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# Early antenatal sonographic assessment of Type III Congenital Pulmonary Airway Malformation

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## ABSTRACT

Prenatal monitoring has led to an increased diagnosis of lung problems during pregnancy. In the past decade, advances in prenatal imaging have transformed the way congenital cystic lung anomalies are identified, evaluated, and treated. Previously, these anomalies were believed to be solely under the jurisdiction of the surgeon, who was authorized to perform surgery on any cystic lung anomalies, regardless of their size or clinical symptoms, in order to decrease the risk of cancer and promote lung expansion, even in newborns that showed no symptoms. However, given that many of these lesions may improve spontaneously over months to years and potentially resolve without intervention, clinicians are now reconsidering this approach and contemplating a more conservative approach for many infants who exhibit minimal or no symptoms in the early months. We present the case of a 28-year-old woman who underwent a TIFFA scan. Antenatal ultrasound detected a single intrauterine live pregnancy at an average gestational age of 22.4 weeks, and the fetus was diagnosed with CCAM.

**Keywords:** Prenatal monitoring, Lung problems, Congenital cystic lung anomalies, Conservative approach, CCAM (congenital cystic adenomatoid malformation)

## 1. INTRODUCTION

Congenital Pulmonary Airway Malformations (CPAM) is multi-cystic masses of segmental lung tissue that display abnormal bronchial growth. These malformations are believed to be one of the various bronchopulmonary foregut abnormalities. They are also referred to as Congenital Cystic Adenomatoid Malformations (CCAM), a name that encompasses both the cystic and adenomatoid subtypes. CCAM has five distinct subtypes, with type I accounting for 70% of cases. This subtype is characterized by giant cysts surrounded by smaller ones (Gross, 1992).

Type II, the second most common subtype, features one or more dominant cysts. Approximately 15% of patients with CCAM also have other

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systemic abnormalities, such as renal agenesis and dysgenesis, pulmonary sequestration, and cardiac anomalies. Type III, which 7 comprises less than 10% of cases is made up of small cysts with a diameter of less than 5 mm that frequently affect the entire lobe and have a poor prognosis. Type IV affects a single lobe, and it is difficult to distinguish from Type I on imaging. The majority of type IV lesions consist of lined cysts. The rarest subtype, type 0, has poor postnatal outcomes, with lung development being completely halted (Beers and Moodley, 2017; Kao et al., 2011).

## 2. CASE PRESENTATION

A 28-year-old woman who is currently pregnant with her second child and has already given birth once visited the antenatal outpatient department for her routine antenatal checkup. The patient reported a gestational age of 21 weeks based on an abdominal examination and 22.4 weeks based on her last menstrual period (LMP). To check for any anomalies, it was suggested that she have an obstetric sonogram called a TIFFA (Targeted Imaging for Foetal Anomalies) scan. During the obstetric sonography, a curvilinear curved array transducer with a frequency range of 1.0 - 5.0 MHz was used to obtain images of the fetus.

The ultra-sonogram indicated a single, viable foetus in the uterus that was 21 weeks and 5 days gestational age and weighed 468 grams on average. The gray scale axial and sagittal images of the fetal thorax showed asymmetrical lung shadows, with the left lung appearing larger and more reflective compared to the right lung. There was also a mediastinal shift to the right side (Figure 1, 2). Multiple small cystic lesions, less than 5mm in size, were present throughout the left lung parenchyma, causing enlargement and hyper-echogenicity. These features are indicative of Type III CPAM (Congenital Pulmonary Airway Malformation).

Other potential diagnoses considered were bronchopulmonary sequestration, congenital diaphragmatic hernia, and congenital lobar emphysema. The fetal heart, however, was showing a normal cardiac axis and cardio-thoracic ratio (Figure 3, 4). The patient was informed about the potential risks and complications of the condition during pregnancy and the potential outcomes of the fetus after birth.

