

## What Do You Know About the Imaging Findings in Congenital Pulmonary Malformations in Children?

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DOI: [10.36347/sjmcr.2020.v08i07.017](https://doi.org/10.36347/sjmcr.2020.v08i07.017)

| Received: 08.07.2020 | Accepted: 16.07.2020 | Published: 28.07.2020

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### Abstract

### Original Research Article

**Aim:** Determine the epidemiological, radiological, therapeutic and progressive profile of bronchopulmonary malformations. **Materials and methods:** This is a retrospective study over 10 years (03/2010 to 10/2019), including 20 children hospitalized in our institution for bronchopulmonary malformation. **Results:** We have identified 7 cases of adenomatoid cystic lung malformations, 3 cases of congenital lobar emphysema, 3 cases of pulmonary sequestration in its extra lobar form, 3 cases of bronchogenic cyst, 1 case of pulmonary sequestration associated with cystic adenomatoid malformations, 1 case of Pulmonary hypoplasia, 1 case of pulmonary agenesis and 1 case of pulmonary arteriovenous malformation. The prenatal diagnosis was carried out in a single patient, other patient benefited from a postnatal diagnosis with an average age of 26 months. All patients were symptomatic with an average age of 14 months. The symptomatology was dominated by respiratory distress, and respiratory infection. The chest X-ray guided the diagnosis in most cases and the CT scan confirmed the diagnosis of all cases. Lobectomy was performed in 67% of the patients. The pathological examination confirmed the diagnosis of the malformation in all cases. The post-operative and long-term outcome, with a follow-up ranging from 3 months to 3 years, was favorable in most of the patients followed. **Conclusion:** Imaging has a fundamental role in the management of congenital pulmonary malformations, in their diagnosis and in the assessment of surgical forms. CT is the modality of choice, thanks to its ease of access, its speed of acquisition, its high spatial resolution that it offers.

**Keywords:** Malformations, lung, imaging.

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## INTRODUCTION

Congenital lung malformations are rare [1, 2]. They are related to abnormalities in the development of the tracheobronchial tree. Their overall frequency is difficult to establish because they can remain asymptomatic and unknown. Since the advent of antenatal ultrasound, the diagnosis has become early, allowing neonatal management [3, 4], however, a number of these malformations can remain pauci- even asymptomatic and of fortuitous discovery during an imaging examination. Chest x-rays play an important role in the incidental discovery of malformities pulmonary diseases. However, sectional imaging is most often required for confirmation, characterization and possible preoperative assessment of these lesions. The purpose of our work is to illustrate the clinical and radiological aspects of these malformations.

## MATERIAL AND METHODS

This is a retrospective, descriptive study, relating to 20 cases of congenital pulmonary

malformations, collected in our institution over a period of 10 years, going from July 2010 to October 2019. A chest X-ray and chest CT scan were performed on all of our patients.

### Inclusion criteria

Age less than 15 years, patients with bronchopulmonary malformation confirmed radiologically or histologically, treated bronchopulmonary malformations.

Data collection was carried out from patient files using a pre-established sheet (see appendices). The data processing was done by software: Excel. The results were expressed as percentages or as means depending on the studied variables.

## RESULTS

During the period from January 2010 to October 2019, twenty cases of congenital lung malformations were collected; the age of our patients

was between 16 days and 9 years, with an average of 2 months with a slight male predominance (sex ratio of 1.25). Clinical symptomatology was dominated by respiratory distress (n = 11), followed by respiratory infections (n = 8) and hemoptysis (n = 2). We found 7 cases of cystic adenomatoid lung malformations (type 1) (35%), 3 cases of congenital lobar emphysema (15%), 3 cases of extra-lobar pulmonary sequestration (15%), 3 cases of cyst bronchogenic (15%), 1 case of pulmonary sequestration associated with cystic adenomatoid malformations (5%), 1 case of pulmonary hypoplasia (5%), 1 case of pulmonary agenesis (5%) and 1 case of arteriovenous malformation (5%).

Imaging data are summarized in Table 1.

