Journal of Neonatal Surgery
ISSN(Online): 2226-0439
Vol. 14, Issue 27s (2025)
https://www.jneonatalsurg.com

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To Study the Management of Congenital Cystic Pulmonary Lesion of patients with its Diagnostic modalities attending a Tertiary Care Centre

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Cite this paper as: Dr. Mayur Shyam Soni, Dr. Edamakanti Swetha Soni, Dr. Shahanwaz Azam, Dr. Akshat Agarwal, Dr. Tarun Gupta, Dr. Santosh Patil, Dr. Sudhakar Jadhav, Dr. Sagar More, (2025) To Study the Management of Congenital Cystic Pulmonary Lesion of patients with its Diagnostic modalities attending a Tertiary Care Centre. *Journal of Neonatal Surgery*, 14 (27s), 478-494.

ABSTRACT

Introduction: Congenital cystic lesions of lung present an interesting spectrum of pathology in the paediatric population. Congenital lobar emphysema (CLE), congenital pulmonary airway malformation (CPAM), Sequestration, intrapulmonary cystic teratomas are rare conditions found in children.

Aim and objective: To evaluate the clinical presentation, investigation modalities, operative management, pathology, outcome (morbidity and mortality) and follow-up of congenital cystic lesions of the lung.

Material and Methods: This was a prospective observation study conducted in the Department of Paediatric Surgery Centre and P.G. Institute. All the cases of congenital cystic lung lesion operated at this institute during the period of August 2018 to January 2021 were studied. A total of 22 patients presented solitary lesions: CCAM, CLE, PS, BC were screeded. The Methods used to detect and confirm CCLL were as chest X-ray, CT scans, and others.

Results: In the present study we included 22 cases of congenital cystic lung lesions & observed for a period of August 2018 to January 2021. In our study the malformations included congenital cystic lesion was distributed in 4 types in which most common was Congenital lobar overinflation (CLO) with 45% of incidence, Congenital pulmonary airway malformation (CPAM) with 32%, Bronchogenic cyst with 18% and Pulmonary sequestration with 5%. Maximum patients were in the age group 0 to 3 months 59% respectively. Slight male preponderance was seen in this study. All patients have undergone surgical excision in terms of lobectomy or excision of the lesion. Post-operative histopathology confirmed the diagnosis.

Conclusion: Congenital cystic lung lesions are typically detected prenatally and managed through observation or surgery, depending on severity. Early diagnosis and treatment generally lead to good outcomes, with ongoing research aimed at improving detection and long-term care.

Keywords: *Congenital lung cyst, congenital lobar emphysema, congenital pulmonary airway malformation.*

1. INTRODUCTION

Congenital pulmonary airway malformations (CPAMs) consist of a spectrum of cystic or non-cystic lung malformations often associated with bronchial atresia during *in utero* development. Several classifications for these lesions have been proposed using radiologic or pathologic criteria based on cyst size and histology, with the Stocker classification being perhaps the most commonly used in the pathology literature.

Congenital cystic lesions of the lung in children are uncommon but potentially life-threatening and warrant an urgent diagnostic work-up. Pulmonary sequestration (PS), congenital cystic adenomatoid malformation (CCAM), congenital lobar emphysema (CLE), and bronchogenic cyst (BC) are the four major congenital cystic lesions, but they share similar embryologic and clinical characteristics.

Cystic lesions of the lungs encompass a wide spectrum of rare lung lesions comprising of congenital cystic adenomatoid malformation (CCAM), congenital lobar emphysema (CLE), bronchopulmonary sequestration, and bronchogenic cyst. CCAM with an incidence of 1 in 10,000 to 25,000 pregnancies is the most common lung cyst.[1] An abnormal antenatal ultrasound scan has now become the most common mode of presentation in Europe and the United States.In 1954, Gross and Lewis, published the first case report of CLE [2]. CLE and CPAM underwent lobectomy, intralober cystic teratoma and extralobar sequestration undergone excision of the lesion.

Congenital cystic adenomatoid malformations (CCAMs) present as cystic or solid lung masses restricted to part of one lung [3]. Most CAMs present with respiratory distress or compromise during infancy or recurrent pneumonias in later years, including adulthood. The pathological feature of CAMs is adenomatoid proliferation of the bronchioles that forms a cyst at the expense of alveoli. Three types of CAMs are recognized based on cyst size, number, and pathology [4]. Some CCAM/CPAMs can be life-threatening if they are not treated, so early and accurate diagnosis is important. The vast majority of CCAM/CPAM lesions are small enough that they will not cause any problems to the baby during pregnancy and the CCAM/CPAM can be removed after birth. However, some large lesions can cause serious and even fatal complications, including fetal heart failure (also called fetal hydrops) or maternal mirror syndrome. These cases may require treatment before birth.

Type 1 consists of multiple large cyst (2-10 cm), at least one cyst will be dominant with smaller cysts seen along its periphery. The cyst wall is lined with ciliated pseudostratified columnar epithelium, contains elastic tissue beneath the epithelium, smooth muscle, and fibrovascular connective tissue, including cartilage.

Type 2 consists of small and more uniform size cysts (0.5-2 cm). They are lined with cuboidal to columnar epithelium and have only a thin fibromuscular wall. This type has associated anomalies – renal, cardiac, skeletal, intestinal, extra-lobar sequestration.

Type 3 lesions are bulky solid lesions that usually involve entire lobe of the lung. There is slight modification of the classification of CCAM with now five types being recognized out of which only three types are identified on imaging as mentioned above. It is now called as congenital pulmonary airway malformations since all the types are not cystic [5] Additional types are, Type 0, which has either no cyst or very small ones (<0.5 cm), and is incompatible with life and Type 4 CCAM which consists of large cysts up to 10 cm in size [6]. In patients under 1 year old, cystic lesions were discovered by respiratory distress; and in patients over 1 year old signs of infection were the most important clinical features.

Congenital lobar overinflation (CLO), previously known as congenital lobar emphysema, is a rare neonatal disease, with a prevalence of 1 in 20,000-30,000 [3]. It is a developmental anomaly of the lung . Males are more frequently affected than females (3:1). In 50% of cases, the cause is unknown. Otherwise, the most common cause, accounting for 25% of cases, is a congenital defect of bronchial cartilage (e.g., malacia, stenosis), and the other 25% are due to a bronchial obstruction (e.g., mucous plugging, redundant mucosal folds/septa, cardiopulmonary vascular anomalies, intrathoracic masses) [1].

Pulmonary sequestration (PS) is a rare congenital malformation characterized by a non-functional mass of lung tissue that does not communicate with the tracheobronchial tree and receives an anomalous vascular blood supply from the systemic circulation. The most widely accepted theory is that PS results from the formation of an accessory lung bud below the normal lung buds [5]. It may occur as an isolated lesion or in association with other malformations. Clinically, PS cases are usually divided into the intralobar and extralobar types. Most children affected by PS have a good prognosis, some of them may experience a high risk of infection, hemoptysis, hemorrhage, and heart failure in rare cases [7].

Bronchogenic cysts represent a rare form of cystic malformation of the respiratory tract. Primarily located in the mediastinum if occurring early in gestation as opposed to the thoracic cavity if arising later in development. However, they can arise from any site along the foregut. They exhibit a variety of clinical and radiologic presentations, representing a diagnostic challenge, especially in areas with endemic hydatid disease.

Early recognition of these relatively rare congenital cystic lung lesions would lead to the immediate, proper surgical intervention. Thus, this observational study was undertaken to know the incidence, presentation, diagnosis, management & follow up of patients who have been operated for congenital cystic lung lesions.

2. MATERIAL AND METHODS

This prospective observation study was conducted in Paediatric Surgery Centre and P.G. Institute. All the cases of congenital cystic lung lesion operated at this institute during period of August 2018 to January 2021.

Inclusion criteria: In this study 22 cases of congenital cystic lung lesions were included . All children from day 1 of life to 15 years age found to be having congenital cystic lung lesion at our institute were included in our study.

Exclusion criteria: Parents not willing to participate in the study.

Sample size estimation: The exact incidence of congenital cystic lung lesion is not available. Based on previous 3-year census roughly 5-6 such cases were expected per year at our institute, so we included all patients satisfying our inclusion and exclusion criteria.

Clinical Presentation: Symptoms, diagnosis, prenatal and postnatal detection, complications, and associated conditions.

Diagnosis/Imaging Techniques: Methods used to detect and confirm CCLL, such as prenatal ultrasound, fetal MRI, postnatal chest X-ray, CT scans, and others.

Management/Treatment: Medical or surgical interventions, follow-up procedures, and prognosis.

