

# CHAPTER 44

## CONGENITAL CYSTIC LUNG LESIONS

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### Introduction

Congenital lesions of the lung are rare with an unknown true rate of incidence. An overall incidence of 1/10,000 to 1/25,000 births (and 2.2% when compared to acquired lesions) has been reported. Presentation varies from life-threatening symptoms at birth to incidental findings at autopsy. Diagnosis is either made in utero or due to complications of the lesion, such as lung abscess, pneumonia, or pneumothorax. At an early stage, there is a good argument for describing these lesions simply as cystic lung malformations because the precise diagnosis may need to await later pathological examination or other postnatal investigations. Embryology and classification of the lesions are attempted. Classic lesions of congenital cystic or pulmonary adenomatous malformation (CCAM or CPAM) of the lung, bronchopulmonary sequestration (BPS), congenital lobar emphysema/overinflation, congenital lung cysts, and other less common anomalies are described. Although described as a number of separate and seemingly distinct entities, these lesions often overlap.

### Embryology

In the third week of the embryonic phase, the lung appears as a ventral out-pouching of the endodermal foregut. At the end of the fourth gestational week, the caudal growth of the respiratory diverticulum becomes separated into the dorsal oesophagus and ventral trachea by the formation of the tracheo-oesophageal septum from the tracheo-oesophageal septum (Figure 44.1). The larynx, which develops from the fourth to the sixth branchial arches, maintains communication with the trachea. Pleural cavities are formed on either side of the foregut by caudal migration of the developing lung bud into the coelomic cavity. At the 6th embryonic week the right lung bud divides into three lobes and the left bud into two. Pulmonary vasculature develops in the mesoderm at 7–8 weeks of gestation. Bronchial division, which forms the conductive airways, is complete in mid-trimester, and the terminal airways and alveoli, which are the sites for gaseous exchange, continue to develop until early childhood (i.e., 8 years of age). Gaseous exchange is possible at 7 months gestation.

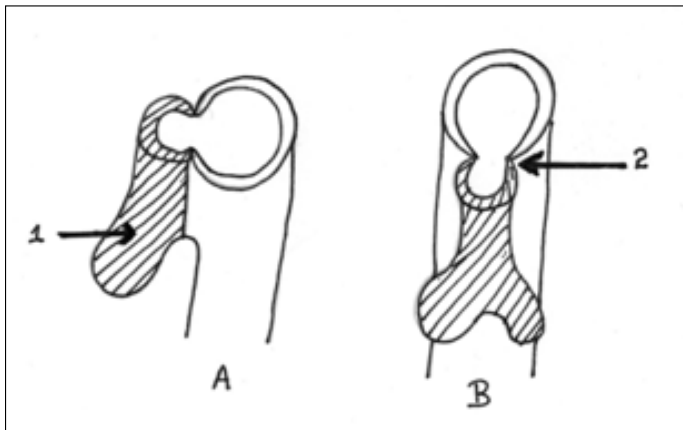


Figure 44.1: Embryology of lung bud: (A) week 3, showing lung bud (1); (B) week 4, showing tracheo-oesophageal septum (2).

### Classification

The historic histologic classification by Stocker (type 0 = total lung involvement; type 1 = >2 cm cyst; type 2 = <2 cm cyst; type 3 = <0.5 cm cyst; type 4 = >5 cm cyst) and prenatal ultrasound classification of Adzick (macrocytic = >5 mm; microcytic = <5 mm) have been replaced by a pathological classification recently proposed by Langston (Table 44.1).

### Pathogenesis

The pathogenesis of congenital cystic lung lesions is controversial, and many theories have been proposed. The historic vascular traction theory, vascular insufficiency theory, vascular maldevelopment theory, accessory bud theory, and the more recent molecular markers and signalling protein theories have not been confirmed.

### Prenatal Diagnosis and Management of Congenital Lung Lesions

Most congenital lung cysts are diagnosed at the 20-week anomaly scan. The diagnostic accuracy is 100% for congenital cysts, but specificity for distinct lesions are variable. Doppler ultrasound may identify an abnormal vessel from the aorta to suspect pulmonary sequestration, but it cannot confirm hybrid (mixed) lesions. Large lesions may cause cardiac compression, resulting in hydrops foetalis and fetal demise. Fetal intervention, fortunately, is indicated in only 10% of prenatally diagnosed lesions that are at risk of developing hydrops foetalis. These are amenable to foetal intervention through thoracentesis, pleuroamniotic shunting, or laser ablation of the feeding artery. Postnatal symptomatic lesions are subject to early surgical treatment; however, the management of postnatal asymptomatic lesions remains controversial. Most centres now propose postnatal computed tomography (CT) scan imaging followed by surgery at 3–6 months of age for asymptomatic lesions due to the risk of infection and malignancy. Small lesions of less than 1 cm on CT scan may be managed conservatively. Proponents of conservative management follow all asymptomatic lesions with serial CT scanning (Figure 44.2).

