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# Congenital Malformations and Developmental Anomalies of the Lung

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**Abstract** Congenital malformations of the lung are a group of diverse, yet related, abnormalities which may involve the lung parenchyma, pulmonary vasculature, or a combination of both. They may be detected in fetal life, produce severe symptoms during infancy, or may not manifest symptomatically until adulthood. The goal of imaging is to demonstrate the various components of the malformation, to facilitate appropriate management. This article discusses the spectrum of congenital and developmental lung anomalies, their etio-pathogenesis, role of various imaging modalities, and characteristic radiological findings.

**Keywords** Congenital lung malformations · MDCT · Focal hyperlucency · Pulmonary vascular malformations · Congenital lobar hyperinflation · Bronchial atresia · Congenital pulmonary airway malformation

## Introduction

Congenital and developmental abnormalities of the lung encompass a heterogeneous group of conditions, which exhibit myriad differences, and yet have many features in common. They may involve different components of the lung—the parenchyma/airway component, arterial supply and/or venous drainage. Congenital lung malformations (CLM) have a reported annual incidence of 30–42 cases per 100,000 population [1, 2]. With emergence of prenatal ultrasonography (US), many cases are now detected in utero. The modes of clinical presentation vary widely—from respiratory distress at birth to incidental detection in an asymptomatic adult.

This article addresses basic concepts regarding the spectrum of CLM, etio-pathogenesis, approach to imaging, radiological findings, and recommended nomenclature and classification.

## Spectrum of Congenital Lung Malformations

CLM can show predominantly parenchymal or predominantly vascular anomalies, or a combination of both (Table 1).They occur as a continuum of abnormalities [3], with the presence of lung parenchymal abnormality and normal vasculature on one hand (in congenital pulmonary airway malformation-CPAM, and congenital lobar hyperinflation-CLH) and abnormal vasculature in the absence of parenchymal abnormality on the other hand (pulmonary arterio-venous malformations—AVMs). Many cases have features overlapping between two or more conditions ('hybrid' lesions)—for instance CPAM-like changes in an area of 'sequestered' lung [4, 5].

## Etiology and Pathogenesis

For a long time, the most widely accepted theory of etio-pathogenesis was that of defective budding, separation, and

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**Table 1** Spectrum of congenital lung malformations [21, 24]

| | |
|---|---|
| Predominantly parenchymal abnormalities | Congenital lobar hyperinflation (CLH) |
| | Congenital bronchial atresia |
| | Congenital pulmonary airway malformation (CPAM) |
| | Foregut duplication cysts (bronchogenic cyst) |
| | Airway abnormalities (laryngeal atresia/stenosis, tracheal atresia/stenosis) |
| Predominantly vascular abnormalities involving | |
| Pulmonary artery | Pulmonary agenesis–aplasia–hypoplasia complex, proximal interruption of pulmonary artery, pulmonary artery sling |
| Pulmonary vein | Pulmonary vein stenosis, venous varix |
| Both pulmonary artery and vein | Pulmonary arterio-venous malformation (AVM) |
| Combination of parenchymal and vascular abnormalities | Pulmonary sequestration (intralobar, extralobar) |
| | Scimitar syndrome/hypogenetic lung syndrome |

differentiation of primitive foregut structures [3, 6, 7]. Langston [8] proposed that many lesions could be explained to occur as a result of airway obstruction leading to secondary pulmonary dysplastic changes. Differences in the level, completeness, and timing of the obstructive event were postulated to result in various anomalies. The etiological role of vascular abnormality has also been suggested. The molecular mechanisms which regulate normal lung development and show altered patterns of expression in CLM have been the subject of research [9–11]. Familial association has also been reported [12]. It is likely that multiple such factors act in concert.

## Approach to Imaging

US is the primary imaging modality for fetal screening. It provides valuable information about the presence and size of a focal lung lesion (lesion volume) [13], mass effect, associated hydrops fetalis, lung hypoplasia, and other organ malformations, all of which affect the prognosis and management [14, 15•]. Fetal magnetic resonance (MR) imaging is helpful—in select cases—for lung volume quantification, in addition to evaluation of the lesion [16•, 17, 18].

Chest radiograph is the first line of investigation in a symptomatic child. Congenital anomalies may exhibit varied appearances [19–21]—like focal hyperlucency, presence of fluid or air-filled cystic (or solid-appearing) lesions, vascular anomaly, airway abnormality or thoracic asymmetry. Radiographic differential diagnosis of few common CLM is given in Table 2. Radiographic findings should be carefully interpreted, compared with previous radiographs, and, when necessary, should prompt further evaluation using cross-sectional imaging techniques like US, multidetector-computed tomography (MDCT) or MR imaging.

US may be very useful in newborns and infants. It is radiation-free, and focal pulmonary lesions can be well assessed. Doppler sonography delineates vascular supply. High-resolution linear transducers are used, and transsternal, parasternal or intercostal approaches may be utilized [22, 23]. However, the US has limited utility in older children.

The benefits of fast speed, high spatial resolution and volumetric imaging with multiplanar and 3D reconstructions make MDCT the technique of choice for evaluation of CLM [21, 24]. It enables high-resolution images of the pulmonary parenchyma, with simultaneous evaluation of the vascular anatomy using CT angiography. The major limitation is radiation exposure. In the pediatric patient, there should be strict adherence to the as low as reasonably achievable (ALARA) principle [25, 26]. Exposure factors (kilovoltage and milliamperage) should be titrated against the age and weight of the child. Evaluation in a single phase (post-contrast) usually suffices, and multiphasic CT examinations should be avoided.

MR imaging scores over MDCT as it does not involve exposure to radiation. It has excellent contrast resolution. However, its spatial resolution is inferior to CT and details of lung parenchyma are not well assessed [21]. Scan times are long. MR imaging may be utilized in select cases of predominantly solid and vascular CLMs and as a follow-up imaging technique.

Digital subtraction angiography (DSA) is not usually required for diagnostic purposes; however, may be performed prior to endovascular embolization for vascular anomalies like pulmonary AVMs.

