Chapter

24

Total Anomalous Pulmonary Venous Return and Heterotaxy Syndrome

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Case Scenario

A 4-month-old female presents with feeding intolerance leading to abdominal distension. She was prenatally diagnosed with heterotaxy syndrome, atrioventricular septal defect, mild left ventricular hypoplasia, severe pulmonary stenosis, and total anomalous pulmonary venous return. Within 48 hours of birth, she demonstrated severe pulmonary venous obstruction and underwent emergent sutureless repair of obstructed pulmonary veins, as well as creation of a modified Blalock–Taussig shunt. At 2 months of age she required cardiac catheterization and balloon dilation of the pulmonary veins due to restenosis. Her current medications include acetylsalicylic acid. Postnatal abdominal imaging confirmed the presence of malrotation; thus a gastrostomy tube was placed and expectant management of malrotation was elected. Her current presenting symptoms are concerning for midgut volvulus necessitating an emergent intraabdominal exploration and a Ladd procedure.

Key Objectives

- Understand the clinical relevance and classification of total anomalous pulmonary venous return.
- Understand the salient features of heterotaxy syndrome.
- Understand that patients with total anomalous pulmonary venous return and heterotaxy can have either single-ventricle or two-ventricle physiology.
- Understand the pathophysiology of pulmonary venous obstruction in the setting of total anomalous pulmonary venous return.
- Describe the preoperative assessment and intraoperative management of a patient with recurrent pulmonary vein stenosis.

Pathophysiology

What is total anomalous pulmonary venous return?

Total anomalous pulmonary venous return (TAPVR) is a rare disorder that occurs in 2% of patients presenting with congenital heart anomalies. It is characterized by the lack of direct connection of the pulmonary veins to the left atrium (LA); instead, the pulmonary veins connect either directly to the right atrium (RA) or indirectly via a vein connected to the RA. This results in oxygenated blood ultimately draining into the RA and mixing with deoxygenated blood, necessitating a right-to-left (R-to-L) atrial shunt to survive.

Total anomalous pulmonary venous return is an isolated defect in approximately two-thirds of cases but can be associated with complex cardiac defects and frequently with heterotaxy syndrome in the remainder of patients. Patients with coexisting heterotaxy syndrome often have significant additional cardiac anomalies and may require single-ventricle palliation, as discussed in the text that follows. Despite improvements in prenatal diagnostics, isolated TAPVR continues to have one of the lowest rates for prenatal diagnosis compared to other CHD lesions (2%–10%). Conversely, TAPVR associated with complex heart disease has a high rate of prenatal diagnosis, up to 100% in those with heterotaxy and TAPVR.

The age at presentation depends on the presence or absence of associated cardiac anomalies. Most patients with isolated TAPVR present as neonates or infants with pulmonary overcirculation and evidence of heart failure due to the large left to right (L-to-R) shunt. However, when severe pulmonary venous obstruction (PVO) exists with limited atrial communication, patients present with hypoxemia, acidosis, and impending circulatory collapse shortly after birth. Total anomalous pulmonary venous return with severe PVO is one of the few congenital cardiac defects requiring emergent surgical intervention in the neonate with congenital heart disease. Because newborns

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with TAPVR associated with heterotaxy are routinely diagnosed prenatally, they should be born at or immediately transported to a hospital with a pediatric congenital heart program.

Clinical Pearl

In TAPVR the pulmonary veins connect either directly to the RA or indirectly via a vein connected to the RA. This results in oxygenated blood ultimately draining into the RA and mixing with deoxygenated blood, necessitating a R-to-L shunt at the atrial level to survive. Total anomalous pulmonary venous return with severe PVO is one of the few congenital cardiac defects requiring emergent surgical intervention in the neonate.

How is TAPVR classified?

There are four major anatomic subtypes of TAPVR and they can be associated with PVO of varying severity. (See Figure 24.1.) Classifications include supracardiac, infracardiac, cardiac, and mixed pulmonary venous return.

