Chapter

42

Pulmonary Hypertension and Prematurity

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Case Scenario

A former 30-week preterm male, birth weight 1500 grams, presents at 40 weeks postconceptual age for a tracheostomy and laparoscopic Nissen fundoplication with gastrostomy tube placement. His history includes bronchopulmonary dysplasia, pulmonary hypertension, an unrepaired atrial septal defect, and failure to thrive. After delivery, he initially required continuous positive airway pressure and was ultimately intubated due to persistent oxygen desaturations below 85%.

A transthoracic echocardiogram revealed a small atrial septal defect, no ventricular septal defect, a dilated right ventricle, and normal biventricular function. Repeat echocardiogram at 7 days of life showed the development of interventricular septal flattening. Inhaled nitric oxide was initiated at 20 parts per million with an increase in SpO_2 to >92%.

Although now successfully weaned from nitric oxide and transitioned to sildenafil, he has failed multiple extubation attempts and is again experiencing frequent severe desaturation events. Current ventilator settings are peak inspiratory pressure 30 mm Hg, positive end-expiratory pressure 8 mm Hg, and FiO_2 0.4 to maintain acceptable tidal volumes and oxygen saturations. He has had poor weight gain with intolerance to increasing nasogastric feeds. His mother has also requested that he be circumcised.

Current vital signs are heart rate 164 beats/minute, respiratory rate 60 breaths/minute, noninvasive blood pressure 70/36 (mean 48), temperature 36.4°C, and SpO_2 93%.

Key Objectives

- Define pulmonary hypertension of prematurity and describe its pathophysiology.
- Describe the significance of the atrial septal defect in this patient.
- Discuss the combined effects of premature lung disease and pulmonary hypertension.
- Discuss anesthetic risk in this patient.
- Describe a perioperative plan including appropriate monitoring.
- Discuss management of a pulmonary hypertensive crisis.

Pathophysiology

How is pulmonary hypertension defined in preterm infants?

According to the 2018 Paediatric Task Force of the 6th World Symposium on Pulmonary Hypertension, pulmonary hypertension (PH) in preterm infants is defined by the same guidelines used for adults and children: mean pulmonary arterial pressure (mPAP) ≥20 mm Hg, pulmonary capillary wedge pressure (PCWP) <15 mm Hg, and pulmonary vascular resistance indexed to body surface area >3 Wood units/m^2 (iWu). Normal mPAP is 15 mm Hg.

What is bronchopulmonary dysplasia?

Bronchopulmonary dysplasia (BPD), the major cause of chronic lung disease in preterm infants, is defined as an ongoing supplemental oxygen requirement at 36 weeks postconceptual age in infants born at or prior to 32 weeks' gestation. It is most common in infants born prior to 28 weeks' gestation and <1000 grams at birth. Bronchopulmonary dysplasia results from a disruption of lung development at an early preterm age due to decreased surfactant supply and/or function, abnormal alveolar growth, and/or abnormal pulmonary vasculature. It is exacerbated by the need for mechanical respiratory support, supplemental oxygen administration, and fluid shifts. Although giving betamethasone to the mother can help reduce the incidence of acute respiratory issues, it has no effect on the development of BPD.

How is PH associated with BPD?

The disruption of growth and function of the pulmonary vasculature observed with BPD contributes to the failure of pulmonary artery pressures to decrease normally in the

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first few months of life. Abnormal pulmonary vasculature leads to a chronic elevation in mPAP and pulmonary vascular resistance (PVR) and over time the arteries become muscularized, hypertonic, and reactive. These alterations impede blood flow through the lungs and worsen hypoxemia, as fewer adequate blood vessels are available to participate in gas exchange. Additional mechanisms associated with persistent PH in the infant with BPD include oxygen toxicity, barotrauma, alveolar hypoxia, cardiac dysfunction, and pulmonary vein stenosis. As many as 20%–40% of patients with BPD have significant PH. The combination is accompanied by significant morbidity and mortality.

Clinical Pearl

The disruption of growth and function of the pulmonary vasculature observed with BPD contributes to the failure of pulmonary artery pressures to decrease normally in the first few months of life. As many as 20%–40% of patients with BPD will have PH. The combination is accompanied by significant morbidity and mortality.

What risk factors are associated with the presence of PH in an infant with BPD?