### Congenital Cystic Adenomatous Malformation

Congenital cystic adenomatous malformation (CCAM) is the commonest congenital lung lesion, accounting for 50–70% of these lesions. It is a hamartomatous malformation characterised by the lack of normal alveoli and an excessive proliferation and cystic dilatation of terminal respiratory bronchioles with varying types of epithelial lining. The lesions are mainly cystic and intrapulmonary, usually unilobar with a slight predilection for the lower lobes of the lung. The side of the lung, race, and gender are equally affected. In the absence of prenatal scanning the presentation may be at birth with respiratory distress. Distinction from diaphragmatic hernia may be assisted by the position of the stomach on chest radiograph (Figure 44.3). Multicystic lesions are usually noted on chest radiograph. Presentation outside the neonatal period includes nonresolving pneumonia, lung abscesses, empyema, reactive lung disease, failure to thrive, and malignancy. At this stage,

Table 44.1: Langston's classification of congenital lung malformations.

**Bronchopulmonary malformation**

- Bronchogenic cyst (noncommunicating bronchopulmonary foregut malformation)
- Bronchial atresia
  - Isolated
  - With systemic arterial/venous connection (intralobar sequestration)
  - With connection to gastrointestinal tract (intra-lobar sequestration/complex or communicating bronchopulmonary foregut malformation)
  - Systemic arterial connection to normal lung
- **CCAM: large cyst type (Stocker type 1)**
  - Isolated
  - With systemic arterial/venous connection (hybrid lesion/intralobar sequestration)
- **CCAM: small cyst type (Stocker type 2)**
  - Isolated
  - With systemic arterial/venous connection (hybrid lesion/intralobar sequestration)
- **Extralobar sequestration**
  - Without connection to gastrointestinal tract (with/without CAM, small cyst type)
  - With connection to gastrointestinal tract (complex/communicating bronchopulmonary foregut malformation)

**Pulmonary hyperplasia and related lesions**

- Laryngeal atresia
- Solid or adenomatoid form of CCAM (Stocker type 3)
- Polyalveolar lobe

**Congenital lobar overinflation****Other cystic lesions**

- Lymphatic/lymphangiomatous cysts
- Enteric cysts
- Mesothelial cysts
- Simple parenchymal cysts
- Low-grade cystic pleuropulmonary blastoma

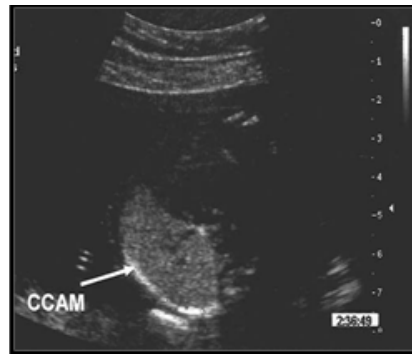


Figure 44.2: Prenatal ultrasound of CCAM.



Figure 44.3: Chest radiographs showing congenital diaphragmatic hernia (CDH) and congenital cystic adenomatous malformation of the lung (CCAM).

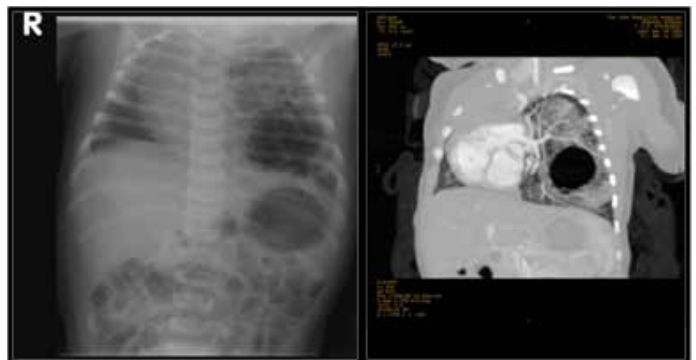


Figure 44.4: CCAM on chest radiograph (left) and CT scan (right).

a distinction should be made from infected pneumatocele, which resolves with antimicrobial treatment. In difficult cases, a CT scan may help in confirming the diagnosis (Figure 44.4).