The ***supracardiac*** variant constitutes about 50% of cases of isolated TAPVR and 30% of those associated with heterotaxy [1]. All pulmonary veins enter the common confluence (a single vessel draining multiple pulmonary veins), which then empties into either the innominate vein or directly into the superior vena cava (SVC). Pulmonary venous obstruction in these patients usually results from compression of the ascending vein between the bronchi and the pulmonary artery or aorta, or from stenosis at the orifice of the vertical vein.

The ***cardiac*** type describes the common confluence draining into the coronary sinus or directly into the RA. Pulmonary venous obstruction is rare in these patients and usually occurs at the point of connection of the confluence with the coronary sinus.

The ***infracardiac*** variant accounts for about 25%–30% of patients, regardless of the presence or absence of heterotaxy. The confluence drains below the diaphragm to enter either the portal vein or the inferior vena cava (IVC) directly; PVO in this variant is very common. There may be stenosis at the point of intersection of the descending vein with the systemic venous system or some degree of flow limitation may exist in the portal venous system.

Mixed TAPVR describes pulmonary venous connections to more than one anomalous location, as discussed earlier.

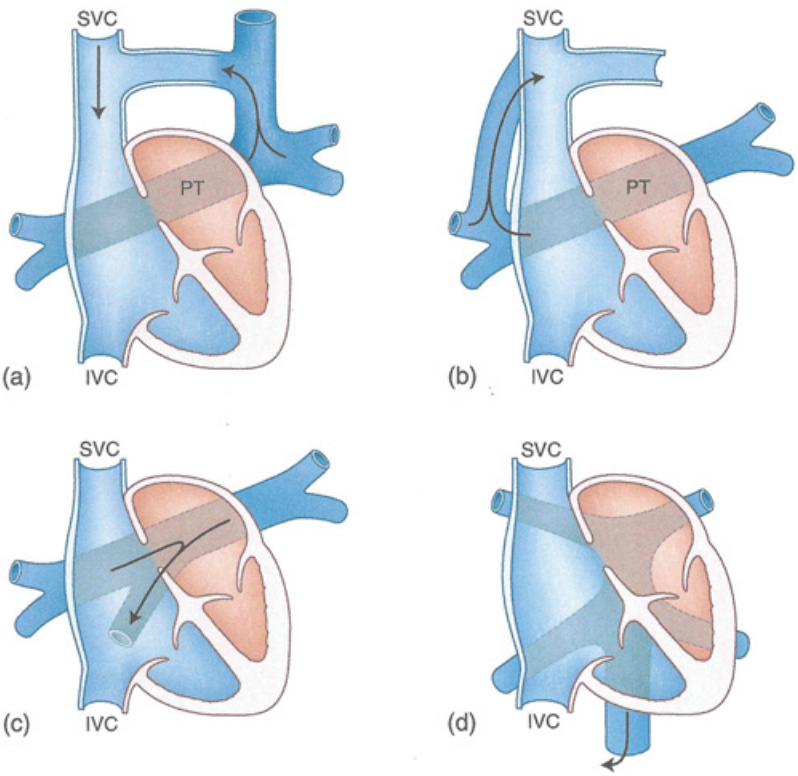

Figure 24.1 Total anomalous pulmonary venous connection (TAPVC) classified based on the site of pulmonary venous drainage (arrows). IVC, inferior vena cava; PT, pulmonary trunk; SVC, superior vena cava. (a) In type I/supracardiac connection, the four pulmonary veins drain via a common vein into the right SVC, left SVC, or their tributaries. (b) In type II/cardiac connection, the pulmonary veins connect directly to the right heart. (c) In type III/infracardiac connection, the common pulmonary vein travels down anterior to the esophagus through the diaphragm to connect to the portal venous system. (d) In type IV/mixed connections, the right and left pulmonary veins drain to different sites, such as left pulmonary veins into the left vertical vein to the left innominate, right pulmonary veins directly into the right atrium or coronary sinus. From G. Ottaviani and L. M. Buja. Congenital heart disease: pathology, natural history, and interventions. In Buja L. M. and Butany J., eds. *Cardiovascular Pathology*, 4th ed. Elsevier, 2015; 611–47. With permission.