## DISCUSSION

### Giant lobar emphysema

a. Definition: This anomaly is the result of a progressive distension of a pulmonary territory (segment, lobe, lung) by a valve mechanism in connection with an incomplete, intrinsic or extrinsic bronchial obstruction, or with a pathology of the tracheal cartilage [5,6]. The prevalence of GLE is estimated at 1 / 20,000 to 1 / 30,000 births [7]. It can be associated with heart defects in 14% of cases [8]. Although a few family cases have been reported [9], none of our patients has a family history of GLE. It affects three times more boys than girls [9]. This male predominance is found in our study.

b. Clinical manifestations: Giant lobar emphysema is rarely diagnosed during the antenatal period; its hyperechoic aspect cannot be distinguished from microcystic CCAM [9].

Most often, it manifests in neonatal period (50% of cases), or in the first months of life (80% before 6 months) and consists of respiratory distress evolving by successive episodes, of increasing severity. The same observation was made in our series; neonatal dyspnea and respiratory distress were the revealing modes of GLE [10].

### c. Imaging Chest X ray

The chest x-ray is the essential diagnostic test. It shows the asymmetry of pulmonary transparency, with a large clear circumscribed pulmonary range. This distended territory represses the adjacent parenchyma, often lowers the diaphragmatic dome, and spreads the intercostal spaces. These compression elements increase on expiration, confirming the trapping. Sometimes the distension is very large, with the appearance of a large, clear hemithorax (Figure 1). In our patients, hyperclarity was the constant radiological sign [11]. The presence of vascular elements within the distended parenchyma is a very important radiological sign, which makes it possible to differentiate with a pneumothorax or a pulmonary cyst [11].

### Computed tomography

It specifies the topography of the pulmonary distension in order to schedule the surgery. The pathological zone appears in the form of a systematized, hypodense, pulmonary zone within which the pulmonary architecture is preserved, more or less distorted [12]. (Figure- 2) The associated signs are atelectasis of compression of the adjacent lung segments or lobes, deviation of the mediastinum contralateral, enlargement of the intercostal spaces and diaphragmatic depression [13].

### d. Treatment

In the majority of cases, surgical intervention is compulsory. The intervention consists in the excision of the emphysematous lesion [11]. The externalization of the lung makes it possible to obtain an often spectacular improvement and lobectomy is the most frequent gesture. Other treatments have been proposed; these are very sporadic cases (bronchotomy, bronchoplasty) [11].

### e. Evolution

After surgical treatment, the evolution is favorable, the growth of the remaining lung can reach 90% [14].

## 2. Pulmonary cystic adenomatoid malformation

a. Definition: This anomaly, long unrecognized, was described by Chin-Tang in 1949, corresponding to the old Craig's disease: parenchymal mass due to adenomatoid (glanduliform) proliferation of the terminal bronchioles, in connection with a localized maturation arrest of the bronchial tree at the glandular stage, and evolution towards a more or less cystic mass. This non-functional structure can communicate with the aerial tree and evolves postnatal to aeric cysts [11]. This anomaly occurred sporadically with an incidence of approximately one in 25,000 to one in 35,000 pregnancies. It affects both sexes indifferently [15].

b. Classification Stocker described the histological aspect, first in three types (1977), then in five types, which he relates to stages of stop of maturation of the bronchial tree [11, 16]

### c. Diagnostic

Antenatal: PCAM is the most common malformation found in antenatal screening [9]. Antenatal ultrasound shows, from the 20th AW, a lung mass made up of several anechoic images of variable size. Classified by Adzick into three groups according to the size of the cysts [17]:

- type I: Macrocystic PCAM: cysts greater than 5 mm in diameter
- type II: Association of large cysts and small cysts
- type III: microcystic PCAM: cysts smaller than 5 mm.

In post-natal: In the neonatal period, this malformation can be responsible for respiratory distress. In children, it is revealed by progressive dyspnea or by recurrent lung infections focalized in the same localization. In our series, it is revealed mainly by respiratory distress [9].

### Imaging

**Chest x ray:** The radiography can show a more or less homogeneous opacity which will gradually aerate with the appearance of one or more cystic lesions of variable size depending on the histological type. It can be responsible for a large clear lung if the walls are thin, imperceptible in chest x ray [18].

