Delivery of anesthesia to patients with severe pulmonary hypertension can be extremely challenging. The profound hemodynamic alterations of the disease can often be exacerbated by alterations in circulatory function brought about by anesthetic and surgical interventions. High perioperative morbidity and mortality rates have been reported. Minimizing adverse outcomes in these patients requires careful perioperative evaluation and planning. Selection of an anesthetic technique suitable for the surgery without causing major hemodynamic alterations, which can lead to cardiac failure and death, is a unique consideration of the anesthesia provider. As shown in this case report, caudal anesthesia, when appropriate, can offer a safe anesthetic for these patients.

Keywords: Caudal anesthesia, pulmonary hypertension, pulmonary vascular resistance.
sure of 100/72 mm Hg and a pulse rate of 81/min in NSR, respiratory rate of 16/min, and SaO₂ of 97% on 6 L/min of oxygen via a nasal cannula. Although venous congestion was noted in the neck veins, the patient denied any respiratory difficulty.

Fentanyl, 50 µg, was slowly given intravenously before caudal block placement. The skin area and subcutaneous tissue above the sacral hiatus were localized with 5 mL of 2% preservative-free lidocaine. An 18-gauge Crawford needle was inserted through the sacral hiatus, and an epidural catheter was threaded 5 cm into the caudal canal without difficulty. After a negative test dose with 45 mg of lidocaine and 15 µg of epinephrine, 15 mL of 0.5% preservative-free ropivacaine was slowly injected through the catheter over a period of 6 minutes. After 15 minutes, there was a small area of sensation around the perineum; another 5 mL of the same local anesthetic was injected. A satisfactory block to the T10 dermatome was established, and the 1-hour surgery proceeded without incident. Blood pressure was maintained within 20% of baseline; heart rate ranged from 76/min to 82/min in NSR, and SaO₂ was 95% to 98% with spontaneous respirations. Blood loss was minimal. The patient received a total of 500 mL lactated Ringer's solution intravenously. No additional sedatives or opioids were administered. He was positioned supine and transferred to the recovery room where he remained in stable condition until discharge.

Postoperatively, the patient recovered uneventfully. Retrospectively, we could have used shorter acting, local anesthetic chloroprocaine because he experienced some urinary retention. However, he was discharged on the same day of surgery without adverse events. An adenosine thallium stress test was performed 3 weeks later. The lower part of the myocardium was noted to have some reversible perfusion defects. The LV ejection fraction was normal. The study findings were otherwise consistent with those of a patient with PHTN.

**Discussion**

- **Classification and Management.** Pulmonary hypertension was reported in autopsy findings in the late 1800s. It has been classified as primary and secondary. Primary PHTN is idiopathic and rare, whereas secondary PHTN is much more common and often caused by cardiac abnormalities (eg, valvular disease, LV dysfunction). In 2003, this classification was revisited at the Third World Conference on Pulmonary Arterial Hypertension in Venice, Italy (Table 1). Most commonly, PHTN is diagnosed in women between the ages of 20 and 40 years, although it can occur in men at any age group.

  Physiologically, the pulmonary vasculature is a high-flow, low-resistant circuit with thin-vessel walls. It is highly compliant, and this resilience plays a critical role in the distention and recruitment of the vasculature in response to high RV output. This feature of the pulmonary circulation is crucial in the maintenance of pulmonary vascular resistance (PVR). Normal systolic and diastolic PAPs range from 20 to 30 mm Hg and 6 to 10 mm Hg, respectively. Mean PAP is about 15 mm Hg. The PVR is determined based on the Ohm law of resistance and flow:

\[ \text{PVR} = \frac{80}{\text{Mean PAP} - \text{LAP}}/\text{CO}, \]

where LAP is left atrial pressure and CO is cardiac output. Clinically, pulmonary arterial wedge pressure instead of LAP is often used. Normal PVR ranges from 50 to 150 dynes·cm⁻⁵·².⁴

In contrast to the peripheral and cerebral vessels, the pulmonary vessels constrict in the presence of alveolar hypoxia (Euler-Liljestrand reflex) and dilate in a high-oxygen environment. Acidosis and hypercapnia also cause pulmonary vasoconstriction; alkalosis and hypocapnia have the opposite effect. Local mediators within the vasculature also affect pulmonary tone. Nitric oxide and prostacyclin play a role in vasodilation, but their counterparts (eg, thromboxane and endothelin) vasoconstrict. Gravity also influences PVR by distributing blood flow to different zones of the lungs. Other determinants of PVR are extremes of lung volumes.