## Parenchymal (Non-vascular) Anomalies

### Congenital Lobar Hyperinflation

CLH was previously known as congenital lobar emphysema, but the term 'emphysema' implies destruction of alveoli, which does not occur in this condition. Though no cause is identifiable in approximately half the cases, this condition is thought to result from bronchial narrowing due to intrinsic (cartilage) abnormality or extrinsic compression, leading to progressive over-inflation due to a 'ball-

**Table 2** Common locations and radiographic differential diagnosis of few common CLM

| Malformation | Common location | Radiographic differential diagnosis |
|---|---|---|
| Congenital lobar hyperinflation (CLH) | Left upper lobe (42 %), middle lobe (35 %), right upper lobe (21 %) | Loculated pneumothorax |
| | (uncommon in lower lobes—<1 %) | |
| | | Congenital pulmonary airway malformation (CPAM) |
| | | Pneumatocele |
| Congenital pulmonary airway malformation (CPAM) | No lobar predilection | CLH |
| | Usually involves single lobe | Loculated pneumothorax |
| | Bilobar/bilateral involvement uncommon (<5 %) | Pulmonary sequestration (if located in lower lobe) |
| | | Congenital diaphragmatic hernia (if located in lower lobe) |
| | | Intrapulmonary bronchogenic cyst/ infected hydatid cyst/ lung abscess (if infected) |
| Pulmonary sequestration | | |
| Intralobar sequestration | 98 % cases occur in lower lobes (left > right), typically in posterior basal segment | Pneumonia |
| | Radiographic appearances are variable | Lung cyst—CPAM, bronchogenic cyst |
| | | Lung mass |
| Extralobar sequestration | Classically in the posterior costodiaphragmatic sulcus between left lower lobe and hemidiaphragm (63–77 % cases) | Basal atelectasis |
| | | Focal eventration of diaphragm |
| Bronchogenic cyst | Mediastinum (65–90 % cases) | Mediastinal lymphadenopathy |
| | Intrapulmonary (10–35 %). More common in lower lobes | Round pneumonia |
| | | Hydatid cyst |
| | | Lung mass |
| | | Infected CPAM/ hydatid cyst (if infected) |

valve' effect. It most commonly affects the left upper lobe (42 % cases), followed by the middle lobe (35 %) and the right upper lobe (21 %) [27, 28]. The lower lobes are uncommonly involved (<1 % cases). Patients present soon after birth with respiratory distress, and presentation after the age of 6 months is uncommon. CLH may occur in a classic form (with normal/reduced number of alveoli) or polyalveolar form (increased number of alveoli).

Focal hyperlucency is usually evident on radiographs, with mediastinal shift (Fig. 1). MDCT enables accurate diagnosis by demonstrating an area of hyperinflation with attenuated bronchovascular markings within. Symptomatic patients are managed surgically (lobectomy), while those without symptoms may be managed conservatively [29].

## Congenital Bronchial Atresia

Bronchial atresia (BA) refers to focal obliteration of a lobar, segmental or sub-segmental bronchus, with normal development of distal airways [24]. Inspissation of secretions in the bronchus distal to the atresia forms a bronchocele. Opening of collateral pathways often leads to air-trapping [27•]. Most cases are asymptomatic. The upper lobe bronchi are commonly involved, with most frequent involvement of the segmental apico-posterior bronchus of left upper lobe [30]. CT is the most sensitive modality for diagnosis, and demonstrates the bronchocele as a branching, tubular opacity, often with air-trapping in the adjacent lung (Fig. 2).

Bronchial atresia may also occur as a component of other CLMs, like PS, CPAM [8].

## Congenital Pulmonary Airway Malformations

CPAMs, earlier known as congenital cystic adenomatoid malformations (CCAMs), result from disorganized proliferation of primary bronchioles, which communicate with the bronchial tree. 95 % cases involve a single lobe, without any lobar predilection [27•]. Bilateral lesions are uncommon. CPAMs have been classified by Stocker et al. [31] into three types—the most common (type 1) shows single/multiple large cysts, size >2 cm (Fig. 3). Type 2 lesions consist of multiple smaller cysts (<2 cm), whereas type 3 lesions (the least common) comprise of microcysts (<0.5 cm) and have a pseudo-solid appearance on imaging. An updated classification by Stocker [32] describes two additional types—type 0 results from acinar dysgenesis or dysplasia of large airways and is incompatible with life, whereas type 4 lesions have large cysts of distal acinar (peripheral) origin [21].

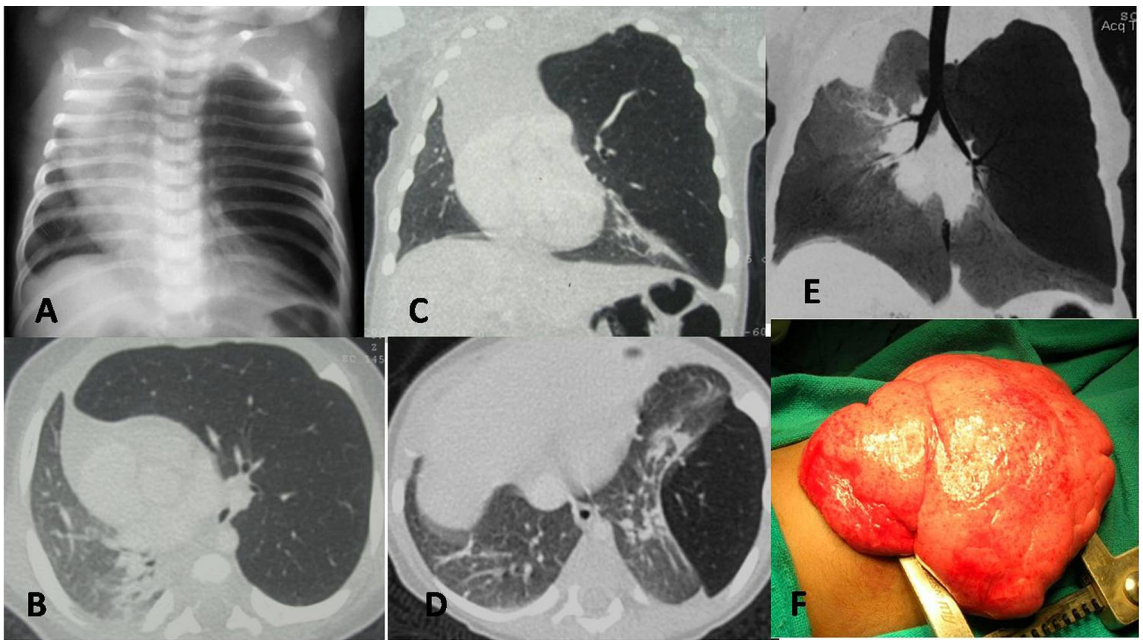


**Fig. 1** Congenital lobar hyperinflation in a 45-day-old male child with acute onset respiratory distress. Chest radiograph (**a**) reveals a focal hyperlucency in the *left upper* and *mid zones*, with mass effect and mediastinal displacement to the right. CT Chest, lung window, axial (**b**, **d**) and coronal (**c**) images, along with minimum intensity projection (MinIP) image (**e**) reveal an over-inflated left upper lobe, showing attenuated bronchovascular markings. Intra-operative photograph (**f**) confirms the findings

Imaging appearances depend upon the type and size of the lesion, and the presence or absence of superadded infection. Radiographs may demonstrate only a hyperlucent lesion (Fig. 3), which, depending on its location, may mimic congenital lobar hyperinflation, pneumothorax, or congenital diaphragmatic hernia (Table 2). On radiographs, infected type 1 CPAMs appear similar to secondarily infected hydatid or bronchogenic cysts (Fig. 4), and may mimic a lung abscess. MDCT depicts air-containing lung parenchymal lesions comprising of cysts of variable sizes (Figs. 3, 5), and may show cyst wall thickening and air-fluid levels within infected CPAMs (Fig. 5). Large lesions usually present in infancy with respiratory distress. Smaller lesions may be detected in older children, incidentally or in association with recurrent chest infections. 'Hybrid' lesions, which overlap with pulmonary sequestration (PS), may demonstrate a systemic arterial supply (Fig. 6).

