricular septal defect with bidirectional flow*
- *Patent central shunt to small neopulmonary arteries with diastolic flow reversal in the descending aorta*
- *Low-normal right ventricular function*
- *Normal left ventricular function*

Key Objectives

- Describe key anatomic and physiologic features of tetralogy of Fallot, pulmonary atresia, and multiple aortopulmonary collaterals.
- Discuss current surgical palliations in this patient population and their impact on subsequent perioperative anesthetic management.
- List hemodynamic and physiologic considerations in perioperative care for a patient with tetralogy of Fallot, pulmonary atresia, and multiple aortopulmonary collaterals.

Pathophysiology

What are key anatomic characteristics of tetralogy of Fallot with pulmonary atresia and aortopulmonary collaterals?

Tetralogy of Fallot with pulmonary atresia (PA) and multiple aortopulmonary collaterals (MAPCAs) represents an extreme variation of tetralogy physiology with an incidence of 0.7 per 10,000 live births. While TOF/PA includes the basic findings of TOF, including an aorta that "overrides" an unrestrictive ventricular septal defect (VSD) and right ventricular hypertrophy (RVH), instead of pulmonary stenosis (PS) there is complete atresia of the pulmonary valve and right ventricular outflow tract. There is variable hypoplasia of the main pulmonary artery, and in severe cases the right and left pulmonary arteries are not confluent.

When complete pulmonary atresia is present, no blood flow path exists from the right ventricle (RV) into the pulmonary arteries. In addition, native pulmonary vasculature is typically hypoplastic, stenotic, or atretic. In TOF/PA/MAPCAs, pulmonary blood flow (PBF) occurs via MAPCAs. Multiple aortopulmonary collaterals are vessels that arise from the descending thoracic aorta or any of its branches (subclavian, bronchial, celiac, or intercostal), often anastomosing proximal to branch pulmonary arteries. Pulmonary blood flow via MAPCAs is variable and nonuniform. Bronchopulmonary segments can have multiple vascular contributions arising from one or more vascular sources, and PBF may vary between segments.

https://doi.org/10.1017/9781108657341.011 Published online by Cambridge University Press

Clinical Pearl

In TOF/PA, instead of pulmonary stenosis there is complete atresia of the pulmonary valve and right ventricular outflow tract. There is variable hypoplasia of the main pulmonary artery, and in severe cases the right and left pulmonary arteries are not confluent.

What is the path of blood flow at birth in this patient?

- Systemic venous blood returns via the inferior vena cava (IVC) and the superior vena cava (SVC) to the right atrium (RA).
- Blood passes via the nonrestrictive VSD from the RV to the LV, mixing deoxygenated systemic venous return with oxygenated pulmonary venous blood return.
- Blood exits via the LV outflow tract.
- Pulmonary blood flow is provided via a PDA (if present) or via MAPCAs from the aorta.

How do patients with TOF/PA/MAPCAS receive pulmonary blood flow?

In TOF with PS some of the systemic venous blood passes through the RV outflow tract to the PAs. In contrast, ***in TOF/PA ALL pulmonary blood flow is from the aorta***: either via flow from a patent ductus arteriosus (PDA), MAPCAs, or a combination of the two.

In more than half of cases the native central pulmonary arteries are well developed and supplied by the PDA. Confluence between the right and left pulmonary arteries is a key differentiator in disease spectrum severity. Confluent branch pulmonary arteries occur in the majority (85%) of patients. In the absence of pulmonary artery (PA) confluence, MAPCAs are always present and lung segments are perfused via individual MAPCAs as well as the nonconfluent PAs.

In TOF/PA, in the absence of a path from the RV to the pulmonary vasculature, presence of a VSD is vital for establishing a path that allows systemic venous return to enter the LV, aorta, and subsequently MAPCAs.

Clinical Pearl

In TOF/PA ALL pulmonary blood flow is provided via the aorta: either from a PDA, MAPCAs, or a combination of the two. Pulmonary blood flow supplied by MAPCAs is variable and nonuniform. Bronchopulmonary segments can have multiple vascular contributions arising from one or more vascular sources, and PBF may vary between segments. In the absence of PA confluence, lung segments are perfused via individual MAPCAs.