Pulmonary hypertension is defined as a mean PAP as high as 25 mm Hg and is associated with right ventricular hypertrophy and dysfunction. When the RV cannot compensate for the increased load, right heart failure results. Although the natural history of PHTN is characterized by a progressive, inexorable decline in RV function, treatment can improve survival and quality of life in most patients.

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Table 1. Classification of Pulmonary Hypertension (PHTN)

<table>
<thead>
<tr>
<th>Class I – Primary PHTN related to:</th>
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<tbody>
<tr>
<td>1. Idiopathic/hereditary conditions</td>
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<tr>
<td>2. Collagen vascular disease</td>
</tr>
<tr>
<td>3. Systemic-pulmonary shunts</td>
</tr>
<tr>
<td>4. HIV infection</td>
</tr>
<tr>
<td>5. Hepatic portal hypertension</td>
</tr>
<tr>
<td>6. Drugs and toxins</td>
</tr>
<tr>
<td>7. Other: thyroid disease, glycogen storage disease, hemorrhagic and blood disorders</td>
</tr>
<tr>
<td>8. Persistent PHTN in neonates</td>
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</tbody>
</table>

**Class II – PHTN related to:**

1. Cardiac/valvular disease

**Class III – PHTN related to:**

1. Lung disease such as chronic obstructive pulmonary disease, interstitial lung disease, and dysplasia
2. Hypoxemia
3. Hypoventilation syndromes and sleep apnea
4. Neonatal lung disease

**Class IV – PHTN secondary to:**

1. Thrombolic/embolic disease
2. Sickle cell disease

**Class V – PHTN as a result of direct pulmonary insult:**

1. Inflammation (schistosomiasis, sarcoidosis)
2. Pulmonary capillary hemangiomatosis
3. Conditions due to pulmonary vessel compression/occlusion
greater than 25 mm Hg at rest (>30 mm Hg during exercise) or, according to Rich, a PVR greater than 300 dynes·s·cm⁻⁵. Patients are considered to have severe PH TN when the mean PAP exceeds 50 mm Hg or the PVR exceeds 600 dynes·s·cm⁻⁵. To confirm the diagnosis, the practitioner must exclude abnormal pulmonary arterial wedge pressure and CO. Cardiac catheterization is the gold standard in the diagnosis of PH TN. The predominant feature of PH TN is a highly constricted pulmonary vasculature. This constriction is influenced by genetic factors, local mediators, and pathological states (e.g., inflammation, coagulopathy, altered blood gases, and endothelial cell injuries). Because of this constriction, there is a strain on the right ventricle, resulting in difficulty in ejecting blood from the right ventricle into the pulmonary system; hence, both RV end-diastolic pressure and volume are increased. The overstretched right ventricle translates into an increase in workload and consequently increases in oxygen demand and consumption. These patients are at a higher risk of RV ischemia during stressful physiologic demands. If PHTN is left untreated, a positive feedback mechanism can lead to progressive RV failure and LV dysfunction, resulting in cardiopulmonary failure (Figure). 

Management of PHTN aims at correcting the underlying disease processes and/or instituting medical therapy with vasodilators, calcium channel blockers, angiotensin-converting enzyme inhibitors, phosphodiesterase inhibitors, diuretics, anticoagulants, cardiac glycosides, oxygen supplement, prostanoids, endothelin receptor antagonists, and inhaled nitric oxide. In severe cases, a heart-lung transplant may be needed. When damage to the pulmonary vasculature has developed, medical treatment can become complex and difficult. Once diagnosed, survival rates depend on the treatability of the underlying cause. According to the Patient Registry for the Characterization of Primary PHTN, patients with a mean PAP greater than 85 mm Hg have a life expectancy of about 1 year after the diagnosis. As reported in National Institutes of Health data, patients with PHTN functional classes I and II (Table 2) usually do not survive past 6 years, whereas those with class III or IV live only 2.5 years and 6 months, respectively. The patient in this report was classified as class II because of the lack of clinical evidence of cardiac decompensation.