Congenital cystic lung lesions (CCLL) are rare developmental abnormalities of the lung, such as Congenital Pulmonary Airway Malformation (CPAM) and bronchopulmonary sequestration (BPS). These lesions are typically detected prenatally through routine ultrasound, with fetal MRI serving as a complementary tool for more detailed imaging. Postnatal diagnosis often involves chest X-rays and CT scans to confirm the lesion's size and structure. Management strategies vary from observation to surgical resection depending on the lesion's severity, with most cases having a favourable outcome when treated early. Continued research focuses on improving prenatal detection methods and long-term prognosis.

Statistical analysis

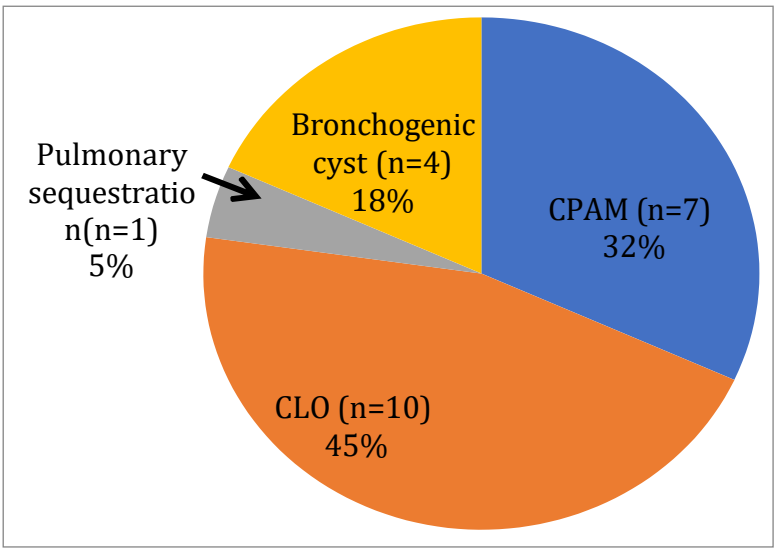
Data was entered on MS-Excel; & analysed using MS-Excel or Statistical software such as SPSS v.16.0 whenever needed. Appropriate descriptive statistics was applied to the data, like Frequency chart, Mean with Standard Deviation with graphical representation and suitable Statistical Test was applied whenever required.

Ethical clearance

The ethical committee clearance certificate was duly taken before starting of study by Institutional Medical Ethical Committee.

3. RESULTS

In our study we include 22 cases of congenital cystic lung lesions & observed from August 2018 to January 2021. In our study the malformations included congenital cystic lesion was distributed in 4 types in which most common was Congenital lobar overinflation (CLO) with 45% of incidence, Congenital pulmonary airway malformation (CPAM) with 32%, Bronchogenic cyst with 18% and Pulmonary sequestration with 5% as shown in Graph no.1. Maximum patients were in the age group 0 to 3 months- 59% respectively as shown in Table no. 1 and Graph no. 2.

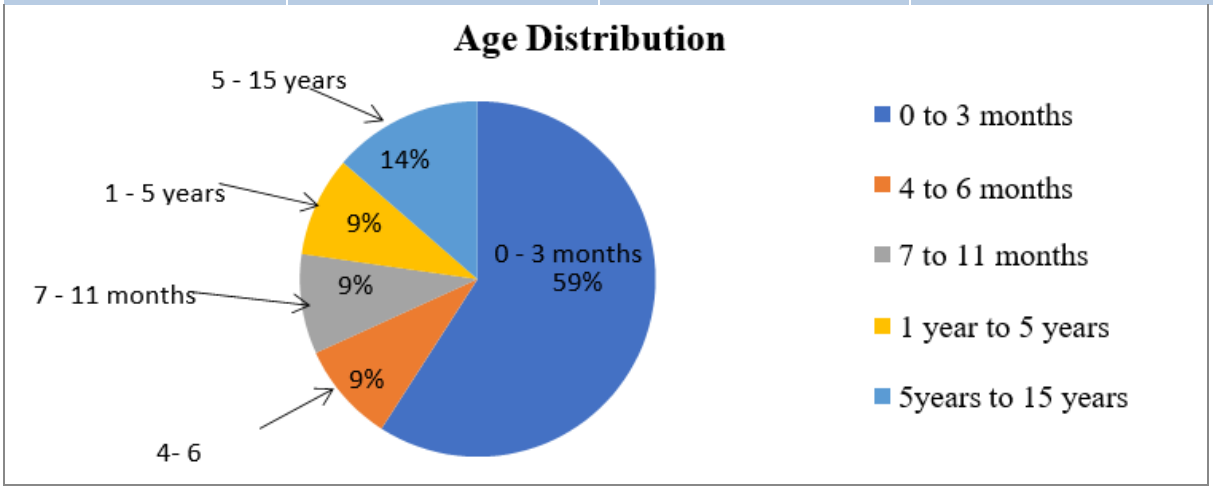


Graph no. 1 Distribution of 4 types of congenital cystic lung lesion in patients.

Most common age of presentation in CPAM, CLO & pulmonary sequestration was 0 to 3 months, while bronchogenic cyst was common in 7 to 11 months & 5 to 15 years age group as shown in Table no.2 and Graph no.3.

Table no.1. Age wise distribution of patients

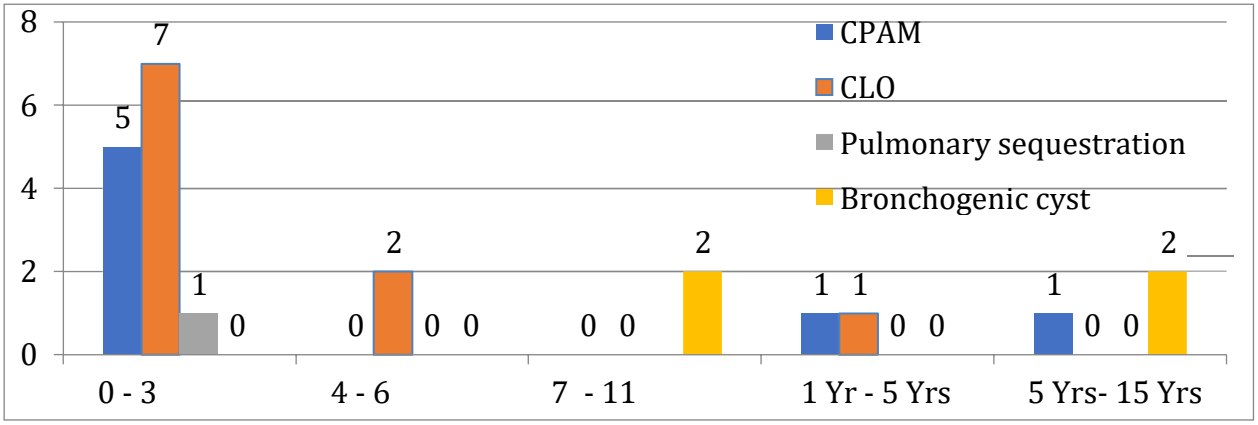
Age Group (n=22)	No. Patients	Percentage	Median Age
0 - 3 Months	13	59.09%	1.5 month
4 - 6 Months	2	9.09%	5 months
7 - 11 Months	2	9.09%	10.5 months
1 Year - 5 Years	2	9.09%	15 months
5 Years – 15 Years	3	13.64%	8 years (96 months)



Graph no 2. Age wise distribution of patients.

Congenital cystic lung lesions	0 - 3 Months (Mean age (45.9 days / 1.5 months)	4 - 6 Months (Mean age 5 months)	7 - 11 Months (Mean age 10.5 months)	1 Year - 5 Years (Mean age 15 months)	5 Years – 15 Years (mean age 7.6 years / 92 months)
CPAM (n=7)	5 (71.42%)	0	0	1 (14.29%)	1 (14.29%)
CLO (n=10)	7 (70%)	2 (20%)	0	1 (10%)	0
Pulmonary sequestration (n=1)	1 (100%)	0	0	0	0
Bronchogenic cyst (n=4)	0	0	2 (50%)	0	2 (50%)

Table no. 2– Age wise distribution in each lesion.



Graph no.3 Age wise distribution in each lesion.

In this study we have found that among 22 patients, 12 (55%) were male patients, and 10 (45%) were female patients. Slight male preponderance was seen in this study as shown in Table no.3 and Graph no. 4 .

Gender Distribution	Male	Female
Congenital cystic lung lesions (n=22)	12 (55%)	10 (45%)

Table no.3 Gender wise distribution.

