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#### Abstract:

Introduction: Congenital Pulmonary Airway Malformation (CPAM) is a rare fetal lung anomaly resulting from abnormal pulmonary development. It is typically detected during prenatal ultrasounds and classified into five types based on cyst size and histological characteristics. Type 2 CPAM presents as multiple small cysts (<2 cm) and is often associated with a favorable prognosis when hydrops is absent. Discussion: This case involves a 36-year-old pregnant woman whose fetus was diagnosed with type 2 CPAM at 19 weeks. Despite delayed referral, the fetus showed no signs of hydrops or hemodynamic compromise. Corticosteroid therapy with betamethasone was administered at 31 weeks, and follow-up imaging revealed complete regression of the lesion. The low CPAM volume ratio (CVR 0.12) and absence of complications supported conservative management. Conclusion: This case highlights the effectiveness of corticosteroid



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# Pulmonary Cystic Adenomatoid Malformation Type 2: First Case Report in Floridablanca, Santander, Colombia

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## Abstract

Congenital Pulmonary Airway Malformation (CPAM) is a rare fetal lung anomaly resulting from abnormal pulmonary development. It is typically detected during prenatal ultrasounds and classified into five types based on cyst size and histological characteristics. Type 2 CPAM presents as multiple small cysts (<2 cm) and is often associated with a favorable prognosis when hydrops is absent. This case involves a 36-year-old pregnant woman whose fetus was diagnosed with type 2 CPAM at 19 weeks. Despite delayed referral, the fetus showed no signs of hydrops or hemodynamic compromise. Corticosteroid therapy with betamethasone was administered at 31 weeks, and follow-up imaging revealed complete regression of the lesion. The low CPAM volume ratio (CVR 0.12) and absence of complications supported conservative management. This case highlights the effectiveness of corticosteroid therapy for type 2 CPAM and underscores the importance of early diagnosis, serial ultrasounds, and specialized perinatal care for optimal fetal outcomes.

**Keywords:** CPAM, malformation; cysts, Stocker's classification; bronchopulmonary sequestration; CPAM Volume Ratio; corticosteroids; hydrops;ultrasound.





## Resumen

La Malformación Adenomatoidea Quística Congénita (CPAM) es una rara anomalía pulmonar fetal causada por un desarrollo pulmonar anómalo. Generalmente se detecta mediante ecografía prenatal y se clasifica en cinco tipos según el tamaño del quiste y las características histológicas. La CPAM tipo 2 se presenta como múltiples quistes pequeños (<2 cm) y suele tener un pronóstico favorable cuando no hay hidrops fetal. Este caso corresponde a una gestante de 36 años cuyo feto fue diagnosticado con CPAM tipo 2 a las 19 semanas. A pesar del retraso en la remisión, el feto no presentó signos de hidrops ni compromiso hemodinámico. Se administró betametasona a las 31 semanas y las ecografías de seguimiento evidenciaron la resolución completa de la lesión. El bajo índice de volumen CPAM (CVR 0.12) y la ausencia de complicaciones respaldaron el manejo conservador. Este caso resalta la eficacia de la terapia con corticosteroides en CPAM tipo 2 y subraya la importancia del diagnóstico precoz, ecografías seriadas y atención perinatal especializada para lograr mejores resultados fetales.

**Palabras clave:** CPAM, malformación, quistes, clasificación de Stocker, secuestro broncopulmonar, índice de volumen de CPAM, corticoides, hidropesia, ecografía





## Introduction

Congenital Pulmonary Airway malformation (CPAM), formerly known as Congenital Cystic Adenomatoid Malformation (CCAM), is a rare pulmonary malformation. It is the most common lung malformation in children and is generally detected in newborns and infants. This defect results from arrested lung development during the early fetal stage (6–15 weeks of gestation), leading to hypertrophy of bronchi, bronchioles, and mesenchymal tissue, and failure in alveolar development<sup>5</sup>. Detection of this malformation is usually based on symptoms such as dyspnea during the neonatal period or later in life. However, it is important to note that prenatal diagnosis is possible through ultrasonography and accurate recognition of the condition.

