Bridging to transplant with a lung assist device following an open lung biopsy complicated by a pulmonary hypertensive crisis

Problem Based Learning Discussion
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Goals:
1. Discuss the perioperative management of a lung biopsy in a patient with pulmonary hypertension
2. Describe the indications and risks of Extracorporeal Membrane Oxygenation [ECMO]
3. Understand how a lung assist device can be utilized in patients with respiratory failure
4. Consider the indications for and limitations of lung transplantation

Case Description:

Preoperative:
An 8-month-old 8.1 kilogram girl with pulmonary hypertension presents for a lung biopsy to evaluate her persistent oxygen requirement. In room air her oxygen saturations drops to 85% within minutes. You are examining her in the preoperative surgery suite.

Her past medical history is significant for being born at 2.8 kilograms at 34 weeks gestation. She had no complications in the immediate neonatal period but was hospitalized at 2 months of age for dehydration caused by diarrhea. At the time of that hospitalization she was noted to have persistent hypoxemia for which she was started on supplemental oxygen.

What are the causes of hypoxemia in an 8-month-old?
What are your preoperative concerns for her open lung biopsy?
Does the presence of pulmonary hypertension modify your concerns?
What would you tell the parents were the risks to their child?

Her preoperative workup included an echocardiogram which revealed an atrial septal defect and systemic right ventricular pressures, a chest X-ray and a chest computerized tomography scan which revealed diffuse bilateral interstitial pulmonary fibrosis. She was on sildenafil, oral prednisone and supplemental oxygen at ¾ liters per minute by nasal cannula. She was also on Zantac and Mylanta for
gastroesophageal reflux disease and Albuterol as needed. She had a regular heart rate at 141, a respiratory rate of 38, blood pressure of 117/62, oxygen saturation of 96% on supplemental oxygen and in the mid to low 80s on room air. She was afebrile at 36.6°Celsius. She was awake, interactive, vibrant and curious.

How would you anesthetize this patient?
  What monitors would you utilize?
  Would you use a regional technique?
  Would you use a lung isolation technique?
  What would be your postoperative management plan?

Early Postoperative Course:
The surgery occurred uneventfully but she developed progressive desaturation and was unable to be extubated and she was therefore admitted to the pediatric ICU for further management. Over the next several days her condition deteriorated and she was placed on venoarterial-ECMO. The results of the lung biopsy were consistent with alveolar capillary dysplasia with misalignment of the pulmonary veins. This is a condition that can only be treated by lung transplantation.

What is ECMO?
What is venoarterial-ECMO?
How does venoarterial-ECMO differ from venovenous-ECMO?
What are the risks associated with being on ECMO?
What are the indications for lung transplantation?
**Intensive Care Unit Course:**

The child is not a candidate for lung transplantation as she is on venoarterial-ECMO. Her therapeutic options are limited and include transition to venovenous-ECMO, conversion to a lung assist device, or withdrawal of care. Extensive discussions occur with the family and an ethics consultation is performed. Following through consideration of their child’s options, and after discussion with the team, the ethics committee and their priest, the family consents to surgery for the lung assist device. She subsequently goes to the OR on venoarterial-ECMO and is converted to a Maquet Quaddrox assist device. The device, which is the oxygenator for an ECMO circuit, was connected by cannulae to the pulmonary artery and the left atrium. This configuration allowed her native right ventricle cardiac output to perfuse the artificial lung, with inflow cannula to the device arising from the pulmonary artery and outflow cannula returning to the left atrium. She is thereafter placed in active status on the lung transplantation list.
What are strategies for hemodynamic management while on a lung assist device?
Would inhaled nitric oxide be any benefit following placement of such a device?
How does one the balance risk of bleeding with risk of device clotting in designing an anticoagulation strategy?
What are the limitations of lung transplantation?

Discussion:

Pulmonary hypertension is an extremely challenging medical condition encountered in pediatric anesthesia practice. Although an uncommon condition, these children undergo anesthetics for diagnostic procedures such as imaging studies or cardiac catheterization and for surgical procedures such as vascular access or open lung biopsy. Children with pulmonary hypertension are at considerable risk for cardiovascular instability, cardiovascular arrest and even death and so the perioperative care must be well organized (Carmonsino, van der Griend). Our patient is typical of such a child, presenting for an open lung biopsy with a persistent unexplained oxygen requirement and echocardiographic evidence of pulmonary hypertension.

Alveolar Capillary Dysplasia (ACD) with misalignment of the pulmonary veins is often diagnosed early in life but many present later in patients with phenotypic variants (Ahmed, Singh). The initial presentation is often within 48 hours of birth and the natural history is progressive lung disease proceeding to respiratory failure and death early in life. ACD can be associated with pulmonary hypertension which may be refractory to therapy. Therapeutic options are limited for these patients, and diagnosis has often led to limitation of futile interventions (Bishop). Ninety percent of the reported cases have been diagnosed on autopsy. A patient with respiratory failure from ACD has been described in the literature that was managed with ECMO but died from refractory pulmonary hypertension (Al-Hathlol).

Our patient underwent lung biopsy to determine the cause of her persistent hypoxemia and postoperatively developed refractory pulmonary hypertension which was stabilized with ECMO. With her diagnosis of ACD the best treatment option was lung transplantation. The Novalung, a lung assist device, has been used to bridge patients to lung transplantation with the longest reported patient maintained for 140 days (Bartosik) and the smallest a 41 kg, 15-year-old (Taylor). For smaller patients such as ours, the Maquet Quaddrox, which is the oxygenator for an ECMO circuit, has a smaller priming volume and has been used in infants with pulmonary hypertension including one who improved sufficiently to avoid lung transplantation (Gazit). The Quaddrox was therefore used to transition this patient off ECMO and bridge to lung transplantation. This represents a novel lifesaving method of stabilizing an infant with refractory pulmonary hypertension until an organ is available for transplantation.
Prior Presentation:
This information was not previously presented at the SPA, ASA or other meeting.

References: