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REVIEW

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Congenital bronchopulmonary foregut malformations: concepts and controversies

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Abstract This article addresses the scope, etiology, important associations and imaging features of congenital bronchopulmonary foregut malformations. Etiologic concepts, including airway obstruction and vascular anomalies, are highlighted. Technical imaging advances, especially CT and MR, have greatly enhanced our diagnostic abilities in evaluating these lesions; however, thorough and careful assessment of all aspects of the malformation is still necessary. Several specific lesions are discussed in more detail, particularly regarding controversial issues in classification, understanding, imaging and management.

Keywords Congenital · Foregut malformations · Chest · Thorax

Part I: Spectrum, etiology, pathology and approach to imaging[1]

Introduction

The congenital bronchopulmonary malformation lesions form an important part of pediatric chest imaging. Numerous classifications and terminologies have been suggested for congenital bronchopulmonary foregut malformations, attempting to incorporate a common origin and association of many of the lesions, including variable foregut, airway, lung and vascular components (Fig. 1) [1–13]. Entities classically included are congenital cystic adenomatoid malformation of the lung (CCAM) [now called congenital pulmonary airway malformation (CPAM)], pulmonary sequestration, bronchogenic cyst, bronchial atresia, and congenital lobar emphysema (CLE) [now called congenital lobar hyperinflation (CLH)] [1–8]. Other anomalies can also legitimately be considered part of the spectrum of bronchopulmonary foregut malformations. These include: pulmonary agenesis, aplasia, hypoplasia, scimitar syndrome, tracheal and esophageal diverticula, tracheal bronchus, bronchial isomerism, esophageal and neurenteric cysts, congenital esophageal stenosis, esophageal and tracheal atresia, tracheoesophageal fistula and other connections between the gastrointestinal tract and lung (Fig. 1).

Etiology and pathologic concepts

Understanding congenital lung/foregut malformations is difficult and confusing. These anomalies have been characterized as lesions of defective budding, differentiation and separation of the primitive foregut [1–13]. In utero, airway maldevelopment with resultant obstruction is a recurring proposed etiology for many of these lesions, especially those associated with hyperlucent or cystic lung changes. These theories are not mutually exclusive; abnormal budding and airway obstruction can readily occur together.

Langston [14] has suggested that many of the bronchopulmonary/foregut anomalies represent an obstruction malformation sequence with secondary pulmonary dysplastic changes. Differences in the level, completeness and timing of airway obstruction are proposed as being responsible for the spectrum of abnormalities that occur. Vascular abnormality is also commonly suggested as an etiologic or associated feature [12].

There is considerable evidence to support an underlying airway obstructive malformation. Among that evidence is the fact that foregut duplication cysts, sequestration and

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[1] Part I, Table 1 and some of the figures (Figs. 5, 8, 10, and 12) were originally published in similar form in the 2005 Postgraduate Course Syllabus, Society for Pediatric Radiology (SPR). Components are reproduced here with the permission of the SPR.

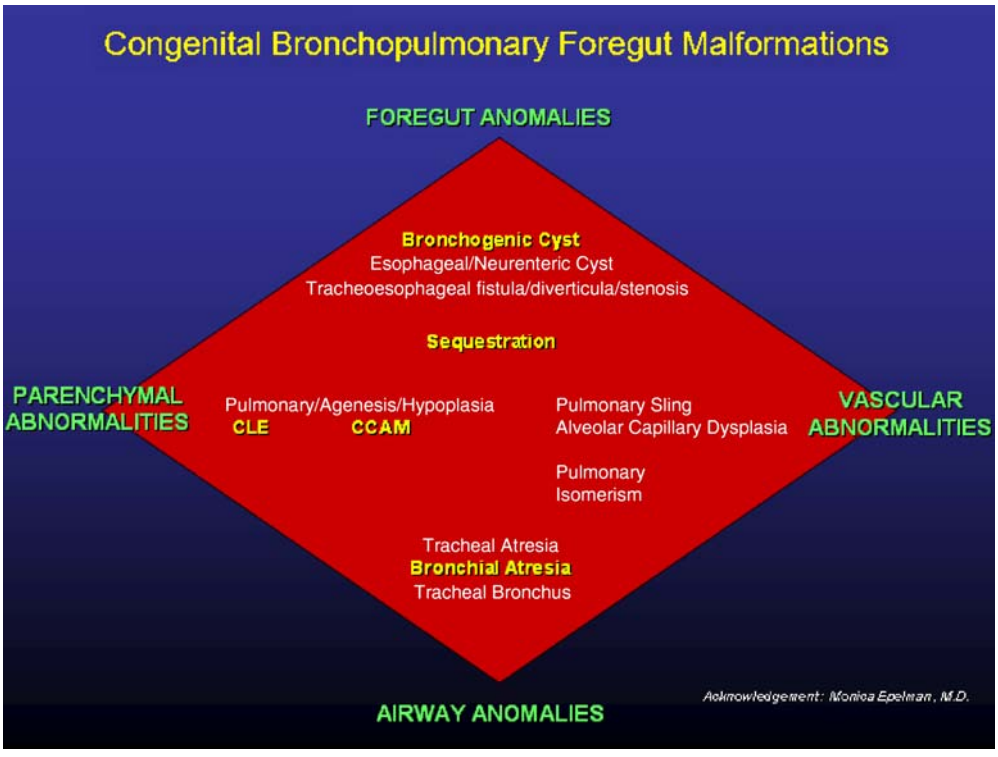

Fig. 1 Diagram of the spectrum of bronchopulmonary foregut malformations including foregut, pulmonary, airway and vascular components. The lesions shown in *yellow* are those more commonly included in the spectrum of these lesions and those in *white* are additional lesions that can be considered part of the group

bronchial atresia usually have lost their connection with the airway, and when they are aerated, air-trapping is typical (Fig. 2). In other lesions such as CLH and CPAM, the supplying airway is often abnormal [1, 3, 7, 14] (Fig. 3).

Langston has described two forms of associated pulmonary pathologic dysplastic findings:

1. Pulmonary hyperplasia (Fig. 4). The lung parenchyma is expanded with air space enlargement with decreased branching airways and vessels. This is the pathologic appearance of the lung in infants with laryngeal or tracheal atresia and is the major pathologic feature of microcystic CPAM (type III). A very similar appearance is found in cases of polyalveolar congenital lobar hyperinflation and also either diffusely or focally in bronchial atresia and pulmonary sequestration [14, 15].
2. Microcystic parenchymal dysplasia (Fig. 4). This resembles the type II (small cyst) CPAM and is a common histologic component in many other bronchopulmonary malformation lesions, including bronchial atresia, pulmonary sequestration, CLH and pulmonary hypoplasia [14, 16].

Corroborating this finding and its association with fetal airway obstruction is experimental data that cystic change occurs in fetal animal lungs after bronchial ligation [15]. This is also a common embryologic theme in other organ systems, such as the genitourinary system, with cystic renal dysplasia a consequence of high-grade urinary obstruction.

I have emphasized this particular etiological concept of the bronchopulmonary foregut lesions representing the continuum of an obstructive malformation sequence because it seems to be useful when faced with the enormously diverse and often confusing and overlapping imaging and pathologic findings.

Approach to imaging

Chest radiographs are often the initial imaging examination on which an anomaly is first suspected. Findings suggesting the presence of a congenital bronchopulmonary/foregut anomaly are manifold and quite varied (Table 1) [4, 16–18]. They include lung asymmetry with a unilateral small or large lung, focal hyperlucency, cystic or solid pulmonary or mediastinal mass and vascular and/or airway abnormality.

The following are important features of bronchopulmonary/foregut malformations that should be considered in the imaging evaluation of these lesions:

- Hybrid lesions (overlap lesions)
- Vascular abnormality
- Communication with the gastrointestinal tract
- Ectopic location
- Other organ system malformations

Hybrid lesions (overlap lesions)

Bronchopulmonary foregut malformations are frequently interrelated, and features of different imaging and pathologic entities can coexist in the same lesion or separately in the same child [12, 15, 16, 19, 20]. Examples include the presence of a foregut cyst medial to a segment of

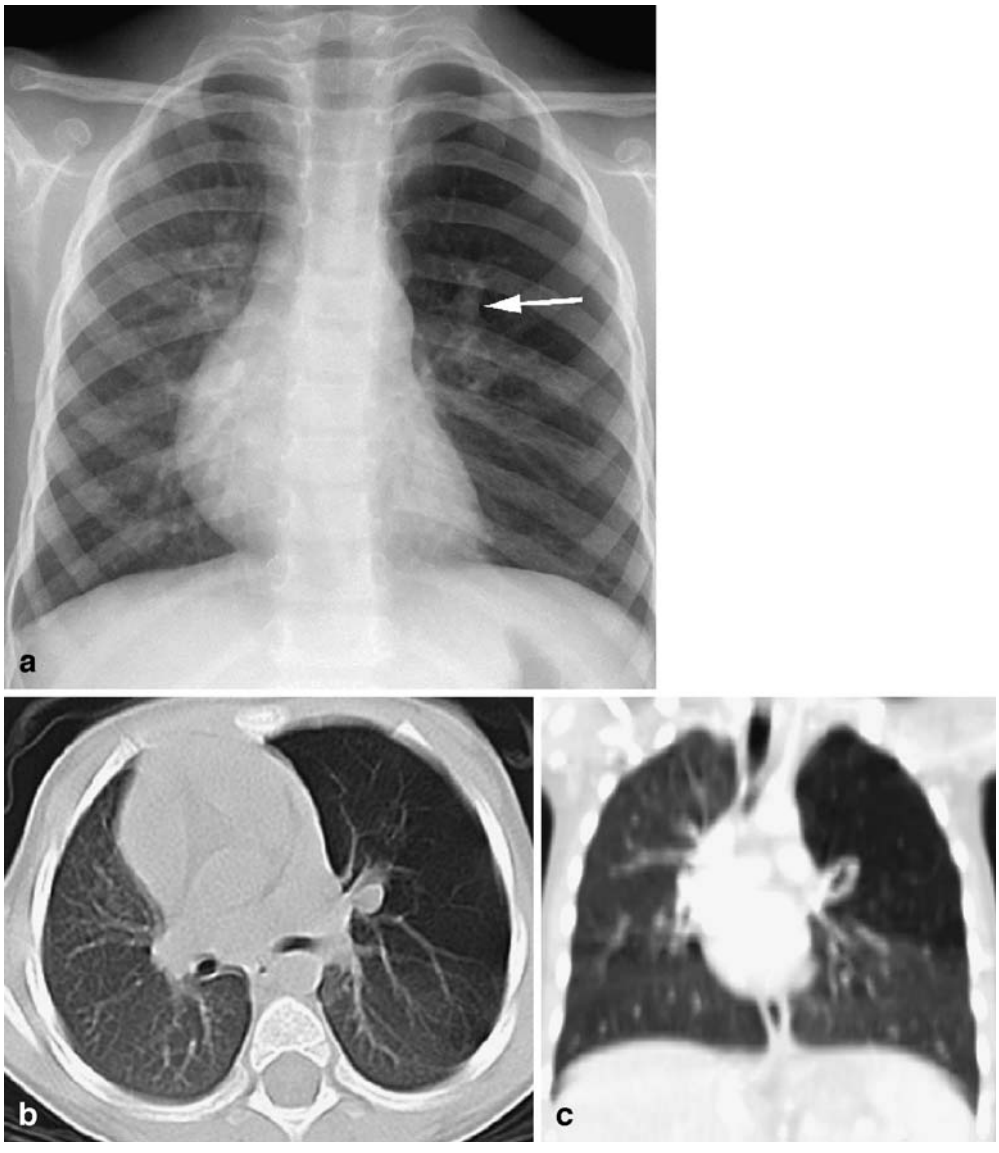

Fig. 2 Bronchial atresia. **a** On the chest radiograph of this 2-year-old, there is left upper lobe hyperinflation and branching linear density near the left hilum (*arrow*) representing mucoid impaction in the upper lobe bronchus distal to the point of atresia. The same findings are depicted on **b** axial and **c** coronal CT reformats (images courtesy of Hospital for Sick Children, Toronto)

sequestration (Fig. 5) or the coexistence of congenital lobar hyperinflation and/or CPAM with bronchogenic cyst, tracheal bronchus or bronchial atresia (Fig. 4) [15, 21]. Probably the most common overlap lesion is that of pulmonary sequestration in association with the small cyst (type II) form of CPAM [8, 12, 22–25] (Figs. 5 and 6). Microcystic parenchymal dysplasia resembling type II CPAM is also often present in the lung in association with bronchial atresia as well as in some cases of CLH and pulmonary hypoplasia [14, 16] (Fig. 4). The theory of a