How do MAPCAs provide pulmonary blood flow?

There is variation in PBF in patients with TOF/PA/MAPCAs due to variability of MAPCA size, origin, and connection to the lungs. Collateral arteries may be classified as ***direct*** when arising from the aorta and ***indirect*** when originating from the brachiocephalic (25%) or coronary arteries (7%). Total number of MAPCAs usually range from two to six with indirect collaterals being less numerous when compared to direct collaterals. Collaterals may course directly into the pulmonary parenchyma to supply a discrete bronchopulmonary segment or toward the lung hilum with direct anastomosis to branches of the pericardial pulmonary arteries at extrapulmonary, hilar, lobar, or segmental levels. Bronchopulmonary segments can contain a dual supply of blood from both intrapericardial pulmonary and systemic-to-pulmonary collaterals in 4%–14% of patients.

What is the physiologic impact of MAPCAs and what are their characteristics?

Multiple aortopulmonary collaterals may induce anatomic and physiologic changes in pulmonary vessels due to chronic endothelial exposure to systemic arterial pressures. They may also induce arterialization of distal low pressure pulmonary arterial tributaries, and the resultant increased pressure and volume burden results in discrete dilations within vascular branch points distal to their respective MAPCA insertions. Long-standing elevated pressures in pulmonary vessels often induce vascular endothelial muscular changes and increased vascular reactivity.

In addition to abnormal PBF, lung parenchymal development is also impacted in TOF/PA/MAPCAs. In utero, diminished PBF contributes to decreased distal pulmonary vessel arborization. As a result of compromised perfusion, lungs may be hypoplastic. Importantly, MAPCAs anastomotic sites may become stenotic over time, negatively impacting PBF.

Clinical Pearl

In addition to abnormal PBF, lung parenchymal development is also impacted in TOF/PA/MAPCAs. In utero, diminished PBF contributes to decreased distal pulmonary vessel arborization. As a result of compromised perfusion, lungs may be hypoplastic. Importantly, MAPCAs anastomotic sites may become stenotic over time, negatively impacting PBF.

What cardiac surgical palliative or repair pathways are available for these patients?

The ultimate surgical treatment goal, if possible, is restoration of two discrete and non-mixing systemic and pulmonary circulations. Complete separation of the systemic and

https://doi.org/10.1017/9781108657341.011 Published online by Cambridge University Press

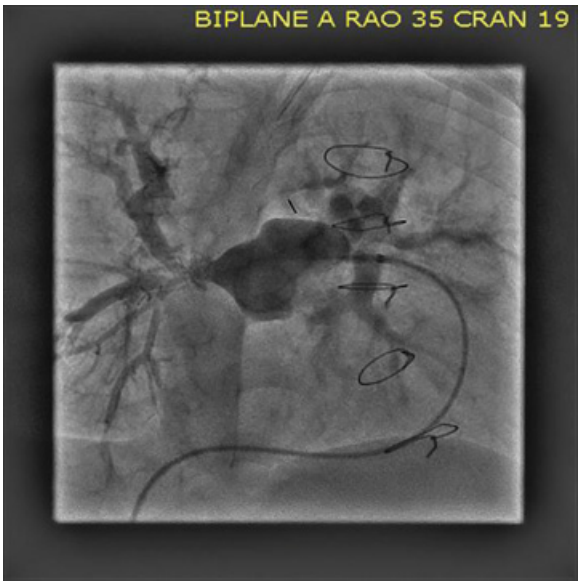

Figure 10.1 Unifocalized right pulmonary artery. An angiogram is performed in the right pulmonary artery in the AP projection. The dilated proximal conduit extending into the proximal right pulmonary artery is noted, with the stenotic, unifocalized right pulmonary artery branches extending distally. Courtesy of Russel Hirsch, MD.

