CONGENITAL PULMONARY ADENOMATOID MALFORMATION / CONGENITAL CYSTIC ADENOMATOID MALFORMATION (CPAM/CCAM)

Surgical Specialty:	Pediatric Surgery
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Background:

- General Considerations
  - Congenital pulmonary adenomatoid malformation (CPAM) or congenital cystic adenomatoid malformation (CCAM) is a rare congenital bronchopulmonary abnormality with an incidence of 1:10,000 to 1:35,000 live births [1,2]
  - CPAM/CCAM accounts for 25% of all congenital lung abnormalities[1]
  - Cystic and adenomatous lesions arise from the bronchopulmonary tree, most often unilaterally, with 41% of cases involving the left upper lobe, 34% the right middle lobe, and 21% the right upper lobe.
  - Abnormalities during embryogenesis result in increased cell proliferation with decreased apoptosis of bronchi without normal alveolar development, resulting in the lesion that can increase in size during development or regress.
  - Microscopic features that differentiate CPAM from normal lung tissue include:
    - Proliferation of terminal respiratory structures forming cysts
    - Polypoid projections from the mucosa
    - Increased smooth muscle and elastic tissue within cyst walls
    - Absence of cartilage
    - Presence of mucous-secreting cells
    - The absence of inflammation
  - Cysts are not involved in gas exchange and are prone to recurrent infections
  - CPAM/CCAM is classified by the Stocker Classification -Types 0-IV according to the location of origin, size, and the histopathological characteristics of the lesion[2]
    - Type 0 – arises from the trachea or bronchi
      - Fatal/Incompatible with life
    - Type I – arises from the bronchi
      - Cysts are usually multiloculated
      - Appears as an air-filled cyst on X-ray
      - > 2 cm in size
      - Cysts lined with columnar epithelium
      - Represents 70% of all CPAM cases

        - Presents with increased respiratory effort, tachypnea, and cyanosis
    - Type II is the second most common variant, representing 10-15% of CPAM
        - arises in the bronchiolar region
        - Presents as multiple small cysts and no mass effect
        - <2 cm in size
        - Presents with respiratory distress, tachypnea, and cyanosis
        - Associated with other congenital abnormalities
    - Type III – arises from the bronchiolar region
        - The lesion is solid rather than cystic/ microcystic
        - Lesion <0.5 cm in size
        - Lined with plumb cuboidal epithelium
        - Enlarges to encompass the entire lung
        - Leads to hydrops and pulmonary hypoplasia
    - Type IV- arises from the acinar structures of the lung
        - Large air-filled cysts
        - Cyst lined with alveolar lining cells
        - Presents with increased respiratory effort, tachypnea, and cyanosis, but also pneumothorax
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  - CPAM can present anytime from the neonatal period to adulthood
  - Presentation varies from fetal hydrops, neonatal respiratory distress, to an incidental finding in an asymptomatic adult
  - Fetal surgery may be necessary if there is a risk for fetal hydrops
- Patient Considerations
  - Symptomatic CPAMs are resected
  - Management of asymptomatic CPAMs is controversial, and no consensus exists on the timing of surgery. Early resection strategies aim to avoid the development of symptoms. Conservative surgical strategies aim to avoid overtreatment and unnecessary surgical exposure.[3]
  - Lobectomy has been the standard for CPAMs confined to a single lobe.
  - Recently, other procedures, such as segmentectomy and wedge resection, when feasible, are equivalent and preserve the unaffected lung.
  - Disease Specific Considerations
    - The timing of resection is dictated by the type and symptom severity
        - Large lesions or the threat of hydrops may require fetal in utero lobectomy in the mid-gestation period
        - EXIT procedures may be undertaken for large malformation, proximal airway compromise, or threat of hydrops
        - The timing of post-natal intervention is dictated by symptoms and the ability to medically optimize respiratory status
  - Associated Comorbidities/Syndromes
    - Type II is associated with other congenital anomalies
        - Renal agenesis
        - Esophageal atresia
        - Congenital diaphragmatic hernia
        - Cardiovascular lesions
        - Skeletal defects
    - Increased morbidity and mortality with other associated anomalies

        - Type II is associated with malignancy, particularly bronchoalveolar carcinoma, and less commonly rhabdomyosarcoma, adenocarcinoma, and pulmonary blastomas.