Risk factors associated with the presence of PH in an infant with BPD include extreme prematurity, birth weight <1500 grams, and respiratory factors including prolonged mechanical ventilation and prolonged oxygen therapy. Cardiovascular abnormalities, oligohydramnios, maternal preeclampsia, and intrauterine growth retardation are also risk factors, as are maternal infection and genetic abnormalities.

What screening is recommended in preterm infants with BPD and PH?

Early identification of PH in infants with BPD serves to improve patient management. Screening for PH with transthoracic echocardiography (TTE) is recommended in symptomatic patients or those with identified risk factors for PH. Patients with BPD can often show signs of PH on TTE as early as 7 days of life. If not previously performed, TTE is recommended at the time BPD is formally diagnosed. It is especially important to screen high-risk infants if they require general anesthesia, as PH is associated with an increased risk of adverse events under anesthesia.

Transthoracic echocardiography is recommended at least every 3 months for infants with BPD. However, if the infant maintains a persistent supplemental oxygen requirement or continues to display signs of respiratory distress such as increased work of breathing, more frequent examinations are indicated.

Laboratory values such as brain natriuretic peptide (BNP) or N-terminal pro-BNP (NT-proBNP) are not sufficient to formally diagnose or exclude PH; however, serial measurements may be used to observe trends which are helpful in assessing a patient's clinical status or response to treatment.

Clinical Pearl

Transthoracic echocardiography is recommended at least every 3 months for infants with BPD.

What information does TTE provide?

Transthoracic echocardiography has the advantage of being noninvasive and can evaluate alterations in the cardiac and pulmonary anatomy and pressures to diagnose or monitor PH. Depending on the quality of the TTE data, the following findings may be observed and monitored:

- Estimated right ventricular systolic pressure >40 mm Hg or mPAP >20 mm Hg
- Right ventricular: systemic systolic blood pressure ratio >0.5
- Intracardiac shunting with bidirectional or right-to-left flow
- Assessment of septal wall flattening

Assuming no RV outflow tract obstruction exists, the systolic PAP and right ventricular (RV) pressures can be estimated using tricuspid regurgitant jet velocity. If imaging results are nonconclusive, it can be difficult to determine if the etiology of persistent oxygen requirement is due to BPD, PH, or a combination of factors. If suspicion exists for undefined disease, providers may choose to utilize cardiac catheterization to assess hemodynamic values and the need for vasodilator therapy such as inhaled nitric oxide (iNO) or selective phosphodiesterase type 5 inhibitors such as sildenafil.

What cardiac catheterization findings signify the presence of PH?

Cardiac catheterization is the gold standard for diagnosis of PH but is less frequently performed in this patient population due to the patient's fragility and the need for general endotracheal anesthesia. For this reason, many providers choose to empirically treat preterm infants with

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iNO and/or oral vasodilators such as sildenafil and to assess treatments with serial TTE.

Cardiac catheterization findings indicating the presence of PH include

- PA systolic: systemic systolic blood pressure ratio ≥ 0.5
- PVR ≥ 3 iWu
- PVR: systemic vascular resistance (SVR) ratio ≥ 0.5
- Normal PCWP or normal LV end-diastolic pressure without significant evidence of pulmonary vein stenosis

Clinical Pearl

While cardiac catheterization procedures are the gold standard for diagnosis of PH, the majority of preterm infants are screened and diagnosed via noninvasive TTE. Clinical response to medical therapy is also monitored with serial TTE.

What are medical management goals in infants with BPD and PH?

The goals for management of PH in infants with BPD are focused on optimizing respiratory support to improve gas exchange, promoting blood flow to the pulmonary vasculature, minimizing further lung injury, and promoting growth of the lung parenchyma. This is accomplished with pulmonary vasodilators, supplemental oxygen, and conservative mechanical ventilation to avoid barotrauma.

Clinical Pearl

Medical management of PH in preterm infants utilizes pulmonary vasodilators such as iNO or sildenafil, supplemental oxygen, and optimizing respiratory support to avoid further lung injury and to promote gas exchange.

How does PH affect normal cardiac function? How do RV changes impact the LV?

Pulmonary hypertension causes increased RV afterload due to elevations in mPAP and PVR. Over time, increased afterload increases right ventricular end-diastolic volume (RVEDV) and right ventricular end-diastolic pressure (RVEDP) leading to RV hypertrophy. If untreated, RV dilation ensues. The morphologic RV changes secondary to increased RVEDV and RVEDP alter normal blood flow through the right coronary artery. In a normal heart the right coronary artery fills in systole and diastole; however, these pathologic changes impede flow, allowing filling only during diastole.

