Chapter 41

Pulmonary Hypertension and Congenital Heart Disease

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Case Scenario

A 7-year-old female with hearing loss is scheduled to undergo a tympanoplasty. Her past medical history is significant for repair of a complete atrioventricular septal defect at age 6 months with residual pulmonary hypertension. She additionally has Down syndrome, hypothyroidism, moderate obstructive sleep apnea, recurrent bronchitis, and gastroesophageal reflux. She is managed with continuous positive airway pressure at night, and her medications include tadalafil, ambrisentan, levothyroxine, and ranitidine. Ambrisentan was added to the tadalafil a year ago to slow the progression of her pulmonary hypertension, and a recent cardiac catheterization was performed to evaluate its effect.
A review of the pulmonologist's notes shows that the child has maintained her baseline level of activity with minimal reported symptoms. She has had no syncopal episodes or complaints of angina.

Her echocardiography report from earlier this year shows:

- *Mild right atrioventricular valve regurgitation*
- *Mild to moderate left atrioventricular valve regurgitation*
- *Mild right ventricular hypertrophy and dilation*
- *No left ventricular outflow obstruction and normal biventricular systolic function*

A review of serial echocardiograms reveals that the pulmonary artery systolic and main pulmonary arterial pressures have slightly increased over time.

Key Objectives

- Describe features of Down syndrome that can contribute to development of pulmonary hypertension.
- Identify common residual lesions in patients who have undergone repair of complete atrioventricular septal defect.
- Describe how to utilize echocardiography and cardiac catheterization reports to assess the severity of pulmonary hypertension.
- Risk stratify patients with pulmonary hypertension undergoing noncardiac surgery.
- Develop a plan for management of perioperative pulmonary hypertension medications.
- Describe intraoperative management strategies and factors to consider when determining postoperative disposition in patients with pulmonary hypertension.

Pathophysiology

Is Down syndrome relevant to this patient's diagnosis of pulmonary hypertension?

Down syndrome increases the risk of developing pulmonary hypertension (PH) ***even in the absence of congenital heart disease*** (CHD). Contributing factors include intrinsic airway and lung abnormalities, multifactorial lung injury, and a tendency toward obesity that in combination often lead to respiratory acidosis and hypoxia. (See Table 41.1.) Patients with Down syndrome also have an increased incidence of hypothyroidism. While the relationship between thyroid dysfunction and PH is unclear, the cardiopulmonary effects of hypo- or hyperthyroidism can also have detrimental effects on pulmonary vascular resistance (PVR) and respiratory function. Due to these comorbidities and the frequent presence of CHD, the incidence of PH in children with Down syndrome may be as high as 28%. The majority of such patients are diagnosed within the first year of life, and while most experience remission over time, nearly 30% can have persistent or recurrent disease. This patient has the contributing factors of obstructive sleep apnea, recurrent bronchitis, gastroesophageal reflux, and hypothyroidism.

Clinical Pearl

Down syndrome alone is a risk factor for developing PH, even in the absence of CHD, due to its multifactorial effects on the airway and lungs.

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Table 41.1 Contributors to the Development of Pulmonary Hypertension in Down Syndrome

Anatomic or Physiologic Contributors	Clinical Manifestations	Diagnostic and Therapeutic Interventions
Airway anomalies		
Midface hypoplasia Macroglossia Narrow nasopharynx Adenotonsillar hypertrophy	Obstructive sleep apnea	Continuous positive airway pressure Tonsillectomy and adenoidectomy
Generalized hypotonia Laryngomalacia Tracheomalacia Bronchomalacia	Chronic lung disease	Laryngoscopy and bronchoscopy
Pulmonary anomalies		
Impaired lung growth Lung immaturity Lung injury Aspiration Recurrent pneumonias Gastroesophageal reflux	Impaired lung compliance and gas exchange	Gastrostomy tube and/or Nissen fundoplication Frequent antibiotic therapy H_2 blockers
Relative immunodeficiency		Frequent antibiotic therapy
T- and B-cell lymphopenia Reduced antibody response to immunization Impaired neutrophil chemotaxis	Exacerbation of airway obstruction and lung injury through frequent and prolonged upper and lower respiratory infections	
Thyroid dysfunction		
Congenital hypothyroidism Hyperthyroidism	Potential adverse effect on pulmonary vasculature	Thyroid replacement therapy

What other risk factors for PH does this patient have?