CPAM is a lower airway malformation with an incidence as low as 1 in 35,000 births<sup>2</sup>. Literature indicates that larger cysts are the most common, accounting for 70% of cases, with an incidence of 8 in 100,000 live births<sup>2</sup>. Studies report that this malformation predominantly occurs in males.

The genetic background of CPAM remains unknown, but associations have been suggested with various genes, including TTF-1, FABP-7, FGF-7, FGF-9, FGF-10, Hoxb-5, and SOX2. Abnormal expression of Hoxb-5 during human pulmonary branching morphogenesis has been implicated in the development of CPAM<sup>17</sup>.

Since 10% to 20% of fetuses with CPAM present with associated congenital anomalies, a comprehensive fetal evaluation is essential. The most common associated anomalies are cardiovascular and urogenital, although others have been reported, such as tracheoesophageal fistula, cleft lip and palate, and anomalies of the diaphragm, central nervous system, bones, and other pulmonary malformations<sup>7</sup>.

These lesions are observed in 80–85% of full-term and preterm neonates, as this condition is frequently diagnosed during the postnatal period. They are rarely seen in children between the ages of 2 and 10, as well as in adolescents and adults. CPAM usually presents unilaterally and is confined to the lower lobe, with rare occurrences in the left lower lobe<sup>4</sup>. A tendency for spontaneous resolution of microcystic types of CCAM has been reported, usually after 28 weeks of gestation. However, postnatal regression of the lesions is unlikely<sup>5</sup>.



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There are five histological classifications according to Stocker, ranging from 0 to 4, categorizing them as malformations related to abnormalities in fetal pulmonary branching morphogenesis<sup>3</sup>. Like most malformations, they originate from an imbalance between cell proliferation and apoptosis during organogenesis.

Stocker's classification (Table 1) is as follows: Type 0: Solid tissue affecting both lungs due to congenital acinar dysplasia, representing only 1% to 3% of cases; Type 1: Single or multiple large cysts (>2 cm), the most common form of CPAM, representing 60% to 70% of cases; Type 2: Multiple small cysts (<2 cm), accounting for 15% to 20% of CPAM cases and receiving systemic blood supply, Type 3: A solid mass, comprising 5% to 10% of CPAM cases, with an acinar origin and consisting of adenomatoid proliferation of distal airways or air spaces; Type 4: A large peripheral cyst also known as pleuropulmonary blastoma (PPB) type 1, with high malignant potential. It accounts for 5% to 10% of CPAM-type malformations. Types 0 and 4 are seen almost exclusively in infants<sup>1</sup>.

Fetal assessments are carried out during the prenatal and postnatal periods. Prenatal evaluations include the CPAM volume ratio (CVR)<sup>8</sup>, the mass-to-thorax ratio (MTR), the lesion-to-lung volume ratio (LLV), and the cardiomediastinal shift angle (CMSA)<sup>9</sup>. On the other hand, postnatal assessments may involve evaluating features through chest X-ray, thoracic ultrasound, and echocardiography, especially in fetuses diagnosed with hydrops. Additional assessments include evaluating the size of cardiac chambers, valvular regurgitation, ventricular contractility and function, and Doppler examination of the inferior vena cava, ductus venosus, and umbilical vein<sup>10-11</sup>.

When these intraparenchymal lung lesions are present, complications such as mediastinal shift to the contralateral side, possibly with cardiac rotation, may occur. Special attention should be paid to mediastinal deviation, as a severe shift that compresses the inferior vena cava (IVC) and the heart can lead to hydrops<sup>12</sup>.