Elevated RV pressures can also displace the interventricular septum toward the LV, changing LV morphology. This is often described on echocardiogram as "septal flattening" and results in impaired LV filling, decreased cardiac output and hypotension. These decreases in systemic pressures will further reduce coronary perfusion and risk complete cardiovascular collapse.

How does iNO affect the pulmonary vasculature?

Endogenous nitric oxide is produced by endothelial cells and released into the vascular system. Nitric oxide aids in the formation of cyclic guanosine monophosphate (cGMP), which allows smooth muscle relaxation in the pulmonary system. This relaxation allows PVR to decrease, potentially improving cardiac output and decreasing RV strain. Therefore, in patients with PH the addition of iNO may be therapeutic.

What is sildenafil and how does it help patients with PH? What side effects can occur with sildenafil?

Sildenafil is a selective cGMP phosphodiesterase inhibitor, thereby blocking the breakdown of cGMP. Cyclic guanosine monophosphate allows for the relaxation of smooth muscle, a primary component of the pulmonary vasculature. Relaxation of the pulmonary vasculature aids in reducing PH. Sildenafil should be continued without interruption throughout the perioperative period. Patients receiving sildenafil or iNO may develop hypotension from smooth muscle fiber relaxation, resulting in decreased brain, gastrointestinal, and renal perfusion.

What other medications are these patients frequently receiving?

Infants with BPD and/or PH also frequently suffer from gastroesophageal reflux and feeding intolerance. Increased airway reactivity may also result from gastroesophageal reflux and require medical treatment. Thus, medications such as ranitidine, omeprazole, furosemide, and albuterol are frequently administered as well.

How is the presence of a shunt significant in a patient with PH?

Intracardiac or systemic-to-pulmonary communications permitting left-to-right (L-to-R), right-to-left (R-to-L) or bidirectional intracardiac shunting are common in preterm infants and can include an atrial septal defect (ASD), patent foramen ovale, and/or persistent patent ductus arteriosus (PDA). A persistent L-to-R shunt leads

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to an increase in pulmonary blood flow (PBF), increasing the volume and pressure load to the vasculature. This results in increased shear stress, smooth muscle hypertrophy, and endothelial dysfunction which increase PVR and further worsen PH. As the RVEDP rises to equal RA pressures, the L-to-R shunt becomes bidirectional; it can eventually reverse to a R-to-L shunt through the ASD, increasing cyanosis. Most commonly the presence of a shunt means that with the abrupt rise of PVR blood can shunt bidirectionally, increasing cyanosis.

Clinical Pearl

The presence of a communication such as a patent foramen ovale, ASD, or PDA means that with an abrupt increase in PVR patients may shunt bidirectionally or R-to-L, maintaining cardiac output but increasing cyanosis.

Anesthetic Implications

What are the preanesthetic considerations for infants with BPD and PH?

The preoperative evaluation of infants with BPD and PH should include a thorough review of their current clinical status, including physical examination, medications, and clinical studies.

The gestational age at birth as well as the current postgestational age are important factors for risk stratification. Preterm infants frequently experience multisystem organ disturbances beyond the cardiac and pulmonary systems. These include the inability to regulate body temperature, an immature central nervous system contributing to apneic spells, a predisposition to or a history of intraventricular hemorrhage (IVH), anemia, electrolyte imbalances, and retinopathy of prematurity. The preoperative examination should include ongoing measures to address these concerns in a systematic fashion.

Temperature: Heat is lost by radiation, conduction, convection, and evaporation. Patient temperature should be continually monitored and maintained above 36.5°C. Hypothermia can promote pulmonary vasoconstriction and worsen PH. Perioperative interventions can include the use of thin plastic wrap, utilizing a hat to reduce cranial heat loss, and forced air warming through warm air blankets and/or heat lamps.

Neurologic:

- *Central nervous system immaturity*: Predisposition to apneic spells can lead to bradycardia and desaturation events requiring intervention. In patients not already dependent on mechanical ventilation this may indicate a need for postoperative mechanical ventilation.
- *Intraventricular hemorrhage*: Patients with various grades of IVH may have a seizure history requiring antiepileptics to be continued perioperatively. If anticoagulant medications are to be given during the procedure, a discussion about potential risks and benefits should occur with the medical teams and disclosed to the family.

Cardiac: Evaluation for the presence or absence of additional congenital heart disease should include an electrocardiogram and echocardiogram in addition to clinical examination. Coexisting congenital heart disease may worsen or even be the primary cause of PH in this patient.