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Clinical Pearl

Pulmonary venous obstruction is most commonly observed in the infracardiac variant of TAPVR. Total anomalous pulmonary venous return with severe PVO is one of the few congenital cardiac defects requiring emergent surgical intervention.

Are there other defects associated with TAPVR?

Other than an atrial communication, two-thirds of patients with TAPVR have no other associated cardiac defects. Of the remaining one-third of patients, most have associated heterotaxy syndrome, and the majority of these will have single-ventricle (SV) physiology [1].

Clinical Pearl

Understanding the intracardiac anatomy is paramount. Approximately one-third of patients with TAPVR have associated major cardiac defects, most of them resulting in single-ventricle physiology.

How is TAPVR surgically corrected?

The goal of surgical intervention is to establish the natural communication that should occur between the pulmonary veins and the LA. An anastomosis is created between either the pulmonary venous confluence or the posterior pericardium and the posterior wall of the LA. The procedure is performed on cardiopulmonary bypass, often with deep hypothermic circulatory arrest. A sutureless surgical technique was developed in an attempt to decrease the incidence of the recurrent PVO at the anastomotic sites. It involves creation of a pericardial well encasing the pulmonary venous confluence; the pericardial walls of this well are then anastomosed to the back of the LA [2].

Surgical considerations in children with heterotaxy syndrome are largely the same. Due to the presence of the atrioventricular septal defect (AVSD), the child in this scenario has a common atrium that therefore already allows mixing of oxygenated and deoxygenated venous return. Because of this, the L-to-R shunt from the TAPVR is irrelevant in the short term. However, due to the likelihood of partial or increasing obstruction to pulmonary venous blood flow in patients with supra- or infracardiac type TAPVR, patients with these anatomic variants are repaired soon after birth, while patients with intracardiac TAPVR may require surgical intervention only if a subsequent complete two-ventricle cardiac repair is pursued for their other cardiac anomalies.

What are possible postoperative sequelae of TAPVR repair?

Recurrent pulmonary venous stenosis occurs in up to 25% of patients with TAPVR, and the rate is thought to be significantly higher in patients with associated heterotaxy. Restenosis can occur at the anastomotic site or as a result of intimal hyperplasia involving the individual pulmonary veins. While the sutureless technique has decreased the rate of anastomotic site stenosis, intimal hyperplasia is a progressive disorder with limited treatment options and a poor prognosis. High long-term mortality is associated with PVO requiring reintervention, especially in patients with associated heterotaxy.

Sinus node dysfunction and other dysrhythmias occur frequently in this patient population. Ongoing follow-up with arrhythmia surveillance is recommended.

Clinical Pearl

Recurrent pulmonary venous stenosis occurs in up to 25% of patients with TAPVR, and the rate is likely to be higher in those patients with heterotaxy syndrome.

What is heterotaxy syndrome and how does it affect perioperative outcomes?

Heterotaxy, also known as ***atrial isomerism***, is an embryologic disruption of right and left laterality of the thoracic and abdominal organs. It results in loss of the normal spatial relationship between the intraabdominal and intrathoracic organs. Constellations of cardiac manifestations are frequently seen in patients with heterotaxy syndrome, and they represent approximately 3% of patients with CHD. Cardiac manifestations are typically severe, leading to considerable morbidity and mortality. Children with heterotaxy and TAPVR have even higher rates of early and late morbidity and mortality. Patients with SV anatomy and TAPVR tend to have the least favorable outcomes because long-term SV outcomes are reliant on a compliant pulmonary vasculature and normal pulmonary vascular resistance (PVR). The presence of PVO elevates both morbidity and mortality rates.

Clinical Pearl

*Heterotaxy, also known as **atrial isomerism**, is an embryologic disruption of right and left laterality of the thoracic and abdominal organs. Constellations of cardiac manifestations are frequently seen in patients with heterotaxy syndrome, representing approximately 3% of patients with CHD.*

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Which organs are involved in heterotaxy syndrome?