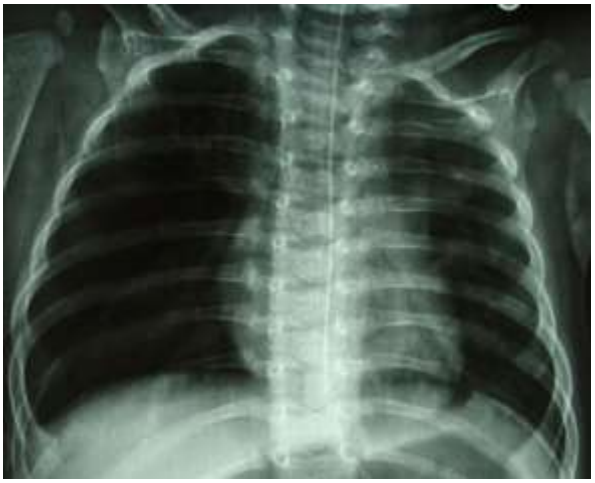
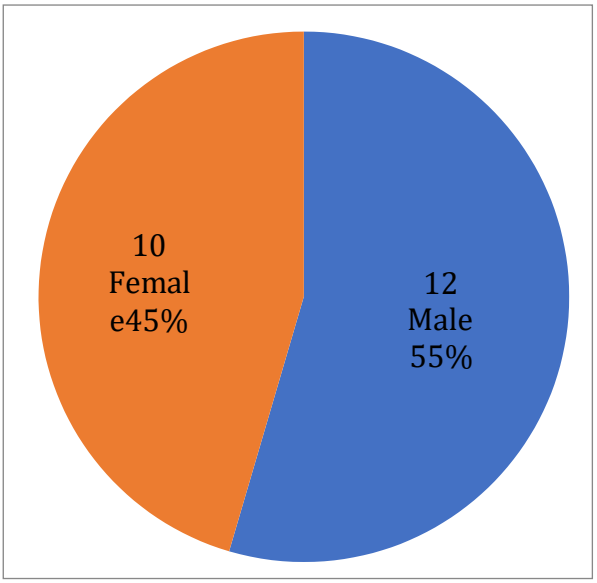


Fig 1: X-ray showing hyperinflation of affected side in a case of CLO.

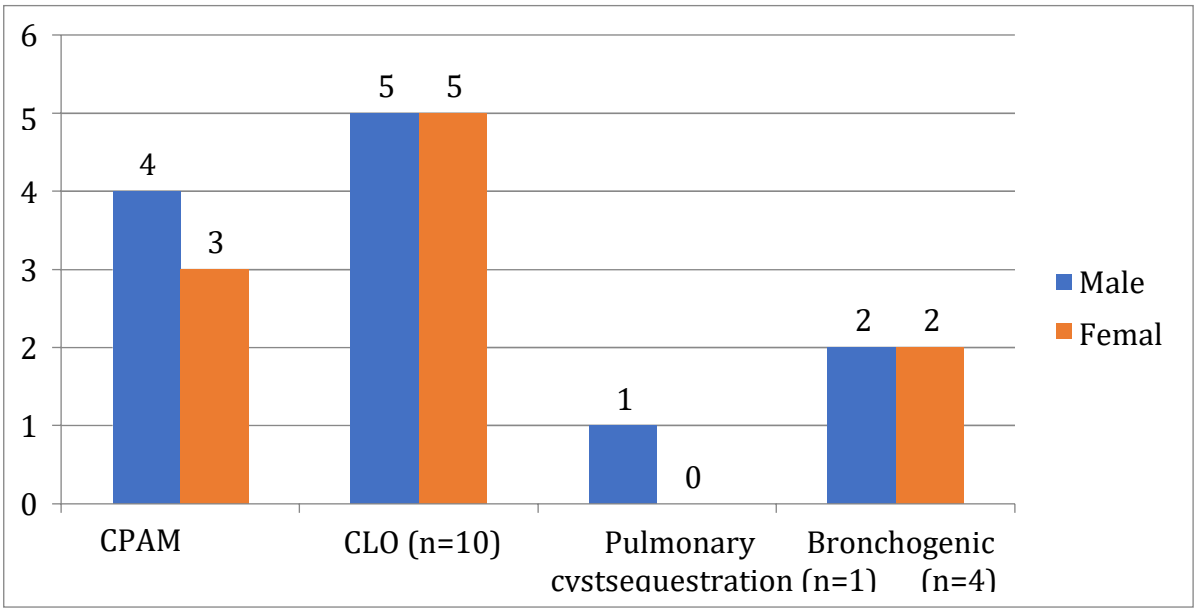


Graph no. 4- Gender wise distribution.

Among 7 cases of CPAM 4 (57%) were male & 3 (43%) were female, in 10 cases of CLO 5 (50%) were male & 5 (50%) were female. Only one case of pulmonary sequestration was found which was male, & in bronchogenic cyst out of 4 patients, 2 (50%) were male & 2 (50%) were female. Among CPAM, maximum was male. In CLO & bronchogenic cyst, both, male & female showed equal distribution as shown in Table no. 4 and Graph no.5.

Congenital cystic lung lesions	Male	Female
CPAM (n=7)	4 (57%)	3 (43%)
CLO (n=10)	5 (50%)	5 (50%)
Pulmonary sequestration (n=1)	1 (100%)	0 (0%)
Bronchogenic cyst (n=4)	2 (50%)	2 (50%)

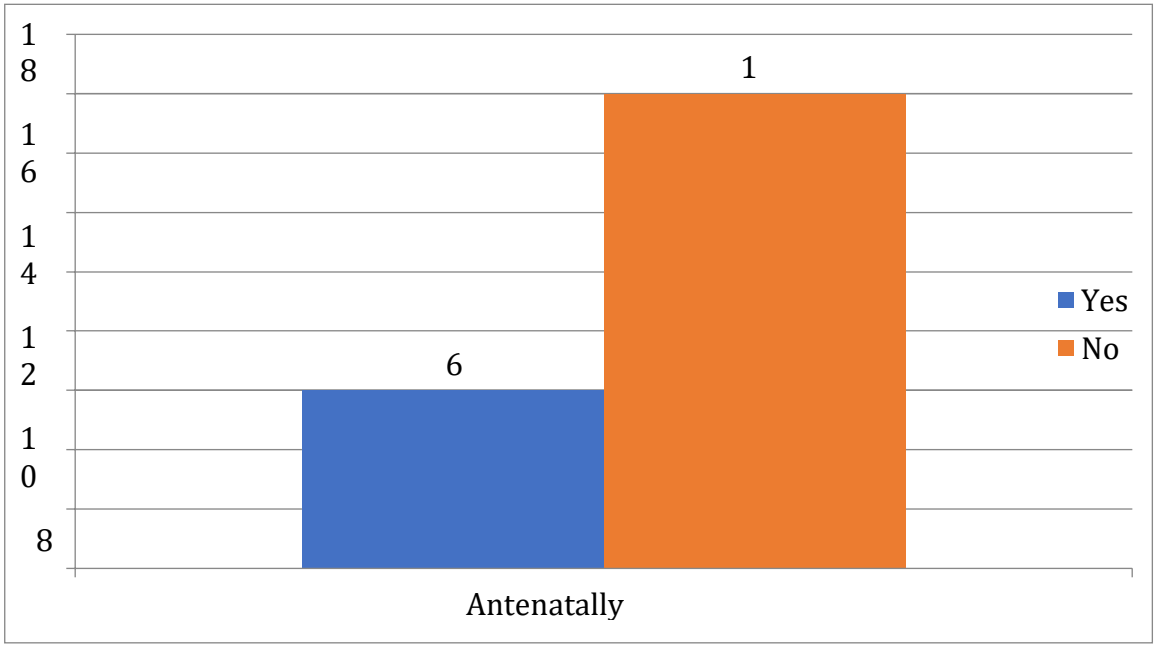
Table no. 4 –Gender Distribution of each lesion.



Graph no.5 Gender Distribution in each lesion.

Antenatally diagnosed	Yes	No
Congenital cystic lung lesions	6 (27.3%)	16 (72.7%)

Table no.5- Antenatal diagnosis of cystic lung lesions.



Graph no.6 Antenatal diagnosis of cystic lung lesions.

In our study out of 22 patients only 6 (27.3%) were diagnosed antenatally, while 16 (72.7%) were not diagnosed antenatally.Thisshows that congenital cystic lung lesions were not commonly diagnosed antenatally.