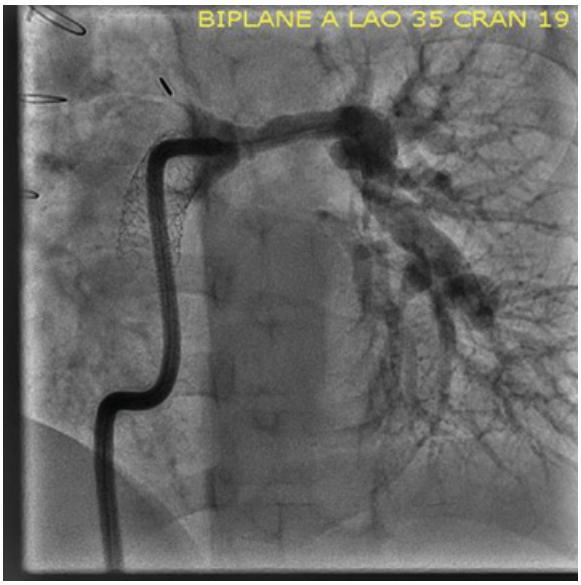

Figure 10.2 Unifocalized left pulmonary artery. An angiogram is performed in the left pulmonary artery in the AP projection. The long segment stenotic proximal conduit is noted with the small unifocalized left pulmonary artery branches extending distally. Courtesy of Russel Hirsch, MD.

pulmonary circuits with the lowest possible RV pressure is the ultimate goal. Because of anatomic complexity in TOF/PA/MAPCAs, single step complete repair may not be achievable, and a staged approach or palliation may be utilized. The feasibility of complete repair is heavily dependent on the arborization of the pulmonary bed, the size and morphology of the pulmonary arteries and MAPCAs, and the amount of PBF.

Complete repair includes the following:

- Collateral pulmonary arteries are "unifocalized" into a neopulmonary artery confluence. (See Figures 10.1 and 10.2.) Aortopulmonary collaterals and pulmonary arteries supplying bronchopulmonary segments are combined into a single source via an augmented and reconstructed PA.
- Right ventricular outflow tract obstruction is corrected and a valved conduit connecting the RV to the PA confluence is placed.
- Intraventricular/atrial connections are closed.

A staged or palliated approach is common in TOF/PA/MAPCAs.

- Complex anatomy may require unifocalization in stages and a surgically created central (aortopulmonary) shunt may be placed. The centrally created shunt or conduit is connected to the small but functional unfocalized neopulmonary arteries to provide a source of PBF and support pulmonary arterial vascular growth.
- A right ventricular to pulmonary artery conduit to the neopulmonary confluence can be placed.
- It may not be possible to close the VSD if the pulmonary arteries and the pulmonary vascular bed are not of sufficient caliber (<50%–75% of normal) to handle the entire cardiac output from the RV. The VSD is left open and acts as a "pop off" to allow for some blood flow from the RV to LV; mixing at the ventricular level of deoxygenated and oxygenated blood means that child will remain cyanotic. When the VSD is closed before the pulmonary vascular bed is able to handle the entire RV cardiac output, the child will exhibit signs and symptoms of RV hypertension and RV failure.

Some institutions utilize a staged procedure via thoracotomy, unifocalizing the pulmonary arterial supply to a single lung, either the left or right lung. The pulmonary blood supply to the contralateral lung is unifocalized at a later date. Correction of the VSD can occur after the complete restoration of pulmonary arterial supply to both lungs during the final staged correction.

Regardless of anatomy, evidence suggests that single stage complete correction may offer advantages and improved outcomes compared to staged unifocalization

https://doi.org/10.1017/9781108657341.011 Published online by Cambridge University Press

in greater than four-fifths of cases. The exception to this rule is noted when confluent central PAs with normal arborization share dual blood supply with MAPCAs and present with cyanosis. Patients undergoing complete unifocalization with intracardiac repair, however, are at risk of developing prolonged postoperative respiratory failure. In either staged or complete repair, pulmonary arteries are often hypoplastic and stenosis occurs over time. Frequent interventional catheterizations for balloon angioplasty and/or stenting are common.

Clinical Pearl

In either staged or complete repair, pulmonary arteries are often hypoplastic and stenosis occurs over time. Frequent interventional catheterizations for balloon angioplasty and/or stenting are common.

What is the physiology of PBF supplied by MAPCAs?

At birth, anatomic variations in MAPCAs, lung development and PVR determine the severity of cyanosis. In patients with TOF/PA/MAPCAs, PBF is often limited by abnormal, hypoplastic pulmonary vessel anatomy and neonatal pulmonary vascular elevation; Q_p:Q_s may be low-normal or low, resulting in low oxygen saturations.