In addition to congenital heart defects of varying types and severity, various intraabdominal organs may be involved including the stomach, intestines, liver, and spleen. Intestines may be malrotated, meaning that they do not loop correctly in the abdomen during fetal development. This can predispose the patient to intestinal volvulus. All babies with heterotaxy should be evaluated to rule out intestinal malrotation. Some children with heterotaxy syndrome can have biliary atresia. Patients may also have asplenia, where the spleen may be missing entirely, or polysplenia, where the spleen is divided into several smaller spleens with variable levels of function. Irregularities of the skeleton, central nervous system, and the urinary tract may also be present, albeit less commonly. (See Figure 24.2.)

How is heterotaxy classified?

Heterotaxy is classified into two types, right and left, based on atrial appendage morphology. Classification is important because right and left isomerisms are associated with markedly different anatomic findings of the affected organs, and these anatomic differences can be clinically significant.

Right atrial isomerism, sometimes referred to as ***asplenia syndrome***, is associated with bilateral morphologically right atrial appendages. Other associated cardiac anomalies may include a constellation of some or all of the following: AVSD, TAPVR, transposition of the great arteries (discordant ventriculoarterial connections), side-by-side rather than spiraling of great arteries, bilateral SVCs, supraventricular tachycardia, bilateral trilobed lungs, central nervous system anomalies, and asplenia.

Left atrial isomerism is associated with bilateral morphologically left atrial appendages, as well as some or all of the following: AVSD, partial anomalous pulmonary venous return, interruption of the IVC, absence of the coronary sinus, mirror-image spiraling of great arteries, atrioventricular block (including complete heart block, necessitating insertion of a permanent pacemaker), bilaterally bilobed lungs, and asplenia or polysplenia.

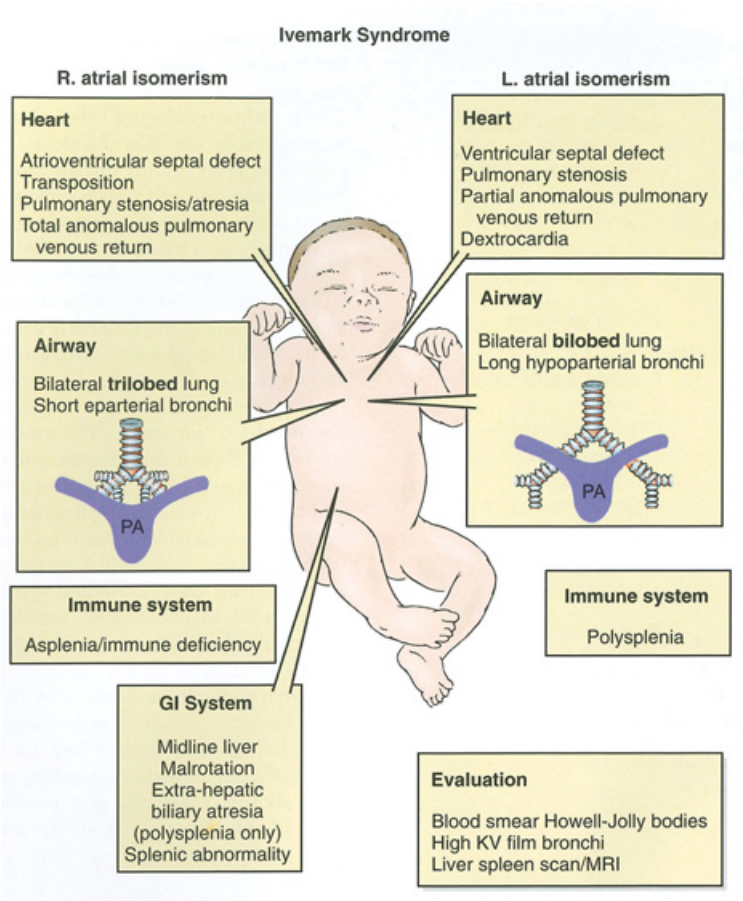

Figure 24.2 Associated defects of heterotaxy/Ivemark syndrome. From B. J. Landis and M. T. Lisi. Syndromes, genetics, and heritable heart disease. In Ungerleider R. M., Meliones J. N., McMillan K. N., et al., eds. *Critical Heart Disease in Infants and Children*, 3rd ed. Elsevier, 2019; 892–904. With permission.