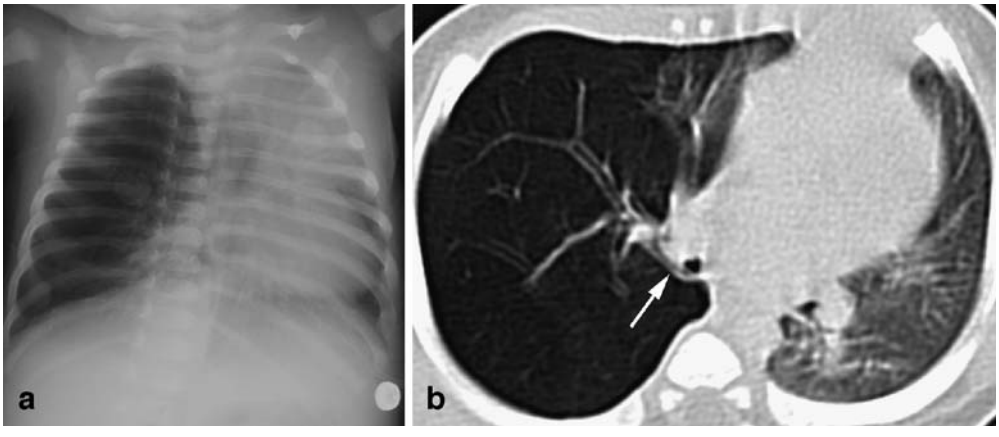

Fig. 3 Right upper lobe congenital lobar hyperinflation. **a** This 7-week-old boy with respiratory distress has a hyperinflated right upper lung and contralateral cardiomediastinal shift with left-side atelectasis on the chest radiograph. **b** CT demonstrates lobar hyperinflation of the right upper lobe with contralateral shift. Note the narrow proximal right upper lobe bronchus (*arrow*) with an unusual posterior origin from the right mainstem bronchus (3-mm slice thickness, single detector scanner)

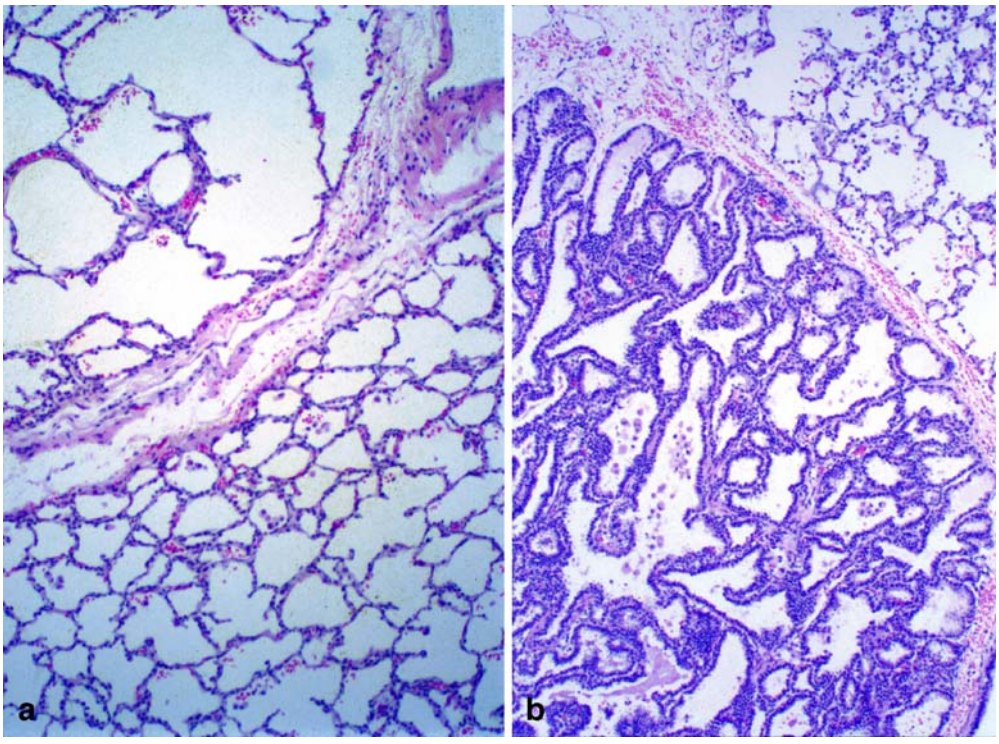

Fig. 4 Pathologic dysplastic changes associated with in utero airway obstruction. **a** Pulmonary hyperplasia. The lower part of the image demonstrates normal lung. On the left upper part of the image in an area of subsegmental bronchial atresia in the left lower lobe there are enlarged alveoli with decreased vessels. **b** Microcystic maldevelopment. The area of abnormality in the left lower part of the image represents a well-circumscribed area of small cyst malformation within a right upper lobe with congenital lobar hyperinflation. There are multiple irregular bronchiole-like structures without intervening alveolar parenchyma (**a**, **b** courtesy of Dr. Claire Langston, Houston, Texas [14])

variably timed obstructive malformation sequence with secondary pulmonary dysplastic changes seems to help in understanding these overlap lesions [14].

With CT and MR angiographic and 3-D techniques, greater anatomic detail is visualized than was possible previously, and it has become increasingly difficult to categorize and separate these lesions. Bush [26] has recommended the use of an all-encompassing term, congenital thoracic malformation, to describe all foregut/bronchopulmonary lesions. Perhaps this term is too all-encompassing. However, I endorse his suggestion that the spectrum of abnormal findings should be described in detail rather than insisting on specific categorization and nomenclature that is often confusing and ambiguous [26].

Vascular abnormality

Vascular and airway development closely parallel each other in utero; abnormalities, likewise, often occur in concert [11–13]. In a number of cases, the vascular abnormality appears to be the primary insult (Figs. 7 and 8). Panicek et al. [12] describe a spectrum of pulmonary malformation ranging from abnormal lung with relatively normal vasculature such as congenital lobar hyperinflation to normal lung with abnormal vasculature such as pulmonary arteriovenous malformation, with bronchogenic cyst, CPAM, sequestration and scimitar syndrome ranging in between. A similar scheme of the various components of these anomalies is represented in Fig. 1.

The retention of a primitive embryonic systemic arterial supply to the lung is typically considered a feature of pulmonary sequestration (Fig. 9). However, lesions that more clearly resemble the current concepts of CPAM and foregut cyst can also be associated with abnormal systemic arterial supply to a segment of lung (Fig. 6). The systemic artery might directly connect with a pulmonary artery or vein without an intervening capillary bed creating a form of vascular malformation, a common feature of scimitar syndrome (Fig. 10).

Pulmonary vascular anomalies are also commonly seen in association with pulmonary agenesis and hypoplasia; often the vascular abnormality appears to be the major anomaly. Unilateral pulmonary hypoplasia is frequently associated with absence of the ipsilateral pulmonary artery or vein [27, 28] (Figs. 7 and 8). In scimitar syndrome there is right-side pulmonary hypoplasia along with ipsilateral anomalous pulmonary venous return and a small or absent ipsilateral pulmonary artery. Systemic arterial flow to the lung with or without a segment of sequestered lung is also common (Fig. 10).

A left pulmonary artery sling has a strong association with tracheal stenosis and also can occur in conjunction with right-side pulmonary agenesis and hypoplasia as well as scimitar syndrome and other foregut lesions [19, 29, 30] (Fig. 11).

Communication with the gastrointestinal tract

The spectrum of tracheoesophageal fistula lesions with or without esophageal atresia and proximal or distal fistula is the most recognized lesion with an airway to gastrointestinal communication (Fig. 12). An esophageal bronchus or segmental communication with the distal esophagus, stomach or biliary tree is also associated with other

Table 1 Chest radiographic findings in congenital bronchopulmonary foregut malformations (Figs. 2, 3, 5, 7, 9, 10, 11, 12, 15, 16, and 17) (*CLH* congenital lobar hyperinflation, *CPAM* congenital pulmonary airway malformation, *AVM* arteriovenous malformation, *TEF* tracheoesophageal fistula)

1. Asymmetry	
	Unilateral small lung (usually right)
	- agenesis
	- hypoplasia
	- scimitar syndrome
	- pulmonary artery sling (type II)
	Focal lung hyperlucency
	- CLH
	- bronchial atresia
	- air trapping due to airway compression, e.g. bronchogenic cyst
	- pulmonary artery sling (type I)
	- pulmonary sequestration (usually older child)
	- CPAM
2. Focal mass/consolidation	
Pulmonary	
	Solid appearance
	- CPAM or CLH in a neonate (retained lung fluid)
	- sequestration (especially medial left lower lobe)
	- mucocele (branching) in bronchial atresia
	- bronchogenic cyst (adjacent to airway)
	Cystic (air filled)
	- CPAM (may be marked mass effect and midline shift)
	- sequestration (older child)
	- bronchogenic cyst (older, air-fluid level)
3. Mediastinal (round mass)	
	- bronchogenic cyst (adjacent to carina)
	- esophageal duplication cyst (posterior)
	- neurenteric cyst (posterior – vertebral defect)
	- dilated air-filled atretic esophagus ± distal gas
4. Vascular abnormality	
	- scimitar vein
	- pulmonary artery sling (abnormal mediastinal contour; mass between trachea and esophagus)
	- small hilum/pulmonary artery
	- unilateral congestion/reticulation
	- AVM
5. Airway abnormality	
	- tracheal bronchus
	- tracheal stenosis
	- T-shaped carina (type II pulmonary artery sling)
	- abnormal bronchial branching/abnormal lung lobation
	- airway compression (e.g. by bronchogenic cyst/pulmonary artery sling)
	- branching mucocele (distal to bronchial atresia)
	- esophageal atresia and TEF
	- ectopic, e.g. esophageal bronchus
6. Other lesions	
	Bone
	- vertebral anomalies
	- rib anomalies
	Gastrointestinal
	- diaphragmatic hernia
	- duodenal atresia/stenosis (double bubble)
	Cardiovascular
	- cardiomegaly
	- left to right shunt
	- pulmonary edema