**Computed tomography:** PCAM are most often unilateral and unifocal, restricted to a pulmonary lobe. They are described indifferently in the right and left pulmonary lobes, and seem to predominate in the lower lobes [19]. Which is confirmed in our series. In macrocystic forms (type 1 and 2), it's characterized by well-defined aeric cystic structures, with thin walls, with at least one lesion of more than 20 mm for type 1 and lesions of homogeneous size, measuring between 5 and 20 mm for type 2. In type 3, the microcysts are indistinguishable and form a heterogeneous mass of condensation with ill-defined contours. Type 4, which usually appears as large cysts, cannot be distinguished from a cystic form of pleuropulmonary blastoma [20] (Figure 3).

Systemic arterial vascularization should be systematically sought, in particular in types 2. The presence of thickened walls, enhanced after injection of contrast medium and / or of a hydro-aeric level can testify to an infection. Pneumothorax is a rare complication but can be revealing [12].

**d. Treatment:** The treatment is surgical and indisputable in the symptomatic forms, sometimes constituting a neonatal emergency. The therapeutic indication is more difficult to specify in the case of an asymptomatic form: monitoring may be adopted for the most minor forms. [11].

**e. Evolution:** The natural evolution leads to air distension, sometimes to pneumothorax, pneumonia by infection, signs of mediastinal compression. Hydatidosis and aspergillosis transplants have been described. Late tumor degeneration has been reported [11].

### Pulmonary sequestration

**a. Definition:** This type of anomaly, defined by Pryce in 1946, corresponds to an abnormal non-functioning pulmonary territory that has lost its connections with the tracheobronchial tree and whose arterial vascularization is systemic, most often of aortic origin. This is the likely persistence of a primitive systemic vessel [11]. The venous return is most often made in the azygos or the inferior vena cava [21]. There

are two types of sequestration: intralobar and extralobar. Intralobar sequestration (80%) is an isolated, non-functional pulmonary segment without communication with the airways or the adjacent normal parenchyma, and irrigated by one or more abnormal systemic arteries [22]. It does not have a clean pleural envelope. The extralobar form is less frequent (20%). It corresponds to an aberrant mesenchyme, which develops autonomously, without connection with the normal lung. It has its own pleural envelope. In practice, there is a possible overlap of the two forms [23]. The two cases of pulmonary sequestration in our series, the one isolated and the other associated with PCAM, were intra-lobar forms.

Extralobar forms are associated more frequently and in 60% of cases with congenital anomalies, in particular cardiac, pulmonary and diaphragmatic. Intralobar forms can also be associated with other types of lung malformations, such as PCAM [23]. In our series, both patients had an associated malformation, PCAM and malformative uropathy. Thus, certain teams propose the realization of a systematic karyotype in the event of confinement.

### b. Diagnostic

- **Prenatal:** Antenatal diagnosis is frequent. Pulmonary sequestration appears, in its typical form, as a homogeneous triangular mass left posterobasal hyperechoic on ultrasound, and hyperintense on MRI. The diagnosis of certainty is made by the identification of the systemic feeding artery in MRI, easily carried out by an experienced operator, or more simply by Doppler in ultrasound [21].
- **Postnatal:** At birth, sequestrations are asymptomatic in 80% of cases. After the neonatal period, sequestrations can remain asymptomatic in 10 to 15% of cases [24]. Regarding the symptomatic forms, the clinical signs are comparable to those encountered during PCAM.

### c. Imaging

**Chest x-ray:** PS can take on several aspects, the most frequent of which is a homogeneous opacity of round, oval or triangular liquid density and perfectly delimited. Elsewhere, it may be heterogeneous opacity with gas clarity, images of segmental atelectasis secondary to inflammatory episodes, or images of pneumonia [24]. In our series, the chest x-ray of the isolated sequestration did not show a specific aspect.

**Computed tomography:** It allows the diagnosis of certainty and the characterization of the anomaly. The pulmonary lesion is localized in the lower lobes in 98% of the cases, the left side being more frequently affected (75%), in general in the postero-basal and paracardiac segments [13]. (Figure- 4). Extralobar sequestrations, most often of intra-thoracic location, between the left lower lobe and the diaphragm, can also

be located in the abdomen, anterior mediastinum or posterior mediastinum [12].

The CT aspect is variable; most often, there is a zone of pulmonary condensation, homogeneous or heterogeneous which can be associated with a peripheral aeric trapping, with a bronchocele, with a liquid bronchogram or, in border forms with PCAM, with air cystic lesions or with fluid content [12]. Computed tomography allows the visualization and characterization of systemic vessels as well as the evaluation of venous return [12].