Retinopathy of prematurity (ROP): A large percentage of preterm infants are at risk to develop ROP. While ROP is a concern in preterm infants, the risk: benefit ratio of supplemental oxygen administration must be considered and inspired oxygen concentration titrated to maintain adequate oxygen saturations.

Respiratory: The extent of respiratory support required (either via nasal cannula, continuous positive airway pressure, or endotracheal intubation) parallels disease severity. Oxygen flow and concentration, peak inspiratory pressure (PIP), inspiratory time, respiratory rate, tidal volume, and positive end-expiratory pressure (PEEP) should be evaluated to determine the optimal intraoperative ventilation management and respiratory support strategies. Intraoperative use of a neonatal intensive care unit (NICU) ventilator should be considered when an infant requires high levels of PEEP or maintains very low tidal volumes with increased PIP, and/or additional concerns exist about adequate ventilation. The goal is to maintain and optimize adequate ventilation and oxygenation. Insufficient ventilation leads to hypercarbia and will worsen PH. Respiratory therapies such as albuterol, sildenafil, and iNO should be continued perioperatively.

Hematologic: Neonates are frequently anemic due to decreased production of red blood cells and frequent blood draws. Determination of limits for allowable blood loss and appropriate transfusion thresholds allows optimization of oxygen delivery in patients with cardiorespiratory illnesses. Target hemoglobin levels may vary between patients with varied comorbidities but maintenance of hemoglobin levels above 9–10 g/dL is recommended.

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Glucose: In preterm infants the liver is still developing; therefore, gluconeogenesis is not always sufficient. Thus, once fasting prior to surgery, infants should receive dextrose-containing maintenance fluids. Recent blood glucose levels and fluctuations in glucose levels should be accounted for and appropriately monitored. Hypoglycemia has significant neurologic affects that may adversely affect preterm infants. Alternatively, high serum glucose concentrations can lead to an osmotic diuresis and hypovolemia.

Vascular access: Vascular access should be adequate to appropriately manage PH and BPD perioperatively. The location and adequacy of peripheral intravenous (IV) access should be evaluated, along with the possible presence of a peripherally inserted central catheter (PICC). Patients with severe disease may benefit from central access in the event of cardiovascular compromise requiring intraoperative resuscitation.

Is anesthesia risk greater in patients with PH? What risks should be discussed with the family?

It is well documented that the presence of PH increases anesthetic risk and the risk of perioperative complications. The risk varies between patients and will correlate with individual disease severity. Patients with PH who are <2 years of age typically have the greatest anesthetic risk. Anesthetic risk increases as PA pressure becomes greater than half systemic pressure and is highest when PA pressure is suprasystemic. For patients with significant PH the potential role of extracorporeal membrane oxygenation (ECMO) rescue should be addressed in the preoperative assessment with the family and medical teams.

Clinical Pearl

Pulmonary hypertension is associated with an increased risk of adverse events under general anesthesia – especially for patients <2 years of age who have PA pressure greater than one-half systemic blood pressure.

Can all procedures be done safely at this time, and is there a suggested order?

The urgency and appropriate timing for each procedure should be considered for preterm infants, particularly in infants with PH. The patient's responsiveness to vasodilator therapies aids in presurgical planning, and continual reassessment of hemodynamic stability intraoperatively will help guide any necessary changes to the intraoperative plan. Clear communication between all surgical teams is essential to clarify parameters for successful completion of all procedures.

Laparoscopic Nissen procedures involve surgical conditions that may be disruptive to a fresh tracheostomy site. Therefore, if adequate ventilation can be maintained it may be best to proceed with the abdominal procedures prior to the tracheostomy. Alternately, significant issues with ventilation could increase the urgency of the tracheostomy and preclude performing the fundoplication during the same anesthetic.

Circumcision should be the last procedure performed providing the patient's cardiorespiratory status is stable. These expectations must be addressed with the parents and medical team in the preoperative period as ultimately patient safety should dictate the ability to proceed with all three procedures. The risk of increased anesthetic duration should be balanced against the risk of necessitating another general anesthetic in the future.

Are additional studies indicated prior to anesthetizing this patient?

As the last echocardiogram on this patient was performed at 1 week of age it is important to obtain an updated TTE to evaluate this infant's current cardiac status – specifically, the degree of PH and assessment of the therapeutic effect of the current medication regimen. If the patient's current cardiac condition has worsened, it would be reasonable to delay nonurgent procedures in order to optimize therapies for his PH. Additionally, other diagnostic or therapeutic interventions such as cardiac catheterization or PDA closure may be considered if PH has worsened in the interim.