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Are there special infection-related precautions in heterotaxy patients?

Patients with the diagnosis of heterotaxy and uncertain splenic function should be started on antibiotic prophylaxis as soon as possible. Amoxicillin should be dosed at 20 mg/kg/day and can be divided into one or two doses per day. At 2 years of age, those with solitary spleen or polysplenia should undergo evaluation of splenic function. Antibiotic prophylaxis can be discontinued if the function is normal. For those in whom antibiotic prophylaxis is continued beyond the age of 2, it can be safely discontinued at the age of 5 years.

Clinical Pearl

Patients with heterotaxy and uncertain splenic functional status should receive ongoing antibiotic prophylaxis until at least the age of 2 years.

Anesthetic Implications

What is malrotation and how is it diagnosed and managed?

Malrotation is a congenital abnormal rotation of the midgut, frequently associated with heterotaxy syndrome. Malrotation increases the risk of midgut volvulus due to twisting of the abnormally fixed small bowel around a narrow-based mesentery and superior mesenteric artery. Presentation can vary from minor gastrointestinal distress and/or feeding difficulties to abdominal distention and bilious vomiting. Infants with CHD, particularly heterotaxy syndrome, have an incidence of malrotation as high as 40% to 90%. The diagnostic test of choice is an upper gastrointestinal bowel study with small bowel follow-through. Patients diagnosed with malrotation not associated with heterotaxy syndrome should be evaluated for intestinal atresia, trisomy 21, imperforate anus, Meckel's diverticulum, and cardiac anomalies.

The recommended surgical procedure is a Ladd procedure, performed via either an open or laparoscopic approach. The open procedure involves a right upper quadrant transverse incision or midline laparotomy. Intestinal contents are eviscerated and detorsed counterclockwise if volvulus is present. Grossly necrotic bowel is resected, and consideration is given to second look laparotomy if bowel viability is questionable. Ladd cecal bands (fibrous peritoneal bands that can cause intestinal obstruction) are released, the small intestine mesentery is broadened, and an incidental appendectomy is performed. The small bowel is placed on the right and the colon on the left, and the abdomen is then closed. With a laparoscopic approach three or four port sites (umbilical, right abdomen, left abdomen, +/− epigastric for liver retractor) are placed. All steps of the Ladd procedure except evisceration are performed. In the setting of acute volvulus, use of the laparoscopic approach is somewhat controversial.

Traditionally, a diagnosis of malrotation was followed by the Ladd procedure irrespective of symptoms. Currently performance of a prophylactic Ladd procedure in an asymptomatic patient is controversial, especially in children with multiple high-risk comorbidities. Many centers proceed with prophylactic Ladd procedure in children with heterotaxy with malrotation; however, this decision and the optimal timing remain controversial in this patient population. Single-ventricle physiology, regardless of the stage of palliation, places the patient at risk for increased perioperative morbidity. On the other hand, significant risk is clearly incurred in an infant with SV physiology who develops acute volvulus. In summary, perioperative risks due to the patient's current cardiopulmonary physiology and status must be weighed against the risk of acute or chronic abdominal pathology.

When a Ladd procedure is to be electively performed in a patient with SV physiology, an argument may be made for waiting until after Stage II palliation (superior cavopulmonary anastomosis, or bidirectional Glenn) has taken place, as surgical and anesthetic risk are lower at this stage than for patients with shunt-dependent physiology. However, this strategy does not address the risk of acute volvulus occurring in the first several months of life prior to the Stage II palliation procedure. (See Chapter 27.)