Left-to-right (L-to-R) shunts can also affect the pulmonary vasculature. Because the shunt in complete atrioventricular septal defect (CAVSD) is generally unrestrictive and occurs at both the atrial and ventricular levels, thereby subjecting the pulmonary bed to both a volume *and* pressure load, increases in PVR can occur rapidly. To decrease this risk, surgical repair is commonly performed within the first year of life, ideally between 4 and 6 months of age. Repairs taking place late in the first year of life or beyond, particularly in a child with Down syndrome, should raise concern for the presence of PH.

Residual or newly acquired defects following surgical repair can also exacerbate existing PH. The most common residual lesion in CAVSD is left atrioventricular valve (LAVV) regurgitation. Left ventricular outflow tract obstruction (LVOTO) may also develop depending on the initial anatomy and surgical technique used for the repair. Both defects affect the pulmonary vasculature by causing an increase in left atrial pressure that is transmitted to the pulmonary veins. If severe and prolonged, these lesions can produce changes in the pulmonary arterial system.

Clinical Pearl

When evaluating a patient with a history of CAVSD repair, the age at surgical repair and the presence and severity of residual lesions – most commonly LAVV regurgitation – can help in assessing the patient's risk of PH.

How common is the combination of Down syndrome and a cardiac lesion including a L-to-R shunt?

Nearly 25% of children with Down syndrome also have an atrioventricular septal defect (AVSD), and of the population of children diagnosed with AVSDs approximately half also have Down syndrome. Therefore, when one of these diagnoses is observed, it is common or even likely for the other to be present as well.

What is the impact of this combination on the pulmonary vasculature?

Because Down syndrome and L-to-R shunts act through different mechanisms to produce PH, their effects on the pulmonary vasculature are synergistic. The risk of

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developing PH is therefore higher than if only one of these conditions were present alone and should raise a red flag when observed together.

Clinical Pearl

The combination of Down syndrome and CHD with a L-to-R shunt is relatively common. As both processes can produce PH through different mechanisms, the risk of PH is higher when they are observed together rather than alone.

In the child with repaired CHD and pulmonary hypertension, what are the determinants of anesthetic risk?

Risk assessment focuses on the following patient- and procedure-specific concerns:

- Patient age
- Severity of pulmonary hypertension
- Residual cardiac lesions of hemodynamic significance
- Presence of other significant comorbidities
- Procedural risk for hypotension or hypovolemia

The severity of PH can be described by the relationship or ratio between pulmonary and systemic arterial pressures. Anesthetic risk increases as pulmonary pressures exceed half-systemic and is highest in those with systemic or suprasystemic pulmonary pressures. The presence of right ventricular (RV) dysfunction in the setting of PH is also indicative of disease severity and reflects increased risk. Due to ventricular interdependence, RV dysfunction can also cause left ventricular (LV) dysfunction in an otherwise normal LV.

Disease severity and risk can also be inferred from the patient's functional status and the number of therapies being used to treat PH. For example, a patient who becomes dyspneic with minimal exertion or who has had syncopal episodes has more advanced PH than one who has minimal limitations in activity. Likewise, a patient on multidrug therapy would generally have more severe PH than one who is on monotherapy.

Younger age is associated with an increased risk of anesthetic complications, and patients with PH who are <2 years of age may be at increased risk compared to those older than 2.

Clinical Pearl

Anesthetic risk increases as pulmonary pressures exceed half-systemic and is highest in those with systemic or suprasystemic pulmonary pressures.

Where should information regarding disease severity and residual cardiac defects be sought?

Common sources of information include:

- Clinic notes from the patient's cardiologist and pulmonologist
- Echocardiography and cardiac catheterization results
- Laboratory values: brain natriuretic peptide (BNP)

The patient's cardiologist and pulmonologist should be intimately involved in preoperative assessment and optimization. For those who have residual defects, the question of whether they should be addressed prior to proceeding with elective surgery should be discussed. Marked elevations in BNP suggest ventricular dysfunction and heart failure and are concerning for severe PH.