What intraoperative monitoring and access should be considered?

Standard recommended American Society of Anesthesiologists monitoring should be utilized in addition to any additional monitoring indicated by the patient's cardiac disease and functional status. A 5-lead electrocardiogram is preferred to monitor for cardiac arrhythmias and ischemia, although in a small infant with multiple procedures leads will need to be carefully placed away from surgical areas. The need for additional vascular access and monitoring will depend on the size and scope of surgery, anticipated blood loss, fluid shifts, and degree of potential cardiac complications. Placement of an arterial line should be considered to allow for close monitoring of both hemodynamic and respiratory status if RV function is significantly compromised or risk of decompensation is anticipated.

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The necessity of central access is dictated by the severity of PH and BPD. Often a PICC is present in this patient population. If present, a chest radiograph should be reviewed to evaluate location of the catheter tip as lines are not always centrally located. Even with a PICC line, a peripheral IV line should be placed if possible, particularly if the PICC line is a single lumen line. Although useful for administration of medications PICC lines are generally not as helpful should administration of blood or fluid boluses be necessary.

Is an uncuffed endotracheal tube acceptable? Should it be exchanged for a cuffed tube?

The presence of an uncuffed endotracheal (ETT) may contribute to difficulty ventilating during a laparoscopic procedure if a significant leak exists. This should be assessed prior to beginning the procedure. Previous documentation should be carefully reviewed for any issues with mask ventilation and recent ease of intubation, including the view on direct laryngoscopy. Options include exchanging the uncuffed ETT for a cuffed ETT or proceeding with the scheduled tracheostomy and rescheduling the laparoscopic Nissen fundoplication after appropriate recovery. Even if endotracheal intubation was previously easily accomplished, exchange of an ETT in a neonate with PH who requires significant ventilatory support carries significant risks even in experienced hands. Rapid desaturation and decruitment with loss of PEEP can at best require time to recover to baseline, and at worst can precipitate a PH crisis. Endotracheal tube position can also migrate as a result of insufflation. Through the insufflation process, frequent adjustments to inspired oxygen concentrations and minute ventilation may be required to prevent increases in end-tidal carbon dioxide ($ETCO_2$) levels or acidosis.

Clinical Pearl

Even if intubation was previously easily accomplished, exchange of an ETT in a neonate with PH who requires significant ventilatory support carries significant risks even in experienced hands. Rapid desaturation and decruitment with loss of PEEP can at best require time to recover to baseline, and at worst can precipitate a PH crisis.

What considerations exist for ventilation of this patient intraoperatively?

Depending on the capabilities of the anesthesia ventilator, consideration may be given to utilizing the NICU ventilator during surgery. This is particularly relevant in patients requiring a high level of ventilatory support when multiple procedures are being performed and the anticipated operating room time may be prolonged. Regardless of whether the anesthesia ventilator or the NICU ventilator is utilized iNO should be available for use if needed during the procedure.

How should anesthetic induction and maintenance be managed?

Performing a safe anesthetic induction in a patient with PH involves minimizing changes in SVR and PVR while ensuring adequate ventilation. Patients with PH are at risk for rapid hemodynamic decompensation on induction of anesthesia due to these effects. Slow titration of IV medications is preferred in preterm infants with PH with the addition of inhaled anesthetics as tolerated within desired hemodynamic parameters. Rescue medications (atropine, epinephrine, and phenylephrine) should be readily available prior to induction of anesthesia in the event of cardiovascular collapse.

Maintenance of general anesthesia can be continued with a balanced technique utilizing narcotics and inhaled anesthetics, with the goal of maintaining adequate analgesia and anesthesia to blunt the surgical stress response and avoid abrupt increases in PVR. As the patient will remain mechanically ventilated postoperatively opioids may form a significant part of the anesthetic plan to blunt stress responses. It is helpful to know the degree of exposure the patient has previously had to sedative and opioid regimens, as some neonates may require higher opioid dosing than expected to achieve the desired level of anesthesia due to prolonged preoperative exposure.