The two types of sequestration are vascularized by an abnormal systemic artery which originates in the majority of cases of the descending thoracic or abdominal aorta or, occasionally, of the celiac trunk, of a splenic, intercostal, subclavian, mammary or even coronary. In 20% of cases, several vessels are present [25].

Venous drainage is variable. In the intra-lobar form, it most often takes place towards the pulmonary venous system, at the level of the lower pulmonary vein. In extralobar sequestration, it can be pulmonary or systemic, generally at the level of the cave system, azygos, less frequently at the portal level, subclavian or even at the level of the mammary veins. The coexistence of the two types of venous drainage is possible [25].

A precise and detailed description of the vascularization is essential in the programming of treatment, whether surgical or interventional radiology, particular attention must be paid to the description of the number of systemic arteries and the existence of early divisions [12,36] (Figure 5).

**d. Treatment:** For persistent sequestrations at birth, the therapeutic choice is made between radiological embolization or excision surgery. Whichever option is chosen, treatment is scheduled and deferred: a chest scanner must be performed around 2 months of life [26] (Figure 8).

**e. Evolution:** The monitoring to be put in place is the same as for other pulmonary malformations, but in the event of a large receiver with a significant flow in the artery which supplies the mass, it is necessary to be wary of the occurrence of a possible "shunts effect". With the onset of heart failure. This complication is rare with a survival rate of sequestrations, all cases taken together, is 13 to 25% due to the rate of associated malformations [27].

#### Bronchogenic cyst

**a. Definition:** These are isolated cystic structures, filled with fluid or mucus [21]. These are infrequent anomalies. They represent 10% of the

mediastinal masses encountered in children. A male predominance is reported [9]. Associated anomalies are frequent, 25%, either of the pulmonary parenchyma or other remote anomalies [11].

#### b. Diagnostic

- Prenatal: Bronchogenic cysts are rarely discovered. The average age of diagnosis is 26 weeks of amenorrhea [9]. Antenatal ultrasound shows an isolated unilocular thin-walled anechoic cystic image, usually posterior mediastinal. On MRI, there is a homogeneous cystic image in T2 hyperintensity [28].
- Postnatal: The bronchogenic cyst does not have its own symptomatology. The possible symptoms are linked to the mass effect of the cyst on the adjacent structures [28] and therefore depend on its size and location.

#### c. Imaging

**Chest x ray:** It appears in the form of a medio-mediastinal or intra-parenchymal mass, of water tone, with clear contours, associated with indirect signs such as an enlargement of the hull, stretching and compression of the stem bronchi, atelectasis or air trapping [26].

**Computed tomography:** Solitary, rounded or oval, well limited lesion, the content of which appears homogeneous and of fluid density, from 0 to 20 Hounsfield units (HU) in 50% of cases or higher due to a protein or calcium composition [28]. The cystic walls are fine or even imperceptible, they can rarely calcify. The content of the lesion does not enhance after injection of contrast medium. Air content in the cyst is rare and indicates communication with the air or intestinal tract system.

A hydro-aeric level, heterogeneous, partitioned or solid content, thickening and intense enhancement of the walls are most often signs of infection.

The computed tomography must analyze the adjacent structures within the framework of the pre-surgical assessment and look in particular for a mass effect on the tracheobronchial tree responsible for pulmonary ventilation disorders such as atelectasis or air trapping.

**d. Treatment:** In case of a pulmonary site, excision of the cyst is possible when it is proximal; on the other hand, in case of a more peripheral localization, a parenchymal resection is often necessary, especially as the BC can take on the appearance of a pulmonary cyst [11].

**c. Evolution:** Superinfection is frequent, sometimes leading to a real abscess and for some fistulization in the bronchus is secondary [11]. The prognosis is

generally excellent, but monitoring is useful, especially if the cyst has caused localized tracheomalacia [11].