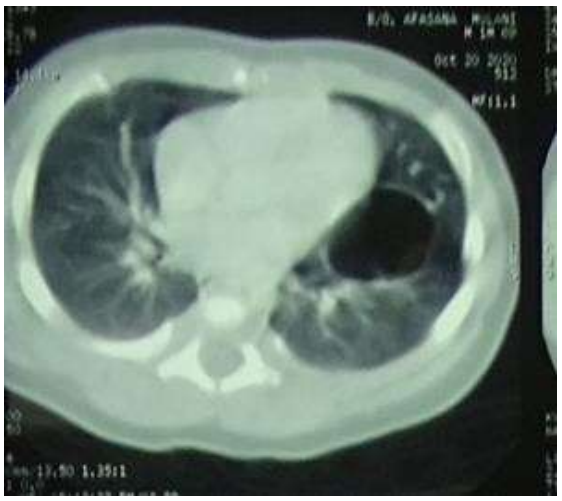


Fig2 :Cysticlesionseen incaseofCPAMon CT

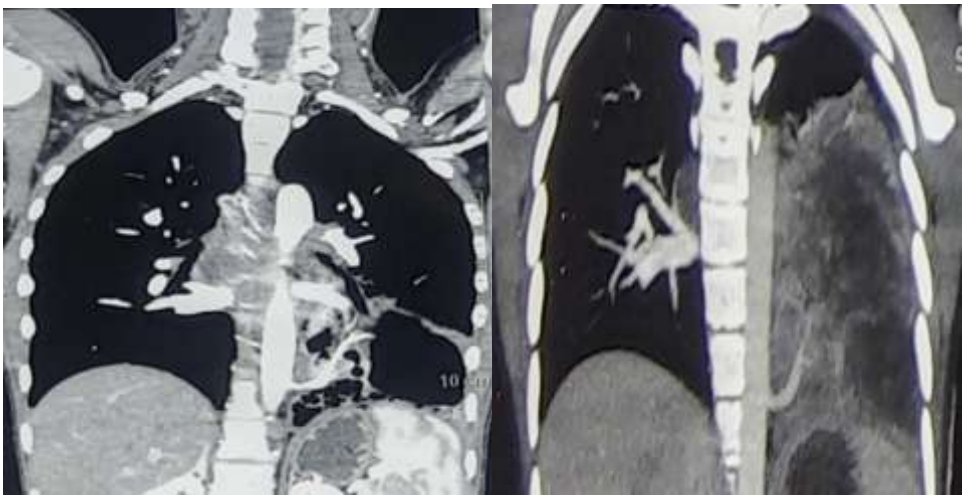
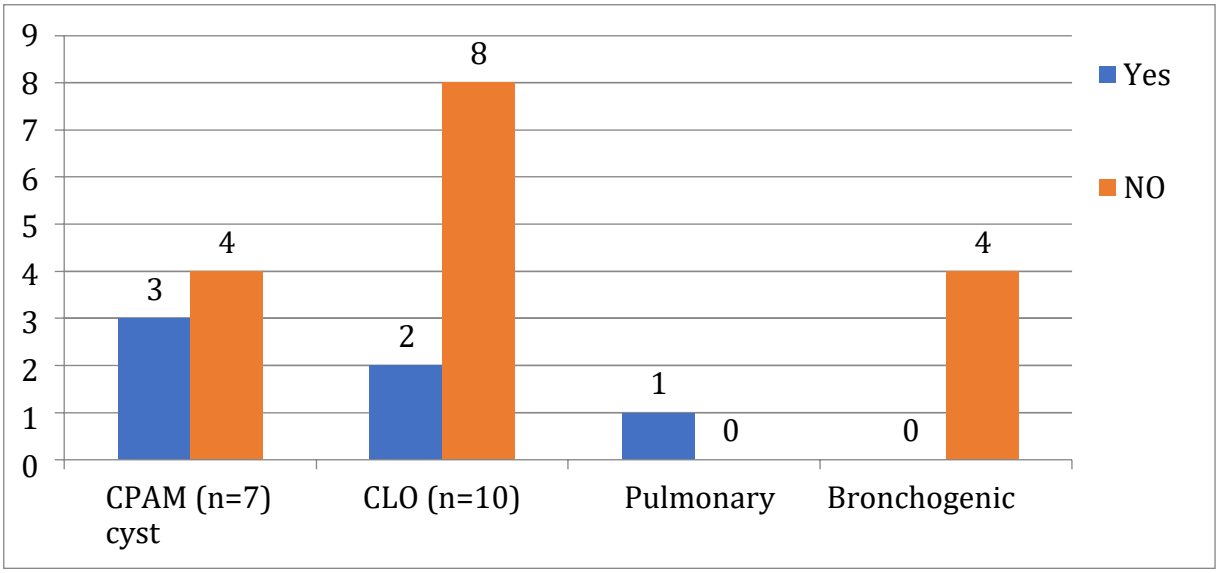


Fig3:pulmonarysequestrationshowingseparate arterial&venoussupply

Antenatally Diagnosed	Yes	No
CPAM (n=7)	3 (42.85%)	4 (57.15%)
CLO (n=10)	2 (20%)	8 (80%)
Pulmonary sequestration (n=1)	1 (100%)	0 (0%)
Bronchogenic cyst (n=4)	0 (0%)	4 (100%)

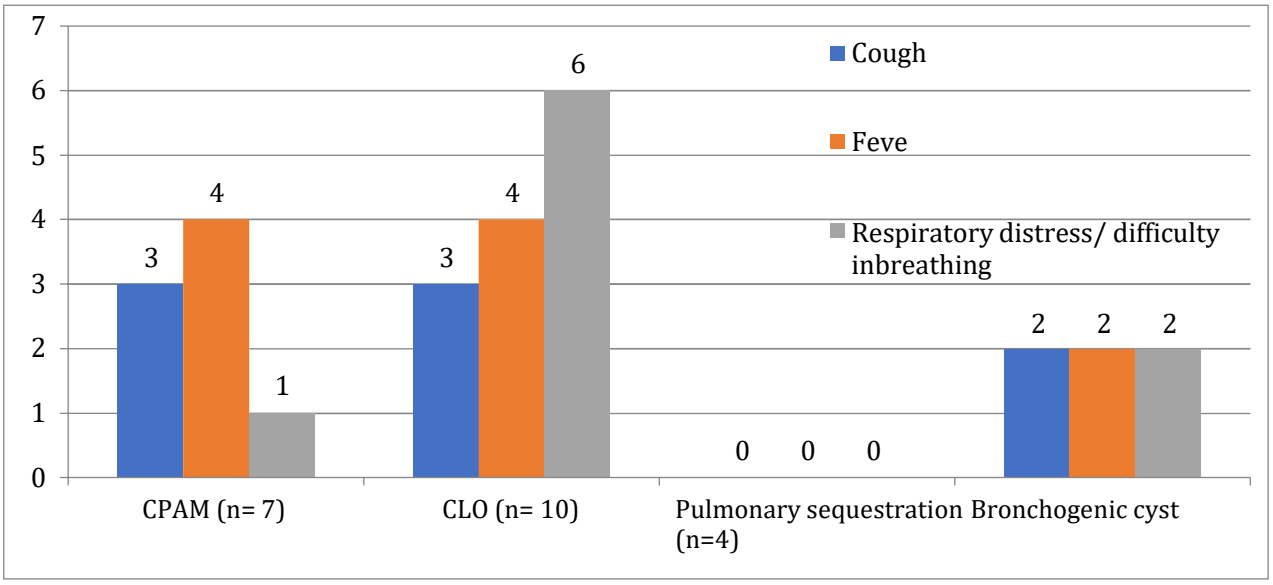
Table no. 6- Antenatal Diagnosis of each lesion.



Graph no. 7 Antenatal Diagnosis in each lesion.

Individually, among 7 patients of CPAM, 3 (42.85%) patients were diagnosed antenatally & 4 (57.15%) patients were not diagnosed antenatally. Among 10 patients of CLO, 2 (20%) were diagnosed antenatally & 8 (80%) were not diagnosed

antenatally. In pulmonary sequestration only 1 patient was found and was diagnosed antanatally (100%), in bronchogenic cyst all 4 patient were not diagnosed antenatally.