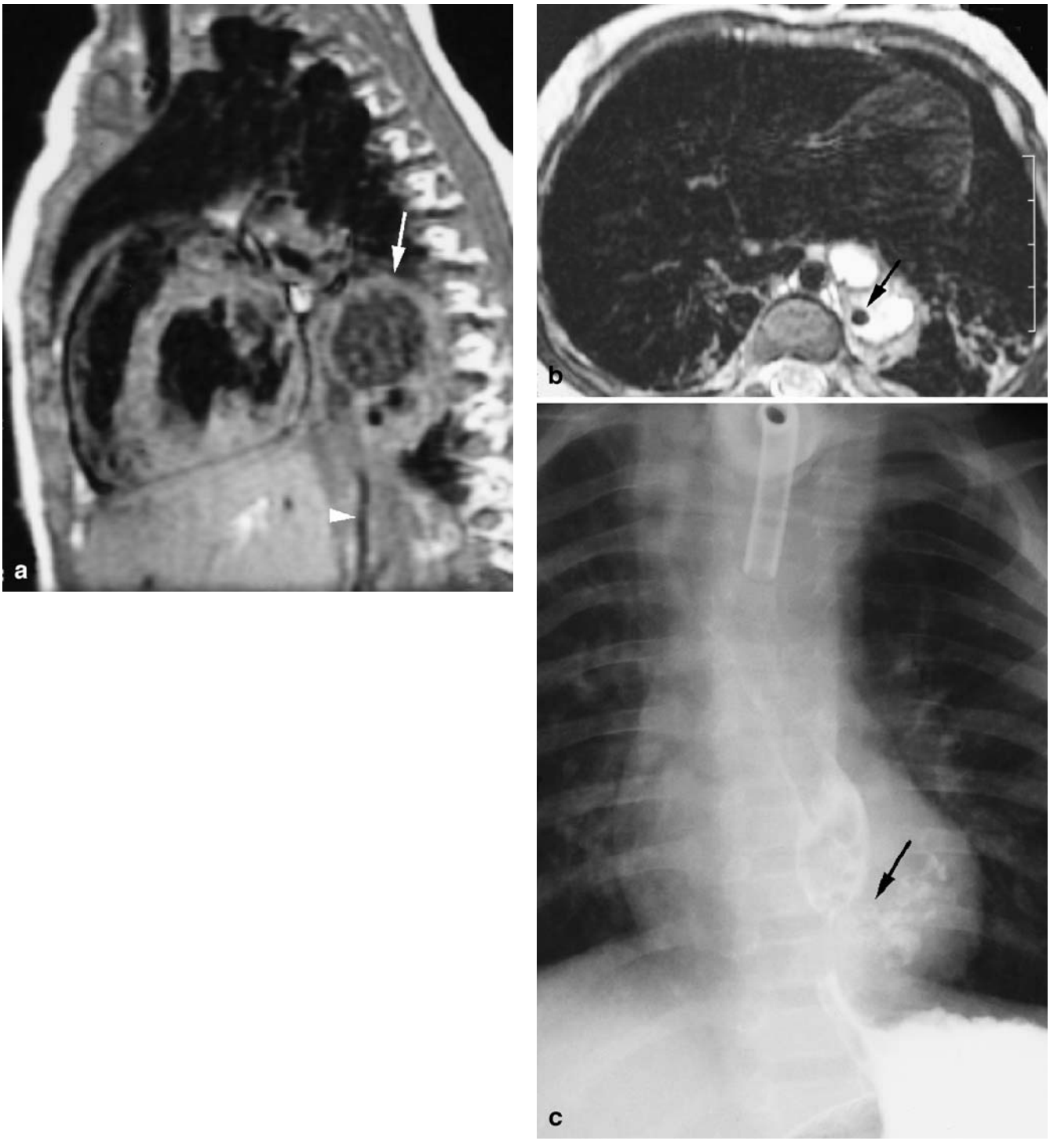

Fig. 5 Foregut malformation (bronchogenic cyst and extralobar cystic sequestration) with gastrointestinal connection. This 5-month-old boy with prior repair of tracheoesophageal fistula had persistent respiratory and feeding problems. An MR image obtained to evaluate for vascular ring demonstrated instead a complex cystic and solid malformation in the left lower lobe. **a** The sagittal T1-weighted image demonstrates a superomedial large cyst (bronchogenic cyst) (*arrow*) and a more inferior more solid mass supplied by a systemic abdominal artery (*arrowhead*) and containing several low signal rounded foci. **b** The T2-weighted axial image demonstrates solid and cystic components of this sequestration lesion. Some of the rounded areas are bright, suggesting fluid, while others remain dark on T2, (and also did not show enhancement after administration of contrast agent – not shown), suggestive of air (*arrow*). **c** On the esophagram there is a mass impression on the distal esophagus by the malformation and an aberrant esophageal bronchus (*arrow*) entering and branching within the sequestered lung

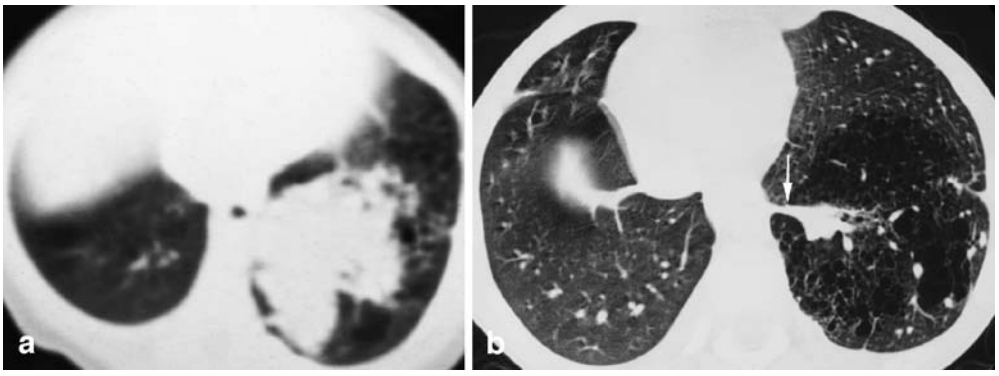

Fig. 6 Intralobar sequestration/CPAM – probable hybrid lesion. **a** CT scan on a newborn girl with a rounded mass in the left lower lobe on chest radiograph (not shown). This appears to be a predominantly solid mass with some possible peripheral aerated cysts (5-mm slice thickness, non-helical scan, lung window). The parents refused surgery. **b** Same child at age 7 years. In the interim, the child had several episodes of pneumonia. The lesion is now completely aerated and consists of multiple small air-filled cysts resembling type II CPAM. A large systemic artery (*arrow*) arising from the descending aorta supplied the lesion (high-resolution CT scan, 1-mm slice thickness, lung window)

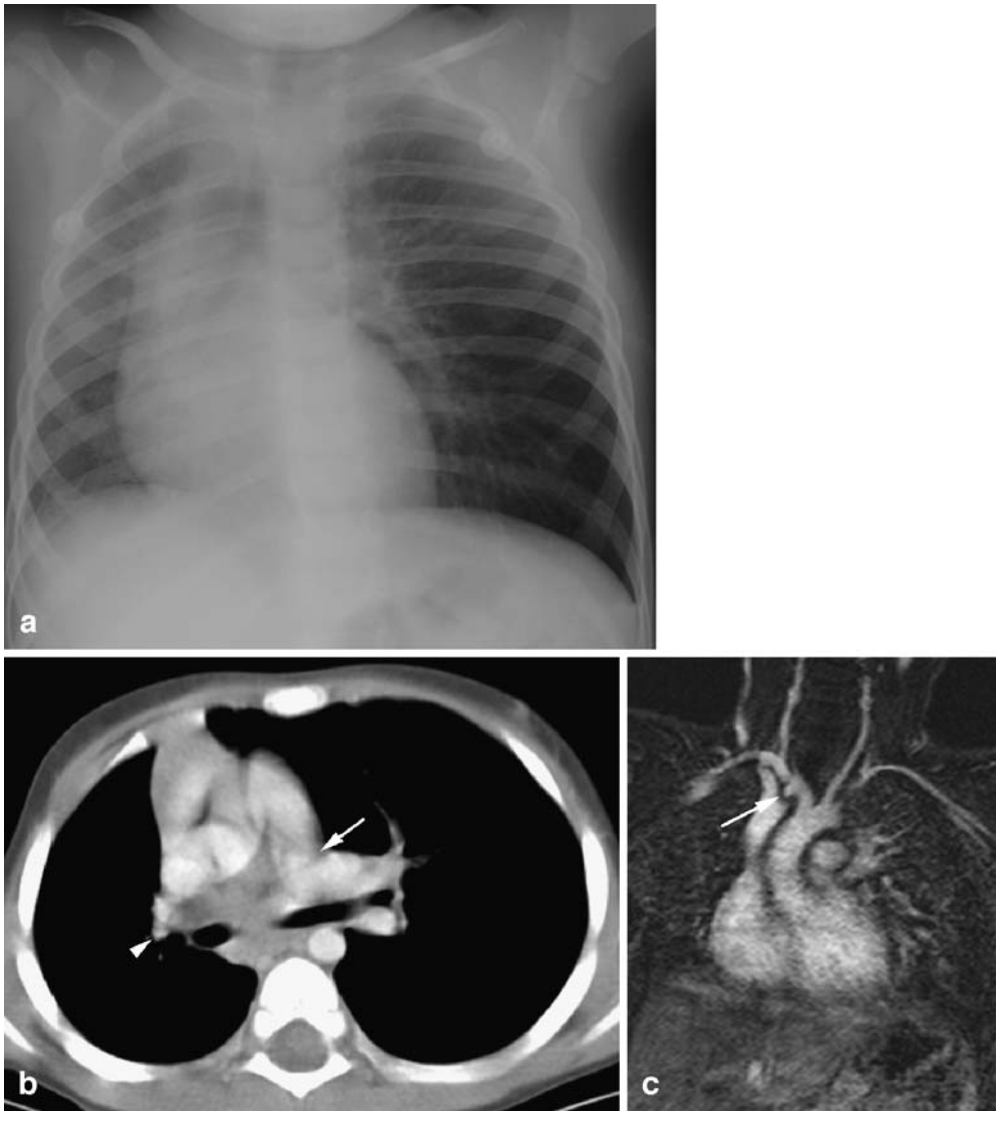

Fig. 7 Absent right pulmonary artery and pulmonary hypoplasia. **a** This 2½-year-old boy had a hypoplastic right lung on chest radiograph. **b** The axial contrasted CT scan (5-mm slice thickness, single helical detector) demonstrates a large left pulmonary artery (*arrow*) and absent proximal right pulmonary artery. A tiny hilar right pulmonary artery is seen (*arrowhead*). Note the small volume right lung and ipsilateral cardiomediastinal shift. **c** Coronal MRA. Thin maximum-intensity projection reconstruction demonstrates a right-side ductal diverticulum (*arrow*) at the base of the right innominate artery, the remnant of the ductal vessel supplying flow to the right lung in utero. The right lung is small with decreased pulmonary vascular branching in comparison with the left lung

foregut lesions. These include tracheal atresia, pulmonary sequestration and foregut duplication cysts [9, 16, 19] (Figs. 5 and 13).