### Pulmonary agenesis/hypoplasia

- a. **Definition:** Pulmonary agenesis is the complete absence of the lung parenchyma, bronchus and lung vasculature while pulmonary hypoplasia is defined by the presence of a bronchus and rudimentary lung parenchyma with a reduction in number and size of airways, alveoli and pulmonary vasculature [28].
- b. **Diagnosis:** The clinical presentation is variable. If the defect is focal and isolated, the infant may not have any symptoms, but usually some mild respiratory distress is present.
- c. **Imaging :** A chest radiograph reveals unilateral lung or lobar collapse with a shift of mediastinal structures, which leads to a suspicion of bronchial or bronchiolar obstruction. Misdiagnosis may subject the infant to the unnecessary risks of bronchoscopy when CT is readily diagnostic (figure 6). Associated anomalies in the cardiovascular, gastrointestinal, genitourinary, central nervous and musculoskeletal systems have all been described [29].
- d. **Treatment:** If the defect is isolated to a single lobe, surgical resection will reduce symptoms and lessen the chance for infection. If the defect is extensive but the fetus is considered salvageable, an ex-utero intrapartum therapy procedure may be performed [29].
- e. **Prognosis:** Prognosis depends on the degree of pulmonary involvement, a history of recurrent pulmonary infections, and the presence of associated anomalies. Bilateral defects are invariably lethal. If the defect is focal, the remaining normal lung tends to hypertrophy to compensate. Still, mortality rates exceed 50%, generally because of the presence of associated malformations, which are common [30].

## CONCLUSION

Imaging has a fundamental role in the management of congenital pulmonary malformations, in their diagnosis, characterization and in the assessment of surgical forms. Thanks to its ease of access, its speed of acquisition, its high spatial resolution and the many possibilities of three-dimensional and voluminal reconstruction it offers, computed tomography is the modality of choice. In addition, the development of iterative reconstructive software will ultimately result in a considerable reduction of the doses of radiation delivered.



Fig-1: Chest X-ray frontal view showing congenital lobar emphysema of the right middle lobe

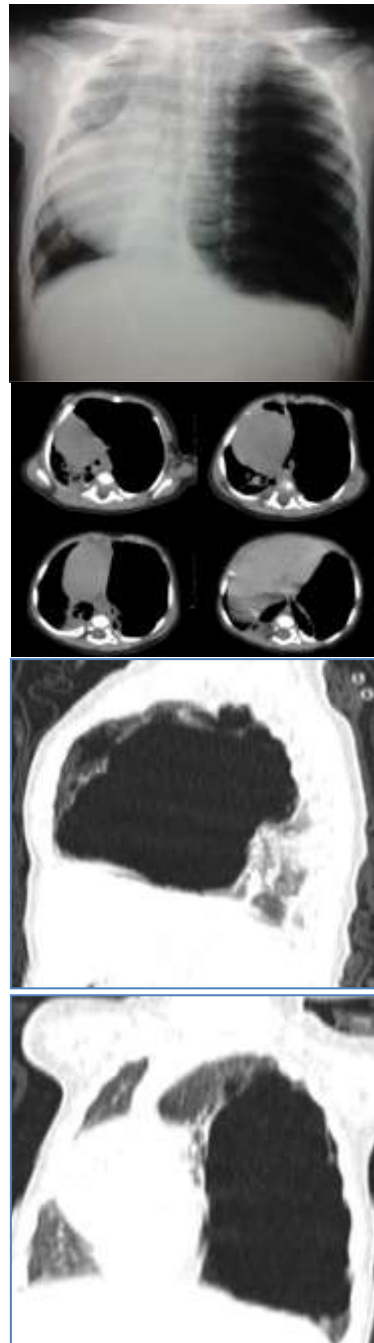


Fig-2: 2-month-old infant, female

A - Front chest x-ray showing hyperclarity of the left lung with repression of the heart and right mediastina and enlargement of the intercostal space.

B- Thoracic CT in mediastinal window in axial sections showing an expansion of the left lung responsible for repression of the mediastinum and contralateral lung with anterior transmediastinal hernia.

C- Thoracic CT in parenchymal window in sagittal section ( right) and coronal (left) showing an emphysema bubble occupying almost the entire left pulmonary parenchyma.

