

## **A Case Report of Congenital Pulmonary Airway Malformation (CPAM) Stocker's Type III: A Diagnostic Dilemma.**

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### **Abstract**

Congenital pulmonary airway malformation (CPAM) is a rare congenital abnormality that can manifest from prenatal period to childhood or uncommonly to adulthood. CPAM results from proliferation of terminal bronchiolar structures forming multicystic lung mass suppressing the alveolar growth<sup>5</sup>. It manifests as neonatal respiratory distress due to progressive expansion of the affected lung. The rarest type of CPAM along with a poorer prognosis is Stocker's type III, as it usually involves the entire lobe of the affected lung. We present a case of an infant who had respiratory distress post-delivery. The clinical presentation and imaging features raise a diagnostic dilemma between CPAM, oesophageal lung and pulmonary sequestration. The baby was eventually diagnosed with type III CPAM, was managed appropriately, and showed clinical improvement afterward. Type III CPAM, the rarest in Stocker's classification, poses a potential threat due to acute respiratory distress. It can be mistaken for other conditions like consolidation or pulmonary sequestration. When faced with persistent consolidation, consider type III CPAM as a possible differential diagnosis.

### **Keywords**

Congenital pulmonary anomalies, Congenital pulmonary airway malformation (CPAM), Stocker's classification, Pulmonary sequestration, Oesophageal lung.

### **Introduction**

Congenital pulmonary airway malformation (CPAM) is a rare congenital abnormality that constitutes about 25% of all congenital pulmonary anomalies, with the incidence reported as 1 in 10000 to 1 in 35000 births<sup>1,2</sup>. In 1977, Stocker et al classified CPAM into three types which further added another two subtypes in 2009. The categories of CPAM are different from others in the origin of the cystic lesion, its size, and its

microscopic appearance. Type III CPAM is the rarest type of this malformation with 8 % frequency<sup>10</sup>. It can occur at different stages of lung development with no known aetiology or maternal factor association.<sup>1</sup> CPAM mostly affects the newborn causing acute progressive respiratory distress being the main reason for early detection of the disease for further appropriate management. As CPAM is a rare congenital disorder, especially type III CPAM, the diagnosis could easily be missed. Therefore, this article aims to raise awareness of this diagnosis and discuss the possible differential diagnosis that may resemble features of CPAM.

### Case Presentation

A 1.0 kg female baby was born as late preterm at 36 weeks via emergency caesarian section due to severe pre-eclampsia and fetal intrauterine growth restriction (IUGR). The baby was delivered vigorously with a good Apgar score. However, she developed acidotic breathing and peripheral cyanosis at 2 hours of life, which required intubation and intensive care admission. Lung surfactant (Survanta) was given at 2 hours of life.

Antenatally, there were suspicions of CPAM features on her initial detail ultrasound scan but were not visualized on her follow-up scan. Her blood TORCHES screening was positive for CMV and Rubella IgG. A serial chest radiograph (Figure 1) showed persistent right homogenous opacity at the right lower and middle lobe with air bronchogram in the right upper lobe. Due to persistent consolidation in the serial chest radiograph and failure to wean down oxygen from the high-flow oscillation ventilator to conventional ventilation, she was then planned for CT Thorax.

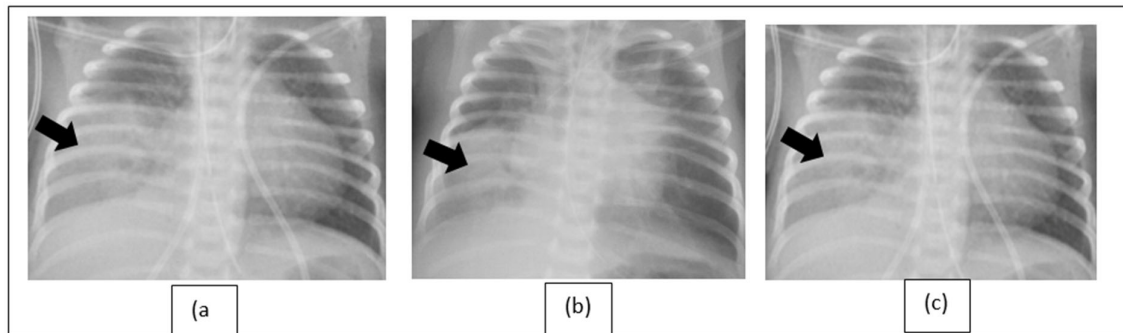


Figure 1: Serial chest radiograph portable AP supine showing homogenous opacity at middle and lower lobe of right lung (black arrow), consistent with consolidation.