An esophagram is usually the most useful imaging tool for evaluating this possibility. Thin-cut CT and MR images with multiplanar reformatting are additional methods of depicting this anatomy (Figs. 5 and 13). Although gastrointestinal communication is an uncommon feature of foregut malformations, it is important to bear this association in mind, as it is often overlooked.

Ectopic location

Foregut malformation lesions are typically intrapulmonary but can occur in other sites including the neck, mediastinum, pericardium and upper abdomen. Lesions described in such ectopic locations include sequestration, bronchogenic cyst and CPAM [14]. Approximately 15% of extralobar sequestrations are found below the diaphragm, usually in the left suprarenal area [22, 31] (Fig. 14).

Other organ system malformations

Other malformations commonly occur in conjunction with congenital bronchopulmonary/foregut lesions. These include musculoskeletal (especially rib and vertebral anomalies); cardiovascular (especially atrial and ventricular septal defects and patent ductus arteriosus); other gastrointestinal lesions (diaphragmatic hernia, duodenal stenosis and atresia, bowel duplication, anorectal malformations); genitourinary anomalies (malposition, obstruction, renal cystic disease); and occasional central nervous system anomalies [2, 19, 32, 33] (Figs. 10 and 12).

This spectrum of anomalies brings to mind the well-known VACTERL association: Vertebral anomalies, imperforate Anus, Cardiac anomalies, TracheoEsophageal fistula, Renal anomalies, Limb anomalies (radial ray). Malformations other than tracheoesophageal fistula are associated with a similar spectrum of systemic anomalies, though the frequency and type of lesions that occur can vary considerably. For example, though the range of VACTERL anomalies is found accompanying tracheal atresia, complex cardiac lesions are more commonly aligned with tracheal atresia, and vertebral anomalies and imperforate anus are more frequently found along with tracheoesophageal fistula [34].

Part II: Specific lesions and issues

In this section special consideration is given to selected congenital bronchopulmonary lesions with emphasis on controversial issues, recognition and understanding as well as imaging and management.

Pulmonary sequestration

Questions and issues:

(1) What should be included in the spectrum of pulmonary sequestration?
(2) Hybrid lesions, particularly overlap with CPAM.
(3) Intra- versus extralobar sequestration.
(4) Is intralobar sequestration a congenital or acquired lesion?
(5) Imaging and management.

Pulmonary sequestration is usually defined as lung with no normal bronchial connection and an aberrant systemic arterial supply [10, 33, 35] (Figs. 5, 6 and 11). There has been disagreement as to whether the spectrum of sequestration should include other forms of abnormal systemic arterial supply such as to abnormal lung that maintains a bronchial connection or even aberrant systemic arterial supply to normal lung [10, 12, 15] (Figs. 9 and 10). I tend to favor an inclusive definition of aberrant systemic arterial

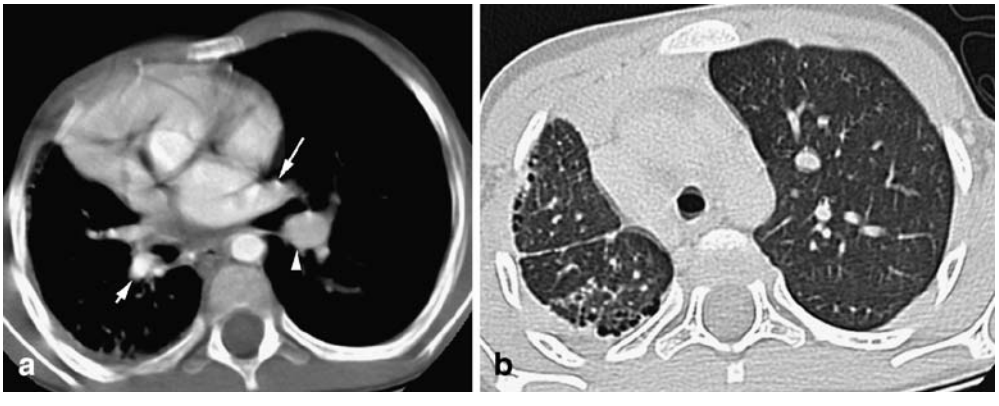

Fig. 8 Unilateral pulmonary vein atresia. A 5-year-old boy with recurrent wheezing and small right lung on chest radiograph. **a** Contrasted CT – (mediastinal window, 5-mm slice thickness). The right lung is hypoplastic with ipsilateral cardiomediastinal shift. Right-side hilar pulmonary arteries (*short arrow*) are smaller than those on the left (*arrowhead*). No central right pulmonary veins are seen. Note normal left superior pulmonary vein (*long arrow*). There is peripheral pleural thickening and scarring on the right. **b** CT lung window (1-mm slice thickness, high resolution). Note bilobed hypoplastic right lung with peripheral reticular and cystic changes indicative of either fibrosis/infarction or pulmonary dysplasia. There were bilateral hyparterial bronchi (not shown)

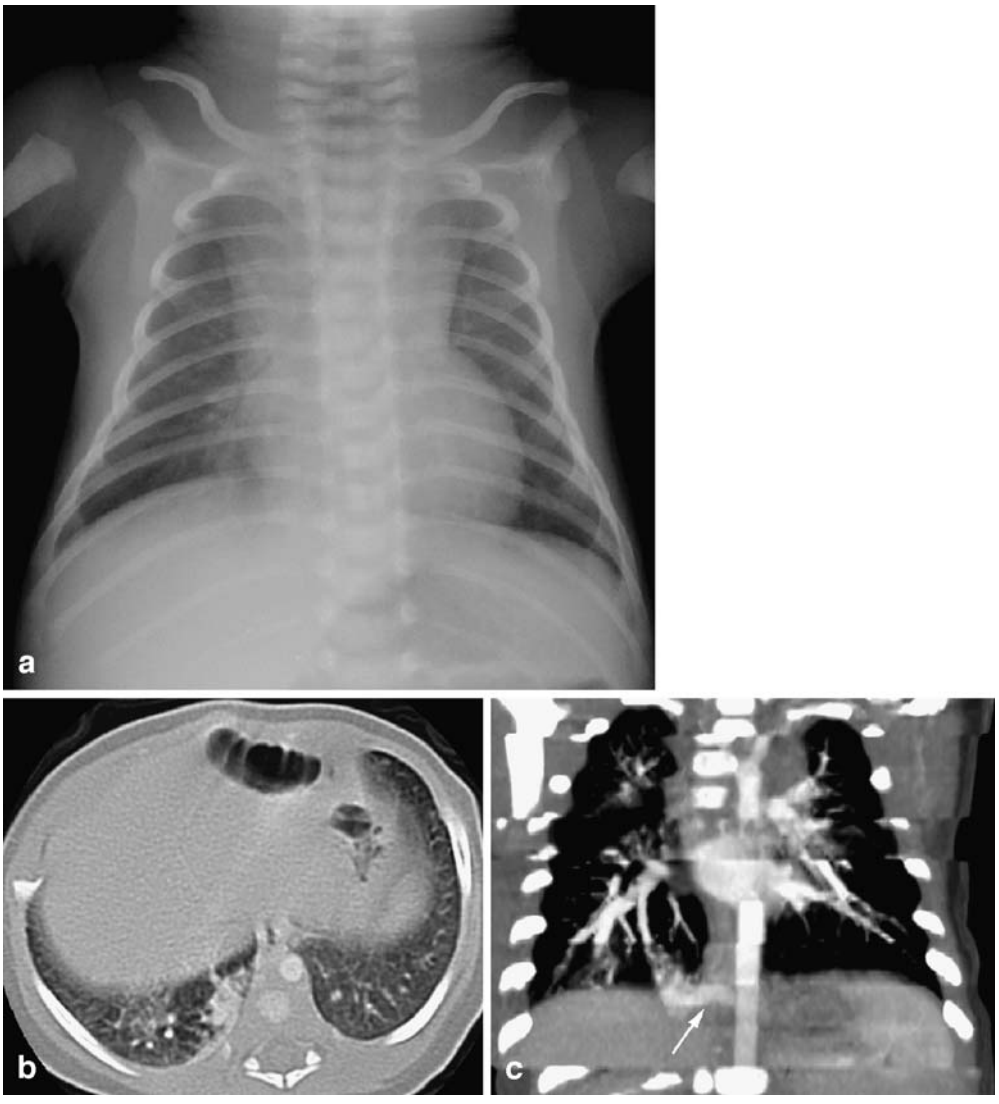

Fig. 9 Intralobar pulmonary sequestration – aerated at birth. This 6-week-old boy was found to have a congenital lung malformation, perhaps sequestration, on prenatal US. **a** The chest radiograph demonstrates subtle linear opacity in the medial right lower lobe. **b** CT axial image (lung window, slice thickness 2.5 mm, four-detector scanner) demonstrates large dense branching vessels surrounded by hyperlucent lung with possible small cystic changes in the medial right lower lobe. **c** Coronal CT (maximum intensity projection reconstruction – axial images revised to 1.25 mm with coronal reformats) demonstrates a large systemic artery from the abdominal aorta (*arrow*) entering the medial right lower lobe sequestration. This lesion was resected; pathology did not demonstrate features of CPAM, but some pulmonary hyperplastic dysplastic changes were present (see Fig. 4)

supply to the lung as part of the pulmonary sequestration spectrum. It is helpful to also include other foregut malformations associated with aberrant systemic arterial supply. With the increasing prevalence of prenatal US and Doppler and progressively more detailed angiographic cross-sectional imaging techniques, many more such hybrid lesions are being described [10, 15, 22, 23] (Figs. 5, 6 and 14). Especially common are mixed imaging and pathologic features of sequestration and small cyst CPAM, present in up to 50% of lesions [10, 23] (Figs. 5, 6 and 14). Consideration of many of these lesions as the spectrum of an in utero obstructive malformation sequence with secondary pulmonary dysplastic changes helps one to understand and integrate seemingly endlessly varied and disparate findings. I endorse the suggestion of more general terminology and then specific systematic description of all abnormal component findings including arterial, venous, lymphatic, airway and pulmonary parenchymal regions [26].

Pulmonary sequestration is typically divided into intralobar and extralobar types. These are differentiated by a separate pleural investment in extralobar sequestration (Figs. 5, 11 and 14) and inclusion of intralobar sequestration within the lung (Figs. 6 and 9). There are many similarities between these entities and there is much overlap. In both types, systemic arterial supply usually arises from the distal thoracic or proximal abdominal aorta, sometimes from the celiac or splenic arteries, reaching the lung via the inferior pulmonary ligament [36]. Occasionally, the systemic vessel originates from intercostal, subclavian or even coronary arteries [37]. These vessels are thought to be persistent, primitive, post-branchial arteries that supplied the lung before the development of the pulmonary artery. Venous drainage is typically to the pulmonary (intralobar) or systemic (extralobar) veins, with overlap and sometimes dual drainage. An abnormal communication with the gastrointestinal tract, especially the esophagus, can occur in both types and is particularly likely when airway elements are present along with vessels in the inferior pulmonary ligament [9, 14, 33] (Fig. 5).