Based on the echocardiography report, does this child have PH?

While the echocardiographic findings presented are reassuring for preserved RV function, they do not comment on the presence or severity of PH. The body of the echocardiography report may be examined for additional data. Four observations are of particular importance:

1. ***Tricuspid regurgitation (TR) peak velocity***: If the patient has TR, its velocity can be used to calculate RV systolic pressure (which should equal PA systolic pressure in the absence of pulmonary stenosis). This value can then be compared to systemic blood pressure, which should be documented on the echo report. A TR velocity greater than 2.8 m/second is highly suggestive of PH. Using the modified Bernoulli equation:

   $$\textbf{PA systolic pressure} = 4(\textbf{TR peak velocity})^2 + \textbf{RA pressure},$$

   where *PA* = pulmonary artery; *TR* = tricuspid regurgitation; *RA* = right atrial.

2. ***Appearance of the interventricular septum during systole*** (a qualitative assessment of RV systolic pressure): The normal contour of the interventricular septum during systole is concave. That is, the septum extends slightly into the RV because LV systolic pressure normally exceeds RV systolic pressure. If RV pressure is elevated relative to LV pressure, however, the interventricular septum may be flattened, or in the case of suprasystemic RV pressures, it may bow *into* the LV during systole. LV filling may be compromised as a result.
3. ***Pulmonary regurgitation (PR) peak velocity***: This diastolic velocity can be used to calculate mean pulmonary artery pressure (mPAP):

   $$\textbf{mPAP} = 4(\textbf{PR peak velocity})^2 + \textbf{RA pressure},$$

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where *mPAP* = mean pulmonary artery pressure; *PR* = pulmonary regurgitation; *RA* = right atrial.

Mean PA pressures >25 mm Hg are concerning for PH.

4. ***Size, thickness, and function of the RV***: RV dilation, hypertrophy, and/or depressed function are all suggestive of elevated PA pressure (assuming there is no pulmonary stenosis). Patients with depressed RV function and PH are at greater risk for periprocedural complications compared to those with preserved RV function.

Clinical Pearl

Echocardiographic findings consistent with PH can include:

- *Tricuspid regurgitant jet >2.8 m/second*
- *Interventricular septal flattening or bulging into the LV in systole*
- *Pulmonary regurgitant peak velocity > 2 m/second*
- *Right ventricular hypertrophy and/or dilation*
- *Diminished RV or LV function*

The patient's echocardiogram reveals the following information: TR peak velocity is 4.2 m/second and PR peak velocity is 1.99 m/second; the interventricular septum is flattened, there is mild RV dilation and hypertrophy with preserved RV function, and a systemic pressure measured during the study by noninvasive blood pressure cuff is 110/80 mm Hg. How can these findings be interpreted?

These findings are consistent with the known diagnosis of PH and can be used to risk stratify the patient.

- A TR peak velocity of 4.2 m/second would estimate a PA systolic pressure of 81 mm Hg assuming a CVP of 10.

$$\textbf{PA systolic pressure} = 4(4.2)^2 + 10 = 81$$

- Given that the noninvasive systolic pressure was 110 mm Hg, this would suggest a PA (and RV) systolic pressure that is nearly 75% systemic. This would account for the interventricular septal flattening observed during systole and the presence of RV hypertrophy.
- The PR peak velocity of 2 m/second would estimate a mean PAP of 26 mm Hg assuming a CVP of 10.

$$\textbf{mPAP} = 4(2)^2 + 10 = 26$$

Which data points are most relevant in evaluating catheterization data in a patient with pulmonary hypertension?

These variables (see Table 41.2) are first assessed at the patient's "baseline": most commonly at room air with normocapnia. Subsequently, these variables are repeated with exposure to 100% oxygen and 40 parts per million inhaled nitric oxide (iNO). These gases are both potent pulmonary vasodilators and should cause a decrease in PVR and mPAP. Such a response can help tailor therapy, and for the anesthesiologist, should underscore the need for iNO to be available when managing future anesthetics. Conversely, if the PVR does not change with these pulmonary vasodilators, then the patient has fixed pulmonary vascular disease. This occurs in the setting of long-standing PH and is often accompanied by significant RV hypertrophy and possibly dilation and decreased function. In these patients, oxygen and iNO provide no significant benefit, and anesthetic management focuses on maintaining cardiac output and avoiding further increases in PVR.