CPAMs are increasingly being detected on prenatal US. They are seen as cystic or solid-appearing lung masses. Their natural course is variable, and most microcystic lesions plateau in their growth by 26–28 weeks' gestation [15•]. Up to 15 % lesions regress in size and may 'disappear' on US, though they can be identified on postnatal CT [33]. Associated hydrops is a harbinger of fetal demise, and an indication for fetal therapy [15•]. Large macrocystic CPAMs have been successfully treated with throacoamniotic shunting [34]. Maternal betamethasone therapy is useful in predominantly solid lesions with hydrops [35]. Patients who do not respond may be candidates for fetal surgery (at <30 weeks). CPAM resection may be planned

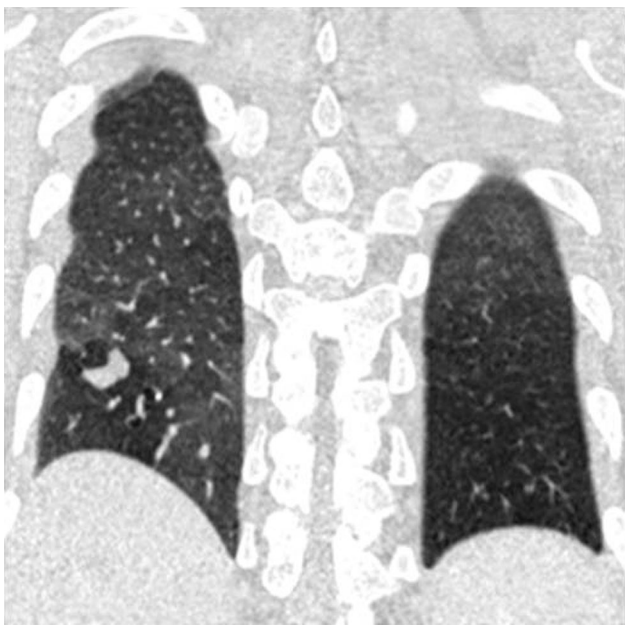

**Fig. 2** Bronchial atresia in an 18-year-old girl being evaluated for fever. CT chest, coronal image (lung window) reveals a bronchocele involving the lateral segmental bronchus of the right lower lobe with associated air-trapping (patient was managed conservatively)

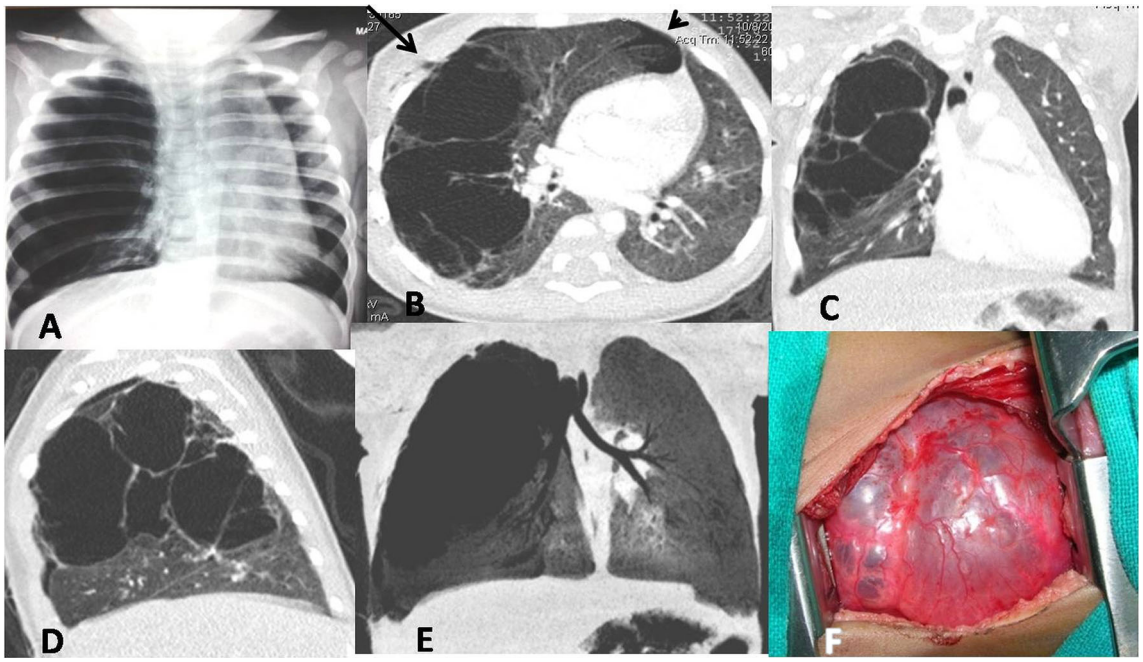


**Fig. 3** A 6-month-old child with respiratory distress. Chest radiograph (**a**) shows a focal hyperlucency in right upper/mid zones, with mass effect. This was interpreted as a pneumothorax, and chest tube was inserted at a primary care center. Symptoms did not significantly improve. Subsequent CT chest, lung window, axial (**b**), coronal (**c**) and sagittal (**d**) images, and coronal MinIP image (**e**) reveals a type 1 CPAM in the right upper lobe, with multiple cysts, many larger than 2 cm. A small right pneumothorax (*arrowhead* in **b**) is also seen, along with chest tube tract (*black arrow*). Intraoperative photograph (**f**) shows the multi-cystic lesion

at the time of delivery by EXIT (ex utero intra partum) procedure [15•].

Expectant management is appropriate for most non-hydropic fetuses. Postnatal management of symptomatic lesions is surgical resection. The management of asymptomatic lesions, however, is controversial [36–38], most notably as CPAMs are associated with increased risk of malignancy, including pleuropulmonary blastoma and bronchioalveolar carcinoma [36•]. Further research is needed into the merits of a surgical versus conservative approach to asymptomatic cases.