Langston [14] refers to the intralobar form of sequestration as a variant of bronchial atresia occurring in the lower

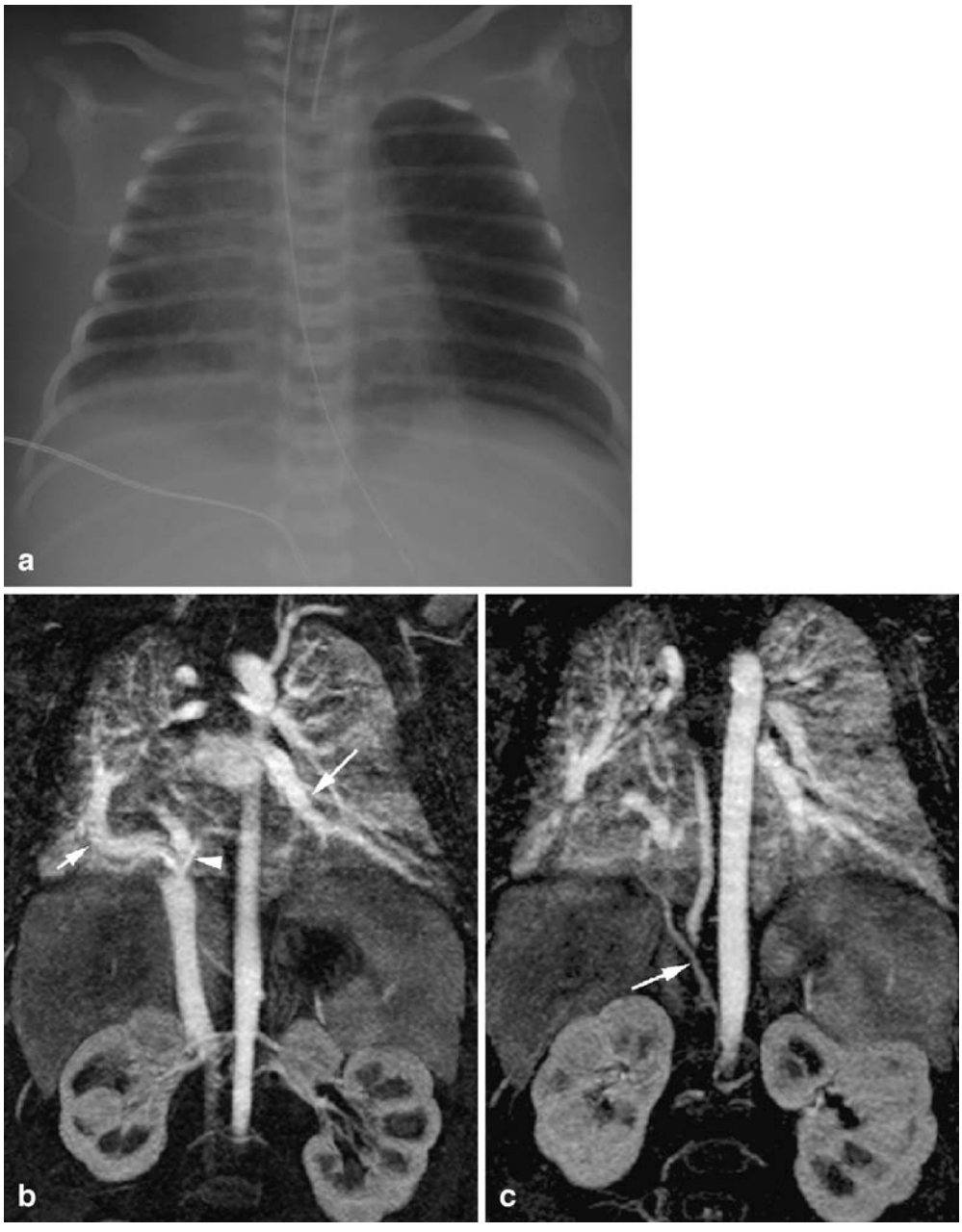

Fig. 10 Scimitar syndrome with unilateral obstructed anomalous veins and horseshoe lung. This newborn with tetralogy of Fallot had severe respiratory distress that required extracorporeal membrane oxygenation (ECMO) for 1 week. **a** Chest radiograph. There is persistent haziness and decreased size of the right lung with ipsilateral cardiomediastinal shift. Pulmonary hypoplasia or scimitar syndrome was suspected, though no scimitar vein was visualized. **b**, **c** Coronal thin maximum-intensity projection reconstructions from the MR angiogram. **b** Multiple dilated anomalous right pulmonary veins drain to the inferior vena cava with stenosis at the junction with the cava. The veins include a scimitar vein (*short arrow*) as well as a separate vein (*arrowhead*) draining a central horseshoe lung segment. There is a normal lower left pulmonary vein draining to the left atrium (*arrow*). **c** There is a moderate-sized systemic artery (*arrow*) arising from the celiac artery and supplying part of the right lower lobe, possibly directly joining a pulmonary vein. No airless sequestered lung segment was seen

lobes. There are indeed many commonalities between the two entities; in both there might be an identifiable atretic bronchus. The pathologic appearance of sequestration closely follows that of bronchial atresia, with dilated, branching, fluid-filled airways. Dysplastic pathologic features including pulmonary hyperplasia and microcystic parenchymal changes are both commonly present in intralobar sequestration, as well as more typical upper lobe bronchial atresia [14]. However, these features, anatomic and pathologic, also occur in extralobar sequestration, including an atretic bronchus that might be ectopic rather than adjacent to vessels. Labeling all intralobar lesions as bronchial atresia and the extralobar form as sequestration seems even more confusing than our current terminology.

The overlap between sequestration and bronchial atresia as well as other bronchopulmonary foregut lesions serves to emphasize their probable common origin as an obstructive malformation complex.

Controversy with regard to the congenital versus acquired nature of intralobar sequestration still exists. It has been suggested that repeated lung inflammation could cause small, normally present systemic pulmonary ligament vessels to hypertrophy, producing an acquired intralobar sequestration [20]. In older series, most of the cases of intralobar sequestration were found in older children and adults, with extralobar sequestration occurring more commonly in infants. However, with the increased frequency of prenatal US, many cases of intralobar sequestration have been described both prenatally and in

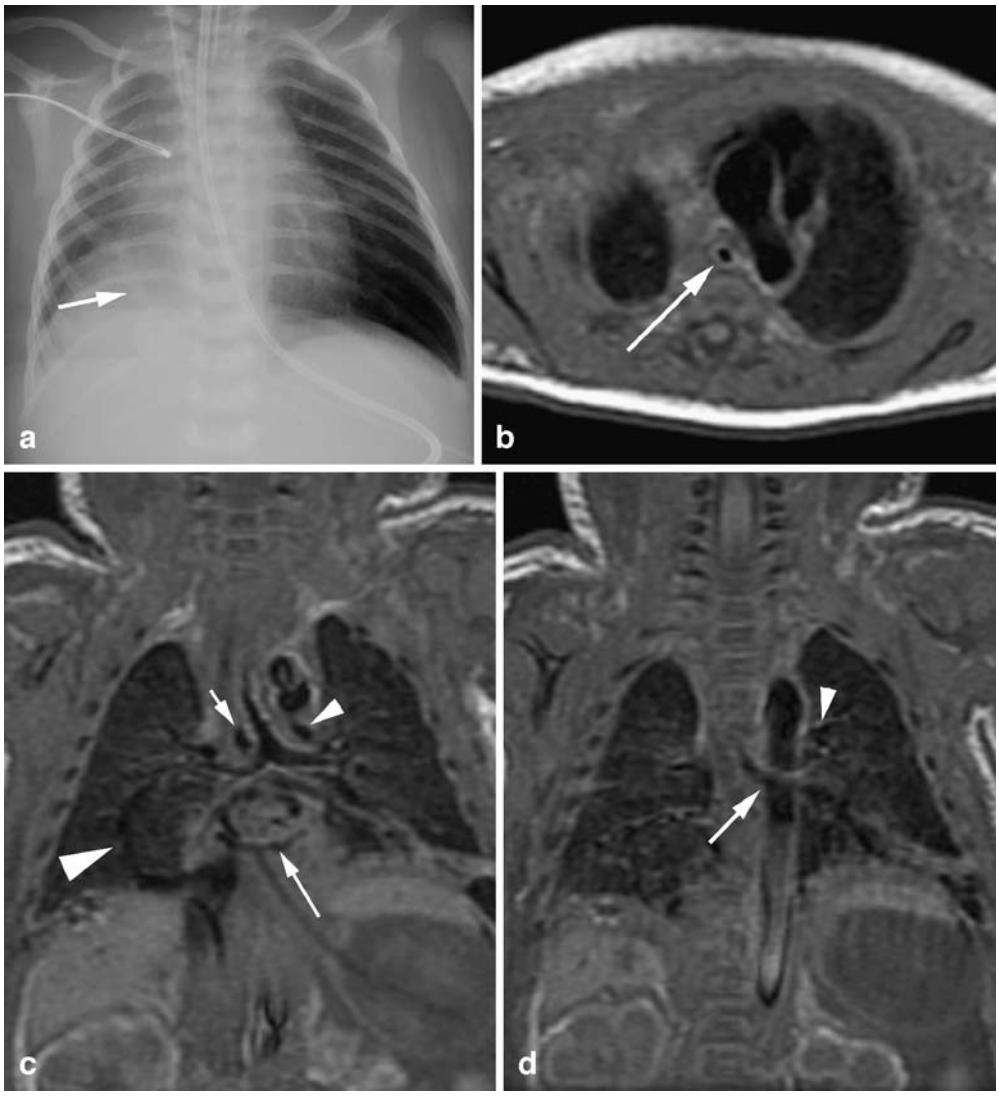

Fig. 11 Hypoplastic right lung, partial pulmonary artery sling, tracheal stenosis, scimitar syndrome, horseshoe lung and pulmonary sequestration in a 10-day-old boy with respiratory distress. **a** Chest radiograph. There is a small-volume hypoplastic right lung with decreased vascular branching and ipsilateral cardiomediastinal shift. There is a curving tubular density in the right lower lung suggestive of a scimitar vein (*arrow*) and moderate right lower lobe opacity. The trachea and bronchi are poorly visualized. **b** Axial T1-weighted MR image at the level of the aortic arch (left side) demonstrates a narrow trachea (*arrow*). The circular configuration and circumferential signal around the airway suggest stenosis with complete cartilaginous rings. **c** Coronal T1-weighted MR image. Note the long segment tracheal stenosis and T-shaped low carina characteristic of type II pulmonary artery sling. Normal right (*short arrow*) and left upper (*small arrowhead*) pulmonary arteries are seen on either side of the airway. Also visualized is a curving right-side scimitar vein draining to the inferior vena cava (*large arrowhead*). Centrally, there is a horseshoe segment of airless sequestered lung (extralobar) containing a tangle of systemic arterial vessels (*large arrow*) that arose from the descending thoracic aorta (not shown). Venous drainage of the segment was to the inferior vena cava. The linear tubular density extending to the stomach represents the nasogastric tube. **d** Coronal MR image immediately posterior to **c**, demonstrating a partial pulmonary artery sling with the left lower pulmonary artery (*arrow*) arising from the right pulmonary artery and crossing behind the trachea. The left upper pulmonary artery (*arrowhead*) arose normally from the main pulmonary artery (not shown)

young infants (Fig. 9), with an incidence similar to extralobar sequestration [20]. In some instances, both intralobar and extralobar sequestration have been found in the same infant [10, 14, 20, 33, 36]. Both types of sequestration have been described in conjunction with other lung anomalies such as bronchogenic cyst, bronchial atresia, CPAM, scimitar syndrome and gastrointestinal communication [10, 15, 20] (Figs. 5, 6, 10, 11 and 14). Data seem to support a predominantly congenital origin for both intra- and extralobar sequestration.