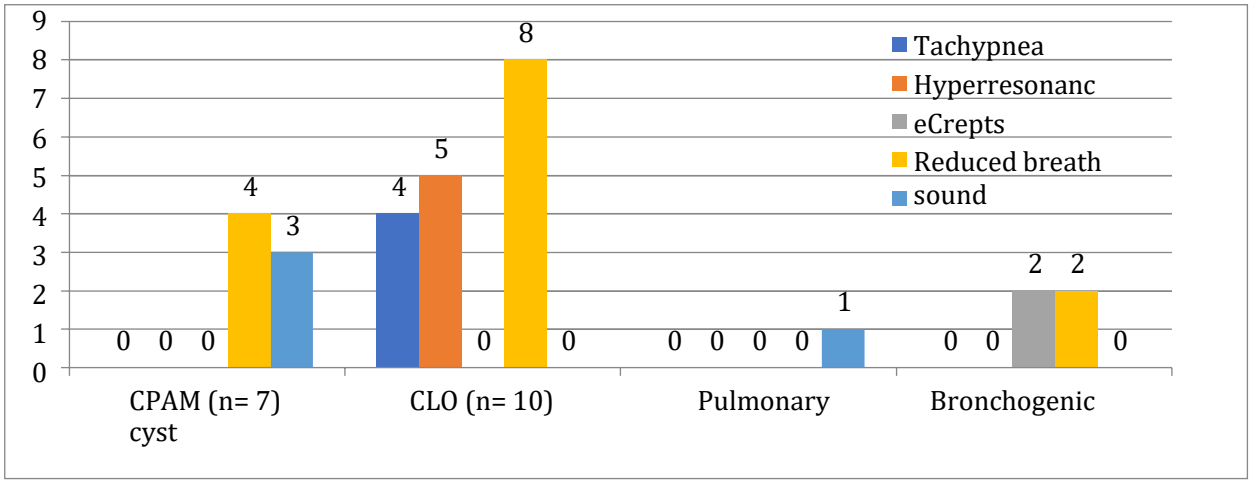


Graph no. 8- Presenting Complaints in each lesion.

In 7 patients of CPAM, 3 (42.85%) presented with cough, 4 (57%) presented with fever & 1 patient presented with difficulty in breathing, 3 patients had both cough & fever and 1 patient had fever with difficulty in breathing. In 10 patients of CLO, 3 (30%) patients presented with cough, 4 (40%) presented with fever, 6 (60%) presented with respiratory distress. Amongst these 3 (30%) patients had both cough and fever, 1 (10%) had cough with respiratory distress, & 1 (10%) had cough, fever with respiratory distress.In a single case of Pulmonary sequestration- patient had no complaints.In 4 cases of bronchogenic cyst, 2 (50%) had cough, 2 (50%) had fever, 2 (50%) had respiratory distress, among them 1 (25%) patient had cough & fever both, 1 (25%) patient had fever with respiratory distress.

Congenital cystic lung lesions	**Tachypnea**	**Hyperresonance**	**Crepts**	**Reduced breath sound**	**Normal breath sounds**
CPAM (n= 7)	0	0	0	4 (57.2%)	3 (42.8%)
CLO (n= 10)	4 (40%)	5 (50%)	0	8 (80%)	0
Pulmonary sequestration (n=1)	0	0	0	0	1 (100%)
Bronchogenic cyst (n=4)	0	0	2 (50%)	2 (50%)	0

Table no.7– Respiratory system examination finding.

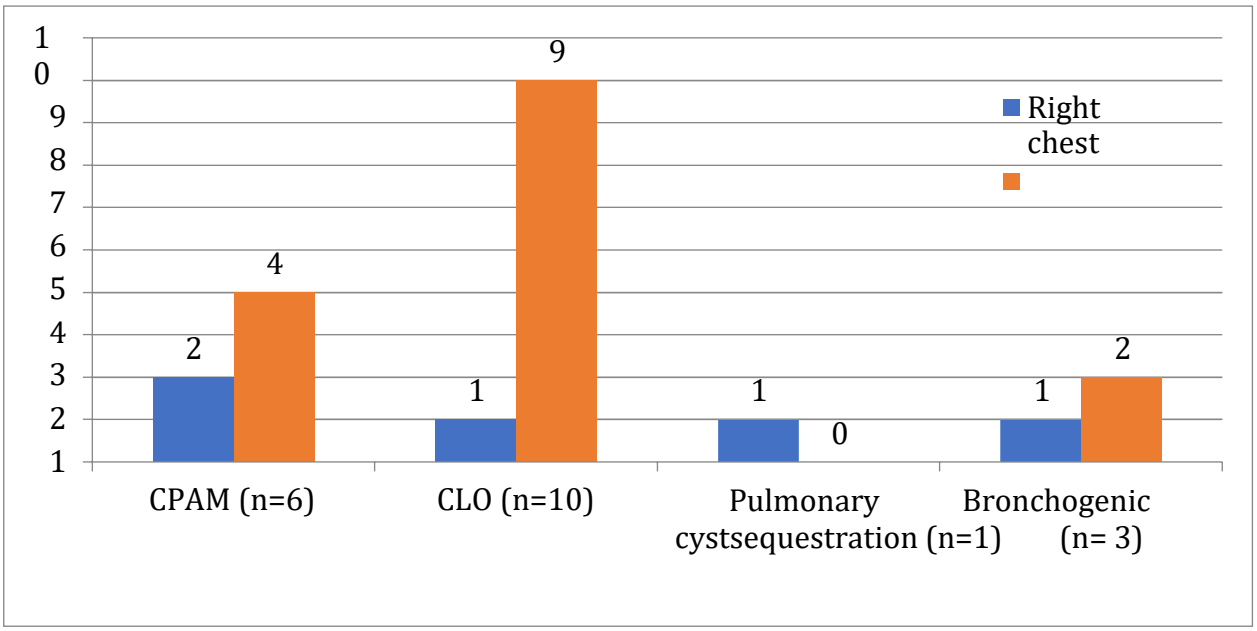


Graph no.9 - Respiratory system examination finding.

On respiratory system examination in patients of CPAM, most common finding was reduced breath sounds on affected side.In patients of CLO, reduced breath sounds on affected side was common finding, followed by hyperresonance on percussion & tachypnea.In patient of pulmonary sequestration, no abnormal respiratory system examination was found.In patients of bronchogenic cyst, crept& reduced breath sound in affected zone of chest was common finding.

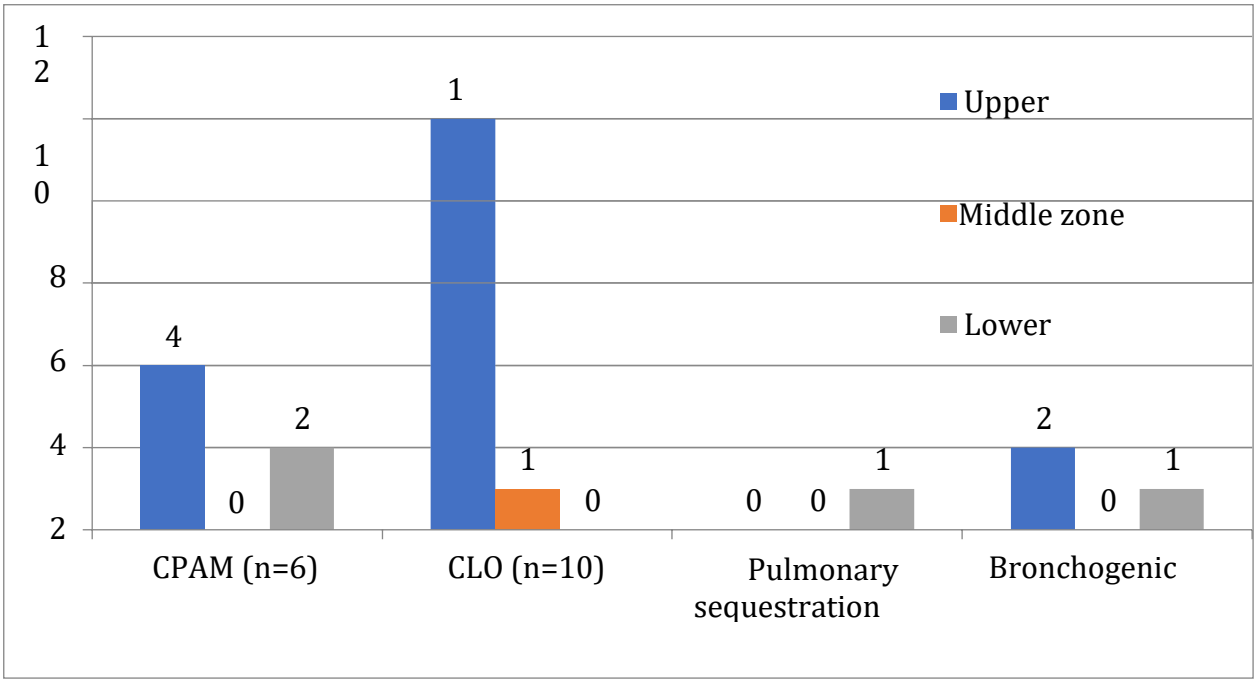
Diagnostic Modalities

Distribution according to X ray finding :