## Bronchogenic Cysts

Bronchogenic cysts are a part of the spectrum of foregut duplication cysts (which also includes enteric and neurenteric cysts) and result from abnormal airway budding/branching during fetal life. They are commonly located in

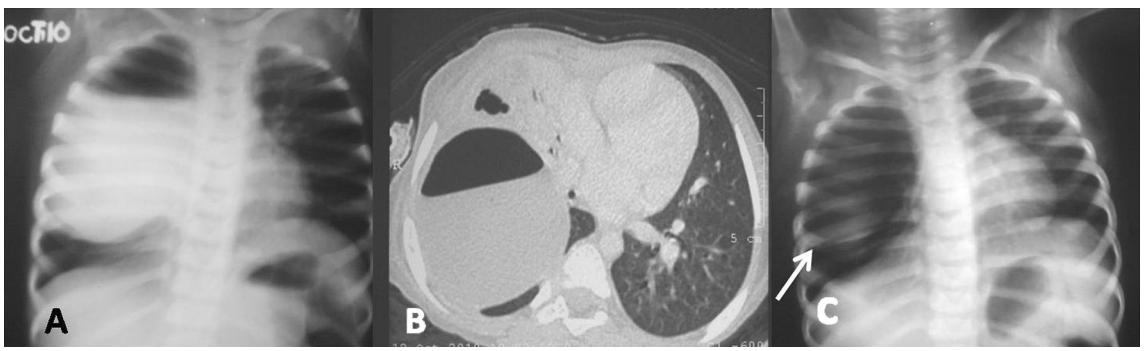


**Fig. 4** A 3 year-old-child presented with fever and cough since 1 week, with a history of two past episodes of chest infection. Frontal chest radiograph (**a**) reveals a large well-defined cystic lesion in the right lung with an air-fluid level. CT chest, lung window, (**b**) confirms its intrapulmonary location and shows adjacent collapse-consolidation. A previous chest radiograph of the same child, done 6 months earlier (**c**), shows a thin walled air-filled lung cyst (arrow). Diagnosis of an infected lung cyst was made. Surgery and histopathology confirmed an infected type 1 CPAM

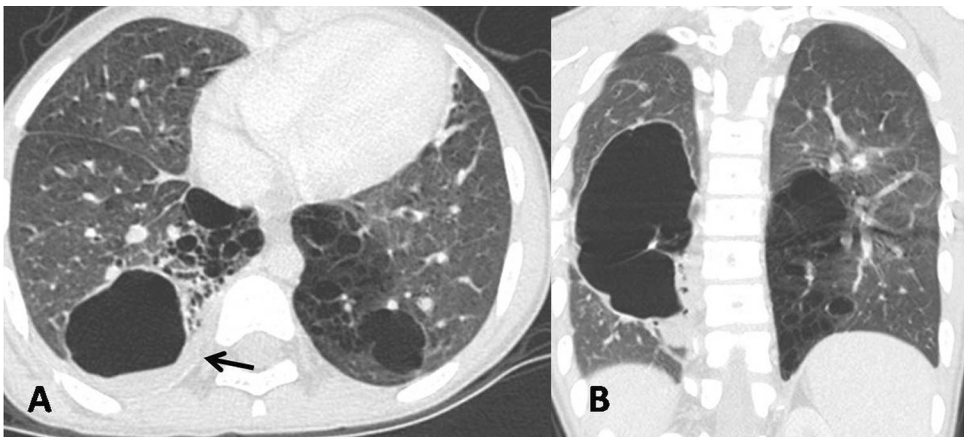


**Fig. 5** A 6-year-old girl with bilateral CPAM presented with difficulty in breathing. CT chest, lung window, axial (**a**) and coronal (**b**) images reveal multi-cystic lesions in lower lobes of both lungs. The right lung lesion (containing cysts >2 cm—type 1 CPAM) shows thick walls and an air-fluid level (*arrow*). This lesion was infected and was surgically resected. The left lung lesion had smaller cysts (<2 cm—type 2 CPAM), and was managed conservatively

the mediastinum—subcarinal, hilar or paratracheal regions (65–90 % cases) [24]. They may present with mass effect on the esophagus or airway (do not usually communicate with the airway). Due to proteinaceous content, their CT attenuation may be higher than water in 50 % cases.

Less commonly, cysts may be intrapulmonary, commonly in lower lobes. These cases may have an airway communication, and may present with infection, with radiological findings resembling those of infected CPAMs (Fig. 7). Final diagnosis is established on pathology by demonstration of respiratory epithelium [39].

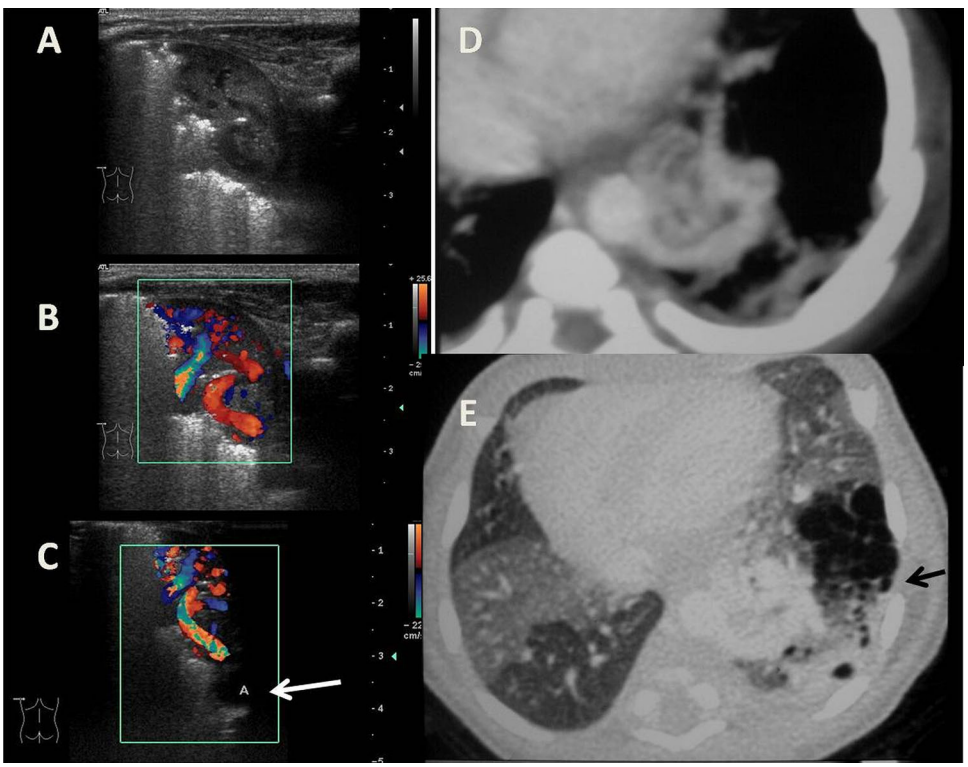


**Fig. 6** An asymptomatic newborn had been diagnosed with a left lung 'mass' on prenatal US. Postnatal grayscale US (**a**) and doppler (**b**, **c**) images reveal left lower lobe consolidation, and an aberrant vessel arising from the aorta (*white arrow* in image **c**). CECT chest, axial section, mediastinal window (**d**) demonstrates the supplying vessel (systemic artery), and axial lung window image (**e**) shows an area of consolidation in left lower lobe, along with multiple small air-filled cystic lesions (*arrow*), suggestive of a component of CPAM. A 'hybrid' lesion—sequestration with CPAM—was proven on post-operative histopathology