**Surgical Procedure**

The surgical procedure is performed via thoracotomy or more recently thoroscopically. Lobectomy is preferred to segmentectomy to avoid recurrence of disease infection and development of malignancy in the incomplete resection. At surgery, abnormal vasculature should be looked for in the event of a hybrid lesion. Hybrid lesions are CCAM with intralobar pulmonary sequestration (see below). Prenatally diagnosed lesions are discussed above.

**Malignant Transformations with CCAM**

Bronchioloalveolar carcinoma (BAC) and rhabdomyosarcoma in association with CCAM have been repeatedly reported in children and adults. Since 1980, more than 25 cases of malignancy have been reported

in children as young as 1 month to 13 years of age. These malignant transformations were noted in primary CCAM lesions and those that were incompletely resected. The long-term malignant potential of in situ CCAMs has been further reported, suggesting postnatal surgical excision by means of lobectomy rather than segmentectomy as well as surgical preference to long-term radiological surveillance. Thus, with the increasing number of reports and case series of malignancy within CCAM, together with the possibility of lung infection, surgical resection in nearly all cases of CCAM is recommended.

**Bronchopulmonary Sequestration (BPS)**

Bronchopulmonary sequestrations (BPSs) make up 10–30% of congenital cystic lung lesions. These are solid, nonfunctioning congenital lung lesions that have a blood supply originating from the aorta rather than the pulmonary artery and an absence of communication with the bronchial tree (Figure 44.5). They can be subdivided into *intralobar* BPS or

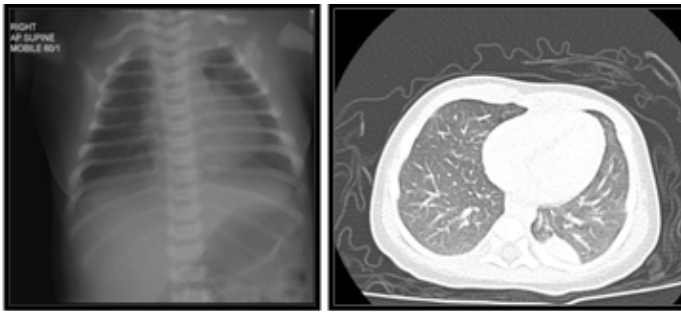


Figure 44.5: Bronchopulmonary sequestration on chest radiograph (left) and CT scan (right).

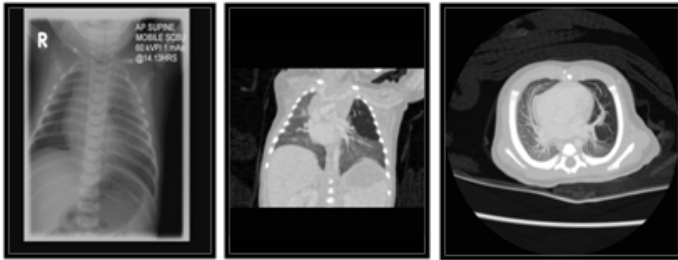


Figure 44.6: Congenital lobar emphysema on chest radiograph (left) and CT scan (right images).

extralobar BPS, with the former enveloped by normal lung parenchyma and the latter are an entirely separate entity with a complete covering of visceral pleura.

### Intralobar Sequestration

Intralobar sequestrations usually affect the left lower lobe with an aberrant vessel from the thoracic aorta; however, branches from the abdominal aorta, intercostal vessels and brachiocephalic vessels have been noted. Presentation includes nonresolving pneumonia, lung abscess, and, rarely, haemoptysis. Chest radiography demonstrates a nonaerated atelectatic mass, and a contrast CT should confirm the mass with an aberrant vessel. Management consist of resection via thoracotomy or thoracoscopy.

### Extralobar Sequestration

Extralobar sequestration is most commonly found in the left lower chest, with 80–90% above the diaphragm and 10–20% below. The aberrant blood supply is mainly from the thoracic aorta, and 20% arise from the abdominal aorta. Associated anomalies such as diaphragmatic hernia, cardiac defects, arterio-venous malformations, and other anomalies are present in 50% of extralobar sequestration. Presentation is often asymptomatic at birth, but due to arteriovenous shunting may develop congestive cardiac failure in infancy or hydrops prenatally. Chest radiography typically depicts a left posterior mediastinal mass; however, further imaging with contrast CT scan is recommended. A prenatal scan with Doppler ultrasound may identify the aberrant blood supply. Resection via thoracotomy/thoracoscopy in early infancy is recommended.