Assessing the importance of the contribution of the PDA to overall PBF is a key in the perinatal period. The PDA may be a significant source of PBF via either MAPCAs or hypoplastic native pulmonary arteries. If saturations fall to unacceptable levels with PDA closure, prostaglandins are utilized to reopen the PDA and the patient will require neonatal intervention to maintain ductal patency. This can be accomplished either via an interventional cardiac catheterization with stenting of the PDA or placement of a surgical aortopulmonary shunt in the cardiac operating room. In the less common setting of abundant or unrestricted MAPCAs, PBF increases as PVR falls after birth resulting in pulmonary overcirculation and congestion.

After the neonatal period, dominant physiology and hemodynamic expectations depend on anatomy of the MAPCAs, Q_p:Q_s, potential lung disease, and pulmonary vascular reactivity.

What are the expectations for oxygen saturation, Q_p:Q_s, and hemodynamics?

Patients whose lesions are completely repaired and who no longer have interventricular or atrial connections should have normal oxygen saturations. Palliated and unoperated patients may present with reduced oxygen saturations depending on the palliation, degree of residual collaterals, intracardiac shunting, and PVR.

Because of the variation in PBF and resultant oxygen saturations that can exist in patients with unrepaired or palliated TOF/PA/MAPCAs, a thorough understanding of each patient's current anatomy and palliative stage is essential in order to determine appropriate hemodynamic and saturation goals. Baseline oxygen saturations should be noted at both rest and with exertion (elevated RV pressure) in order to better define current physiology. In order to minimize over-circulation of both pulmonary and systemic arterial systems, an optimal oxygen saturation should ideally reflect a balanced Q_p:Q_s on room air, resulting in oxygen saturations of 75%–85%. The combination of abnormal pulmonary vessel anatomy, hypoplastic lungs, and variable pulmonary vascular reactivity often creates a resultant physiology that typically has low or low-normal Q_p:Q_s and may present challenges with oxygenation and ventilation. Oxygen saturations may be 70%–80% with physical and physiologic sequelae of chronic hypoxemia such as clubbing and polycythemia. Patients with elevated pulmonary vascular reactivity may be chronically taking pulmonary vasodilators.

Clinical Pearl

A thorough preoperative assessment and cardiology consultation is recommended for understanding of the individual child's dominant physiology, as a wide spectrum of variation can exist in PBF and resultant oxygen saturations in patients with TOF/PA/MAPCAs.

What long-term complications may occur with TOF/PA/MAPCAs?

After repair or palliation, patients with TOF/PA/MAPCAs can suffer from short- and long-term respiratory insufficiency. Immediate postoperative respiratory complications following complete repair with unifocalization are common. Prolonged postprocedural intubation, longer exposure to anesthetics, intricate surgical dissection, vascular reconstruction, and extensive suture lines contribute to increased postoperative respiratory compromise. Prolonged intubation may result in tracheal mucosal and/or vocal cord injury as well as worsen preexisting or new bronchomalacia. Dissection within the thorax may increase the risk of iatrogenic unilateral or bilateral recurrent laryngeal nerve damage resulting in postoperative stridor, airway obstruction, and potential respiratory failure.

https://doi.org/10.1017/9781108657341.011 Published online by Cambridge University Press

Airway obstruction or malacia due to changing intrathoracic anatomy is possible. Left and right reconstructed pulmonary arteries may cause compression of posterior airway structures including the bronchi. Intrathoracic obstruction secondary to compression of bronchi may go unnoticed in these patients until chest closure is attempted, resulting in the need for anastomosis revision, intraoperative bronchoscopy, or surgical fixation of structures anterior to the site of compression to the chest wall to relieve the obstruction.

Clinical Pearl

Airway obstruction or malacia due to changing intrathoracic anatomy is possible. Left and right reconstructed pulmonary arteries may cause compression of posterior airway structures including the bronchi.