### Figure. Positive Feedback Mechanism of Untreated Pulmonary Hypertension (PHTN)

RV indicates right ventricular; LV, left ventricular; †, increased; ‡, decreased.

### Table 2. World Health Organization Functional Classification of Pulmonary Hypertension

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>I</td>
<td>the patient has no signs of fatigue, shortness of breath, chest pain, or presyncope during normal physical activities.</td>
</tr>
<tr>
<td>II</td>
<td>the patient exhibits mild fatigue, dyspnea, chest pain, or presyncope with normal physical activities and has some limitation in performing activities.</td>
</tr>
<tr>
<td>III</td>
<td>the patient shows overt fatigue, dyspnea, chest pain, or presyncope with limited activities and has marked limitation in performing activities.</td>
</tr>
<tr>
<td>IV</td>
<td>the patient experiences discomfort, fatigue, dyspnea, chest pain, or presyncope at rest and is unable to perform any physical activities.</td>
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PVR during stressful events. Indeed, this acute change is always the likely cause of the sudden and acute deterioration of cardiac function and/or death.12

Because of the high morbidity and mortality rates, providing anesthesia in these patients is extremely challenging. It is essential to appreciate and understand the hemodynamic alterations associated with PHTN to avoid catastrophic outcomes. Careful evaluation before surgery and anesthesia is mandatory. Exertional dyspnea is the most common complaint.3,5 Other clinical symptoms include fatigue, chest pain, hypotension, acid-base disturbances, low SaO₂, signs of RV failure (hepatomegaly, peripheral edema), and syncope. The prognosis is grave when patients experience syncope.5,7 Preoperative preparation should include optimization of the patient's condition. Elective surgery is postponed should there be any evidence of cardiopulmonary decompensation. Anti-PHTN medications should be continued up to the morning of surgery.7,12 At a minimum, a chest x-ray film, an ECG, and an echocardiogram should be obtained. Other diagnostic tests such as pulmonary function tests and a ventilation-perfusion scan can be useful.4,7

The overall anesthetic goals are to improve the cardiopulmonary performance by maintaining CO and coronary perfusion pressure, optimize RV function, and reduce oxygen consumption.2 Anesthetic options for excision of anal condyloma in the prone jackknife position include general anesthesia with airway instrumentation, local anesthesia with sedation (monitored anesthesia care), and neuraxial blockades (lumbar epidural, spinal/saddle, or caudal). In the author's institution, monitored anesthesia care with a propofol infusion and supplemental intravenous fentanyl and/or ketamine is often chosen for the excision of anal condyloma. A local infiltration block by the surgeon is often inadequate unless accompanied by additional intravenous sedatives and/or opioids given by the anesthesia provider. Sedatives and opioids can put the patient at risk of respiratory depression, causing hypoxia and producing further increases in PVR. Ketamine is a potent analgesic while maintaining airway reflexes. On the other hand, this drug is known to increase PAP by as much as 40% at high doses14 and is frequently not a good choice in patients with severe PHTN.

Induction of general anesthesia poses the greatest risk because of the abrupt changes in hemodynamic parameters. Because of fixed RV output, patients with PHTN can be intolerant of the decrease in preload occurring immediately after induction. Höhn et al15 reported a case of cardiac arrest after induction in a patient with severe PHTN (PAP 125/59 mm Hg) due to systemic hypotension and decreased CO. These effects were also accentuated by the application of positive-pressure ventilation. If general anesthesia is necessary, a high-dose opioid induction might be preferable because opioids tend to maintain RV outflow, blood pressure, and CO with little influence on PVR. Nitrous oxide may augment pulmonary vasoconstriction because of its sympathomimetic effects, and its use is not advocated.14 Hyperventilation with 100% fraction of inspired oxygen, low positive end-expiratory pressure, and avoiding hypoxia and extremes of lung volumes are recommended12 if general anesthesia is used.