## Congenital Lobar Emphysema

Congenital lobar emphysema (CLE), also known as congenital lobar overinflation (CLO), is characterised by air trapping and overdistention of one or more pulmonary lobes, possibly secondary to a defect in the bronchial cartilage. Less frequent aetiology is extrinsic compression from lymphadenopathy, anomalous vessels, and masses. Antenatal diagnosis of CLE has been reported, and spontaneous regression in the third trimester may occur. The typical postnatal presentation is of respiratory distress, which may necessitate excision of the affected lobe. Asymptomatic patients may be managed expectantly. Diagnosis is confirmed on chest radiograph, and echocardiogram is recommended for the 15–20% associated with congenital heart disease (Figure 44.6).

## Bronchogenic Cyst

Bronchogenic cysts contain a lining of respiratory epithelium and have smooth muscle, glandular tissue, and cartilage in the wall and cysts. Bronchogenic cysts may share an embryological origin with that of foregut duplications. Such lesions are found in the mediastinum in up to two-thirds of cases, lying adjacent to the major airways, heart, or oesophagus, with the remainder found within the lung parenchyma. Most cases present postnatally, usually with pulmonary infection, but a proportion are diagnosed incidentally. Surgical excision is curative and can be achieved by segmentectomy, lobectomy, or simple cyst removal (peripheral lesions). Malignant transformation has not been reported in relation to these lesions.

## Hybrid Lesions

Many lateral series have shown that some cystic lung lesions have features of both CCAM and BPS, suggesting that they share the same developmental ancestry and perhaps represent two ends of a broad spectrum of pathology. Any time a congenital lung lesion is approached surgically, the surgeon should be aware of the possibility of an aberrant arterial supply, even if this is not demonstrated by advanced imaging techniques. The possibility of separate coexisting lesions should also be considered.

## Communicating Bronchopulmonary Foregut Malformations

Congenital bronchopulmonary foregut malformations are sequestrations that communicate with the upper digestive tract, usually the oesophagus. Diagnosis is suspected with recurrent chest infection and air-filled mass in the mediastinum. Upper gastrointestinal contrast may confirm the diagnosis.

## Surgical Technique

Detailed surgical technique is beyond the scope of this book; however, salient points for a left lower lobectomy are described as follows:

1. The patient is positioned in the right lateral decubitus position, left side up, with the arm extended and placed over the head and a rolled towel under the chest for support.
2. A posteriolateral incision is performed over the fourth or fifth intercostal space marked just lateral and below the nipple.
3. Latissimus muscle is divided along the line of incision with serratus anterior muscle spared if possible.
4. Rib space is identified by elevating the scapula to count the ribs and the space entered by dividing the intercostal muscles on the upper border of the lower rib to avoid the intercostal neurovascular bundle.
5. The pleura are entered without damaging the underlying lung, and access to the chest cavity is obtained by using a rib spreader.
6. The interlobar fissure is exposed and the lingular artery identified before the lower lobe artery is ligated and divided.
7. The lung is retracted anteriorly to expose the pulmonary ligament in the basilar region. The ligament is divided to expose the inferior pulmonary vein, which is again ligated and divided.
8. The bronchial attachment to the lobe is divided and sutured with non absorbable suture using the interrupted suturing technique. Air leaks are checked with saline test and lung inflation.
9. Damage to the adjacent structures such as pericardium, aorta and phrenic nerve is avoided.
10. A chest drain with an underwater seal is placed for drainage, and the wound is closed in layers following rib approximation with absorbable sutures. The drain is removed after 24hours provided no air leak is demonstrated.

## Outcome

The outcome for lung surgery is excellent except where previous infection existed. Air leaks, residual disease, infection, and damage to adjacent structures are noted morbidities.

## Evidence-Based Research

<b>Title</b>	Management of congenital lung lesions.
<b>Authors</b>	Stanton M, Davenport M
<b>Institution</b>	King's College, London UK
<b>Reference</b>	Early Hum Dev 2006; 82(5):289–295.
<b>Problem</b>	Conservative versus surgical management of prenatally diagnosed lesions
<b>Intervention</b>	Surgery
<b>Comparison/control (quality of evidence)</b>	Review
<b>Outcome/effect</b>	Surgery recommended to avoid long-term complications.

## Key Summary Points

1. Cystic lung lesions are rare.
2. Accepted classification is now descriptive.
3. Symptomatic lesions require surgical intervention.
4. Management of asymptomatic lesions is controversial; however, most institutions recommend operative treatment.
5. A chest radiograph may diagnose most symptomatic lesions. A CT scan, where available, should provide better imaging of the lesion.
6. Lobectomy or cystectomy is the treatment of choice in most conditions.
7. Morbidity is due to air leaks and infection, but overall outcomes are good.

## Suggested Reading

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