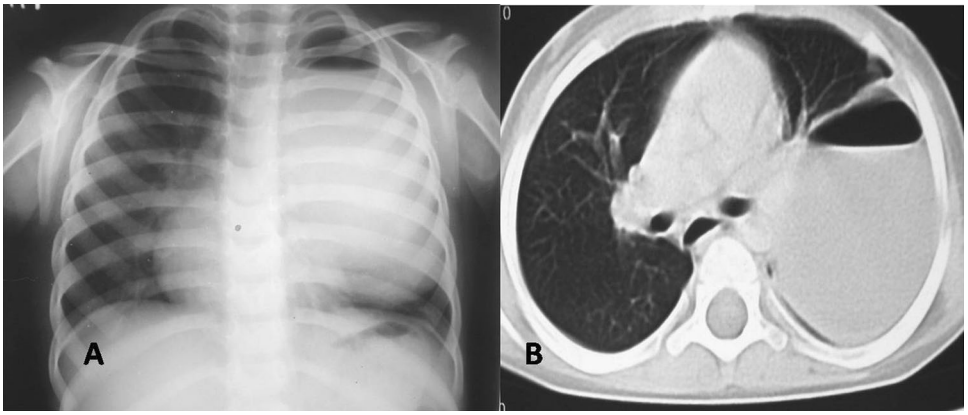


**Fig. 7** A 2-year-old boy presented with fever of 2 weeks' duration. Frontal chest radiograph (**a**) reveals a large cyst containing an air-fluid level in the left lung, also seen on chest CT, lung window (**b**). This was an infected lung cyst, confirmed at surgery and histopathology (which revealed respiratory epithelium). Final diagnosis was intrapulmonary bronchogenic cyst

## Airway Anomalies

Congenital high airway obstruction syndrome (CHAOS) is a rare anomaly caused by laryngeal or tracheal atresia [40]. Pulmonary hyperplasia develops secondary to obstruction to the outflow of fetal lung fluid [41]. Prenatal US findings are characteristic, with enlargement and increased echogenicity of both lungs, inversion of hemidiaphragms and

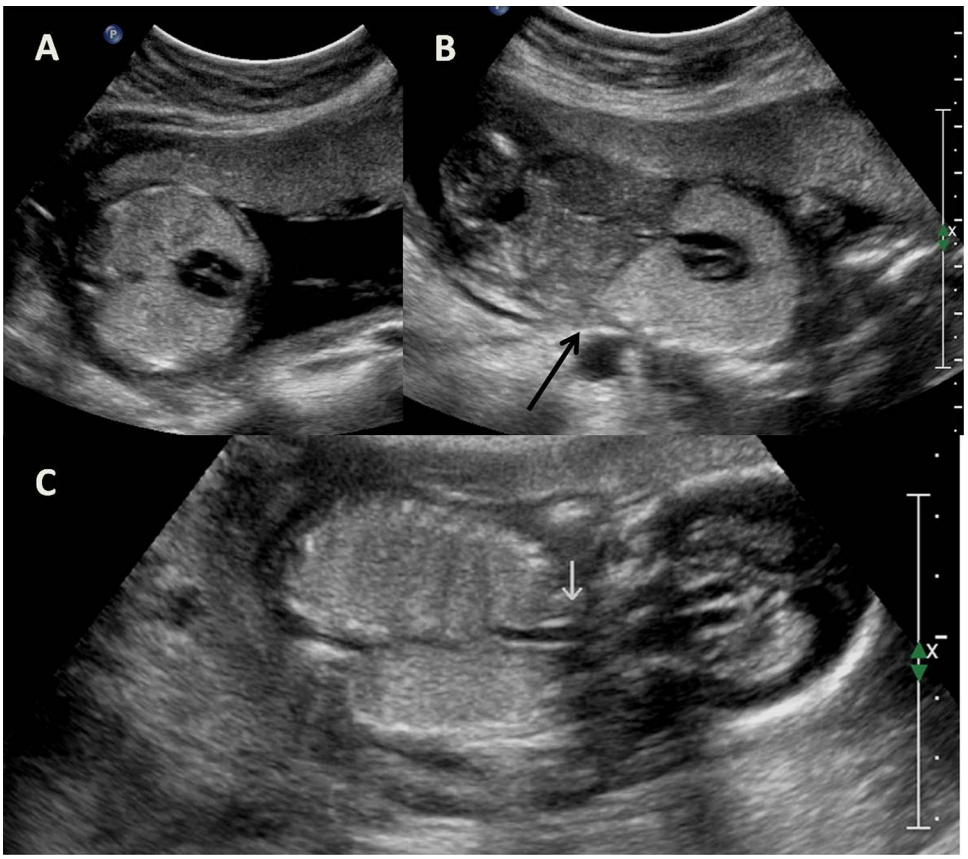


**Fig. 8** Prenatal US of a 19-week-old fetus. Axial (**a**) and coronal oblique (**b**) scans through the fetal thorax reveal enlargement and increased echogenicity of both lungs, with mild compression of the fetal heart and flattening of diaphragm (*black arrow*). Coronal oblique section through the fetal neck/upper chest (**c**) reveals a dilated, fluid-filled trachea (*white arrow*). Diagnosis of congenital high airway obstruction syndrome (CHAOS) was made, and subsequent fetal autopsy confirmed laryngeal atresia

compression of the fetal heart (Fig. 8). Distal to the site of obstruction, fluid-filled dilated trachea and bronchi are seen. This condition is fatal.

## Vascular Anomalies

### Pulmonary Agenesis–Aplasia–Hypoplasia Complex

Pulmonary agenesis (complete absence of lung tissue, bronchus and pulmonary artery) is part of a spectrum of pulmonary underdevelopment which includes pulmonary aplasia (absence of lung and pulmonary artery with rudimentary main bronchus) and hypoplasia (hypoplastic pulmonary artery and bronchus with variable amount of lung tissue). Abnormal blood flow in the dorsal aortic arch during fetal development has been suspected to play a role in pathogenesis [27•]. Patients may present as newborns with respiratory or feeding difficulty, or may remain asymptomatic till adulthood. Pulmonary agenesis, aplasia, and hypoplasia appear similar on chest radiographs, which reveal a small opaque hemithorax with ipsilateral shift of mediastinum. MDCT can differentiate these conditions by demonstration of hypoplastic bronchus and/or pulmonary artery (Fig. 9). Congenital anomalies involving other organ systems are often associated, and influence the prognosis. Bilateral pulmonary agenesis or aplasia proves fatal.

### Agenesis of Pulmonary Artery

Agenesis or proximal interruption of the pulmonary artery is a rare anomaly which occurs due to embryological failure of development of the proximal portion of the main pulmonary artery, with presence of pulmonary artery at the hilum and distally. It is more common on the right side, and leads to lung hypoplasia.