The differences that appear to remain between extralobar and intralobar sequestration include:

1. Ectopic location. Ectopic locations in the neck, mediastinum, pericardial and intraabdominal (especially left suprarenal) are seen with extralobar and not intralobar sequestration [22, 23, 38] (Fig. 14). By definition, intralobar sequestration is in continuity with lung tissue. Ectopic intraabdominal sequestrations are typical in appearance, with uniform echogenicity with or

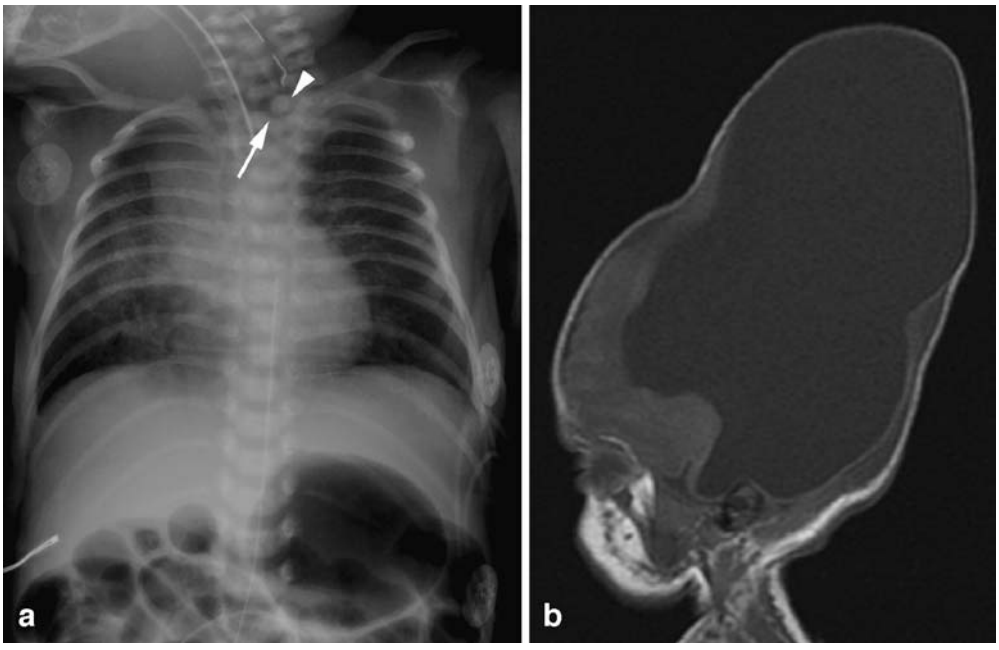

Fig. 12 Tracheoesophageal fistula, vertebral anomaly and encephalocele in a 2-day-old infant with skull mass and respiratory distress. **a** The frontal chest radiograph demonstrates location of the nasogastric tube in a dilated, air-filled esophageal pouch suggesting esophageal atresia (*arrow*). A distal fistula is inferred from the presence of abdominal gas. Patchy right lung opacity likely represents aspiration. A left hemivertebra is noted at the level of the esophageal pouch (*arrowhead*). **b** A sagittal T1 MR demonstrates a large parietal meningoencephalocele

without cysts evident on sonography and hypodensity on CT [22, 38] (Fig. 14).

2. Extralobar sequestration has a higher association with other systemic malformations such as congenital diaphragmatic hernia, pulmonary hypoplasia, pulmonary sling and other foregut cysts [16, 33] (Figs. 5 and 11).

Both types of pulmonary sequestration might be associated with development of in utero hydrops fetalis. This is thought to be related to large lesion size with compression of the esophagus and thoracic venous structures and resultant polyhydramnios, congestive failure, hydrothorax and fetal hydrops [39]. In these cases, intervention such as resection, drainage, or early delivery is necessary to prevent fetal demise [10, 14].

Pulmonary sequestrations are often asymptomatic at birth and found either incidentally at surgery or imaging or after recurrent infection. Early symptomatology and prognosis is related to the presence of a large systemic arterial supply with a large left-to-right shunt as well as the severity of associated anomalies such as cardiovascular lesions [10] (Figs. 10, 11).

The imaging appearance of sequestration is quite varied. In infants it is most often described as a solid mass lesion, most commonly in the medial left lower lobe [10] (Fig. 6). However, intralobar sequestration can be aerated at birth, appearing as a hyperlucent lesion. This appearance is being reported more frequently in follow-up of lesions found on prenatal US [39] (Fig. 9). In older children and adults, hyperlucent areas with small cysts are often seen, best demonstrated on high-resolution CT [35] (Fig. 6). These lesions overlap with CPAM both on imaging and pathologic evaluation. Development of collateral air channels has been postulated as leading to aeration of sequestration

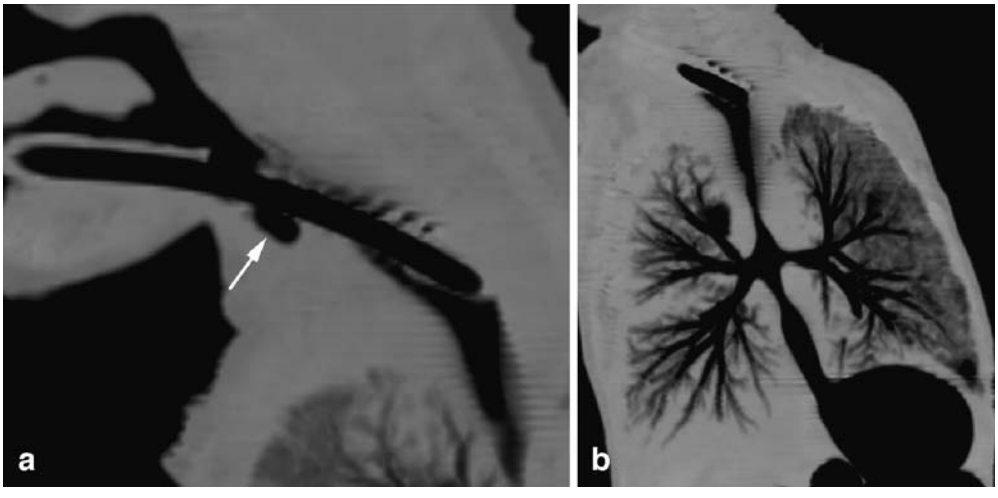

Fig. 13 Newborn with tracheal atresia. Sagittal (**a**) and coronal (**b**) minimum-intensity projection reconstructions from an axial CT scan (four-detector scanner, 2.5-mm thick slices reconstructed to 1.25 mm). **a** There is an endotracheal tube in the esophagus; the trachea could not be intubated. A short blind segment of proximal airway is seen (*arrow*). **b** The right and left bronchi arise independently from the mid esophagus (type III tracheal atresia). Note stenosis of the esophagus above and below the bronchial origins. The intrathoracic trachea was entirely absent

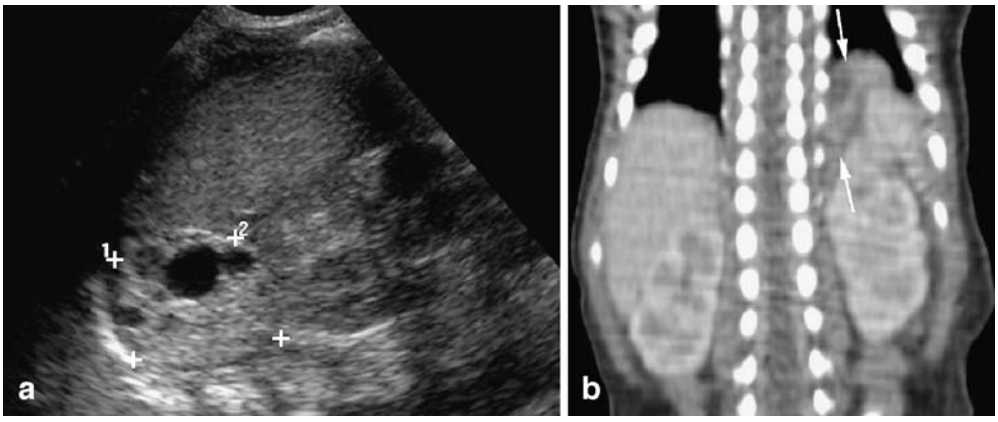

Fig. 14 Abdominal sequestration/CPAM hybrid in a 5-day-old girl with a left suprarenal mass seen on prenatal US. **a** Sagittal US image demonstrates an echogenic mass containing small cysts in the posterior left suprarenal subdiaphragmatic region (mass outlined by measurement calipers). This appeared separate from the adrenal (not shown). **b** Coronal contrasted CT reconstruction demonstrates a posterior low-attenuation mass (*arrow*) with associated focal eventration of the left hemidiaphragm (images courtesy of Hospital for Sick Children, Toronto)

lesions. Some authors have suggested that these are facilitated by recurrent infection. However, the presence of partially or completely aerated sequestrations in young infants suggests that infection might be a result rather than the cause of aeration in pulmonary sequestration.

The presence of air in an extralobar sequestration implies a gastrointestinal connection [9] (Fig. 5).

The vascular components of pulmonary sequestration, including systemic arterial supply and venous drainage, are increasingly well-demonstrated with CT and MR angiographic techniques with almost no need for conventional angiography for diagnosis [10, 33, 35] (Figs. 6, 7, 8 and 11).

Most recognized pulmonary sequestrations are removed surgically. In some cases, embolization or ligation of the systemic artery has been performed rather than resection. With this approach, there might be complications related to lung infarction or infection. There is also a questionable small risk of development of malignant neoplasm, especially in hybrid lesions with components of CPAM [21, 40, 41].