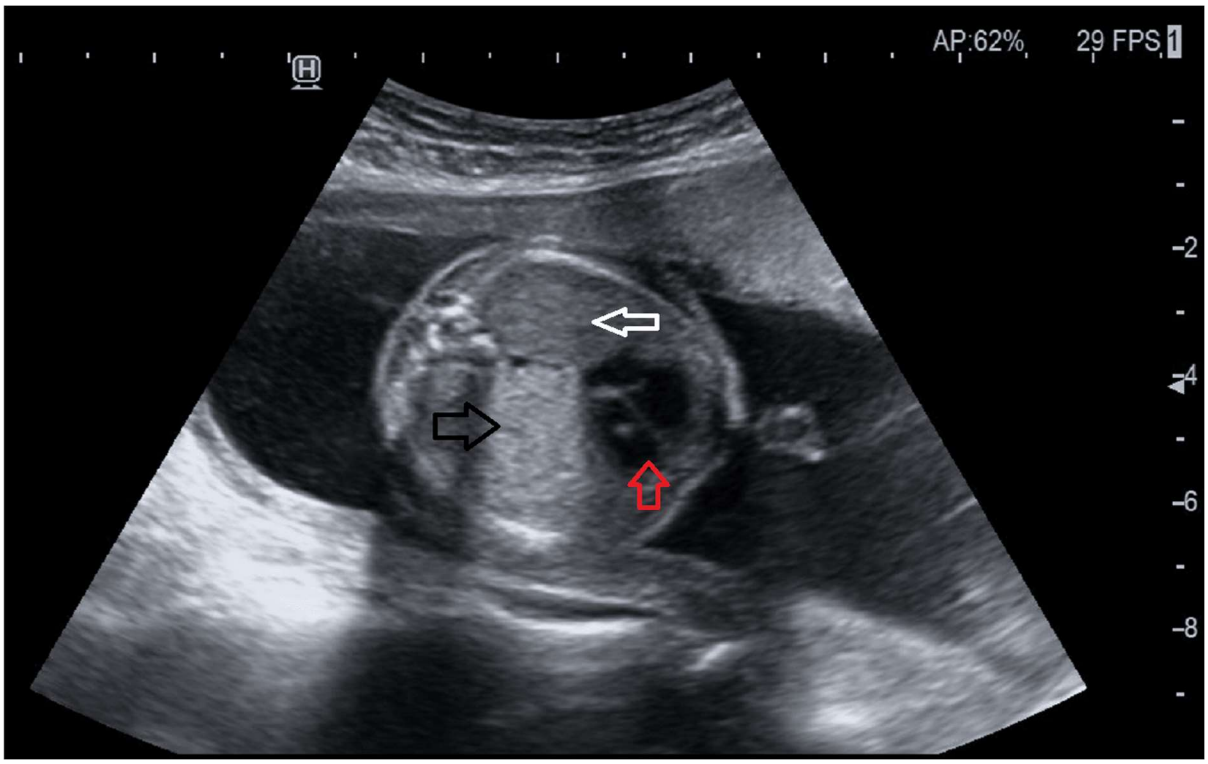


**Figure 1** Transabdominal antenatal ultrasonographic axial section at the level of thorax showing asymmetric lung parenchyma, left lung appearing hyperechoic and large (black arrow) compared to right lung (white arrow) with mediastinal shift to the right side (red arrow)