The patient's catheterization diagram and data are presented in Figures 41.1 and 41.2. What are the essential points?

Comparison of the pre- and post-iNO and hyperoxia cardiac catheterization data reveals that the child still has significant PH by mPAP and pulmonary vascular resistance indexed to body surface area (PVRI) criteria but is responsive to iNO and oxygen.

Clinical Pearl

Concerning findings on cardiac catheterization include

- *mPAP >25 mm Hg*
- *PVRI >3 Wood units/m^2*
- *<20% decrease in mPAP following exposure to iNO and FiO_2 1.0*

Table 41.2 Catheterization Parameters to Aid in Assessment of Pulmonary Hypertension

Variables	Abnormal Findings
mPAP	>25 mm Hg
PVR indexed to body surface area	>3 iWu
Pulmonary capillary wedge pressure (PCWP)	>18 mm Hg indicates left-heart disease as a contributor to PH while <15 mm Hg suggests a pre-capillary cause or pulmonary veno-occlusive disease
Response to increased FiO_2 and iNO: acute vasoreactivity testing	Lack of response pulmonary vasodilators defined as <20% decrease in mPAP

FiO_2, fraction of inspired oxygen; iNO, inhaled nitric oxide; iWU, indexed Wood units; mPAP, mean pulmonary artery pressure; PCWP, pulmonary capillary wedge pressure; PVR, pulmonary vascular resistance.

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Given this functional information and the objective data from the echo and catheterization reports, how can the severity of this patient's PH be described?

See Table 41.3 for assessment of PH severity and anesthetic risk for this patient.

Using the framework noted previously and based on her moderate degree of PH, this patient is at increased risk for perioperative complications due to RV ischemia and/or PH crisis. While her functional status, surgical repair, and preserved biventricular function are all reassuring, they should not obscure this fact. The presence of moderate obstructive sleep apnea is also concerning for the potential for hypoventilation in the immediate postoperative period. Further assessment would be needed to ensure that the child's thyroid levels are normal and that the last episode of bronchitis was remote prior to proceeding. Of note, catheterization findings demonstrated that the patient still responds to iNO and oxygen, which should alert the

Table 41.3 Assessing Pulmonary Hypertension Severity and Anesthetic Risk for This Patient

Pulmonary hypertension severity	*Moderate:* PA pressures are 75% systemic in the setting of preserved right and left ventricular function; (+) response to vasoreactivity testing
Residual cardiac lesions of hemodynamic significance	No significant LAVV or RAVV regurgitation or stenosis, no LVOT obstruction
Presence of other significant comorbidities or end-organ dysfunction	OSA, GERD, hypothyroidism, and recurrent bronchitis
Procedural risk of hypotension or hypovolemia	Minimal expected hemodynamic fluctuations or fluid shifts for tympanoplasty
Age <2 years	No

GERD, gastroesophageal reflex disease; LAVV, left atrioventricular valve; LVOT, left ventricular outflow tract; OSA, obstructive sleep apnea; PA, pulmonary artery; RAVV, right atrioventricular valve.