There are differential diagnoses, among the most important are mediastinal cystic teratoma, pleuropulmonary blastoma (PPB), congenital peribronchial myofibroblastic tumor, and fetal pulmonary interstitial tumor (FPIT)<sup>14</sup>. Congenital diaphragmatic hernia can also mimic a multicystic and heterogeneous pulmonary mass, and may cause mediastinal and cardiac deviation. Pulmonary sequestration, also known as bronchopulmonary sequestration (BPS), differs from CPAM in that BPS does not communicate with the tracheobronchial tree. Another distinction is that BPS has anomalous systemic feeding and draining vessels, which can often be identified







through color Doppler imaging, whereas blood flow to CPAM occurs via the normal pulmonary vasculature<sup>12-13</sup>

The prenatal course of CPAM lesions has been reported to show that most exhibit rapid progressive growth in most cases from approximately 20 to 26 weeks of gestation, peaking around 25 weeks, and subsequently stabilizing and often regressing<sup>15</sup>. It has been determined by ultrasonography that before birth, in 50% of cases, the lesion may resolve, usually in those with a microcystic/solid appearance and a low CPAM volume ratio (CVR)<sup>16</sup>.

Treatment is primarily conservative in most cases; however, in some instances, the use of betamethasone<sup>18</sup>, cyst drainage, or thoracoamniotic shunting may be necessary, especially when the mass causes hydrops. First-line therapy includes steroids before 32 weeks of gestation in hydropic fetuses or in fetuses at risk of developing hydrops due to a CPAM volume ratio (CVR) >1.6. The therapeutic mechanism for the resolution of hydrops is unknown, but it may be related to accelerated lung maturation or steroid-induced involution of the mass<sup>19</sup>. Second-line in utero therapy is considered in fetuses <32 weeks with hydrops or at risk of developing it (CVR >1.6) that do not respond to steroids, invasive intervention is the next option. The choice of the best invasive approach depends on the type of lesion (macro- or microcystic), and the approach may involve cyst aspiration<sup>19</sup>, thoracocentesis<sup>20</sup>, thoracoamniotic shunting<sup>21</sup>, ethanol ablation, open resection<sup>22</sup>, or the ex utero intrapartum therapy (EXIT) procedure<sup>23-24-25</sup>, which has a survival rate of approximately 90%, although maternal bleeding risk is higher and 13% of mothers require transfusion<sup>26</sup>. On the other hand, postnatal therapy prioritizes surgical intervention as the first-line treatment, depending on the severity of symptoms<sup>27</sup>.

The initial evaluation of CPAM should include the assessment for associated congenital anomalies. Although its genetic origin is uncertain, there may be links with cancer-related genes. Microarray testing is recommended, and in complex cases, whole exome or genome sequencing may be indicated. Echocardiography and magnetic resonance imaging (MRI) are useful tools for evaluating fetal anatomy and prognosis. Serial ultrasounds every 1 to 4 weeks are suggested, depending on the CVR index and gestational age. Most masses grow until week 25, after which they tend to stabilize or regress. If hydrops does not occur before week 28, the prognosis is usually excellent.





## **Clinical case**

36-year-old female patient, with no significant medical history, obstetric formula G4P1C2V3, with a last menstrual period (LMP) dated August 18, 2024, the current pregnancy is with a new sexual partner, the patient weighs 80.9 kg, is 1.58 meters tall, with a body mass index (BMI) of 32. She attended her prenatal check-up at 6 weeks of gestation, where a positive B-HCG confirmed the pregnancy, she began taking multivitamins with good adherence. The first trimester TORCH panel results were: HIV negative, HBsAg negative, non-reactive syphilis test, and blood type O positive. Her first ultrasound, dated November 28, 2024, reported: a single live fetus, in a longitudinal breech position with the back on the left side, showing frequent spontaneous body and respiratory movements. The umbilical cord contains two arteries and one vein. The placenta is located posteriorly, classified as grade II/III, and the amniotic fluid index is 9 cm. No abnormalities were identified in the central nervous system, genitourinary tract, or gastrointestinal tract. Biometry for 15 weeks of gestation estimated a fetal weight of 109 grams.

The patient was scheduled for a second detailed anatomical ultrasound on December 30, 2024, with a gestational age of 19 weeks based on the last menstrual period (LMP).

The ultrasound result showed a single live fetus with biometric measurements corresponding to 19.4 weeks, with an estimated fetal weight of 303 grams, growing at the 46th percentile. The fetus is male. The thorax presented a mixed echogenic, heterogeneous area, predominantly solid with anechoic regions, located at the base of the right lung, measuring 15 x12 mm (*Picture 1*). No color Doppler signal was observed, and there was no mass effect or mediastinal shift. No findings of diaphragmatic hernia or other additional anomalies were noted. The amniotic fluid index was 13.3 cm, corresponding to the 42nd percentile.