Following that, CT Thorax was done and revealed consolidative changes in the right upper lobe (Figure 2). The arterial supply for the solid right lower lobe was from the pulmonary artery (Figure 3). CT findings suggested the right bronchus intermedius was not visualized with dilated middle and lower lobe airways with soft tissue appearance of right lower lobe which might be suggestive of esophageal lung. However, in the Upper GI study, no communication between the esophagus and bronchopulmonary tree was noted.

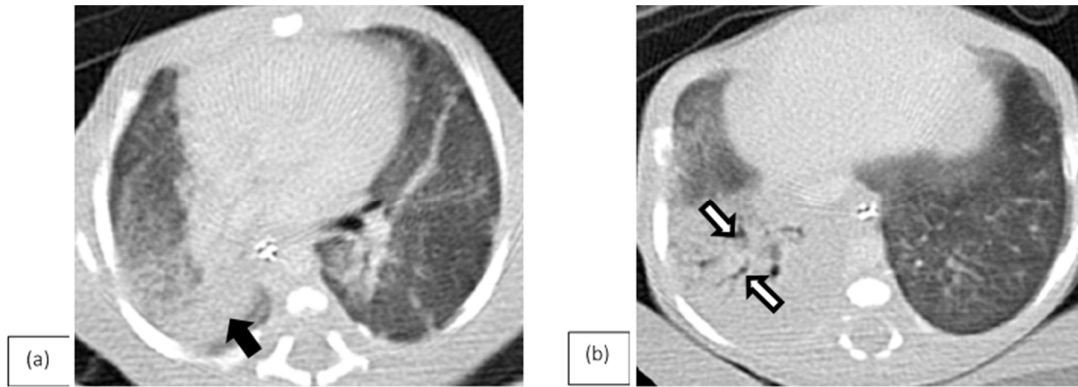


Figure 2: Axial section of CT Thorax. (a) Collapse consolidation (black arrow) in the right upper lobe, mainly involving apical and posterior segment. (b) Reduced volume of right lower lobe with soft tissue density/attenuation, with presence of air bronchogram (white arrow).

The respiratory team was consulted and CPAM was put as the provisional diagnosis with the differential including pulmonary sequestration and oesophageal lung. A consensus diagnosis of CPAM was made and the patient was subjected to right thoracotomy and right upper lobe lobectomy. Intraoperative findings showed right upper lobe consolidation, firm to touch with no obvious cystic appearance, that was not expanding on manual bagging. Right upper lobe lobectomy was done. The sample was sent for HPE and demonstrated features of type III CPAM, complicated with moderate to severe acute and chronic inflammation and exudation. Post-operatively the baby showed steady progressive recovery and was eventually discharged with oxygen support. Periodic follow-up has been carried out since then to monitor the patient's condition.

### Discussion

Congenital pulmonary airway malformation (CPAM) has been categorized from type 0 to type IV, by Stocker's criteria in 2009.<sup>2</sup> Type 0 involves the primary bronchi or trachea. It is very rare and incompatible with life. Type I is associated with large (up to 10 cm) cystic lesions of the bronchi, being the most prevalent and most favorable prognosis, contributing to about 60–70% of cases. Type II is characterized by bronchiolar involvement with many small cysts (<2 cm), involving the entire lobe. Type III is the rarest with a prevalence about 8 % of all cases and is associated with the poorest prognosis.<sup>10</sup> It is associated with large lung masses which may contain very small cysts (<5 mm) with no discernible cystic spaces, involving distal bronchiolar and alveolar regions. Finally, type IV is associated with the most distal acinar region of the lung, with large cysts similar to type I. Type I, II and III of CPAMs are distinguished at imaging. Type IV usually appears as a large cyst and is indistinguishable from type I in imaging per se.

The diagnosis of CPAM can be suspected from prenatal ultrasound, postnatal CT or MR imaging, however, the mainstay of diagnosis is from histopathology. Diagnosis of CPAM can be established prenatally, most often in the second trimester. It can vary from an isolated cystic or solid intrathoracic mass appearance. The lesion may remain stable in size, or even regress upon follow-up ultrasound.<sup>4</sup> The prenatal ultrasound in our case initially showed features of CPAM, however, it was not seen on subsequent ultrasound which made the diagnosis a bit challenging. This could be due to partial regression which occurs in about 41% of cases, that causes changes in appearance during subsequent ultrasound follow-up. However, the operator factor can be one of the reasons why the lesion was not detected upon follow-up scan. Postnatally, the chest radiograph could help in achieving the diagnosis. However, in our case, the radiograph showed features of

