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Delilah Penn
Otterbein University, delilahannpenn@gmail.com

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Type 1 Congenital Pulmonary Airway Malformation: The most common lung lesion in newborns.

Delilah A. Penn, MSN, RN
Otterbein University, Westerville, Ohio

Introduction
- Congenital pulmonary airway malformation (CPAM), previously known as congenital alveolar sac malformation (CASM), is a rare developmental anomaly in the respiratory tract.
- Identified in 1949 by O’Fallon and Tipp, they described a “solid-appearing mass consisting of an adenomatoid or gland-like proliferation of terminal respiratory structures occurring in infants or premature neonates” (O’Fallon-De Cheseau & Stocker, 1991).
- CPAM represents 25% of congenital lung malformations and 60% of congenital malformations (Azizkhan & Tipp, 2002).
- The 1997 Stocker Classification system was created to identify the different types of CPAM. Currently the Stocker Classification system has identified Types 0-4. Although rare, CPAM is the most common congenital lung lesion detected prenatally.
- Due to the effectiveness of prenatal ultrasound screening, CPAM is commonly identified between 16-21 week gestation (Kauratyma & Imam, 2015).
- There are CPAM lesions that are not diagnosed during routine ultrasounds. Infants with CPAM detected antenatally may have their diagnosis confirmed during postnatal management (Azizkhan & Limperis, 2015).
- Large fetal CPAM specimens that were resected displayed “increased cell proliferation and decreased apoptosis compared with gestational age-matched normal fetal lung tissue” (Azizkhan & Limperis, 2000).
- Three genes have been implicated in the pathogenesis of CPAM. The HoxB1, Ptgf and PDGF-B play a critical role in “branching morphogenesis, induction of growth factors, mesenchymal proliferations, and mesenchymo-epithelial interactions” (Azizkhan & Limperis, 2000).

Type 1 CPAM
- composed of single or multiple large cysts, ranging from three to ten centimeters surrounded by smaller cysts
- Predominant cystic type. They are C offenders to one lobe, and are filled with air or fluid.
- Some lesions to communicate with bronchi associated anomalies are rare in this subtype.
- Present during the first week to month of life
- Larger cysts are lined with pseudostriated column epithelium
- Smaller cysts are lined with cuboidal to-epithelial, typically seen in clusters

Pathophysiological Processes
- During pregnancy lung development occurs over the course of 5 distinct stages.
- Embryonic stages, during weeks 4-7, initiate the development of lung buds, branching and vascular development.
- Pseudoglandular stage, during weeks 7-17, consists of the growth and branching of the tracheobronchial tree.
- Terminal branching occurs in the CURNUR stage, weeks 16-21.
- Gas exchange is initiated during the Saccular stage during weeks 24-38.
- Lastly, in the saccular stage terminal alveoli form, and this stage starts at 38 weeks and continues until age 2 (McBride, 2016).
- CPAM develops during the early stages of lung development. Although the exact pathogenesis is unknown, it is suggested that “abnormal events during organogenesis result in the formation” of lung lesions (McBride, 2016).
- Gajewska-Knap & Limperis, 2015, state “there is a failure of maturation of bronchial structures during pseudoglandular stage of lung development. It results in overgrowth of the terminal bronchioles without corresponding alveoli. CPAM have a normal blood supply from the pulmonary arteries and communicating with the tracheobronchial tree (Gajewska-Knap & Limperis, 2015).
- Large fetal CPAM specimens that were resected displayed “increased cell proliferation and decreased apoptosis compared with gestational age-matched normal fetal lung tissue” (Azizkhan & Limperis, 2000).
- Three genes have been implicated in the pathogenesis of CPAM. The HOXB1, Ptgf and PDGF-B play a critical role in “branching morphogenesis, induction of growth factors, mesenchymal proliferations, and mesenchymo-epithelial interactions” (Azizkhan & Limperis, 2000).

Signs and Symptoms
- Antenatal presentation
- CPAM may manifest in a mother who displays polyhydramnios. It is “been proposed that impregnated fetal swallowing due to mass effect on the esophagus, increased fluid production or decreased fluid absorption by the lesion, and secretion of antidiuretic hormone” (Rosaos De Cheseau & Stocker, 1991). Nomineuse Hydros is seen in CPAM and is thought to be due to impaired cardiac contractility and impaired venous return to the heart due to compression of the heart and inferior vena cava by the lesion (Rosaos De Cheseau & Stocker, 1991).
- Postnatal presentation
- The majority of newborns present with respiratory distress immediately following delivery. The infant may display grunting, tachypnea, retractions, and cyanosis. Upon auscultation, the healthcare provider may notice diminished breath sounds and distant or shifted heart sounds (Rosaos De Cheseau & Stocker, 1991).

Clinical Implications
CPAM are increasingly being diagnosed during pregnancy. Accurate diagnosis of lung lesions is important as the treatment of lesions may differ depending on the type. Although most lung lesions are detected during antepartum, some are not, therefore it is imperative that healthcare providers are informed on the presentation and presentation of patients with CPAM. Infants presenting with respiratory distress will require healthcare providers who are trained in stabilization and resuscitation of the newborn. Mechanical ventilation may be required if respiratory distress is severe. Health care providers must be aware of the risk of pneumothorax, pneumopericardium, and pneumomediastinum to patients receiving mechanical ventilation. Frequent assessments of respiratory status is required as well as serial x-rays to monitor the need and level of support (Puligith & Vaupe, 2002).

References

Infant displaying subcutaneous retractions during respiratory distress. (Julee 2008, from newborns.stanford.edu. Photographed by Dr. Janelle Abey

Infant developing subcutaneous retractions during respiratory distress. (Julee 2008, from newborns.stanford.edu. Photographed by Dr. Janelle Abey

CT scan of Type 1 CPAM in right lobe Case courtesy of Dr. Praven Jha, Radiopaedia.org, rID: 8277. http://radiopaedia.org/articles/congenital-pulmonary-airway-malformation

Type of CPAM in right lobe Case courtesy of Dr. Praven Jha, Radiopaedia.org, rID: 8277. http://radiopaedia.org/articles/congenital-pulmonary-airway-malformation