What anesthetic risks should be discussed with the perioperative care team and the child's family?

The care team and family should understand that there is considerable risk for a child with shunt-dependent or SV physiology undergoing a lengthy abdominal surgery that can be associated with significant fluid shifts. A child with SV physiology with a systemic artery-to-pulmonary artery shunt (such as a modified Blalock–Taussig [mBT] shunt) providing pulmonary blood flow (PBF) is at risk for imbalances in both pulmonary and systemic perfusion. As the SV works at near maximal capacity to provide both pulmonary and systemic output in the setting of the inherent inefficiencies of a mixing lesion, the infant is at risk for tenuous perfusion of the coronary arteries and thus myocardial ischemia, which in turn can result in decreased heart

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function acutely and in the long term. Surgical interventions also induce derangements in coagulation, making patients more prone to clotting. A systemic-to-pulmonary shunt is prone to thrombosis, and that risk is exacerbated by the surgical procedure and the inflammation caused by the intestinal malformations. The clinical risk associated with the Ladd procedure and systemic-to-pulmonary shunt occlusion in a patient with SV physiology and heterotaxy has been reported to be as high as 20% [3]. However, the anesthetic risk must be weighed against the risk presented by the abdominal pathology. In the setting of impending volvulus and resultant bowel ischemia, surgery must proceed, whereas proposed elective surgery necessitates a careful discussion of the optimal surgical time to mitigate perioperative risks. The patient in this stem is presenting with possible volvulus and must proceed to surgery.

How should this patient be evaluated preoperatively?

A focused history, including review of pertinent medical records, is necessary to appreciate the child's precise anatomy, physiology, disease course, interventions, and most recent test results. Records of all previous surgical interventions and details of prior anesthetics should be sought.

On physical examination, the current hemoglobin–oxygen saturation should be compared to recent historical numbers prior to the current illness. Decreases in hemoglobin–oxygen saturations from the expected baseline can be indicative of mBT shunt narrowing, respiratory insufficiency, and/or low cardiac output. The lungs should be auscultated, and work of breathing observed. Hypotension can signal the presence of significant abdominal pathology and sepsis.

Given the complexity of the cardiac anomalies associated with heterotaxy syndrome in this child, a recent echocardiogram should be available; if not, one should be obtained. Echocardiography should evaluate palliative shunts and ventricular function, valvular pathology, and evidence of intracardiac shunting. Echocardiography can also screen for the presence of PVO, which would be evidenced by turbulent flow at the anastomotic site as well as abnormal, high velocity, nonphasic flow on Doppler. In this scenario, it is also important to rule out progressive mBT shunt stenosis, worsening atrioventricular valve function, or reduced function of the predominant ventricle (in this case, the RV).

A cardiac catheterization is not usually required preoperatively, and certainly not prior to urgent or emergent surgery. However, if surgery is elective and there is evidence of PVO, then consideration must be given to cardiac catheterization with pulmonary vein angioplasty with or without stent placement. While neither of these interventions have been found to have a high long-term success rate, they may delay the need for cardiac reoperation and, if recently performed with success, may decrease the degree of PVO at the time of the anesthetic for the Ladd procedure.

Given the high incidence of conduction anomalies associated with heterotaxy syndrome, an electrocardiogram (ECG) should be done. If the patient has congenital heart block and an epicardial pacemaker system, the pacemaker must be interrogated preoperatively, and intraoperative considerations for children with pacemakers must be considered. (See Chapters 23 and 49.)

The need for laboratory studies is guided by the child's history and physical exam and the surgery itself. An infant with complex heart disease should have at a minimum a complete blood count, electrolyte profile, and type and cross for packed red blood cells prior to abdominal surgery. A coagulation panel and liver function tests should also be considered.

What monitoring is appropriate for this patient?