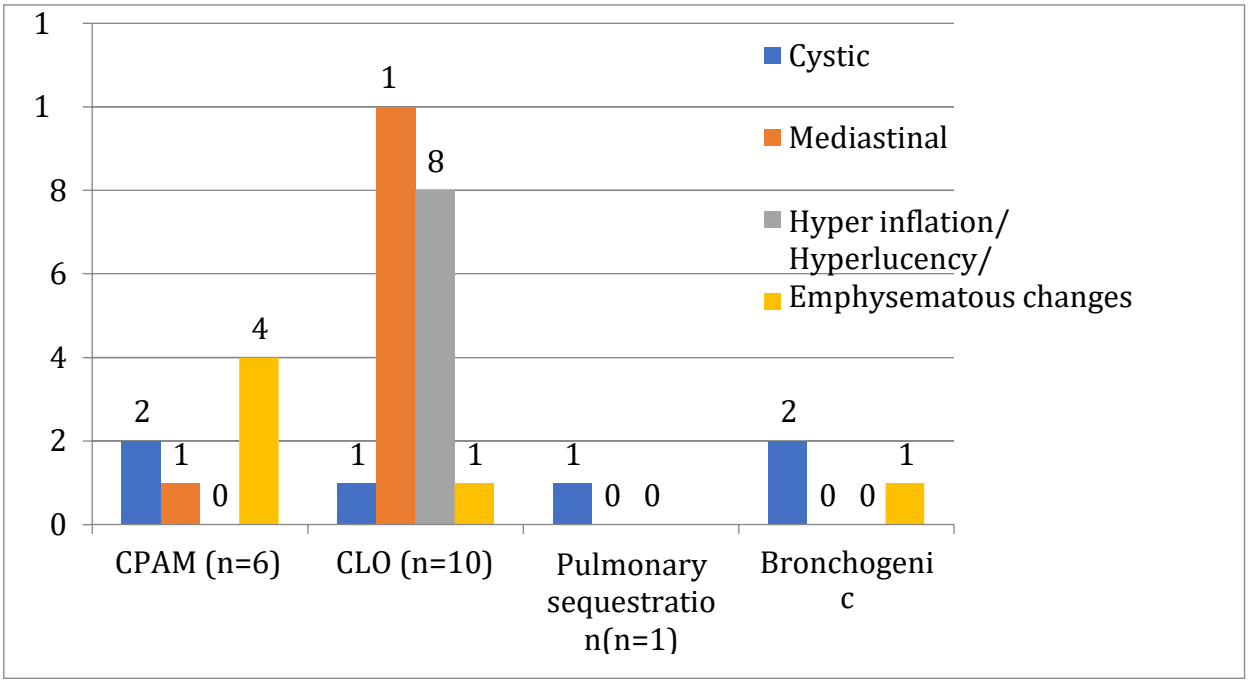
Graph no. 10 - X-ray Chest - Side involved.

Left side involvement was found more common in CPAM, CLO & bronchogenic cyst, while right side involvement was seen in pulmonary sequestration on X-ray.



Graph no.11- X-ray Chest- Lobe involved.

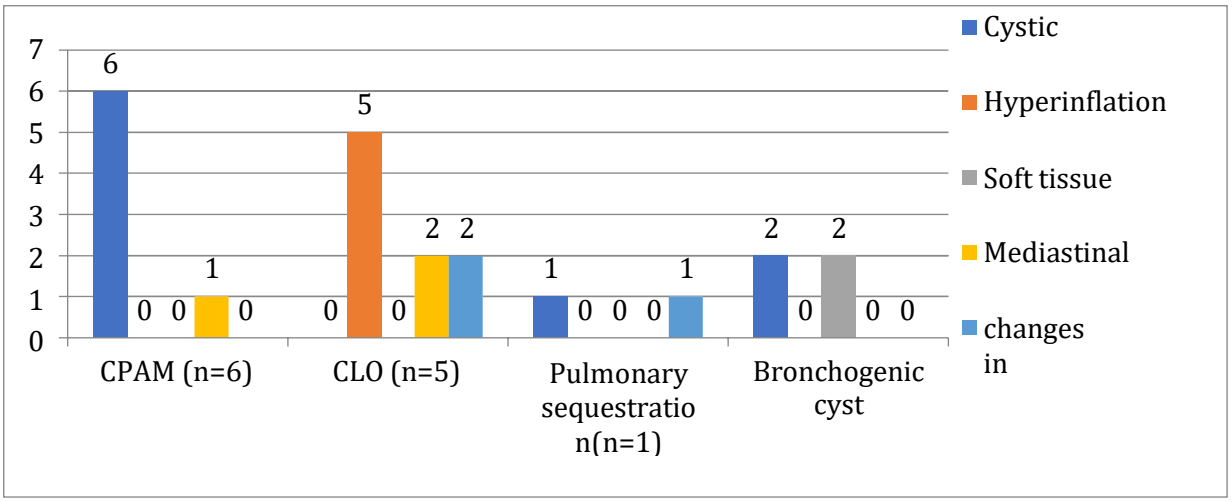
Upper zone involvement was more common on X-ray in CPAM, CLO & bronchogenic cyst, followed by lower zone involvement.Middle zone involvement was seen only in CLO.Pulmonary sequestration showed lower zone involvement.



Graph no.12- X-ray Finding in each lesions.

In CPAM- most common finding on X-ray was opacities, followed by cystic lesions & mediastinal shift.In CLO- most common finding on X-ray was mediastinal shift to opposite side of lesions, followed by hyperlucency or hyperinflation changes on the same side, others were cystic lesions, ground glass opacities with consolidation.In pulmonary sequestration, ill- defined cystic areas were seen on X-ray.In bronchogenic cyst, cystic lesions & opacity was common on X-ray.

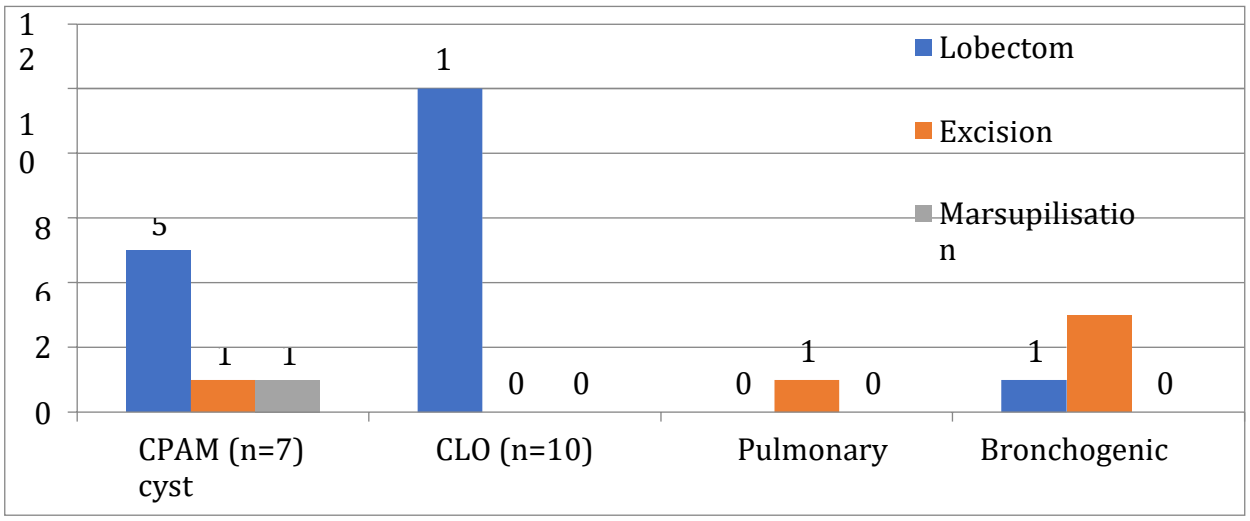
Distribution according to CT scan finding:



Graph no.13- CT scan chest finding.

In our study Cystic lesions were found most common in cases of CPAM.In CLO most common was hyperinflation of lung.In pulmonary sequestration cystic air spaces with separate arterial & venous supply was seen on CT scan.In bronchogenic cyst CT scan showed cystic lesions & soft tissue density.

Surgical management



Graph no. 14 Operative procedure.

In cases of CPAM & CLO, lobectomy was the operative procedure of choice. In pulmonary sequestration & bronchogenic cyst, excision of the sequestration or cyst was the operative procedure of choice.