Congenital pulmonary airway malformation

Questions and issues:

(1) Terminology, classification and overlap lesions
(2) Imaging
(3) Natural history and management

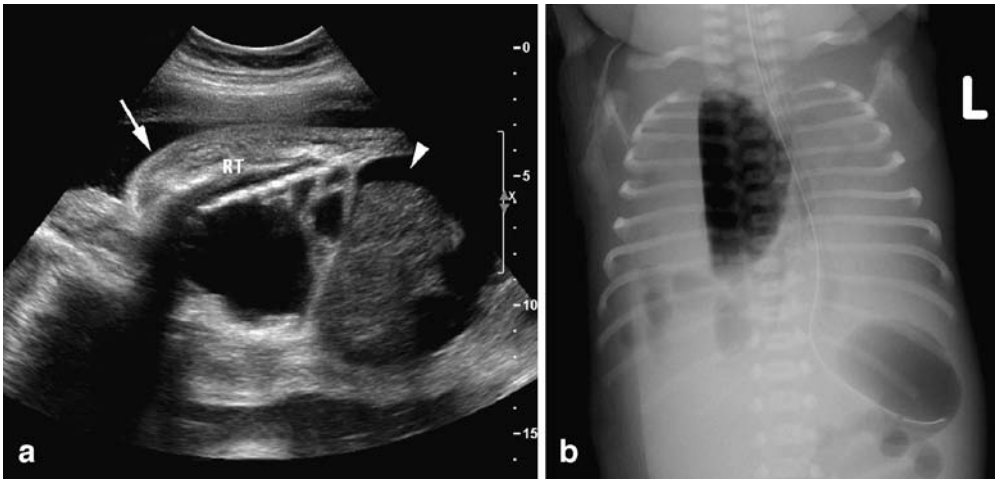

Fig. 15 Type I/III CPAM of the lung. **a** An in utero longitudinal US of the chest at 28 weeks demonstrates multiple large right-side pulmonary cysts occupying most of the chest. Note marked edema of the body wall (hydrops) (*arrow*) and ascites (*arrowhead*). In utero cyst aspiration was performed. The baby was born at 30 weeks by emergency cesarean section. **b** Right lateral decubitus chest radiograph at birth. In the right lung, there is a single, very large cyst and multiple smaller cysts with air-fluid levels denoting incomplete removal of fetal lung fluid. In spite of the decubitus positioning, there is marked mediastinal shift to the left. The left lung is opacified, probably because of atelectasis, fluid, and/or respiratory distress syndrome of the newborn. Progressive expansion of the cystic malformation precipitated surgical resection of the right lower lobe. Pathology revealed findings consistent with both type I and type III CPAM

CPAM is characterized by overgrowth of bronchial structures at the expense of alveoli associated with an abnormal supplying airway (lacking cartilage) [1, 3, 5, 8].

There is considerable controversy over classification and nomenclature. Congenital pulmonary airway malformation has recently been recommended as a preferred term to congenital cystic adenomatoid malformation because not all of the lesions are cystic and only type III is adenomatoid [25, 41]. Stocker [25] originally classified these lesions into three types: type I, macrocystic (>2 cm); type II, multiple smaller cysts; and type III, solid form (microscopic cysts). An expanded classification (types 0–4) has been proposed representing malformations of larger through smaller airways with some overlap with cystic pleuropulmonary blastoma [25, 41]. Simple separation into large cyst and small cyst types has also been suggested as a more practical alternative [5, 14, 42], and this approach works well with regard to imaging.

Both large and small cyst types can be associated with abnormal systemic arterial supply, more often in the small cyst form [14]. Langston [14] suggests that the small cyst (type II) and solid types (type III) represent the dysplastic consequences of airway obstruction in utero, hence the frequent imaging and pathologic overlap with tracheal atresia, bronchial atresia, pulmonary sequestration, bronchogenic cyst and CLH [12, 14–16, 36, 38] (Figs. 4, 5, 6 and 14). If one accepts this theory, CPAM, especially types II and III, might not be a separate entity.

The large cyst form is most common and often presents in early infancy with respiratory distress and air-trapping (related to an abnormal bronchial connection). There is compression of normal lung with contralateral mediastinal shift (Fig. 15). Emergent surgical resection is often required [43]. In the early postnatal period, the affected lung might appear opaque, secondary to slow clearing of fetal lung fluid, the hallmark of a lesion with an abnormal supplying airway [3] (Fig. 15). Beyond the newborn period, the most common presentation of CPAM is with superimposed pneumonia or as an incidental finding on a chest radiograph obtained for other reasons [40, 43].

Both large and small cyst lesions are identified prenatally with increasing frequency, and most have a good prognosis [29, 39]. Microcystic lesion (type III), bilateral lung involvement and hydrops are poor prognostic features [43]. Fetal hydrops and stillbirth are rare; when present they are associated with a large lesion likely related to the same sequence of esophageal and venous compression as seen in large pulmonary sequestrations (Fig. 15). These usually require emergent intervention [13, 24, 36, 42, 43] and are most commonly type I (large cyst) or type III (microcystic) lesions. Occasionally the lesion is large enough to produce pulmonary hypoplasia with subsequent pulmonary hypertension and significant morbidity and mortality even after surgical removal [42–44].

Many of the lesions, including those that overlap with pulmonary sequestration, are followed in utero, peak in size during the second trimester and then progressively decrease in size or even disappear sonographically in the third trimester [13, 36, 39]. Even though these infants might be asymptomatic at birth with a normal-appearing chest radiograph, a residual lesion is often found on postnatal CT imaging [13, 36, 40, 42, 43] (Fig. 9). This suggests that in the past there were many unrecognized small lesions present in newborns. Other lesions are stable over a long period of time in utero and asymptomatic at birth [31, 36], with recognizable cystic changes on postnatal chest radiographs. The natural history, i.e. further regression and disappearance or persistence, is incompletely known.

The question is repeatedly raised as to whether surgical resection of these asymptomatic lesions is appropriate. Recurrent infection, pneumothorax, hemorrhage and a small risk of malignant neoplasia have been cited as reasons for elective resection [10, 14, 31, 39, 40]. It has been suggested that most of the neoplasms, at least those found in association with large cyst malformations, might represent unrecognized low-grade cystic pleuropulmonary blastoma, essentially indistinguishable on imaging [40, 41] (Fig. 16). Laberge et al. [40] have likened the relationship between CPAM and pleuropulmonary blastoma to that of nephrogenic rests and Wilms tumor. There are clearly a small number of tumors that occur within previously unresected or incompletely removed CPAM lesions and even at the site of lesions that were thought to have been completely resected [4, 31, 41, 44–46]. These are usually pleuropulmonary blastomas in infants and bronchoalveolar carcinomas in older children and adults [13, 40, 45].

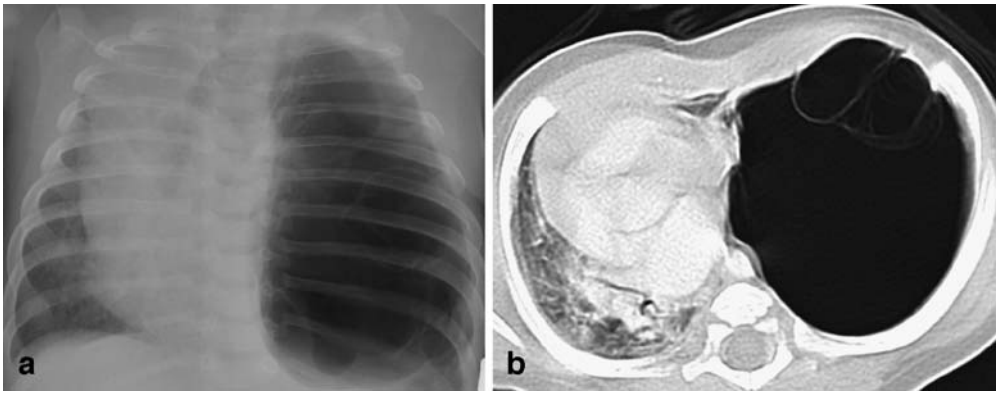

Fig. 16 Cystic pleuropulmonary blastoma (pathologically proven) in a 1-month-old with respiratory distress. **a** Chest radiograph. There is marked overinflation of the left lung with multiple cysts producing contralateral cardiomediastinal shift. **b** CT mediastinal window. Multiple large cysts with mass effect are noted within the left lung, suggesting type I CPAM

Currently, most centers still recommend elective surgical removal of congenital cystic lung lesions [13, 24, 36, 40, 42–44]. This might change, however, as increasing numbers of small asymptomatic lesions are recognized because of prior prenatal US.

Pulmonary hypoplasia

The many variations of pulmonary hypoplasia typify the frequent coexistence of vascular and airway anomalies. This association is often poorly understood or appreciated on imaging.

Pulmonary hypoplasia is rarely primary or idiopathic in origin. Extrinsic or intrinsic lung compression is often implicated as an underlying condition, these include [4, 17, 27, 28, 47–49]:

(1) Intrinsic space-occupying lesions, e.g. congenital diaphragmatic hernia, large CPAM or sequestration and large pleural effusions
(2) External uterine compression associated with oligohydramnios, as in renal absence or severe dysfunction or prolonged amniotic fluid leak
(3) Chest wall compression caused by malformations such as kyphoscoliosis and some skeletal dysplasias
(4) Neuromuscular or chromosomal disorders probably related to decreased fetal breathing movements
(5) Decreased pulmonary vascular perfusion, including cardiac lesions such as tetralogy of Fallot, pulmonary atresia, hypoplastic right heart and primary pulmonary vascular abnormalities

Persistent pulmonary hypertension is a life-threatening condition that is frequently present in babies with pulmonary hypoplasia, whether primary or secondary. This appears to be related to both an overall decreased pulmonary vascular bed as well as increased peripheral arterial muscularization and labile vasoconstriction of pulmonary arterioles. Treatment is very difficult and includes nitric oxide, oscillating ventilation and extracorporeal membrane oxygenation (ECMO) [17].

Radiographically, bilateral pulmonary hypoplasia might be difficult to recognize. Radiographic features include: small-volume, hazy lungs; unexpected or spontaneous air leak phenomena; and a bell-shape thorax or elevated diaphragm. Specific findings such as mass, hernia, effusion or chest wall or diaphragmatic abnormality might be present, depending on the underlying cause [4, 17].

With unilateral hypoplasia, the heart and mediastinum are usually shifted to that side, varying in degree with the size of the affected lung (Figs. 7, 8, 10 and 11). The hypoplastic lung is often hyperlucent (unless venous obstruction or sequestration is present) (Fig. 10), with a smaller hilum. Pulmonary hypoplasia is associated with a similar spectrum of anomalies that is found in other congenital bronchopulmonary lesions. These include musculoskeletal, diaphragmatic, renal, cardiovascular, bronchopulmonary and gastrointestinal tract anomalies [4, 17].

In a child with a unilateral small lung, an acquired airway lesion such as Swyer James (bronchiolitis obliterans) might be considered a differential diagnostic possibility. Patchy air-trapping on expiratory CT or ventilation lung scan serves to differentiate this from congenital pulmonary hypoplasia [4].