### Pulmonary Artery Sling

Pulmonary artery sling is a vascular anomaly in which the left pulmonary artery has an aberrant origin from the

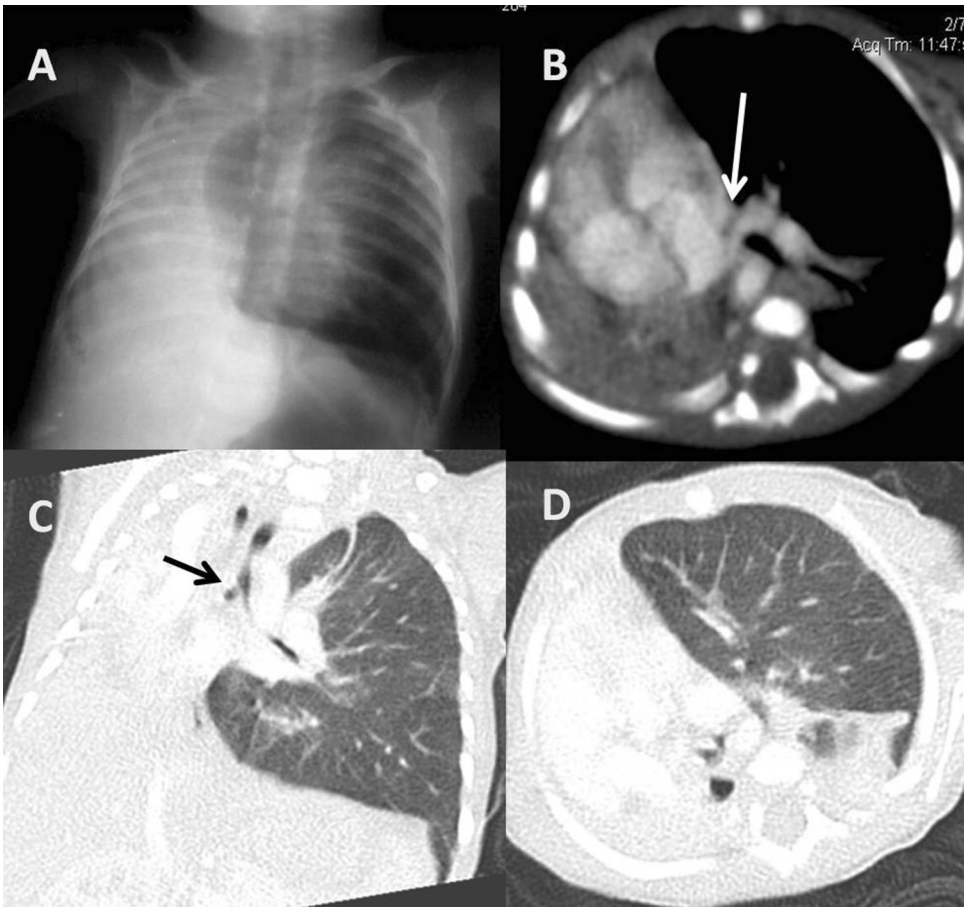


**Fig. 9** A 3 month-old-female child presented with respiratory distress. Frontal chest radiograph (**a**) reveals a small, opaque right hemithorax with ipsilateral shift of mediastinal structures. CT chest, axial mediastinal window (**a**), reveals a hypoplastic right pulmonary artery (*white arrow*). Lung window sections in the coronal (**c**) and axial (**d**) planes reveal a rudimentary right bronchus (*black arrow*) and a small amount of right lung parenchyma. This was a case of hypoplasia of right lung

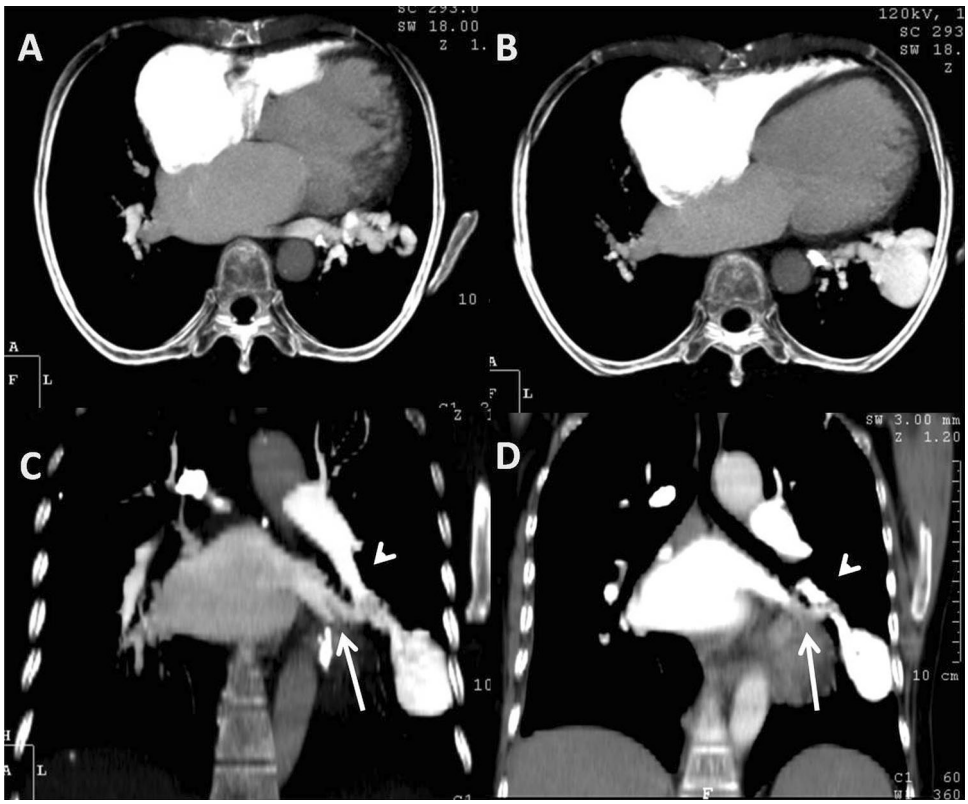


**Fig. 10** A pulmonary AVM detected on CT chest of a 60-year-old woman being evaluated for tubercular lymphadenopathy. Axial MIP sections (**a**, **b**) reveal a cluster of abnormal, dilated pulmonary vessels in the left lower lobe. Coronal MIP sections (**c**, **d**) reveal a feeding artery arising from the left lower lobar pulmonary artery (arrowheads) and a vein draining the lesion via the inferior pulmonary vein (*long white arrows*) into the left atrium

proximal right pulmonary artery, and crosses the mediastinum between the trachea and the esophagus, thus forming a 'sling' around the distal trachea [27•]. It may result in tracheo-bronchial compression. There are two types of pulmonary artery sling—type 1, in which carina is normal in position (T4-5), and type 2, which is associated with a low-lying (T6) carina and horizontal course of main bronchi. Type 2 lesions are often associated with long-segment tracheal stenosis, bridging bronchus, horseshoe lung, lung agenesis, and congenital heart disease.