In addition to standard American Society of Anesthesiologists recommended monitors, one should consider 5-lead ECG to enhance surveillance for arrhythmias, as well as ischemia during potential periods of hypoxemia and fluid shifts. The ECG can be adjusted to show pacer spikes in patients with pacemakers. Invasive arterial blood pressure monitoring can be of value both in open and laparoscopic procedures, but the decision to place an arterial line should depend on patient status. Adequate peripheral venous access is mandatory regardless of the surgical approach, but the need for central venous access depends on the overall status of the patient and the preference of the care team. The presence of a peripherally inserted central catheter can be invaluable should administration of inotropic medications become necessary. The monitoring of cerebral and systemic near-infrared spectroscopy can be beneficial for following trends in systemic oxygen delivery during anesthesia and surgery.

What methods are appropriate for induction and maintenance of anesthesia in this child?

In the setting of acute abdominal pathology from the child's malrotation, an intravenous (IV) induction should be performed. Depending on the degree of gastrointestinal symptomatology consideration should be given to a rapid or modified rapid sequence intubation. The child's current ventricular function and baseline hemoglobin–oxygen saturations will aid in determining the induction strategy.

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The use of ketamine along with a narcotic (e.g., fentanyl) and an appropriate neuromuscular blocking agent would be appropriate.

This patient, palliated with an mBT shunt, currently has parallel circulations, and thus maintaining balance between the pulmonary and systemic circulations is essential. All anesthetic interventions should take into account their potential effects on both pulmonary and systemic vascular resistances in order to maintain this balance. Decreases in SVR will result in "steal" from the mBT shunt and reduce pulmonary blood flow, which may already be limited to some lung segments due to the presence of existing PVO. Inspired oxygen concentration (FiO_2) should be utilized as needed to maintain the infant's "normal" preoperative baseline saturation, and therefore the appropriate FiO_2 will likely vary during the course of the surgery with surgical manipulation, particularly if a laparoscopic approach is utilized.

Clinical Pearl

In a patient who is palliated with an mBT shunt, anesthetic interventions should take into account potential effects on both pulmonary and systemic vascular resistances in order to maintain this balance. Inspired oxygen concentration (FiO_2) should be utilized as needed to maintain the infant's "normal" preoperative baseline saturation.

How should this patient be ventilated?

Endotracheal intubation and the use of positive pressure ventilation (PPV) are necessary to perform the Ladd procedure. Maintenance of normal PVR is essential in order to maintain appropriate PBF.

- ***If PVR is significantly decreased*** via hyperoxygenation or hyperventilation, the child with PVO is at risk for pulmonary edema due to increased PBF in the setting of downstream obstruction. This is true for patients with either single- or two-ventricle physiology.
- ***If PVR is increased***, this will compound the inherent resistance secondary to the PVO.
  - In a patient with biventricular anatomy this will result in right heart strain.
  - In a patient with an mBT shunt, significantly elevated PVR will reduce shunt flow and thus PBF, leading to hypoxemia.
- ***Ventilatory goals*** include the use of moderate tidal volumes (6–8 mL/kg) and short inspiratory times, favoring low mean airway pressures and PVR.
  - Positive end expiratory pressure (PEEP) should be used in moderation; at optimal levels it helps to maintain functional residual capacity but at high levels can decrease PBF.
  - Inspired oxygen concentrations should be titrated to maintain the patient's "normal" baseline oxygen saturations.

What other intraoperative concerns exist in patients with repaired TAPVR and PVO?