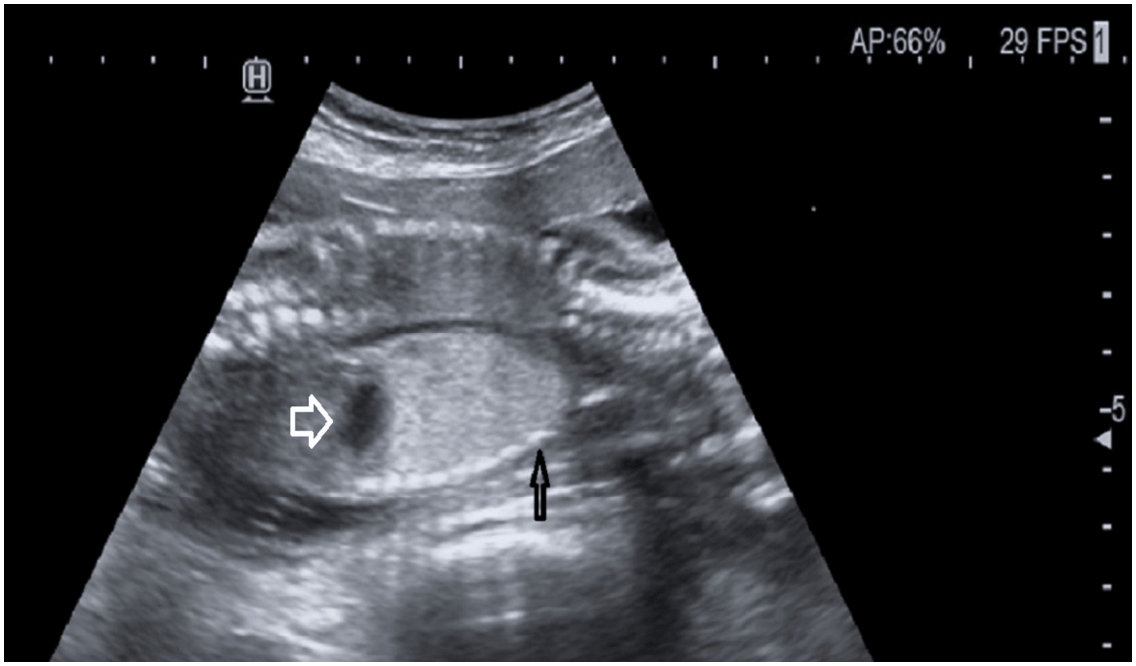


**Figure 2** Antenatal sonography sagittal image of the thorax and upper abdomen showing hyperechoic and enlarged left lung (suggestive of microcysts <5mm) (black arrow) with stomach bubble inferiorly (white arrow)

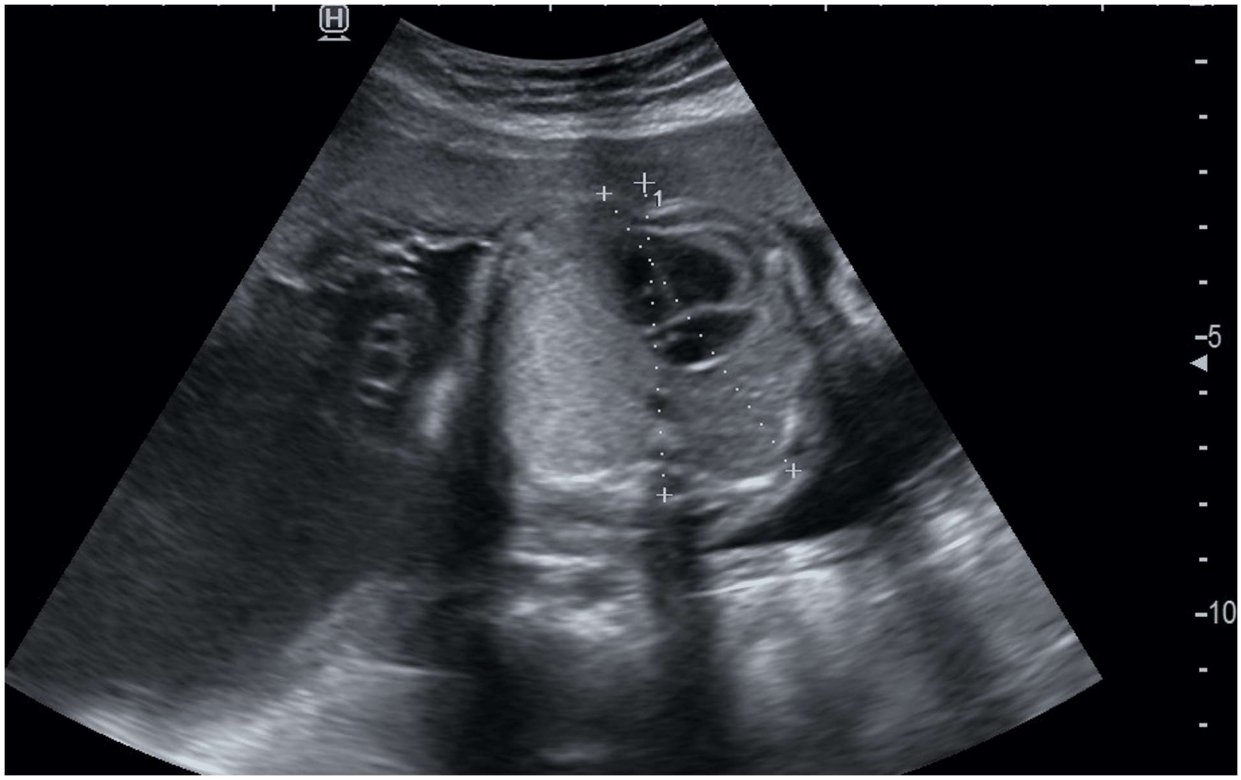


**Figure 3** Axial Ultrasound image at the level of four-chamber view of the heart showing a normal fetal cardiac axis of 23 degrees