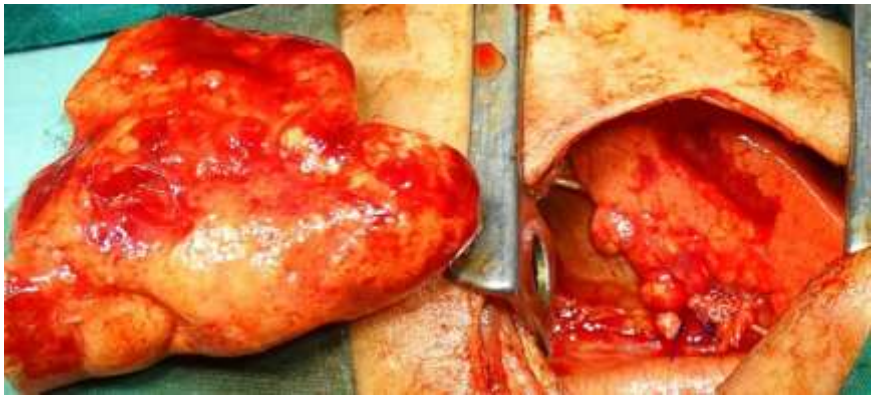


Fig 4: Lobectomyina case of CPAM

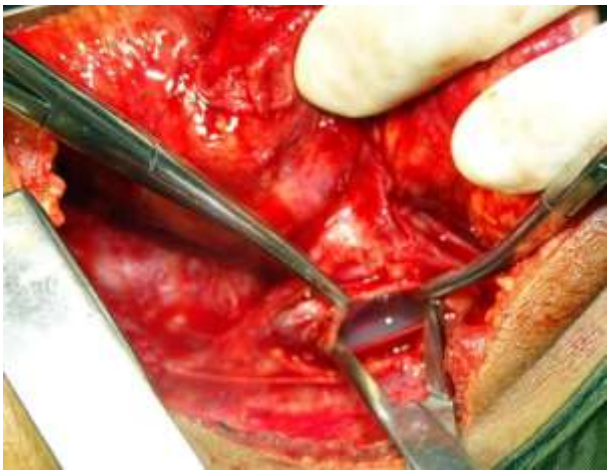


Fig 5 :Bronchogenic cyst showing white mucoid content–excision of cyst done.

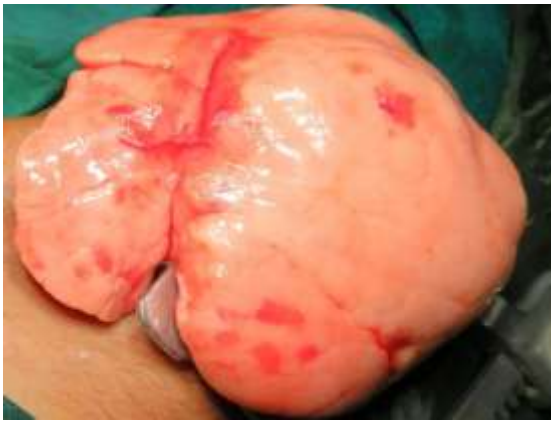


Fig 6:Overinflated lunglobein case of CLO

Distribution according to Histopathological finding:

In our study, out of 7 patients of CPAM, in 4 (57.14%) patients histopathology showed congenital pulmonary airway malformation type-1, out of which 1(14.2%) patient suspected to be bronchogenic cyst/ CPAM on investigation. In 3(42.85%) patients histopathology showed congenital pulmonary airway malformation type-2, out of which 1(14.2%) patient was diagnosed to be infected lung cyst on investigation. Out of 10 patients of CLO, in 9 (90%) histopathology showed pan acinar or congenital lobar emphysema of left lung upper lobe, 1 (10%) patient showed pan acinar or congenital lobar emphysema upper & middle lobe of right lung. In our study we had only one patient of pulmonary sequestration,

sequestration of lung on histopathology showed presence of pulmonary tissue. Out of 4 patients of bronchogenic cyst, in all 4 (100%) patient histopathology confirmed presence of bronchogenic cyst.

Congenital cystic lung lesions	Fever	Higher antibiotic	Ventilatory support
CPAM (n=7)	3 (42.8%)	3 (42.8%)	1 (14.2%)
CLO (n=10)	2 (20%)	2 (20%)	4 (40%)
Pulmonary sequestration (n=1)	0	0	0
Bronchogenic cyst (n=4)	1 (25%)	1 (25%)	0

Table no.8 Post operative period.

In post operative period, out of 7 cases of CPAM 3(42.8%) had fever, 3(42.8%) required higher antibiotics in post operative period & 1 (14.2%) required ventilatory support. Fever was most common post operative complication in CPAM & was managed on higher antibiotics. In CLO, out of 10 patients, 2 (20%) had fever, 2 (20%) required higher antibiotics, 4(40%) required ventilator support from minimum 12 hours post operative to 2 days post operative. Requirement of ventilator support was more common in CLO patients. Pulmonary sequestration patient showed smooth post operative recovery. In Bronchogenic cyst out of 4 cases, 1 (25%) developed fever & 1 (25%) required higher antibiotics.

Congenital cystic lung lesions	<3days chest drainage	>3 days chest drainage
CCAM (n=7)	4 (57.14%)	3 (42.86%)
CLO (n=10)	7 (70%)	3 (30%)
Pulmonary sequestration (n=1)	1 (100%)	0
Bronchogenic cyst (n=4)	3 (75%)	1 (25%)

Table no. 9 Duration for which chest tube drainage

In majority of patients of CPAM, CLO, pulmonary sequestration & bronchogenic cyst chest tube drainage was required for less than 3 days.

Congenital cystic lung lesions	Fever	Cough	Pneumothorax	Stitch abscess	Poor weight gain	Hospital admission
CPAM (n=7)	3 (42.8%)	3 (42.8%)	0	1 (14.2%)	0	0
CLO (n=10)	2 (20%)	4 (40%)	0	1 (10%)	1 (10%)	0
Pulmonary sequestration (n=1)	0	0	0	0	0	0

Bronchogenic cyst (n=4)	1 (25%)	1 (25%)	1 (25%)	0	0	1 (25%)

Table no. 10 Complications on follow up.

On follow up most common complication was fever and cough which was managed on oral medication in most of the cases. Stitch abscess required debridement on OPD basis. In case pneumothorax, patient required readmission & insertion of ICD tube was done.

4. DISCUSSION

Congenital cystic lesions of lung in children are a spectrum of anomaly. Early diagnosis, investigations, and management of these potentially life-threatening anomalies rewarded with good result. Congenital cystic lung lesions are a rare but significant cause of morbidity in infants, children and young adults presenting with respiratory distress and repeated chest infections. They consist of congenital pulmonary airway malformation (CPAM) previously known as congenital cystic adenomatoid malformation (CCAM), congenital lobar overinflation (CLO) previously known as congenital lobar emphysema (CLE), pulmonary sequestration, and bronchogenic cyst.

Hence, there is a high possibility that the diagnosis of congenital lung malformations would be missed, unless specifically kept in mind and sought for by advanced imaging studies as pneumonia is the most common cause of respiratory morbidity in children, especially in developing countries [2].

In the present study 22 cases of congenital cystic lung lesion were included & observed from August 2018 to January 2021. The demographic features, clinical presentation, clinical examination, investigations, management & outcome and follow up were studied and analyzed.

In the current study it was observed that the most common age group was between 0 to 3 months with 13 cases, and the median age was 1.5 months. In our experience, congenital cystic lung lesion were most commonly affects 0 to 3 months age group with only few cases affected in older age groups, these findings were similar to a study done by Shibsankar Barman et.al[8]and Lima, M et al [9] where 50% cases presented within 3 months of age.

In our study out of 22 cases, 12 (55%) were male and 10 (45%) were female, thereby indicating slight male preponderance. But equal sex preponderance was seen in a study done by Shibsankar Barman et al.[8] In another study by Lima, M et al [9] and Costa Junior et al.[10] similar results of male predominance was seen which was in accordance to the current study.

In the present study 22 patients were divided into 4 groups in which 10 patients (45%) in CLO, 7 patients (32%) in CPAM, 4 patients (18%) in bronchogenic cyst & 1 patient (5%) in pulmonary sequestration.This study was in accordance with the study performed by the other research investigator Costa Junior et al.[10]and Mohamed ElShabrawy Saleh et al. [11] and krishna Mohan Golla et al [12].

In our study out of 22 cases only 6 (27.3%) cases were diagnosed antenatally, among which 3 cases were CPAM, 2 cases were CLO & 1 case was of pulmonary sequestration, showing that less cases were diagnosed antenatally, which is contradictory to current situation where lot of studies shows that these cystic lung lesions are early picked up on antenatal sonography, mostly because we get patients from periphery where expert radiologist are not available, hence these lesions were missed to be picked up on antenatal scan. In study by Mohamed ElShabrawy Saleh et al.[11] similar finding of not getting diagnosed antenatally in majority of cases was seen.