The patient was referred for evaluation by the Perinatology service at the emergency department of a Level IV complexity clinic. However, she only attended on March 27, 2025. During the assessment, high obstetric risk factors were identified due to advanced maternal age and primipaternity. Additionally, the patient did not have a complete second-trimester TORCH panel. An anatomical detail ultrasound with Doppler was performed, revealing a single live fetus with biometry corresponding to 31.1 weeks and an estimated fetal weight of 1787 grams, growing at the 37th percentile, with an amniotic fluid index of 12 cm and a posterior fundal placenta. A





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hyperechogenic, heterogeneous mass with anechoic areas was visualized in the right hemithorax, located at the pulmonary base, measuring 24 x 18 mm (*Picture 2-3-4*), without visible feeding vessels (*Picture 5*). A CPAM Volume Ratio (CVR) was also calculated, with a result of 0.12. The TORCH panel was requested for the next follow-up visit. Medical management was initiated with corticosteroids, administering 12 mg intramuscular betamethasone in two doses separated by 24 hours.

From the perinatology perspective, the cystic mass was considered to correspond to Type 2 Congenital Pulmonary Airway Malformation (CPAM) based on ultrasound characteristics specifically, a small mass (diameter <2 cm) with small anechoic areas, located at the base of the right lung. Given the epidemiology of the condition and the absence of fetal hydrops at the time, the perinatology specialist scheduled a follow-up appointment for April 8, 2025. During this visit, a new detailed obstetric ultrasound was performed, revealing a cephalic fetus with left dorsal orientation, biometry consistent with 33.2 weeks of gestation, and an estimated fetal weight of 2116 grams. The placenta was located in the left anterior fundus, grade II/IV, and the amniotic fluid index was 20 cm. No findings suggestive of pulmonary malformations were visualized during the anatomical survey (*Picture 6*). A cervical length of 42 mm was measured, with a cervical consistency index of 71%. Laboratory tests included negative HBsAg, negative IgG and IgM for toxoplasmosis, and a normal glucose tolerance test, with no other significant findings.

Based on the above, including the ultrasound findings, it was concluded that the malformation had resolved successfully following medical management with corticosteroids, which were administered at 31 weeks of gestation. Nearly two weeks later, at the time of ultrasound follow-up, the fetal lesion was no longer present.

## **Discussion**

It is of great importance to recognize that the initial management of this condition is the administration of corticosteroids, according to the medical literature, serial administration of these medications is a feasible approach within the fetal lung maturation protocol. This treatment was administered to the patient and successfully contributed to fetal lung development, with the malformation no longer visible on the most recent ultrasound performed at 32.2 weeks of gestation.



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The clinical case presented corresponds to a 36-year-old pregnant woman whose fetus was diagnosed during the second trimester with a pulmonary mass compatible with Type 2 CPAM, as evidenced by ultrasound findings of a small (<2 cm), hyperechoic, heterogeneous lesion with anechoic areas and no mediastinal shift. These findings are consistent with the Stocker classification for this CPAM variant. The calculated CPAM volume ratio (CVR) was 0.12, which indicates a low risk of complications such as fetal hydrops, given that the critical threshold is generally established at 1.6. This correlates with the ultrasound findings and the absence of hydrops.

It is noteworthy that, despite the time elapsed between the initial ultrasound diagnosis and the perinatology evaluation (approximately three months), the clinical picture did not show significant progression or the development of signs of fetal hemodynamic compromise. This favorable evolution allowed for the implementation of a conservative medical intervention with betamethasone, administered at 31 weeks of gestation, which has been shown to be effective in selected cases of CPAM. The therapeutic mechanism of corticosteroids in this context is not entirely clear, but it is hypothesized that they may induce regression of the mass and promote pulmonary maturation.