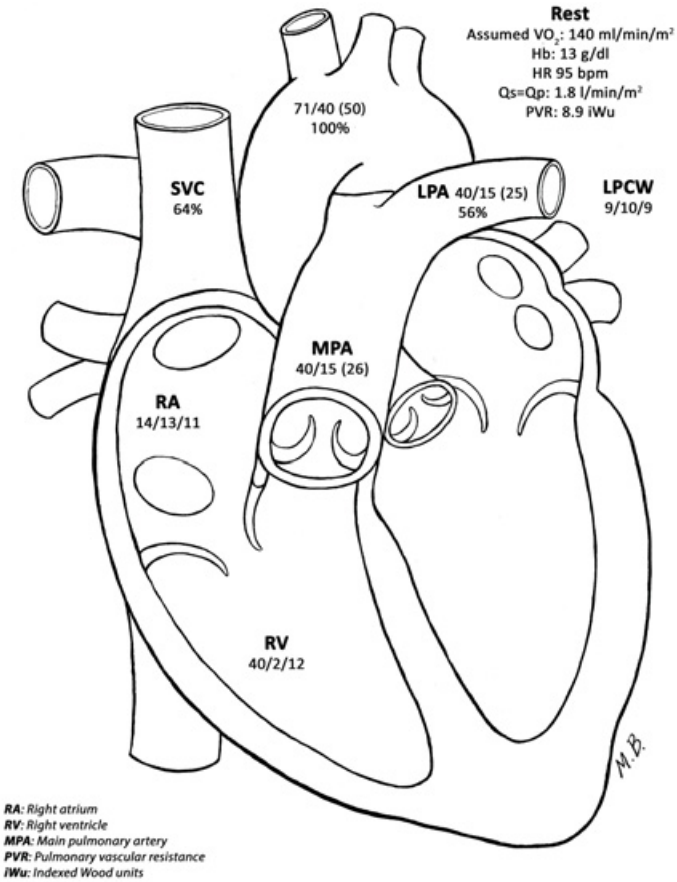

Figure 41.1 **Baseline cardiac catheterization data pre-iNO and hyperoxia.**

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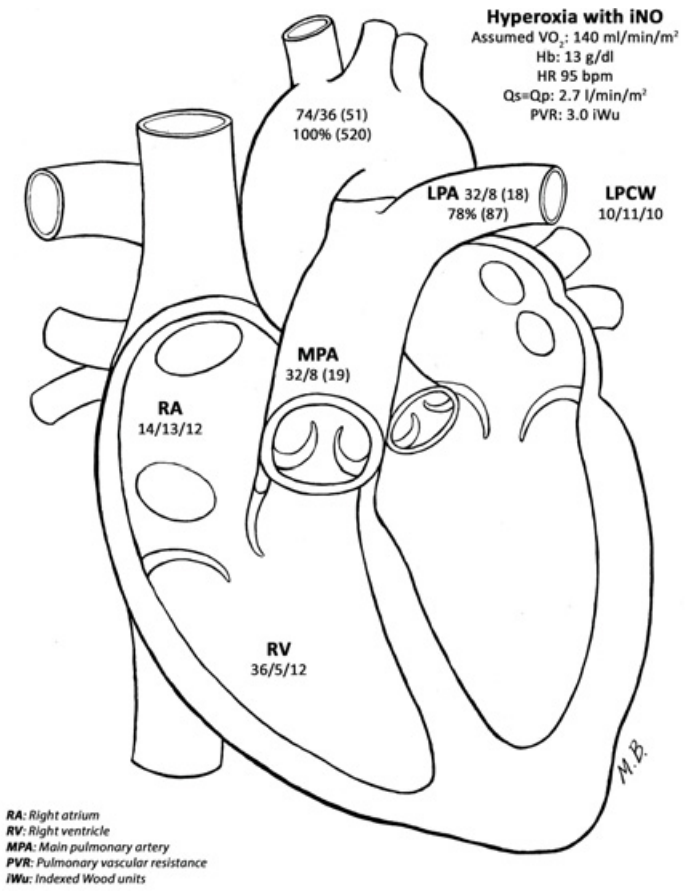

Figure 41.2 Cardiac catheterization data post-iNO and oxygen.

anesthesia team to have iNO readily available in the operating room when managing such a patient.

If the patient's most recent cardiology and pulmonary evaluations are a year old, would it be reasonable to proceed without updated information?

Contacting the patient's medical providers and discussing these issues is never wrong. While it is often assumed that the patient's medical team is aware of an upcoming procedure or anesthetic, that may not be the case. Factors that may suggest the need for an updated evaluation include the patient's risk (based on his or her history and the planned procedure) and any interval changes in clinical status that may have occurred since the last visit. For these patients, preoperative screening is critical not only for planning a safe intraoperative course but also an appropriate post-anesthetic disposition. At many institutions, surgeons refer potential high-risk patients to a preoperative screening clinic so that these issues may be identified and the appropriate consultants can be notified in advance of the procedure.

What other comorbidities or perioperative issues may be present in children with Down syndrome?

See Table 41.4 for associated perioperative concerns in children with Down syndrome.

What additional information would be helpful prior to proceeding with surgery?