Although most collateral arteries are present at birth, evidence suggests that more collaterals may develop postnatally and post-palliation resulting in "acquired" or unmasked collateral arteries. Origins for these acquired collateral arteries include intercostal, bronchial, or systemic arteries. "Aggravated" acquired collateral arteries develop after a surgically created systemic-to-pulmonary shunt. Residual MAPCAs can be unmasked following complete repair when initial anatomic location or vessel size prevented discovery at the time of repair. Increased collateral flow or persistent MAPCAs can contribute to pulmonary congestion, systemic hypoperfusion, and gradually increased PVR over time due to collateral flow.

Patients with TOF/PA/MAPCAs often require repeated diagnostic and interventional cardiac catheterization procedures. Stenosis of the newly reconstructed pulmonary vasculature after complete repair may require reintervention in up to one-third of patients in order to maintain the lowest possible systolic PA pressures. Diagnostic pressure measurements and coiling of persistent MAPCAs or newly developed collaterals may be indicated.

Heterografts, homografts, and synthetic conduits are often used in palliation or complete repair of TOF/PA/MAPCAs. Right ventricle-to-pulmonary artery (RV-PA) conduits are subject to the development of progressive stenosis, regurgitation, and calcification. Homograft deterioration is progressive, with only 30% being intact at 15 years. Prolonged conduit regurgitation and/or stenosis increases RV strain, resulting in ventricular fibrosis, restriction, dilation, and increased risk for arrhythmias and failure. Additionally, conduits are nonnative and do not grow with the pediatric patient. Therefore, conduits need periodic catheter-based reintervention or surgical replacement.

Clinical Pearl

Follow-up recommendations following partial or complete repair include routine catheterization 1 year after repair. It is not uncommon for balloon angioplasty to be performed at that time, even for minor stenoses, in an effort to maintain the lowest total resistance to pulmonary flow possible. Surgical reintervention on the pulmonary arteries is not uncommon and generally occurs at the time of conduit replacement.

Anesthetic Implications

Are there associated genetic syndromes to consider? Would they affect the perioperative plan?

Multiple syndromes can be associated with TOF/PA/MAPCAs including chromosome 22q11 microdeletion (DiGeorge syndrome) and Alagille syndrome. Between 17% and 40% of patients with TOF/PA/MAPCAS have chromosome 22q11 microdeletion. In affected patients a 14% incidence of airway abnormalities including laryngeal web, subglottic stenosis, and laryngomalacia has been noted. In these instances, direct laryngoscopy can provide upper airway evaluation but chest computed tomography (CT) and bronchoscopy may be necessary to provide lower airway evaluation. Preoperative genetic testing and counseling are helpful as these syndromes have associated noncardiac manifestations that can impact anesthetic planning. Issues may include potential difficult airway management, immunodeficiency, hypocalcemia, and liver dysfunction.

Clinical Pearl

Between 17% and 40% of patients with TOF/PA/MAPCAS have chromosome 22q11 microdeletion. In affected patients a 14% incidence of airway abnormalities including laryngeal web, subglottic stenosis, and laryngomalacia has been noted.

What imaging modalities may be utilized to guide surgical intervention and outcomes in patients with TOF/PA/MAPCAs?

Annual transthoracic echocardiography should be performed with a focus on both size and function of the right and left ventricles, including pressure gradient estimates through the right ventricular outflow tract that may

https://doi.org/10.1017/9781108657341.011 Published online by Cambridge University Press

rule out or diagnose new or worsening obstruction. The presence of aortic valvulopathies or history of previous aortic balloon dilation places patients at risk of developing progressive aortic insufficiency. Poor visualization of pulmonary arteries within the adult population relative to their pediatric counterparts can hinder adequate echocardiographic assessment of these vessels.

Computed tomography and angiography (CTA) may be utilized when detailed interrogation of the complex pulmonary circulation is needed. Improved spatial resolution enhances visualization of preexisting stents and the extent of collateralization with the added benefit of assessment of ventricular function via cine viewing. Computed tomography/angiography is also useful in identifying the proximity of conduit location relative to the sternum that may warrant peripheral cannulation in patients that have undergone multiple sternotomies. Unfortunately, the use of CT presents patients with the risks of both radiation and contrast exposure. Patients with abnormal CT scans should reflexively undergo catheterization for hemodynamic quantification and potential intervention to correct any newly discovered lesions.