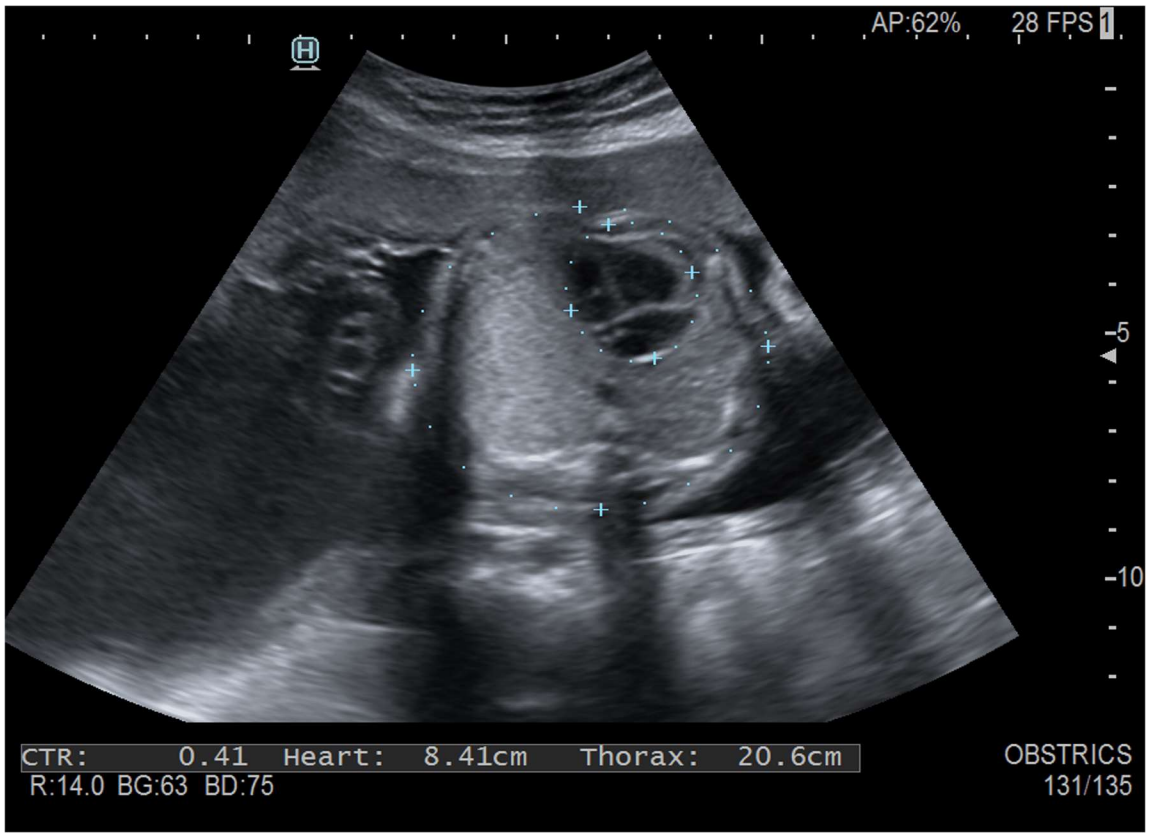


**Figure 4** The sonographic image displays a four-chamber view of the fetal heart in an axial plane, demonstrating a healthy cardiothoracic ratio of 0.4

## 3. DISCUSSION

The development of lungs can be classified into two distinct phases: Initial development and ongoing development. These phases can be further subdivided into five stages, namely embryonic, pseudo glandular, saccular, canalicular, and alveolar stages. The respiratory tract emerges from the primordial gut tube during embryonic development. During the early embryonic stage, lateral folding occurs, resulting in the formation of the gut tube. In the fourth week, an outpouching from the gut tube gives rise to the respiratory diverticulum (Moore et al., 2018).

A septum then develops between the gut tube and respiratory diverticulum, referred to as the tracheoesophageal diverticulum. The respiratory diverticulum continues to divide and produces two buds that eventually form the left and right main bronchi. These, in turn, divide further to form the secondary and tertiary bronchi (Skandalakis, 1994). During the pseudo glandular phase of fetal lung development, occurring between 8 and 16 weeks, bronchopulmonary segments are generated in conjunction with a tertiary bronchus. Nevertheless, at this stage, the lungs are incapable of oxygenating blood since alveoli have not yet formed.

However, the lungs have the ability to acquire oxygen by virtue of the ductus arteriosus, which diverts blood to the aortic arch from the pulmonary artery. Moving on to the canalicular stage, which occurs between 16 and 25 weeks, terminal bronchioles are formed and begin to transition into respiratory bronchioles. Finally, during the terminal stage, alveoli start to develop and two types of cells are formed. The first type, known as type I pneumocytes, are thin, flat epithelial cells, while the second type, type II cells, produce surfactants (Beers and Moodley, 2017).