While the risk of bleeding with tympanoplasty is relatively low, clotting or coagulation defects could have a negative impact on surgical outcome given the confined space of the ear. As children with Down syndrome and PH may have abnormalities in platelet count or coagulation, a complete

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Table 41.4 Perioperative Concerns in Children with Down Syndrome

Comorbidities or Perioperative Issues	Anesthetic Implications
Atlanto-axial instability	Cervical spine precautions or neurosurgical evaluation may be warranted in those who have clinical concerns for spinal compression Routine preoperative radiography is not recommended if asymptomatic
Difficult vascular access	Ultrasound may be helpful
Subglottic stenosis and/or small trachea	Smaller than expected endotracheal tube placement Potential for post-extubation stridor
Tendency toward profound bradycardia with inhalational induction	Close attention to heart rate and sevoflurane concentration during induction Rapid intravascular or intramuscular administration of anticholinergic if hemodynamically significant
Gastrointestinal anomalies Duodenal atresia Tracheoesophageal fistula Hirschsprung's disease Imperforate anus	Potential increased surgical time and risk of bleeding for subsequent abdominal procedures Potential risk of perianesthetic aspiration
Hematologic anomalies Increased risk of leukemia Thrombocytopenia	Potential increased risk of anemia and operative bleeding

blood count and coagulation panel would be reasonable to order. Specific to this patient who has had a CAVSD repair, an ECG would be helpful to identify any atrioventricular or intraventricular conduction delays that may influence the choice of anesthetic drugs used. First-degree atrioventricular block is particularly common in this subgroup. The patient's history of hypothyroidism also warrants thyroid function studies if not recently performed.

Clinical Pearl

Children with Down syndrome and PH may have abnormalities in platelet count and coagulation. This may warrant laboratory evaluation prior to surgery. In children who have undergone CAVSD repair, some element of heart block may exist and can be identified with an ECG.

Anesthetic Implications

How should the child's PH medications be managed perioperatively?

Though there are no pediatric studies to guide management, the tendency is to continue these medications perioperatively as their benefits exceed the risk. That is, they are not held preoperatively, and their scheduled administration is continued during surgery if an intravenous (IV) equivalent exists. A potential risk of pulmonary vasodilators such as the phosphodiesterase inhibitors (sildenafil or tadalafil) or the calcium channel blockers (nifedipine, diltiazem, or amlodipine) is that they may also cause systemic vasodilation. Theoretically, then, they may cause hypotension when administered in proximity to an anesthetic. While this is not commonly observed, one should be prepared to treat this outcome.

Special consideration should be made for those patients receiving treprostinil (Remodulin®). This drug is delivered subcutaneously as a continuous infusion via a pump (similar to an insulin pump in diabetic patients). Depending on the procedure for which a patient is scheduled, conversion to IV treprostinil may be advisable to maintain a steady plasma concentration. At many institutions, the pulmonary service facilitates this transition preoperatively.

Clinical Pearl

The benefits of perioperative continuation of PH medications likely exceeds the risks of the systemic vasodilation that they may cause.

In addition to oxygen and iNO, what other rescue medications can be helpful to acutely reduce PVR intraoperatively?

For patients with severe PH or highly labile pulmonary pressures, the use of inhaled prostacyclin (Iloprost®) can be beneficial. It can be administered through an anesthesia circuit with the appropriate adapters and acts quickly due to its direct delivery to the pulmonary vasculature.

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What vasoactive drugs should be immediately available?

Vasoactive drugs commonly used in the management of PH include epinephrine, norepinephrine, 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

When managing a child with PH available adjuncts should include:

- *Inhaled nitric oxide*
- *Inhaled prostacyclin for patients with severe or labile PH*
- *Vasoactive agents to increase systemic blood pressure*

Are there any anesthetic drugs that should be avoided in this patient?