Remarkably, the follow-up ultrasound performed two weeks after corticosteroid therapy showed no evidence of the previously described mass, suggesting apparent resolution of the lesion a behavior consistent with reports in the literature for low-volume microcystic and solid CPAM lesions. This finding supports the hypothesis that, in the absence of hydrops and with favorable ultrasound characteristics, conservative corticosteroid management may be successful, thus avoiding invasive prenatal procedures such as thoracoamniotic shunting or thoracocentesis.

This case highlights the importance of serial ultrasound monitoring, CVR calculation, and comprehensive fetal assessment as key tools in clinical decision-making. Furthermore, it underscores the need for timely and specialized prenatal care, which in this case may have been affected by delayed and infrequent prenatal visits. Nevertheless, the clinical outcome was favorable, thanks to the appropriate response to medical treatment.







## Conclusions

Congenital Pulmonary Airway Malformation (CPAM), although rare, represents a significant entity in prenatal diagnosis due to its potential implications for fetal health. This case, the first documented in Floridablanca, Santander, Colombia, highlights the importance of serial ultrasound monitoring, risk stratification using the CPAM Volume Ratio (CVR), and comprehensive evaluation by a perinatology team.

The effectiveness of corticosteroid treatment, specifically betamethasone, in the spontaneous resolution of type 2 CPAM lesions without the need for invasive interventions is emphasized. The successful management of this type 2 CPAM with corticosteroids reinforces the value of conservative therapy in selected cases and underscores the importance of ultrasound and perinatal surveillance. This report contributes to the growing body of evidence on the variable clinical course of CPAM and the role of steroids in prenatal management.

Moreover, this case underscores the need for timely and specialized prenatal care, as early diagnosis and treatment may further optimize maternal and fetal outcomes. Finally, the importance of generating and disseminating local case reports is highlighted, as they contribute to the recognition of CPAM in regional clinical practice and support evidence-based decision-making.

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## **Attached files:**

Туре 0	Development arrested at the stage of trachea/bronchia formation; tracheal epithelium. Cysts smaller than 0.5 cm; tracheal epithelium, presence of cartilage
Туре 1	Development arrested at the stage of bronchia formation; bronchial epithelium. Large 4– 10 cm cysts, cartilage rarely present, squamous-like epithelium
Type 2	Development of the bronchial tree arrested at the glandular stage. Multiple cysts < 2.5 cm, covered with columnar epithelium
Туре 3	Development of the bronchial tree arrested at the glandular stage, typical adenomatoid malformation. Multiple cysts < 1.5 cm, covered with columnar epithelium
Туре 4	Development arrested at the stage of bronchia formation; acinar epithelium. Cysts size 2–4 cm(pleuropulmonary blastoma – PPB), cartilage absent

#### Table 1: Stocker histopathological classification of CPAM (2002)<sup>5</sup>.



Picture 1: Ultrasonography performed at 19 weeks of gestation. Longitudinal section showing a hyperechogenic (blue arrow), heterogeneous, predominantly solid mass located at the base of the right lung, measuring 15 x 12 mm.





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Picture 2: Ultrasonography performed at 31.1 weeks of gestation. Cross-sectional view: hyperechogenic (blue arrow) area measuring 24 x 18 mm at the base of the right lung.



Picture 3: Ultrasonography performed at 31.1 weeks of gestation. Longitudinal section: hyperechogenic area measuring 24 x 21 mm at the base of the right lung.





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## **About the Author**

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**Carlos Andrés Gómez Arias** is a medical student at the University of Caldas (Colombia). He has a particular interest in clinical surgery and actively participates in academic spaces focused on research, clinical practice, and interdisciplinary work. His training is characterized by a commitment to lifelong learning, critical thinking, and a vocation for service in hospital and community settings.

**Daniela Lopez Londoño** is a physician graduated from the Universidad Libre in 2021. She completed her medical social service in Cali in an outpatient clinic, providing life cycle care, prenatal care, family planning, and postpartum care, completing it in 2022. She lived in New York between 2023 and 2024, where she studied English at Kaplan Language International, achieving an upper-intermediate level. At the end of 2024, she worked at a telecommunications company providing international medical consultations. Daniela is currently studying with the goal of entering the postgraduate program.