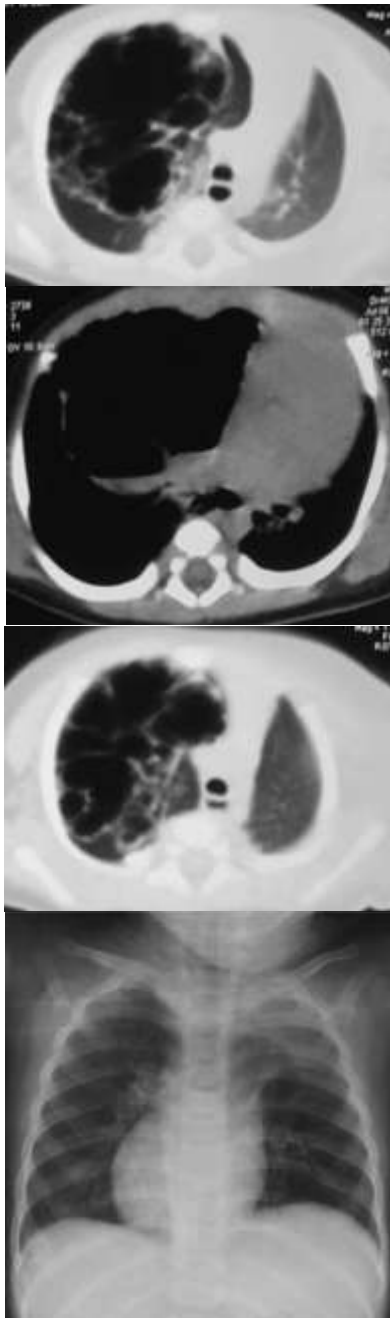


Fig-3: Newborn, (7 days old), who has respiratory distress

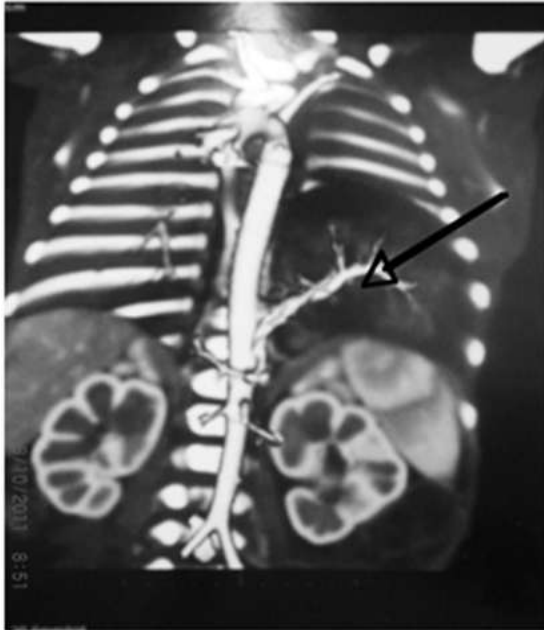
A - Chest x ray showing hyper clarity of the right upper lobe.

B- Thoracic CT in mediastinal window in axial section showing an expansion of the right lung driving the elements of the mediastinum towards the contralateral side with transmediastinal hernia.

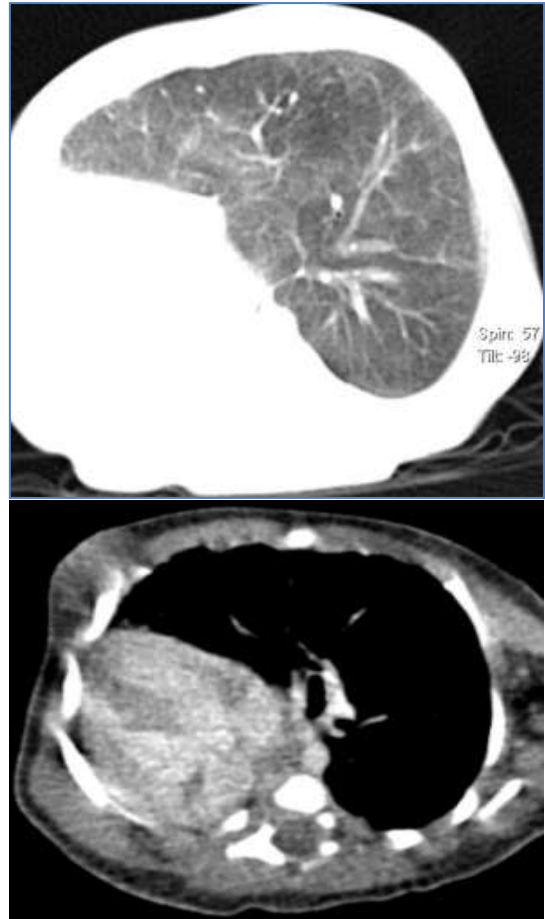
C- Chest CT in parenchymal window in axial section showing bubbles of upper and middle lobar emphysema driving elements of the mediastinum towards the contralateral side with anterior transmediastinal hernia



Fig-4: 7-year-old male child with a history of malformative uropathy operated at one year of age and asthma under treatment who presented to pediatric emergency departments for recurrent respiratory infections complicated by episodes of hemoptysis. A -Front chest x ray objectifying a diffuse bronchial condensation B-Thoracic CT: objectifying a focal zone of triangular condensation of the left vertebral groove, presenting a systemic vascularization starting at the level of the aorta evoking an intra-lobar pulmonary sequestration.