### Pulmonary Vein Stenosis

This condition usually occurs in association with congenital heart disease or anomalous pulmonary venous return [42]. It can also occur in isolation, usually at the veno-atrial junction.

### Pulmonary Varix

A pulmonary varix is an enlarged pulmonary vein in the absence of a feeding artery or nidus, and usually occurs near the pulmonary venous—left atrial junction. It is usually asymptomatic, and can mimic a pulmonary nodule on radiographs [24].

### Pulmonary AVM

A pulmonary AVM is an abnormal communication between pulmonary artery and vein branches, without normal intervening capillaries. It may be congenital or acquired (post-trauma or infections). Hereditary hemorrhagic telangiectasis (HHT), or Rendu-Osler-Weber syndrome is an autosomal dominant condition in which up to 35 % of cases have pulmonary AVMs, frequently multiple [21, 43].

Pulmonary AVMs show lower lobe predominance in 50–70 % cases [24, 42]. Lesions smaller than 2 cm are usually asymptomatic. Larger lesions result in right-to-left shunts, and can present with pulmonary hemorrhage or paradoxical embolization. MDCT angiography is the technique of choice for imaging [24] (Fig. 10), and management of symptomatic lesions is endovascular coil embolization.

## Combination of Parenchymal and Vascular Anomalies

### Pulmonary Sequestration

The word 'sequestration' (derived from Latin 'sequestrare': to separate) is defined as non-functioning lung tissue that is

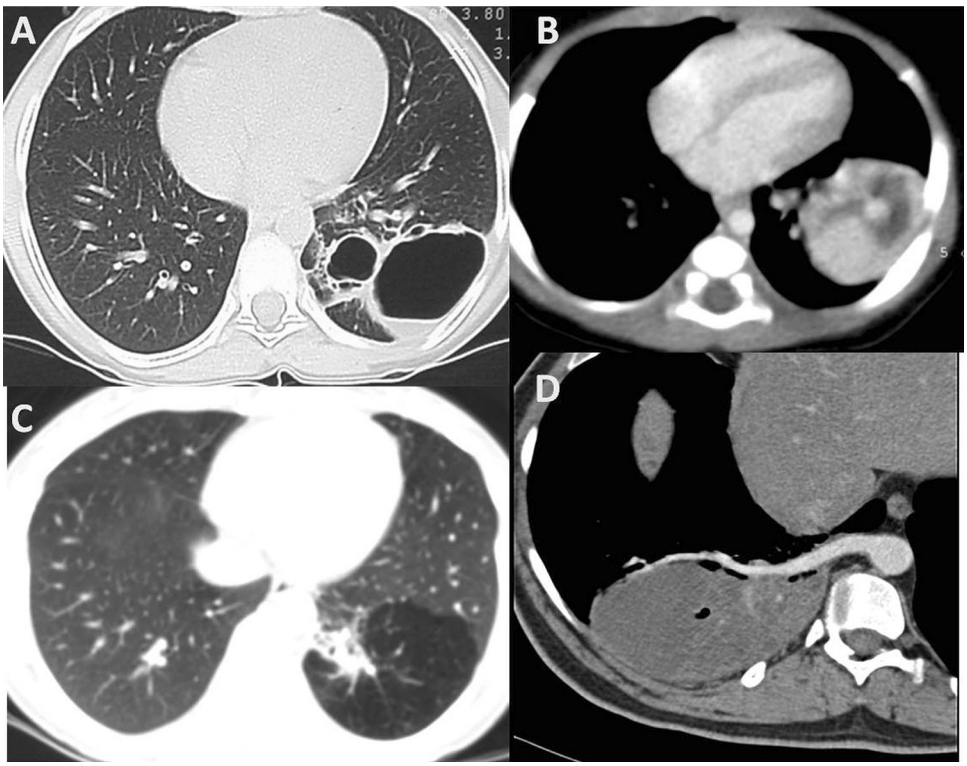


**Fig. 11** CT appearances of pulmonary sequestration (four different patients). **a** Axial CT chest, lung window, shows a multi-cystic sequestered lung lesion in left lower lobe with presence of air, and fluid levels within. **b** Axial CT chest, mediastinal window, shows a heterogeneously enhancing sequestered lesion in the left lower lobe, with few non-enhancing areas, but no air within. **c** Incidentally detected intralobar sequestration in a young woman. Axial CT chest, lung window, reveals fibrotic lung parenchyma within the area of sequestration, with emphysematous changes in the surrounding lung. **d** Axial oblique image, mediastinal window, of CT chest in an adolescent male reveals sequestration within the right lower lobe, supplied by an aberrant systemic artery arising from the descending thoracic aorta

not in normal continuity with the tracheobronchial tree and derives its arterial supply from systemic vessels. Classically, two forms of pulmonary sequestration—intralobar and extralobar—are described. Though intralobar sequestrations were earlier believed to be acquired lesions, increasing evidence supports a congenital origin [44, 45]. Intralobar sequestrations (ILS) are contained within the visceral pleura of normal lung, whereas extralobar lesions have a separate pleural covering. Approximately 75 % cases are intralobar, 98 % cases occur within the lower lobes (left > right), classically involving the posterior basal segment [46, 47]. Most ILS drain via pulmonary veins, while extralobar sequestrations (ELS) show a systemic venous drainage in 80 % cases, through the azygous–hemiazygous system or superior vena cava [48]. An ELS classically occurs between the left hemidiaphragm and the lower lobe (63–77 % cases), though it may occur below the diaphragm or in the mediastinum.

More than half of ELS are associated with other congenital anomalies, like congenital diaphragmatic hernia (seen in 20–30 % cases) [49]. Cases present commonly during infancy. ILS, on the other hand, usually present in adolescence or adulthood [50], often with recurrent infections.

Prenatal US may detect PS as a 'lung mass', which may be indistinguishable from CPAM on gray scale imaging. Doppler sonography is useful to depict the systemic arterial supply. For postnatal evaluation, chest radiograph is the initial imaging modality; however, findings on chest radiographs may be varied and, sometimes, subtle. PS should be suspected in all cases showing persistent radiographic opacity in the lower lobes of the lungs. MDCT can demonstrate the sequestered lesion, as well as its vascular supply (Fig. 11). The sequestration contains fibrotic and consolidated parenchyma, frequently containing cystic areas. ILS lesions may sometimes be air-filled, whereas ELS do not contain air unless it has a foregut-communication. Lung parenchyma surrounding an ILS may show emphysematous changes. The aberrant systemic artery supplying the lesion usually arises from the descending thoracic or upper abdominal aorta, and less commonly from intercostals, internal thoracic, subclavian or even

coronary arteries. Multiple supplying vessels may be present. The most important goal of imaging is to provide a 'vascular roadmap'.

The definite treatment is surgical, though endovascular embolization has been attempted, especially for ELS [51, 52]. Thoracotomy or thoracoscopic surgery may be done, and while ELS can be managed by sequestrectomy, lobectomy is usually needed for intralobar cases.