What priorities exist for preoperative assessment of a patient with TOF/PA/MAPCAs?

The initial preanesthetic evaluation should elicit a complete history including original cardiac anatomy and current palliation stage. Detailed surgical palliative histories including both surgical and anesthetic courses should be reviewed. Recent echocardiographic and catheterization reports for patients undergoing noncardiac surgery should be reviewed. Consultation with cardiology preoperatively is recommended.

Exercise tolerance should be queried, and the most recent echocardiogram and imaging studies reviewed. Airway assessment including any history of prior prolonged intubations, tracheostomy, presence and severity of bronchomalacia, mass effect of the vasculature on airway segments, vocal cord integrity, and recurrent nerve injury is paramount for these patients.

Physical examination may be significant for systolic ejection murmurs with a loud palpable S2 component secondary to preexisting conduits. "To and fro" murmurs over a preexisting conduit location may indicate prominent conduit regurgitation. Classic diastolic murmurs indicating aortic regurgitation are not uncommon. Persistent, continuous murmurs noted laterally or around the posterior thorax may raise concern for the presence of new or persistent MAPCAs or previous surgical shunts that should be clinically correlated. The liver should be palpated to assess for congestion because of increased right heart pressure. A superficial examination of the skin may be notable for previous posterolateral thoracotomy scars on the trunk from prior systemic-to-pulmonary artery shunt procedures.

Preoperative instructions for cyanotic patients should emphasize the importance of minimizing fasting time. American Society of Anesthesiologists fasting guidelines should be followed, and clear liquid intake should be encouraged until 2 hours prior to surgery. If the patient is not the first case of the day, consideration should be given to starting an intravenous (IV) line with administration of maintenance fluids until the start of surgery. The patient's level of preoperative anxiety should be assessed as well; the use of distraction techniques, child life preparation, or oral premedication may all be considered if needed to allay anxiety.

What considerations are important during induction and maintenance of anesthesia?

Anesthetic induction and maintenance management goals for patients with TOF/PA/MAPCAs center on two goals:

- ***Balancing Q_p:Q_s in palliated patients*** can be achieved by knowing the patient's baseline oxygen saturation at rest, utilizing fraction of inspired oxygen concentration (FiO_2) during the procedure to maintain saturations in this range, and avoiding triggers that may increase PVR.
- ***Decreasing the hemodynamic burden experienced by the RV*** associated with changes in preload and afterload; a knowledge of baseline RV function and maintenance of adequate preload are useful for accomplishing this goal.

The choice of IV or inhalational induction and maintenance should be made based on the patient's previous palliation(s), current physiology, and degree of RV dysfunction. Palliated patients presenting without overt signs of severe congestive heart failure or cyanosis may undergo inhalational induction with sevoflurane with titration of the FiO_2 based on the patient's Q_p:Q_s. In this patient, with baseline oxygen saturations of 82% and low-normal RV function, an inhalation induction of anesthesia could be performed, or if the patient had undergone a prolonged period of fasting, placement of an IV preinduction would allow for a more controlled induction of anesthesia.

During induction and intubation an increased FiO_2 is often utilized and then adjusted once the patient is intubated. Ideally, the FiO_2 is adjusted to maintain an oxygen saturation between 75% and 85% to reflect a balanced Q_p:Q_s. Depending on the number and size of MAPCAs,

https://doi.org/10.1017/9781108657341.011 Published online by Cambridge University Press

changes in PVR may have significant impact on systemic and pulmonary flow. Decreasing PVR may cause considerable runoff from the systemic system into the lower pressure pulmonary arterial system and may result in systemic hypoperfusion and subsequent acidosis. Similarly, children with TOF/PA/MAPCAs are at risk for increased pulmonary vascular reactivity and may experience decreased PBF if PVR is acutely increased.

Patients with TOF/PA/MAPCAs are at risk for RV dysfunction. Right ventricular pressure overload may result from stenosis within the conduit, branch pulmonary arteries, previously unifocalized MAPCAs, hypoplastic pulmonary arteries, or other vascular arborization anomalies within the pulmonary system that may increase afterload experienced by the RV. In the setting of prolonged RV pressure overload, the right heart may dilate with subsequent development of tricuspid regurgitation, causing a volume and pressure loaded failing ventricle. Inhalational induction of anesthesia and the increased RV afterload associated with positive pressure ventilation (PPV) may cause worsening RV function or failure.