infective lung changes as the lesion appears as a consolidation of the right lower zone. Features of type III CPAM in chest radiographs usually will appear as large and homogenous intrathoracic opacity, sometimes mistakenly considered as consolidation or a mass.<sup>5</sup> It is difficult to differentiate between type III CPAM consolidation from infective consolidation in radiographs per se. CT scan can depict the internal structure of the affected lung to differentiate between type III CPAM and infective consolidation. In type III lesion, it will show a solid homogeneous mass involving the lobe or even the entire lung, leading to mediastinal shift and compression of adjacent lung in some cases.<sup>6</sup> The involvement of mediastinal shift and compression of adjacent lung usually favors type III CPAM instead of infective consolidation. In this case, the CT scan showed collapsed consolidation of the right upper lobe, however, no mediastinal shift was reported. There was also a non-visualization of the right bronchus intermedius with the solid right lower lobe mass, giving suspicions of bronchopulmonary foregut malformation (oesophageal lung and pulmonary sequestration).

Pulmonary sequestration may appear as type III CPAM in terms of radiological findings, however, in this condition, there is no lung tissue connection with the bronchus, which can be evaluated in CT scan. Evaluation of blood supply to the masses helps to distinguish between these two entities. Pulmonary sequestration is characterized by a portion of the lung that does not connect to the tracheobronchial tree and has a systemic arterial supply, usually from the thoracic or abdominal aorta.<sup>7</sup> Unlike in our case where the arterial supply for the solid right lower lobe mass is from the pulmonary artery (Figure 3). There was a case of pulmonary sequestration reported where the radiological findings were similar to CPAM type III but different in blood supply origin of the solid intrathoracic mass.<sup>8</sup>

Oesophageal lung also falls into bronchopulmonary foregut malformation and is a very rare congenital malformation. It manifests as a connection between oesophagus and bronchial tube. In a condition where the main bronchus originates from the oesophagus, it is termed an oesophageal lung, and if the lobar bronchus arises from the oesophagus it is described as an oesophageal bronchus. An Upper GI study can be done to reveal the connection between oesophagus and bronchial tree.<sup>9</sup> In our case, the upper GI study was able to rule out oesophageal lung as there was no communication demonstrated. Despite differences in pathogenesis, all these three conditions, namely CPAM, pulmonary sequestration and oesophageal lung are traditionally managed by surgical resection.

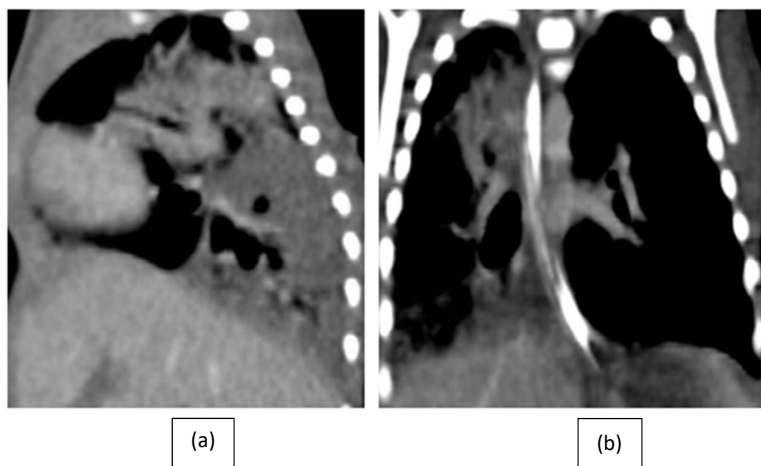


Figure 3 (a&b): Coronal and sagittal plane CT Thorax demonstrate arterial supply for solid right lower lobe is from the pulmonary artery (white arrow).

Achieving the diagnosis of type III CPAM is challenging for physicians as it can mimic other conditions such as pulmonary sequestration or oesophageal lung as in our case. Type III CPAM also may manifest as consolidation which can mislead the diagnosis and cause hindrance in treatment. Knowledge regarding these conditions is crucial in differentiating CPAM from others. By that early diagnosis and prompt management will be executed.

The prognosis of CPAM essentially depends on the type of lesion. The type III lesion carries a poorer prognosis, as it is usually large and presents early with respiratory distress and cardiovascular compromise. The involvement of bilateral lungs, association with other congenital anomalies, and presence of hydrops become the factors that contribute to the poor prognosis of CPAM.<sup>7</sup> Survival rates have been shown to increase after surgical resection of the associated lesion.<sup>1</sup> Satisfactory recovery in our case is likely due to single lobe involvement and no association with other congenital anomalies.

### Conclusion

Type III CPAM being the rarest type in Stocker's classification is a potentially life-threatening condition as it frequently presents with acute progressive respiratory distress. Fortunately, it is reversible if detected early and prompt treatment is carried out. When encountering a case that presents with persistent consolidation, type III CPAM can be considered as one of the differential diagnoses as it can also manifest as consolidation.

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