Neuraxial anesthesia may offer good anesthetic choices for procedures below the diaphragm. Spinal anesthesia can lead to rapid sympathectomy, resulting in failure to maintain preload and systemic blood pressure in patients with PHTN and fixed RV ejection fraction. Epidural anesthesia, on the other hand, may offer more stable and better control of hemodynamics because of its slow onset of sympathectomy. It has been used successfully in obstetric patients with PHTN.7 Despite its slow onset, lumbar epidural anesthesia, especially when it is spread cephalad, can impair venous return.16 Moreover, it might spare the sacral segments unless an extensive block is achieved, which could produce profound sympathectomy, resulting in a dangerous loss of vascular tone. High thoracic epidural anesthesia has little influence on PVR but can block the T1 to T4 cardiac sympathetic accelerator fibers, resulting in decreases in heart rate, contractility, and blood pressure. Caution should be taken when initiating epidural anesthesia to avoid a dramatic drop in venous return.7

In the case report, lumbar epidural anesthesia was considered but rejected because, in order to provide analgesia in the sacral dermatomes, an extensive spread of the local anesthetic would have been needed. Further consideration was a saddle block, which would have required the patient to remain in the sitting position during the onset of the block. Any sympathectomy in the sitting position would tend to decrease venous return and possibly induce syncope from failure to maintain blood flow through the highly resistant pulmonary circuit.

In contrast to the techniques mentioned above, caudal anesthesia would provide several advantages. First, the patient was allowed to assume the surgical position comfortably before commencing the block. The nerve roots that pass through the sacral canal innervate primarily the sacral dermatomes. As the surgery was performed on sacral dermatomes, the deposition of local anesthetics within the sacral canal led to an intense block of the surgical site. Like any other techniques, caudal anesthesia does have risks, such as systemic injection of local anesthetic. There is also up to a 15% chance of an unsatisfactory block.17 Nevertheless, given the case scenario and substantial risks associated with the other anesthetic techniques, it was thought that caudal anesthesia would offer hemodynamic stability and was the best choice for this patient. Moreover, the patient also presented with normal sacral anatomy and the ability to tolerate the prone jackknife position relatively well.
Intraoperatively, hemodynamics should be maintained as close to baseline as possible. Goals are similar to those patients with aortic stenosis: adequate preload; normal to high systemic vascular resistance and contractility; avoidance of bradycardia, tachycardia, and arrhythmias; and maintenance of myocardial oxygen balance. More importantly, further increases in PVR must also be avoided. Standard monitors are required. Patients with PHTN are exquisitely sensitive to fluid volume underload and overload. It is ideal to measure intracardiac filling pressures in major abdominal or thoracic surgeries. Central venous and arterial lines might also be necessary in these cases. Excision of anal condyloma is a relatively minor surgery not associated with fluid shifts; hence, invasive monitoring is unnecessary.

Dobutamine, milrinone, and amrinone have all been used to improve CO. Phenylephrine, epinephrine, and norepinephrine can also be administered alone or in combination to improve coronary perfusion pressure and pump function. In severe or refractory hypotension associated with RV failure, inhaled nitric oxide is the therapy of choice because of its direct pulmonary vasodilation effect. Patients with PHTN often experience sudden death within the first few days after surgery relating to pulmonary and systemic vasoconstriction, embolism, myocardial dysfunction, and fluid shifts. Postoperative care should include adequate pain control, avoidance of hypoxia, and maintenance of euvolemia. It is often appropriate to leave these patients intubated and slowly weaned from mechanical ventilation a few days after major surgeries. Furthermore, intravenous vasodilators can serve as bridges during the weaning process.

Conclusion
This article presents a case report of a patient with PHTN and an overview of its pathogenesis. Pulmonary hypertension is a severe disease that can cause profound hemodynamic alterations in the body. If left untreated, it can quickly progress to cardiopulmonary failure and death. Even with advanced medical therapy, the prognosis is dismal. Patients with PHTN have high morbidity and mortality rates when undergoing surgery and anesthesia. Indeed, it is crucial to perform a careful preanesthetic evaluation and select a safe anesthetic technique. Although challenging, all anesthetic techniques have been employed with success. Despite careful preoperative planning, patients can still potentially deteriorate rapidly related to acute increases in PVR. The anesthesia provider needs to be vigilant and recognize the unique anesthetic considerations in order to avoid catastrophic outcomes. Pain management and avoidance of hypoxia and hypotension are issues that must be addressed in the postoperative period. For procedures in the perineum, caudal anesthesia can be safely performed and avoidance of adverse outcomes can be accomplished. Although caudal anesthesia is used today primarily in the pediatric population, this case report emphasizes the importance of maintaining the skills necessary to perform caudal anesthesia in adults.

REFERENCES

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