The severity of the patient's disease should guide drug selection and dosing. Because ***propofol*** has the potential to cause myocardial depression as well as systemic vaso- and venodilation, hypotension can result with even small bolus doses in the high-risk patient. As such, it should be used with caution, if at all, in those with depressed RV function and/or moderate to severe PH. ***Ketamine***'s effects on PVR and PAP have been the subject of controversy, but the drug is well tolerated assuming hypercapnea is avoided and the patient is exposed to pulmonary vasodilators such as oxygen or sevoflurane. Hemodynamically, ketamine is ideal in that it maintains blood pressure and therefore coronary perfusion pressure while also providing analgesia and sedation. ***Dexmedetomidine*** is thought to have a minimal impact on PVR. Caveats with this drug include its potential to decrease cardiac output by slowing the heart rate significantly or worsening the degree of heart block in a patient with existing conduction delays. Further, in patients with Down syndrome who are already prone to bradycardia, dexmedetomidine may cause hemodynamically significant decreases in heart rate.

Is invasive monitoring excessive for this procedure?

In this child with moderate PH, it would be reasonable to place an arterial line in order to monitor blood pressures more closely and draw blood gases to assess the adequacy of ventilation and oxygenation. A central line should not be needed for this procedure but could be considered if the hemodynamics were concerning following induction. If use of a vasoactive infusion appears likely, it is prudent to obtain central venous access.

How does a PH crisis present?

One of the common intraoperative triggers for a PH crisis is a painful stimulus, most commonly due to intubation or surgical incision. As a result, lung compliance may acutely worsen and blood pressure may fall in the absence of an atrial or ventricular level defect. If an atrial or ventricular level defect is present and provides a "pop-off" (the ability to shunt R-to-L when a PH crisis occurs), hemoglobin–oxygen desaturation could occur with resultant cyanosis but cardiac output would be maintained. If the PH crisis is sustained, ECG changes consistent with RV and LV ischemia often develop, leading to further hypotension and ultimately cardiac arrest.

Equally concerning in a PH crisis is the development of hypotension due to any combination of decreased SVR and/or hypovolemia. Hypotension results in a lower coronary perfusion pressure, and for a patient who already has an elevated right ventricular end diastolic pressure (RVEDP), this lessened gradient can result in RV ischemia.

RV myocardial perfusion = Aortic diastolic pressure – RVEDP

Clinical Pearl

Factors that can acutely increase PVR and precipitate a PH crisis include

- *Painful stimuli (intubation or surgical incision)*
- *Hypoxia*
- *Hypercapnea*
- *Acidosis*

What drugs are most helpful if such an event were to occur?

The main goals in this setting are dual and intertwined: decrease PVR and support RV perfusion. Depending on the etiology, the following drugs/maneuvers should be considered in rapid succession.

Treatment of a PH Crisis

- Phenylephrine to increase systemic pressure and decrease RV ischemia
- Administration of 100% oxygen and initiation of iNO
- Consider inhaled prostacyclin if available
- Analgesia and/or increased anesthetic depth
- Initiation of inotropic and pressor support

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- Boluses of code dose epinephrine if no improvement with the above
- Rapid call for extracorporeal life support if these measures fail to resuscitate the patient

What issues specific to tympanoplasty are relevant in this patient?

The increased incidence of postoperative nausea and vomiting (PONV) associated with tympanoplasty may adversely affect respiratory dynamics as well as volume status. If PONV becomes severe, it may also affect the ability to continue oral PH medications in the postoperative period. Common antiemetics such as dexamethasone and ondansetron can be used to the decrease this risk. A low-dose continuous infusion of propofol may also be considered for PONV prophylaxis assuming the patient's blood pressure is being closely monitored.

Utilization of a pain control strategy that is not overly reliant on opioids can also aid in mitigating the risk of PONV. A multimodal plan including acetaminophen, ketorolac (in the setting of a normal platelet count and surgeon agreement), and ketamine can be useful.

What factors should be considered when determining postoperative disposition?

Perhaps the most important consideration is the patient's preoperative risk based on the aforementioned factors. If the patient is considered moderate- to high-risk, postoperative admission and monitoring in an intensive care setting may be warranted regardless of the stability of the intraoperative course. Discharge to the floor or home can be considered on postoperative day 1 if the patient is progressing as expected. For those who are lower risk, the decision point between floor admission versus discharge home after a period of observation in the recovery room may be considered. Any concerns regarding PONV, pain management, intraoperative hemodynamic instability, or the ability to resume medications should tilt the scales toward admission.

Suggested Reading

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