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

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

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Introduction

- Congenital pulmonary airway malformation (CPAM), previously known as congenital adenomatoid malformation (CCAM), is a rare developmental anomaly in the respiratory tract.
- Identified in 1949 by Ch'in and Tang, where they reviewed ten previously reported cases of a "solid-appearing mass consisting of an adenomatoid or gland-like proliferation of terminal respiratory structures occurred in stillborn or premature neonates with hydrops"(Rosado-De-Christenson & Stocker, 1991).
- CPAM "represents 25% of congenital lung malformations and 95% of congenital lung lesions" (Fulghum & Vasquez 2002).
- In 1977 The 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 diagnosed between 18-21 week gestation (Katarzyna & Impey, 2015).
- There are CPAM lesions that are not identified during routine ultrasounds and present later in life.
- According to Katarzyna & Impey, "this lesion occurs more often in males and is primarily unilateral, but may occur bilaterally.
- Most CPAMS are manageable with early diagnosis and intervention. CPAM Stoker type 1 is the most common subtype, represents 60-70% of lesions, and this I why I have chosen to focus on this lesion.

Pathophysiological Processes

- During pregnancy lung development occurs over the course of 5 distinct stages.
- Embryonic stage, during weeks 4-7, initiates 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 Cannicular stage, weeks 16-23.
- Gas exchange is initiated during the Saccular stage during weeks 24-38.
- Lastly, in the Aveolar 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 suspected that "abnormal events during organogenesis result in the formation" of lung lesions (McBride, 2016).
- Gajewska-Knapik & Limpey 2015, state "there is a failure of maturation of bronchiolar 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 communicate with the tracheobronchial tree (Gajewksa-Knapik & Limpey, 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 & Crombleholme 2008).
- Three genes have been implicated in the pathogenesis of CPAM. The HOXB5, Fgf7 and PDGF-B play a critical role in "branching morphogenesis, induction of growth factors, mesenchymal proliferations, and mesenchymal-epithelial interactions"(Azizkhan & Crombleholme 2008).

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 Confined 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 pseudostratified columnar epithelium
- Smaller cysts are lined with cuboidal to cells, typically seen in clusters

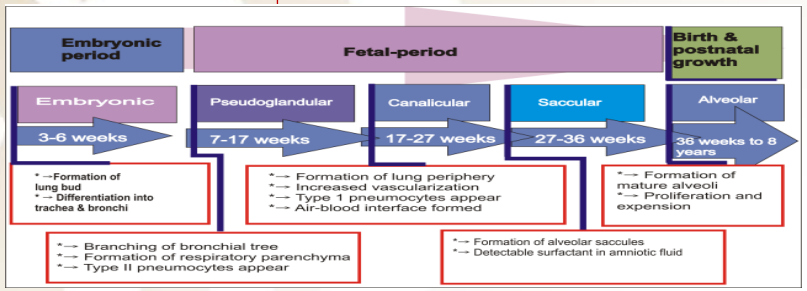


Figure 1 Stages of lung development (Hameed, et al 2013)

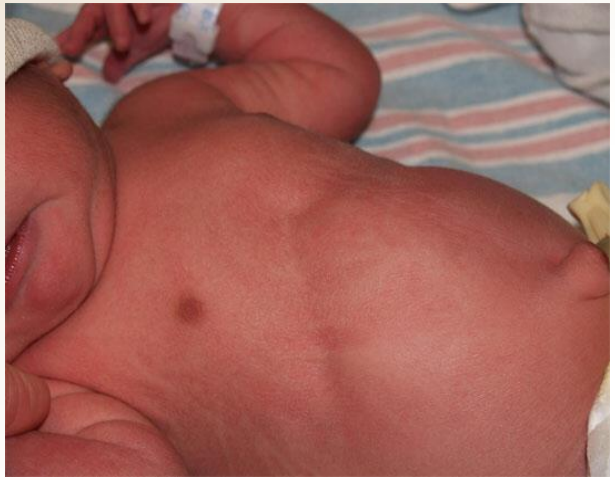
Signs and Symptoms

❖ Antenatal presentation

- CPAM may manifest in a mother who displays polyhydramnios. It is "been proposed that impaired fetal swallowing due to mass effect on the esophagus, increased fluid production or decreased fluid absorption by the lesion, and secretion of antidiuretic hormone (Rosado-de-Christenson & Stocker, 1991). Nonimmune Hydrops 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 (Rosado-de-Christenson & Stoker, 1991).

❖ Postnatal presentation

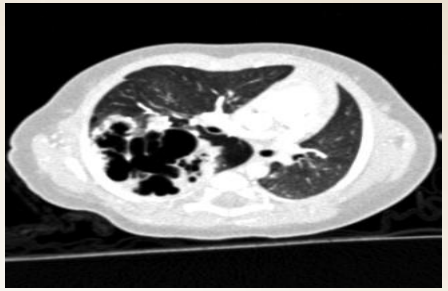
- The majority of newborns present with respiratory distress immediately following delivery. The infant may display grunting, tachypnea, retractions, and cyanosis. Upon auscultation, the healthcare provider may notice diminished breath sounds and distant or shifted heart sounds (Rosado-de-Christenson & Stoker, 1991).



Infant displacing subcostal retractions during respiratory distress. Retrieved July 25th, 2016 from newborns.standford.edu. Photographed by Dr. Janelle Abey

❖ Early Infancy/Childhood/Adulthood

- Infants present with respiratory distress, vomiting, failure to thrive, and recurrent pneumonia. Children and adults typically present with recurrent infection localized to the involved lobe of the lung.(Rosado-de-Christenson & Stoker, 1991).



CT scan of Type 1 CPAM in right lobe Case courtesy of A.Prof Frank Gaillard, Radiopaedia.org, rID: 8277 http://radiopaedia.org/articles/congenital-pulmonary-airway-malformation

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 health care providers be informed of the pathogenesis 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, pnuemopericadiam, and pneumomediastinum in 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 (Fulghum & Vasquez, 2002).

Conclusion

CPAM is a rare congenital anomaly that typically presents in the neonatal period during lung development. Early detection of CPAM usually occurs during routine ultrasounds. Infants with CPAM may present with respiratory distress at birth and the need for prompt respiratory support and immediate surgical intervention. The prognosis of infants who present with CPAM is dependent upon the size of the lesion, the development of healthy lung tissue, and the presence of other anomalies.

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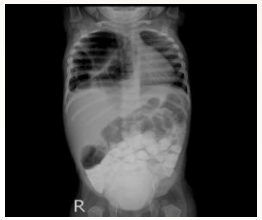

Xray of Type 1 CPAM in right lobe Case courtesy of Dr Praveen Jha, Radiopaedia.org, rID: 2998 http://radiopaedia.org/articles/congenital-pulmonary-airway-malformation

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