**Fig-5: Thoracic CT Angioscan showing an intra-lobular sequestration**



**Fig-6: Newborn 3 weeks old, who has respiratory distress. Front chest X-ray (A): Right opaque and retracted hemi-pulmonary field, deviation of the mediastinal structures towards the affected side, hyperclary left lung with trans-mediastinal hernia. Thoracic CT in axial sections: (injected mediastinal window B and parenchymatous C): Right pulmonary agenesis with an aplastic root bronchus and increase in the left pulmonary volume with right anterior trans-mediastinal hernia and total repression of the elements of the mediastinum on the right**

**Table-1: Imaging data of different lung malformations**

| <b>Pulmonary malformation</b>  | <b>Chest X Ray</b>   | <b>Thoracic CT</b>  |
|--------------------------------|--|---|
| PCAM                           | -Left basithoracic opacity (3 cases) / right basithoracic (1 case), well limited and with polylobed contours (1 case)<br><br>-Avascular hyperclarity of the right lung (2 cases) / left lung (1 case), pushing the elements of the mediastinum towards the contralateral side. | -Voluminous multi-cystic right pulmonary mass (4 cases) / left (3 cases), with clean walls, responsible for a displacement of the mediastinum towards the contralateral side.<br><br>-A trans-mediastinal hernia was associated in one case.  |
| Congenital lobar emphysema     | -Hyperclarity in the right (2 cases) and left (1 case) pulmonary field.<br><br>-A transmediastinal hernia was associated in one case<br><br>Homolateral atelectasis was found in 1 case.   | -CLE of the upper left lobe in 1 case, of the right upper lobe in 1 case and of the middle lobe in 1 case: Parenchymal hyperclarity, rarefaction and small aspect of the vascular structures in the affected lobe (s).<br><br>-Trans-mediastinal hernia has been associated in one case.  |
| Pulmonary sequestration        | -Left retrocardiac dilated vascular structure (1 case)<br><br>-Posterior basi-thoracic opacity, homogeneous and well limited (2 cases).  | -Solid mass of the left costo-vertebral gutter left (1 case)<br><br>-Multiple cystic images juxtaposed with thin walls and with air content (2 cases)<br><br>-Arterial vasculature made by arterial branches from the thoracic aorta, venous drainage by inferior vena cava (2 cases) / pulmonary vein (1 case)                                 |
| Bronchogenic cyst              | -Bullous image of the right pulmonary base (2 cases)<br><br>-Posterior mediastinal fluid tone opacity associated with a grayish appearance of the left pulmonary hemichamps evoking a pleural effusion (1 case).   | -Unilocular bronchogenic cyst of the right lower lobe (1 case) / middle lobe (1 case), confirmed histologically.<br><br>-Cystic mass with homogeneous liquid content and thickened wall and intensely enhanced after injection of contrast medium associated with ipsilateral pleural fluid effusion (1 case).                                  |
| PCAM + Pulmonary sequestration | -Large cavity of air density of the left lower lobe with nodular images in the rest of the pulmonary parenchyma.   | -Multiple cystic cavities of the left lung of air density thin wall, seat of a thin partition.<br><br>-Parenchymatous rounded Condensation under pleural compared to the left costo-vertebral angle, seat of clarity with individualization of a vascular structure within it in relation to a systemic branch arising from the abdominal aorta |
| Pulmonary agenesis             | -Right opaque and retracted lung<br><br>-Deflection of mediastinal structures towards the affected side.<br><br>- Hyperclear left lung with trans-mediastinal hernia.  | -Right pulmonary agenesis with a bronchial aplastic strain and a total repression of mediastinal elements to the right.   |
| Pulmonary hypoplasia           | -  | Reduced Left hemithorax with deflection of the mediastinum to the affected side.  |

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