- ***Fluid management***: In the setting of PVO, judicious IV fluid management is necessary. With obstruction in the pulmonary veins, an increase in intravascular hydrostatic pressure can lead to pulmonary edema. However, insensible fluid losses can be significant during the open Ladd procedure due to the sizable abdominal incision and evisceration of intestines and their exposure to ambient air. Replacement of those insensible losses must be performed diligently and may need to conservatively deviate from the recommended standard estimations and formulas. If a blood transfusion is required during the procedure, one should consider concurrent administration of a diuretic after consideration of the patient's overall volume status. Any change in pulmonary compliance as noted by trends in ventilator settings is important in assessing the development of pulmonary edema that could require additional PEEP to slow the alveolar exudative process.
- ***Ventricular function***: Attention to ventricular function is paramount. As mentioned earlier, the predominant ventricle is already working at high capacity to maintain both pulmonary and systemic outputs (parallel circulations) with each stroke volume. Perturbations that acutely increase afterload, directly reduce contractility or heart rate, or reduce coronary blood flow risk the development of inadequate cardiac output to meet the demands of critical organs, including the heart. Inotropic support should be instituted if the ventricle is struggling to meet demands, while potentially causative mechanisms such as volume status, adequate hematocrit, and optimal oxygenation and ventilation are addressed.
- ***Surgical approach***: Laparoscopic techniques often introduce additional challenges. Insufflation of CO_2 with resultant systemic absorption can increase ventilation requirements and negatively impact efforts to prevent hypercarbia and acidosis.

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Increased intraabdominal pressures will require increased ventilatory pressures, which can have negative effects on venous return and ventricular function. Clear communication with the surgical team is essential should insufflation prove detrimental to the patient's ventilatory status and hemodynamics.

- ***Electrolytes*:** Patients with PVO are frequently treated with diuretics preoperatively as part of their medical therapy. This chronic use of loop or thiazide diuretics can lead to hypokalemia and hypochloremic metabolic alkalosis. One must be aware of the electrolyte shifts that can occur from intraoperative controlled ventilation in patients with acid–base derangements. For example, hyperventilation can further exacerbate hypokalemia and lead to an increased risk of dysrhythmias.
- ***Airway:*** Venous congestion in the airways can predispose patients to increased risk of endotracheal bleeding with suctioning. Suctioning should be performed gently and judiciously. If bleeding occurs, saline lavage should be performed, and the PEEP should be increased. One should then watchfully wait, as over time these maneuvers will frequently decrease and eventually stop the bleeding.

Clinical Pearl

In the setting of PVO, intraoperative goals include maintaining normal PVR to facilitate PBF and reduce right ventricular strain, and judicious fluid management to minimize the risk of pulmonary edema.

What is the most appropriate venue for postoperative management of this patient?

Patients with recurrent PVO are at high risk for postoperative complications, including mortality. This is especially true for patients with variants of SV anatomy in the setting of TAPVR and heterotaxy syndrome. Furthermore, major abdominal surgery in any infant requires inpatient management in a monitored bed due to the need to carefully manage nutrition, fluid shifts, and pain. A patient such as the one in this scenario requires care by a provider who understands their anatomy, physiology, and perioperative risk factors; therefore admission to the critical care setting postoperatively is most appropriate for this patient. Institutional preferences may vary between postoperative care in a cardiac intensive care unit setting versus a pediatric intensive care unit. The likely need for postoperative ventilation also warrants a planned intensive care bed.

When should the patient be extubated?

Timing of extubation depends on patient's clinical status as it relates to the underlying pathology and the intraoperative course. In the setting of PVO and SV physiology with an mBT shunt, excessive intraoperative fluid administration and/or the need for an intraoperative transfusion can lead to the development of pulmonary edema. The risk of pulmonary edema continues into the postoperative period, as third spacing of intravascular fluids continues, necessitating further fluid resuscitation. Patients successfully extubated in the operating room may be at risk for postoperative respiratory failure. Overall, it may be wise to keep any child intubated who has undergone a long surgery with major fluid shifts when concern exists for potential ongoing cardiopulmonary compromise. On the other hand, an older infant with two-ventricle physiology, or a patient with SV physiology who has been palliated with a superior cavopulmonary anastomosis (Glenn shunt) may be extubated if surgery was minimally invasive, fast and well tolerated from a cardiopulmonary standpoint.

How can postoperative pain management be optimized in this patient?

Regional or neuraxial techniques and nonopioid analgesics are encouraged. Both can facilitate early extubation and decrease the risk of hypoventilation and hypercarbia related to opioid administration.

Ileus is a frequent complication of the Ladd procedure regardless of surgical approach, and the use of opioid sparing techniques helps to limit opioid induced constipation and gut motility issues.

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