The condition is attributed to the aberrant growth of terminal respiratory units in a hamartoma-like configuration, impeding the normal development of bronchoalveolar structures. Histologically, these growths exhibit an adenomatoid proliferation of bronchiole like structures and the presence of macro- or micro cysts lined with columnar or cuboidal epithelium, lacking cartilage and bronchial glands. In contrast to bronchogenic cysts, these lesions exhibit connections to the tracheobronchial tree and possess intra-cystic connections. During ultrasound examination, the fetal lungs typically manifest as homogeneous structures with lower echogenicity compared to the liver.

However, the echogenicity of the lungs progressively increases with advancing gestational weeks. Notably, the lung parenchyma may exhibit cystic formations or localized areas of heightened echogenicity indicative of a mass. On the four-chamber scan, the heart can be seen in the left anterior quadrant, positioned to the left of the midline, where it takes up about 25% to 30% of

the thoracic cavity. The orientation of the heart is determined by the interventricular septum, which forms a 45° angle with the midline (Goldstein, 2006).

A cardio mediastinal shift can often identify a unilateral chest mass or diaphragmatic hernia for the first time. Therefore, all foetuses should undergo routine obstetric imaging throughout the second and third trimesters to get a four-chamber image of the heart (Walker et al., 2001). Fetal lung volumes can be evaluated using three- and four-dimensional ultrasound (US) techniques to determine residual lung capacity in fetuses with lung abnormalities (Achiron et al., 2008).

CPAMs can manifest in various forms depending on their type. CPAMs, or isolated cystic or solid intrathoracic masses, can be observed during prenatal ultrasound studies. A thoracic mass typically indicates a solid, hyperechoic type III CPAM. A mass effect could result in the heart appearing to have moved to the opposite side. Due to the increased use of prenatal sonography, CPAMs are frequently identified before birth. The CVR (head circumference to lesion size ratio) is computed during sonography to determine the probability of fetal hydrops and death.

A CVR > 1.6 significantly increases the risk of fetal hydrops. The lesion's shape is used to calculate the fetal lung volume and validate the diagnosis. Fetal MRI is routinely employed alongside sonographic examinations to confirm the diagnosis, determine the lesion's morphology and assess the volume of the developing lungs (Kotecha et al., 2012). Alternatively, the lesion has the potential to either increase in size or remain unchanged. Additional sonographic features that can be detected through ultrasonography include the emergence of hydrops fetalis and polyhydramnios.

Type I and Type II CPAMs may show an air-filled multicystic lesion on a plain radiograph. In cases of large lesions, a mass effect may occur, leading to diaphragmatic inversion, depression, or mediastinal displacement. The cysts inside the lesion may be totally or partially filled with fluid during the early neonatal period, giving the lesion a solid look or the existence of air-fluid levels. Interval imaging can reveal any changes in the size of the lesion over time. Type III lesions typically exhibit a solid composition. CT imaging plays a significant role in the management of CPAM, both in childhood and adulthood, serving various purposes. Initially, the precise localization and measurements of the lesion are ascertained.

In the scenario of patients eligible for surgery, the recognition of a systemic arterial supply holds paramount importance, and this can be accomplished via CT angiography. It is noteworthy that consolidation may arise due to an underlying type III lesion, as the visual representation mirrors the underlying type. Prenatal MR imaging reveals that sizable cystic congenital pulmonary airway malformations exhibit hyperintense unilocular or multilocular abnormalities with well-defined boundaries on T2-weighted images (Daltro et al., 2010).

## 4. CONCLUSIONS

The capability to identify thoracic anomalies during pregnancy is improving thanks to developments in fetal Ultra-sonogram (USG) and magnetic resonance imaging (MRI). This enhanced diagnostic capability enables medical personnel to anticipate potential management challenges during birth or during the neonatal stage, as well as facilitates parents' understanding of the prognosis. Consequently, it is imperative for the healthcare providers involved in the care of the expectant mother to possess knowledge regarding the imaging findings related to various intrathoracic conditions in the fetus. Timely identification of congenital pulmonary airway malformation (CPAM) and access to comprehensive information regarding its characteristics and severity also empower parents and healthcare professionals to make well-informed decisions regarding the fetus's future.

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**Informed consent**

Informed consent was obtained from the patient.

**Author's contribution**

All the authors contributed equally to the case report

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**Conflict of interest**

The authors declare that there is no conflict of interests.

**Data and materials availability**

All data sets collected during this study are available upon reasonable request from the corresponding author.

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