Factors that can negatively impact PVR such as hypoxia, hypercarbia, hypothermia, and acidosis should be avoided. Abrupt increases in PVR can reverse the shunt across the ASD, creating a R-to-L shunt and resulting in a drop in the systemic arterial oxygen saturation. Optimizing PVR via increased inspired oxygen concentration and initiation of iNO will facilitate L-to-R shunt flow and encourage PBF, enhancing cardiac output and coronary perfusion.

Clinical Pearl

It is helpful to know the degree of exposure the patient has previously had to sedative and opioid regimens as some neonates may require higher dosing ranges than expected to achieve the desired level of anesthesia.

What are the potential issues with a laparoscopic surgical approach?

Laparoscopic Nissen and gastrostomy tube (g-tube) procedures involve the imposition of surgical conditions that may not be well tolerated in small infants, particularly those with PH and BPD. Abdominal insufflation during laparoscopy and g-tube placement often significantly compromises ventilation.

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This reduction in ventilation combined with insufflation of CO_2 leads to an increase in $ETCO_2$, thus necessitating a significant increase in ventilatory requirements. Prior to the start of surgery, laparoscopic surgical planning should be discussed. Utilization of the lowest acceptable intraabdominal insufflation pressure is recommended. A contingency plan should also be discussed should the patient fail to tolerate insufflation with consideration for expeditious conversion to an open surgical approach. (See Chapter 26 for discussion of laparoscopic surgery in an infant.)

Shortly after abdominal insufflation begins oxygen saturations drop to the mid-70s: what are the considerations?

Should gradual desaturation occur following insufflation, consideration should be given to manipulating ventilatory settings and decreasing insufflation pressures. With rapid desaturation the differential should also include secretions or a plug in the endotracheal tube, an endobronchial intubation, or the onset of a PH crisis. It is imperative to communicate openly with the surgical team for appropriate patient management and to stop surgical manipulation until the patient returns to his baseline oxygen saturations.

What is a pulmonary hypertensive crisis?

A pulmonary hypertensive crisis is an abrupt increase in PVR resulting in acute right heart failure and inadequate cardiac output.

A PH crisis is defined as two or more of the following changes in <10 minutes:

- Decrease in systolic blood pressure >20% from baseline
- Decrease in oxygen saturation to <90% in acyanotic patients and decline >10% in cyanotic patients in the absence of other causes
- Increase in central venous pressure >20% from baseline
- Change in baseline heart rate < or >20%

During a PH crisis hypotension develops due to impaired preload along with decreases in coronary blood flow and cardiac output. If the PH crisis is not treated expeditiously, bradycardia develops and can progress to cardiac arrest. Bradycardia can also be an ominous early sign accompanying or preceding oxygen desaturation. The goal of PH crisis management is to decrease PVR and increase SVR such that LV output and coronary perfusion are maintained.

Clinical Pearl

A pulmonary hypertensive crisis is an abrupt increase in PVR causing acute right heart failure and inadequate cardiac output; it may precipitate cardiovascular collapse.

How is a pulmonary hypertensive crisis treated?

Management of a PH crisis involves reducing RV afterload and increasing SVR expeditiously.

Although the following steps can be performed in the order listed, it is optimal to perform all actions in an expeditious fashion. (See also Chapter 40, Table 40.2.)

- ***Respiratory interventions***
  - Increase FiO_2 to 100%
  - Increase minute ventilation
  - Avoid excessive PEEP, high inspiratory pressures and long inspiratory times
  - Initiation of iNO
- ***Hemodynamic interventions***
  - Ensure adequate preload and inotropic support for LV
  - Maintenance of SVR to ensure coronary perfusion
  - Sedation or anesthetic strategies: consider neuromuscular blockade for intubated and mechanically ventilated patients
  - Correction of metabolic acidosis
- ***Early ECMO activation if the patient is a candidate***

Vasoactive drugs commonly used in the management of a PH crisis include epinephrine, vasopressin, and phenylephrine. Epinephrine helps maintain or enhance RV function, while vasopressin and phenylephrine help maintain diastolic pressures to allow adequate RV perfusion.

Clinical Pearl

Successful management of a PH crisis includes early recognition and treatment including oxygen, increased minute ventilation, iNO, and vasoactive medications (epinephrine, vasopressin, and phenylephrine), along with correction of any acidosis.

What are the postoperative considerations for this patient?

Anesthetic related risks for patients with PH continue postoperatively. It is imperative to maintain adequate pain control as well as appropriate ventilation and oxygenation to avoid acute elevations in mPAP and PVR following the procedure. Close monitoring is recommended so that adverse events may be averted, and in this case the patient would recover in the NICU.

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