## Scimitar (Hypogenetic Lung) Syndrome

Scimitar syndrome (hypogenetic lung or venolobar syndrome) is a type of partial anomalous pulmonary venous return affecting the right lung, with associated abnormalities. The anomalous vein drains most commonly into the inferior vena cava (IVC), and less commonly into the right atrium, superior vena cava, portal vein or hepatic veins. Common associations include hypoplasia of the lung and pulmonary artery, cardiac dextroversion, and abnormal systemic arterial supply to the right lung (Fig. 12). Other congenital anomalies may be associated in 25 % cases; these include atrial and ventricular septal defects, patent ductus arteriosus, tetrology of Fallot, diaphragmatic defects and horseshoe lung [21]. The name 'scimitar' (meaning curved Turkish sword) refers to the curved appearance of the anomalous vein as it courses toward the IVC.

Symptoms depend upon the degree of left to right shunting, and vary from none to heart failure in infancy. In patients with shunt ratios >2:1, treatment is reconnection of the anomalous pulmonary vein to the left atrium, and embolization of the systemic arterial supply [24, 53].

## Nomenclature and Classification

In 1946, Pryce coined the term 'sequestration' [54], and subsequently described three forms of intralobar sequestration [55]. Many variants were later described, and the concept of 'sequestration spectrum' was given by Sade in 1974 [56]. Since then, many terms and schemes of classification have been proposed to describe CLMs. It was recognized that

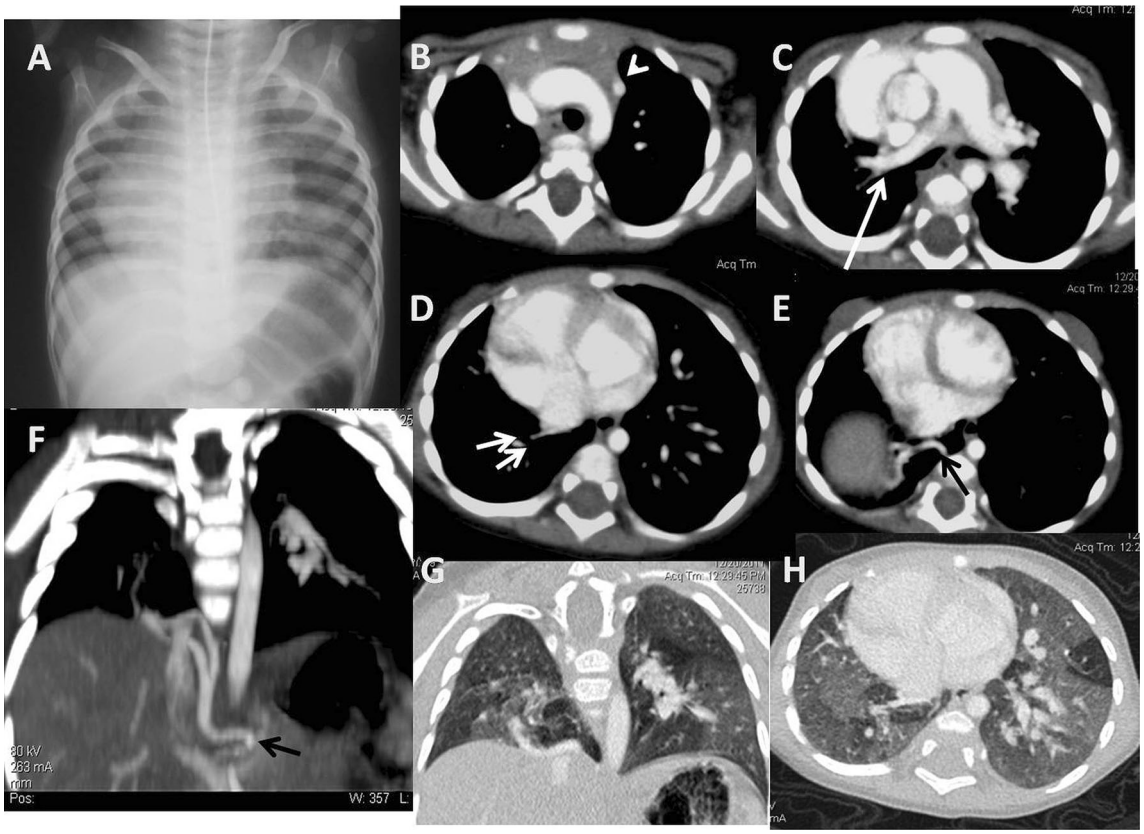


**Fig. 12** An 8-month-old girl presented with recurrent chest infections since 4 months. Chest radiograph (**a**) reveals a small right hemithorax, with ipsilateral shift of the mediastinum. CT chest, axial sections, mediastinal window (**b**–**e**) are shown. There is a double SVC—the left brachiocephalic vein does not cross the midline and continues as left SVC (*arrowhead*), and a small right pulmonary artery (*long arrow*). Also seen is anomalous drainage of right inferior pulmonary vein into the right atrium (*double arrow* in **d**), along with an aberrant systemic artery supplying the right lower lobe (*black arrow* in **e**). Coronal image (**f**) depicts the origin of the anomalous systemic artery from the celiac trunk (*black arrow*). Coronal (**g**) and axial (**h**) lung window images show areas of mosaic attenuation. This was a case of hypogenetic lung syndrome

lesions are often complex, show overlapping features, and involve different components—the tracheobronchial airway, arterial supply, venous drainage, lung parenchyma [57]. The term 'congenital bronchopulmonary foregut malformation' has been used for lesions with a connection with the gastrointestinal tract [58]. Langston proposed a classification system based on pathologic features. The term pulmonary 'maliosculation' ('mal'-abnormal, 'osculum'-mouth) has also been proposed to describe the abnormal connections or openings of different components of the bronchopulmonary–vascular complex [57, 59].

In order to tackle the confusion regarding CLM classification, Bush highlighted the need for a more practical approach [60]. He proposed using simple language to describe 'what is actually seen', and keep clinical and pathological descriptions separate. He stressed upon a systematic approach to evaluate different components of the lesion, as well as other associated abnormalities. There is a need for radiologists and clinicians to concentrate on careful assessment and elucidation of the various components of a CLM, in order to optimize management. Pathological terms and descriptions should be avoided in radiological reporting of these cases.

## Conclusion

CLM involve different components of the lung and its vascular supply. They comprise a continuum of abnormalities, with existence of 'hybrid' lesions. It is important to evaluate all components of the lesion and exclude the co-existence of other anomalies. Imaging plays a vital role in diagnosis and is especially valuable while planning surgical management.

### Compliance with Ethics Guidelines

**Conflict of Interest** Dr. Pooja Abbey, Dr. Mahender K. Narula, and Dr. Rama Anand each declare no potential conflicts of interest.

**Human and Animal Rights and Informed Consent** This article does not contain any studies with human or animal subjects performed by any of the authors.

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