As volatile anesthetics decrease mean arterial blood pressure (MAP), systemic vascular resistance (SVR), and cardiac output (CO) in a dose-dependent manner, it may be prudent to limit the total dose of inhalational anesthetic or consider a total intravenous anesthetic for patients with severe RV dysfunction. Combinations of fentanyl and dexmedetomidine infusions may be used along with a long-acting nondepolarizing neuromuscular blocker if needed. Prior studies demonstrated hemodynamic stability when using high-dose fentanyl, either in bolus dosing or as a continuous infusion. Fentanyl in combination with midazolam leads to a decrease in MAP and CO but an increase in heart rate and SVR. Dexmedetomidine is an α_2-agonist with sedative properties; when used in combination with fentanyl, it minimally decreases heart rate and MAP, while potentiating the analgesic properties of fentanyl.

In older patients with long-standing severe disease, biventricular heart dysfunction and arrhythmias may be present. Left heart failure resulting from the presence of residual MAPCAs may lead to aortic insufficiency from progressive root dilation. Supraventricular arrhythmias (atrial fibrillation, atrial flutter, or atrial tachycardia) related to progressive pathology of the right heart and previous surgical insult to myocardial conduction tissue may be present. Patients can be at increased risk for ventricular arrhythmias due to prior ventriculostomies, abnormalities of the native myocardium, or progressive ventricular dilation and dysfunction.

Clinical Pearl

Anesthetic induction and maintenance management goals for patients with TOF/PA/MAPCAs center on the goals of balancing Q_p:Q_s in palliated patients and decreasing the hemodynamic burden experienced by the RV.

What is the appropriate postoperative disposition for this patient undergoing elective surgery?

Patients with complex congenital heart disease presenting for noncardiac surgery are at high risk for complications in the perioperative period. Postoperative observation in an intensive care unit may be warranted to permit rapid assessment and treatment of potentially fatal complications inherent to this unique subset of patients after complex surgeries. Depending on the complexity of surgical procedure and degree of preoperative morbidity, perioperative complications can include arrhythmias, cardiac ischemia or failure, and acute respiratory compromise. Assuming a stable intraoperative course this patient should be able to recover in the post-anesthesia care unit followed by overnight observation with monitoring of pulse oximetry. As with all patients, adequate pain relief, control of nausea and vomiting, maintenance of normothermia, and adequate volume status are essential for a stable postoperative recovery.

Suggested Reading

Ho S. Y., Catani G., and Seo J. W. Arterial supply to the lungs in tetralogy of Fallot with pulmonary atresia or critical pulmonary stenosis. *Cardiol Young* 1992; **2**: 65–72.

Huntington G. S. The morphology of the pulmonary artery in the mammalia. *Anat Rec* 1919; **17**: 164–201.

Jefferson K., Rees S., and Somerville J. Systemic arterial supply to the lungs in pulmonary atresia and its relation to pulmonary artery development. *Br Heart J* 1972; **34**: 418.

Mainwaring R. D., Reddy V. M., Peng L., et al. Hemodynamic assessment after complete repair of pulmonary atresia with major aortopulmonary collaterals. *Ann Thorac Surg* 2013; **95**: 1397–402.

Quinonez Z. A., Downey L., Abbasi R. K., et al. Anesthetic management during surgery for tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries. *World J Pediatr Congenit Heart Surg* 2018; **9**: 236–41.

Rossi R. N., Hislop A., Anderson B. H., et al. Systemic-to-pulmonary blood supply in tetralogy of Fallot with pulmonary atresia. *Cardiol Young* 2002; **12**: 373–88.

Webb G., Mulder B. J., Aboulhosn J., et al. The care of adults with congenital heart disease across the globe: current assessment and future perspective: a position statement from the International Society for Adult Congenital Heart Disease (ISACHD). *Int J Cardiol* 2015; **195**: 326–33.

https://doi.org/10.1017/9781108657341.011 Published online by Cambridge University Press