Anesthetic Planning:

- Pre-Anesthetic evaluation
    - Detailed history and physical including: evaluation of baseline respiratory function, including evaluation for recurrent or ongoing infection, baseline hemodynamics, birth history, and possible concurrent disease
    - Additional labs/tests indicated during work-up: CBC, arterial blood analysis if in respiratory distress, coagulation and electrolyte studies, particularly in the setting of feeding difficulty or ongoing infection, blood group type and screen versus crossmatch
    - Imaging studies: CXR, CT scan, MRI to localize the lesion, Echocardiogram- to evaluate for concurrent cardio-respiratory conditions
    - Discussions to have with the surgeon/family
        - Code Status
        - ECMO Candidacy
- Specific or Unique Room Set-Up Requirements
    - Airway- tailored to patient anatomy; One-Lung Ventilation (OLV) may be necessary for resection and can be achieved with various devices
        - Endobronchial intubation versus bronchial blockers may be utilized in younger patients.
        - Double-lumen endotracheal tubes (DLT) are an option for larger patients
    - Monitors
        - ASA standard monitors
        - Pre- and post-ductal oxygen saturations to detect shunting and desaturation
        - Consider an arterial line if extensive blood loss or extensive resection is expected
        - Temperature monitoring as well as radiant light and forced air warmer
    - Blood Availability (if indicated)
    - ICU bed Availability (if indicated)
    - Other Indicated Resources (ex. ECMO standby)

Intraoperative Considerations:

- General
    - Intraoperative management will be dictated by surgical timing and severity of presurgical symptoms, and presence of concurrent disease; for example, scheduled elective resection will differ from urgent resection of a neonate in respiratory distress. Here we provide broad guiding principles.
- Induction
    - The choice between inhalation induction and intravenous induction is patient-dependent
    - A rapid sequence intravenous induction may be beneficial to prevent air trapping from a gasping or crying child, minimize positive pressure ventilation, and
    - Should inhalation induction be required/selected, a delayed onset may occur secondary to lung pathology.
    - Nitrous oxide can accumulate in cysts, causing distention, and should be avoided.

    - Positive pressure should be minimized during induction to prevent cyst distention and air trapping.
- Positioning
    - The patient is typically positioned in the lateral decubitus position; this will be accompanied by changes in respiratory physiology and functional residual capacity.
    - Soft padding to avoid nerve injury
    - Difficult access to the patient after surgical draping
    - ETT and endobronchial blockers may be displaced during positioning
    - Repositioning increases the risks of cross-lung contamination
- Maintenance
    - Nitrous oxide can accumulate in cysts, causing distention, and therefore should be avoided.
- Hemodynamic/Physiologic goals
    - One lung ventilation (OLV) is often required, and high peak pressures should be avoided to prevent barotrauma, insufflation of the operative lung, and the risk of cystic inflation.
    - Proximal lesions may require mediastinal manipulation and care must be taken to closely monitor effects on cardiac output.
- Surgical Considerations
    - The degree of resection will be dictated by the cyst's location, while lobectomy might be necessitated for proximal lesions, distal cysts can often be managed by lobectomy or wedge resection.
- Emergence/Disposition
    - Timing of emergence and extubation is dictated by intraoperative and postoperative respiratory parameters; many patients can be extubated immediately after surgery.
    - Early extubation is preferred to minimize PPV to prevent iatrogenic bronchial stump dehiscence.
- Post-op Care
    - Perioperative regional anesthesia, thoracic epidural, ESP block, or cryoablation in supplement to post-operative pain control.
    - Prolonged or substantial postoperative positive pressure ventilation increases the risk of bronchial stump dehiscence.

Case-Specific Complications/Pitfalls

- Positive pressure ventilation can cause rupture of cysts and should be used cautiously or completely avoided with a giant CPAM.
- Large cysts may cause issues with oxygenation and ventilation during positioning.
- Cysts may contain fluid or infectious material, and cross-lung contamination can occur prior to lung isolation.
- Postoperative positive pressure ventilation can result in bronchial stump dehiscence.
- Vigilance for hemodynamic instability, particularly in the setting of intrathoracic surgery in neonatal patients, where large vessels can briefly be compressed or manipulated

References

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2. Mehta PA, SharmaG. Congenital Pulmonary Airway Malformation. StatPearls. https://wwwncbi.nlm.nih.gov/books/NKB551664. [Updated 2022 August 8]. Accessed 4/25/2023.
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