PBLD – Table #15

Title: **Bowel obstruction and CDH recurrence after VEPTR insertion on a child with thoracic insufficiency and pulmonary hypertension.**

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Objectives:

1. Review neonatal chronic lung disease and pulmonary hypertension
2. Review pathophysiology of congenital diaphragmatic hernia
3. Review strategies to avoid pulmonary hypertensive crisis and right ventricular failure
4. Review thoracic insufficiency and surgical management options: Vertical Expanding Prosthetic Titanium Rib (VEPTR)
5. Review utility of transthoracic ultrasonography to evaluate pulmonary pathology
6. Conversion of complication to rescue (failure-to-rescue)

Case history:

A 28 month, 11.9 kg child presents with increasing abdominal girth, nausea, vomiting and desaturation for exploratory laparotomy for bowel obstruction from recurrence of a right-sided diaphragmatic hernia.
Pertinent medical history includes 30 week gestation, s/p congenital diaphragmatic hernia repair with patch, chronic lung disease of prematurity, neonatal ECMO, pulmonary hypertension without right ventricular compromise, thoracic insufficiency and recent placement of a vertical prosthetic expanding titanium rib (VEPTR).
A chest radiograph demonstrates visible loops of bowel in the right chest above the diaphragm. On exam, the child is uncomfortable but interactive. On exam, he is tachypneic with visible retractions, has a prominent abdomen and is tachycardic.
O2 saturation is 92% on non re-breather mask with 100% FiO2, respiratory rate is 60

Past surgical history:

Right-sided Congenital diaphragmatic hernia repair at 2 months of age
Right heart catheterization at 3 months of age
Bilateral strabismus repair at 15 months
Exploratory laparotomy, lysis of adhesions, Nissen/G-tube at 15 months of age
Right VEPTR insertion 2 days prior to presentation.

Medications:

Sildenafil and 0.25 L/min O2 by nasal cannula at baseline, lansoprazole, flovent, albuterol.
Previously he had been on treprostinil and inhaled nitric oxide.

Preoperative studies:

Weight 11.9kg (15th percentile)
Hemoglobin 12.4 g/dL.

BNP 18

Echo summary (6 months prior to surgical intervention):
- Mildly dilated right atrium
- Atrial septal bulge to left, suggestive of elevated right atrial pressure
- Mild dilatation of right ventricle and right ventricular hypertrophy
- Normal right ventricular systolic function
- Ventricular septum is mildly flattened in relation to left ventricle.
- Septal position suggests right ventricular pressure likely 1/3 to 1/2 systemic.

Questions:
- What is the appropriate course of pre-operative optimization?
- Should ECMO be available for backup?
- Management during induction? Rapid sequence? Agents?

Case Progression:

After rapid sequence induction with propofol and succinylcholine, airway was secured.

Intraoperative Care:
- Would you delay procedure to place an arterial line?
- Vascular access strategy? What is sufficient vascular access? Would you rely on lower extremity intravenous lines? Central line placement?
- Ventilation strategy?
- Therapeutic options for managing pulmonary hypertension?

Case Progression:

Surgeon performed exploratory laparotomy and was not successful in releasing adhesions in right thorax. Plan to convert to exploratory thoracotomy on the right side.

Questions:
- Would you require an arterial line at this point?
- Discuss options and implications for one-lung ventilation?

Post-operative Care:
- Post-operative analgesia?
- Ventilation strategy?

Case Progression:

The surgical procedure was completed after 8 hours, including a large abdominal and large thoracotomy incision. Fluid intake included 1400mL lactated ringers and 185 mL red blood cells. Estimated blood loss was 50mL and gastric decompression yielded 1000mL fluid. Intraoperative hemodynamics remained stable without evidence of

tachycardia or hypotension.

Upon arrival to the ICU, the patient remained on mechanical ventilation. Liver remained enlarged and BNP = 200.