In the present study it was found that , on X-ray done in 6 cases of CPAM out of 7 cases in bronchogenic cyst in 3 cases out of 4, because these patients had CT done from the reffering centre. X-ray was sufficient in diagnosing the side & zone involved in most of cases, but in diagnosing the nature of lesion X-ray was not sufficient in CPAM, pulmonary sequestration & bronchogenic cyst, while in CLO X-ray showed sufficiently enough signs to reach exact diagnosis. This study was in support with the study by Mohamed ElShabrawy Saleh et al [11],

D.H. Parikh et al·[13] and by Shibsankar Barman et al [14] where similar finding of left upper lobe being predominant lobe involved was found.

CT scan was helpful in identifying the exact lesion in almost all the cases, according to D.H. Parikh et al.[13] and Aditi Jain et al [14]CT scan is considered as gold standard for diagnosis of congenital cystic lung lesion.

Lobectomy was surgical procedure of choice in cases of CPAM & CLO while excision of cyst or lesion was preffered in cases of pulmonary sequestration & bronchogenic cyst. In a study done by Shibsankar Barman et.al and D.H. Parikh et al.

Intraoperatively, in cases of CPAM cystic lesion involving the lobe completely were seen, in 2 cases concomitant infection of the cyst was seen, in one case poor inflation of the involved lobe, & in one case hyperinflated lobe containing cyst was seen. In cases of CLO emphysematous, hyperinflated lobe were seen. This finding was similar to the study by D.H. Parikh

et al.[13] and Faten Limaïem et al[15]. intrapulmonary bronchogenic cyst were predominant than mediastinal cyst.

On histopathology of specimen done after surgery showed that, 45% (10) cases were of CLO, 32% (7) cases were of CPAM, 18% (4) were of bronchogenic cyst, 5% (1) was of pulmonary sequestration. This study is similar to James Cook et.al [16] study.

Chest tube drainage was required for less than 3 days in majority of cases in postoperative period, 57% in CPAM, 70% in CLO, 100% in pulmonary sequestration & 75% in bronchogenic cyst. Drain days varied from 2-12 days in a similar study done by Mohamed ElShabrawy Saleh et al [11].

Congenital pulmonary airway malformation (CPAM), one of the congenital lung diseases discussed under the umbrella term 'congenital thoracic malformations,' others being a bronchogenic cyst and pulmonary sequestration, is rare, but the most common developmental congenital anomaly of the lung. The malformation is due to abnormalities during embryogenesis and can occur at different stages during lung development, leading to anomalous bronchial morphogenesis. With new advancements and technological progress, CPAM is frequently diagnosed antenatally, and managed by a pediatric surgeon from early on [17].

On follow up persistant fever & cough was most common complaints which were managed on oral medication in majority of cases, 1 case required repeat hospital admission for spontaneous pneumothorax within 1 week of discharge & insertion of ICD tube was done & was treated uneventfully.

Congenital cystic adenomatoid malformation (CCAM) is an anomaly consisting of benign, non-functioning multicystic masses that typically appear in a single lobe of the lung and result from overgrowth of the terminal bronchioles without differentiation of the epithelial and mesenchymal components. The prenatal detection of congenital cystic adenomatoid malformation (CCAM) has significantly improved with advancements in ultrasonography and fetal imaging techniques, enabling early diagnosis and risk stratification. These diagnostic capabilities have also facilitated the development of antenatal therapeutic approaches aimed at managing life-threatening complications, such as hydrops and mediastinal compression, which can arise in severe cases [18,19].

5. CONCLUSION

Congenital cystic lung lesions are rare but significant developmental anomalies that are often detected prenatally through routine ultrasound and further evaluated with advanced imaging techniques such as fetal MRI and postnatal CT scans. Early and accurate diagnosis is crucial for determining the appropriate management, which ranges from conservative observation to surgical intervention, depending on the lesion's size and impact on lung function. While the overall prognosis for patients with CCLL is generally favorable, especially with timely treatment, continued research into early detection and the long-term outcomes of affected individuals remains essential for optimizing care and improving quality of life. This emphasizes the importance of early detection, appropriate management, and future research directions.

Declarations:

Conflicts of interest: There is not any conflict of interest associated with this studyConsent to participate: There is consent to participate.

Consent for publication: There is consent for the publication of this paper.

Authors' contributions: Author equally contributed the work.

REFERENCES

[1] Laberge JM, Flageole H, Pugash D, Khalife S, Blair G, Filiatrault D, et al. Outcome of the prenatally diagnosed congenital cystic adenomatoid lung malformation: A Canadian experience. Fetal Diagn Ther. 2001; 16:178–86.

[2] David M, Lamas-Pinheiro R, Henriques-Coelho T. Prenatal and Postnatal Management of Congenital Pulmonary Airway Malformation. Neonatology. 2016; 110(2):101-15

[3] Qu Y, Liu D, Jia H, Zhou X. Expression Analysis of ACSL5 and Wnt2B in Human Congenital Pulmonary Airway Malformations. J Surg Res. 2018 ;232:128-136

[4] Maneenil G, Ruangnapa K, Thatrimontrichai A, Janjindamai W, Dissaneevate S, Anantaseree W, Suntornlohanakul S. Clinical presentation and outcome in congenital pulmonary malformation: 25 year retrospective study in Thailand. Pediatr Int. 2019 ;61(8):812-816.

[5] Stocker JT. Congenital pulmonary airway malformation. A new name for an expanded classification of congenital cystic adenomatoid malformation of lung. Symposium : Non neoplastic lung disease. Histopathology. 2002; 41:424–31.

[6] al-Salem AH, Adu-Gyamfi Y, Grant CS. Congenital lobar emphysema. Can J Anaesth. 1990; 37:377–9.

[7] Pan F, Li J, Lou H, Li J, Jin Y, Wu T, et al. Geographical and socioeconomic factors influence the birth

prevalence of congenital heart disease: a population-based cross-sectional study in Eastern China. Curr Probl Cardiol. 2022; 47(11): 101341.

[8] Barman, Shibsankar et al. "Congenital cystic lesions of lung in the paediatric population: A 5-year single institutional study with review of literature." African journal of paediatric surgery :2015; AJPS vol. 12,1: 66-70.

[9] Lima, M et al. "Clinical spectrum and management of congenital pulmonary cystic lesions." La Pediatria medica e chirurgica : Medical and surgical pediatrics . 2008; vol. 30,2 : 79-88.

[10] Costa Junior et al. @- Costa Júnior A, Perfeito J, Forte V. Tratamento operatório de 60 pacientes com malformações pulmonares: o que aprendemos?. Jornal Brasileiro de Pneumologia. 2008; 34(9):661-666.

[11] Mohamed ElShabrawy Saleh et al. @ Saleh M, Beshir H, Awad G, ElDerie A, Sanad M.Surgical outcomes for pediatric congenital lung malformation: 13 years' experience.Indian Journal of Thoracic and Cardiovascular Surgery. 2020; 36(6):608-618.

[12] Gulla, K.M., Parihar, M.S., Jat, K.R. et al. Congenital Lung Malformations: Experience From a Tertiary Care Center in India. Indian Pediatr . 2021; 58: 129–133

[13] parikh - @ - Parikh D, Auldist A, Crabbe D, Rothenberg S. Pediatric Thoracic Surgery. London: Springer-Verlag London; 2009.

[14] Jain, Aditi et al. "Congenital cystic lung diseases." Journal of clinical imaging science . 2013; vol. 3 5. 30

[15] Faten Limaïem et al. - @- Limaïem F, Ayadi-Kaddour A, Djilani H, Kilani T, Mezni

[16] F.Pulmonary and Mediastinal Bronchogenic Cysts: A Clinicopathologic Study of 33 Cases. Lung. 2007; 186(1):55-61.

[17] natural history of cong cystic lesion james cook- @- Cook J, Chitty L, De Coppi P, Ashworth M, Wallis C. The natural history of prenatally diagnosed congenital cystic lung lesions: long-term follow-up of 119 cases. Archives of Disease in Childhood. 2017; 102(9):798-803.

[18] Pooja A. Mehta; Girish Sharma. Congenital Pulmonary Airway Malformation. 2023.

[19] Marcos Mestas Nuñez. Congenital Lung Anomalies in Adults. 2024.

[20] Anna Luiza Vidal França et al. Antenatal Therapies in Congenital Pulmonary Cystic Adenomatoid Malformation: Current Knowledge. Women . 2025; 5(1), 7.