Follow up transthoracic echocardiogram:
1. Mildly diminished right ventricular systolic shortening
2. Ventricular septum mildly bowing in relation to the left ventricle
3. Atrial septum bowing right to left
4. Trivial tricuspid regurgitation
5. Normal left ventricular size and qualitatively normal systolic shortening.

Pt. was treated with inhaled nitric oxide, dopamine and furosemide infusions. Despite therapy, the patient continued to develop hypoxemia.

Questions:
- Differential diagnosis for hypoxemia in this setting?

Case Progression:

Sepsis workup initiated and yielded positive test for adenovirus. Chest X-ray continued to show opacification of right lung. Upon bronchoscopic evaluation, the patient was noted to have conformational changes in the right mainstem bronchus, possibly contributing to obstruction/hypoventilation.

Discussion:

Thoracic insufficiency syndrome leads to physical constraints that limit the development and growth of intrathoracic organs and the vertebral column. The affected lung can remain hypoplastic and will lead to the development of elevated pulmonary vascular resistance and ultimately pulmonary hypertension. (1) In addition, the thoracic deformity contributes to the development of restrictive lung disease that progresses with age. (2) Children may suffer frequent respiratory infections and failure to thrive. Children with infantile scoliosis are ideally managed by therapies that promote continued growth of the spine and thoracic cavity, leading to maturation of the spine and lungs. Spinal fusions are not recommended on children younger than 8-10 years of age. (3) Treatments available include non-surgical (brace, casts) as well as surgical options such as VEPTR. VEPTRs involve insertion of an expandable device that is fixated to the ribs as well as the pelvis or spine on the affected side. The child is managed by regular follow up and interval expansion of the device to keep up with growth. (4) The procedure has been associated with various complications including neurologic, skin problems, infectious, device migration and fractures. (5)

Patients presenting for insertion or expansion of VEPTR devices may have complex medical history similar to the case discussion. Thoracic insufficiency is common and may be associated with pulmonary hypertension and cardiovascular compromise. Furthermore, the patients may be more susceptible to the respiratory

depressive effects of opioids that are typically used for post-operative analgesia. Patients at risk for pulmonary hypertension should be closely monitored to avoid acute increases in pulmonary vascular resistance. (1) Acute pulmonary hypertensive crisis is associated with a high incidence of mortality and may present one of the most challenging scenarios for resuscitation in any setting. (6) Initial management should focus on optimizing oxygenation and ventilation as well as lung perfusion. Acute interventions may include initiation of inhaled nitric oxide therapy and vasopressor therapy. It is important to note that systemic arterial blood pressure may be reduced in the acute pulmonary hypertensive crisis as a result of limited venous return from the lungs. The failing right ventricle will be susceptible to increased stress in this setting, and will not tolerate aggressive fluid resuscitation. Intraoperative echocardiography may play a crucial role in guiding therapeutic interventions in this setting. Patients who fail to respond to initial management should be evaluated for ECMO cannulation if clinically appropriate. The progression to acute hemodynamic decompensation can be quite rapid, and resuscitation with CPR may not be adequate due to the elevated pulmonary vascular resistance.

This patient has three conditions that place him at higher risk for severe chronic lung disease of prematurity: 30 week gestation with lung prematurity, congenital diaphragmatic hernia affecting the right lung and infantile scoliosis leading to thoracic insufficiency syndrome affecting the right hemi thorax. Upon insertion of the VEPTR, his diaphragmatic hernia recurred and resulted in an acute bowel obstruction that further compromised his respiratory function due to abdominal competition. Diaphragmatic hernias have a recurrence rate of 3-22% and may not always require surgical intervention. (7) In this case, the acute bowel obstruction and respiratory decompensation led to the emergent reoperation to address the recurrent hernia. Bowel obstruction after CDH repair is not common, but has been reported. (8)

Ultrasound devices are becoming increasingly available due to lower cost and increase in providers experienced with its use. Recently, there have been reports on pulmonary evaluation with ultrasound to detect atelectasis, pneumothorax or even endotracheal intubation. (9) (10)

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