Pulmonary hypoplasia associated with vascular anomalies

This association is most common with unilateral hypoplasia and is most often right-sided. There might be an ipsilateral decrease in size, number and branching of vessels (arteries and veins) that occurs as a secondary phenomenon to pulmonary hypoplasia [27]. More common, however, is an association between what appears to be a developmental abnormality of the pulmonary vascu-

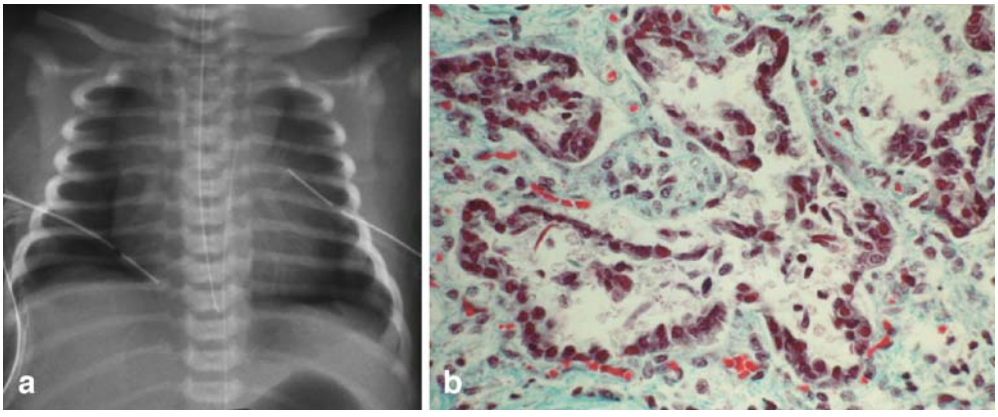

Fig. 17 Alveolar capillary dysplasia. A 2-hour-old full-term girl with severe respiratory distress and persistent pulmonary hypertension. **a** The chest radiograph demonstrates clear lungs with bilateral pneumothoraces and chest tubes. The infant died at 18 hours of age in spite of aggressive therapy. **b** Autopsy biopsy of the lung; high-power view (x600) trichrome stain. Interstitial and fibrous tissues are increased; vessels are sparse. No alveoli are present; there are large branching airways lined by cuboidal/columnar epithelium (**a**, **b** reprinted with permission from *Pediatric Radiology* [50])

lature with consequent pulmonary hypoplasia. These entities include:

- Alveolar capillary dysplasia
- Pulmonary artery sling
- Absent pulmonary artery
- Absent pulmonary vein
- Scimitar syndrome

Alveolar capillary dysplasia This extremely rare condition affects both male and female infants equally. Most cases are sporadic, with a genetic component suggested by some occurrence in siblings [50]. Babies with this lethal disorder present at or shortly after birth with pulmonary hypertension and persistence of the fetal circulation, but without the usual predisposing factors, i.e. meconium aspiration, perinatal asphyxia and sepsis. The lungs are usually clear or mildly hazy, with air-leak phenomena occurring commonly [51, 52] (Fig. 17). The clinical course is typically an unrelenting downhill progression.

Normally all preacinar bronchial and arterial divisions occur together and are completed by 16 weeks of gestation. After this, acinar structures and saccular distal air spaces develop and become vascularized. True alveolar development and multiplication begins at 32 weeks and continues until 8 years of age [51].

In babies with alveolar capillary dysplasia, there appears to be a primary arrest of both vascular and lung development with lack of formation of terminal air spaces and thickened interstitial tissues, with sparse vessels poorly related to the distal airways [25, 50, 51]. In many cases, there is misalignment of lung vessels with pulmonary veins contained in a central common sheath along with distal bronchioles and arterioles instead of their normal location in the periphery of the pulmonary lobule [52].

Other congenital anomalies are seen in 50% of cases. These include gastrointestinal tract anomalies (aganglionosis, imperforate anus, malrotation, tracheoesophageal fistula, duodenal stenosis or atresia), cardiovascular lesions (left-to-right shunts or complex congenital cardiac anomalies) and genitourinary anomalies [50].

Alveolar capillary dysplasia should be considered in the setting of a full-term infant with severe respiratory distress and only mild radiographic pulmonary changes (Fig. 17). It differs from other causes of neonatal pulmonary hypertension in that there is frequently a curiously variable period after birth (hours, days and even weeks) before symptoms begin. The diagnosis is confirmed by lung biopsy.

No effective treatment is available, with the potential exception of lung transplantation, though it is usually not possible to support the infant long enough for an organ to become available.

Pulmonary artery sling In this entity, the left pulmonary artery arises anomalously from the posterior right pulmonary artery and passes between the trachea and esophagus

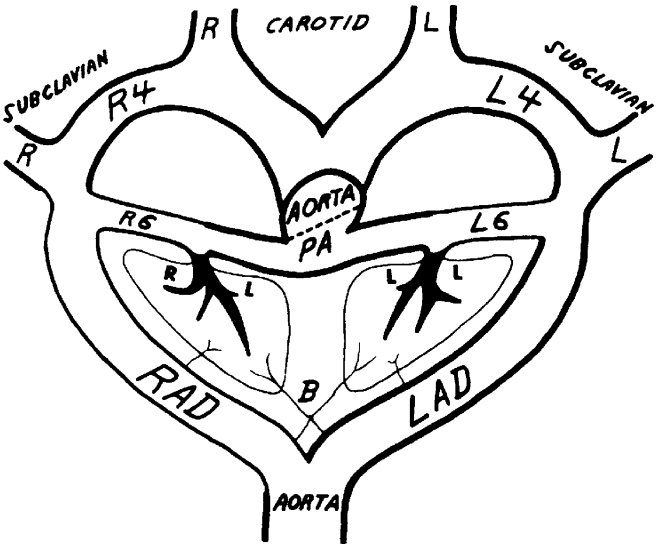

Fig. 18 Diagram of development of the great arteries. Normally the right dorsal aorta (*RAD*) and duct regress early in gestation, leaving a right innominate artery, left arch (fourth arch) and left ductus (distal sixth arch). When the ductus arteriosus persists on the side opposite the arch, it arises from the ipsilateral innominate or subclavian artery (*R4*/*L4* right/left fourth arches, *R6*/*L6* right/left sixth arches, *PA* pulmonary artery, *RAD*/*LAD* right/left dorsal aorta, *RL*/*LL* right/left lungs) (reprinted with permission, AJR [56])

to reach the left lung. There are two major types of left pulmonary artery sling [53, 54]. In type I the airway is usually intrinsically normal and there might be a tracheal bronchus. The anomalous left pulmonary artery might compress the distal trachea and right mainstem bronchus, producing malacia and right-side air-trapping.

Type II (Fig. 11) is more common and is associated with a more inferiorly located left pulmonary artery sling and significant tracheobronchial anomalies, including low carina, long segment airway stenosis with complete cartilaginous rings and abnormal bronchial branching including right bridging bronchus. Other right lung anomalies also occur, including hypoplasia, agenesis, small or absent distal right pulmonary artery, scimitar syndrome, horseshoe lung and pulmonary sequestration [29, 30, 54] (Fig. 11). Cardiovascular and gastrointestinal malformations, including atrial septal defect, ventricular septal defect, patent ductus arteriosus, imperforate anus and Hirschsprung disease, might coexist.

Radiographic findings include a low T-shape carina, narrow or poorly visualized airway and bilateral overinflation or right-side volume loss caused by atelectasis, hypoplasia or agenesis (Fig. 11). In both types, a soft-tissue density might be visualized between the trachea and esophagus occasionally on plain films and more commonly on an esophagram [53].

Left pulmonary artery sling is usually symptomatic because of airway issues or recurrent infection. Cardiovascular anomalies with left-to-right shunting might also cause symptoms. Occasional cases are found incidentally in asymptomatic children or adults. Morbidity and mortality, still about 50%, are primarily related to the

tracheobronchial anomalies, especially long-segment airway stenosis [55]. CT and MRI, especially with newer technology, have proved invaluable in delineating the vascular and airway anomalies and interrelationships (Fig. 11). Echocardiography is the predominant modality for evaluation of associated cardiac defects [55]. Surgical correction needs to address both the vascular and airway anomalies, the latter of which is often much more difficult to manage [53].

Unilateral absence of the pulmonary artery The absent pulmonary artery is typically on the side opposite the arch, more common on the right [27, 56] (Fig. 7). The proximal pulmonary artery is absent, with a hilar artery almost always reconstituted (Fig. 7). In utero, the lung is typically supplied by a persistent ipsilateral ductus. Embryologically, the proximal sixth arch gives rise to the proximal pulmonary artery and the distal sixth arch to the ductus [56, 57]. Normally, the ductus contralateral to the arch disappears very early in development. When the distal right arch regresses, the embryologic position of this duct would originate from the base of the right innominate artery (Fig. 18) or occasionally from an aberrant right subclavian artery [55, 56]. As a result, a residual ductal diverticulum is often seen in this location (innominate artery), reflecting the embryologic pulmonary arterial supply (Fig. 7). Surgically, a residual fibrous cord might be found connecting to the hilar pulmonary artery [57].

This entity might present very acutely in the neonate because of abrupt closure of the supplying ductus with respiratory distress, pulmonary infarcts and effusion [56]. More typically, neonates are asymptomatic, with bronchial collateral vessels enlarging after birth. Patients might remain asymptomatic, but clinical symptoms can develop including pulmonary hypertension, hemoptysis and recurrent infection. Congenital cardiac anomalies such as patent ductus arteriosus, coarctation and tetralogy of Fallot are frequently associated. When large left-to-right shunt lesions are present, early clinical symptoms are much more likely [56, 58].

Radiographically, there is a small lung, small hilum and ipsilateral spidery vascular markings (Fig. 7); peripheral small cystic and possibly dysplastic changes might be present. There is ipsilateral cardiomediastinal shift. Airway branching and pulmonary lobation might also be abnormal [27]. There might be pleural thickening and rib notching related to bronchial collateral vessels [56]. This diagnosis is readily made using angiographic CT/MR images (Fig. 7). Early and accurate diagnosis is important because there is a surgical trend toward early reanastomosis or graft reattachment of the main and hilar pulmonary artery. By providing adequate blood flow and including the lung in the effective pulmonary circuit, it is hoped that both vascular and lung growth will proceed [59].

Less commonly, an absent pulmonary artery occurs on the same side of the arch, invariably left-sided. All such cases have had severe pulmonary outflow obstruction and a left ductus that descends vertically from the underside of the arch (characteristic of lesions dependent on reversed in utero ductal flow, i.e. aorta to pulmonary artery) [56, 59]. Surgically, these cases have a persistent pericardial segment of pulmonary artery present with a shorter gap between main pulmonary artery and left pulmonary artery than with pulmonary artery atresia that occurs on the contralateral side to the arch. This is thought to be an entity acquired at birth as a result of narrowing or closing of the ductus, perhaps with ectopic ductal tissue extending into the left pulmonary artery, similar to etiologies proposed for coarctation or arch interruption [56, 57, 59].

Pulmonary vein atresia In this entity, there is absence of long central segments of the pulmonary veins, typically unilateral, occasionally affecting only one lobe. The etiology is thought to be congenital lack of incorporation of the common pulmonary vein into the left atrium [28]. Approximately half of these patients have associated congenital heart anomalies [60].

There is a spectrum of radiographic findings. The affected lung is usually small with decreased vascularity, probably related to preferential arterial flow to the contralateral lung (Fig. 8). There is impaired growth of the ipsilateral pulmonary artery and lung. Histologically, there might be dysplastic ipsilateral lung tissue [28]. There are often pleural thickening and ipsilateral linear reticular opacities resembling interstitial pulmonary edema (Fig. 8). These findings are thought to be secondary to dilated lymphatics, dilated bronchial veins and patchy parenchymal fibrosis caused by pulmonary infarcts [28, 60]. Nuclear medicine perfusion scans show decreased or no flow on the affected side [28, 60].

Patients might be asymptomatic or present with pulmonary hypertension, hemoptysis and recurrent infection. Surgical removal of the lung might be necessary [28].

Scimitar syndrome The spectrum of what should be considered scimitar syndrome is controversial. The most consistent features are right lung hypoplasia and ipsilateral anomalous pulmonary venous return, usually to the inferior vena cava at or below the diaphragm (Fig. 10). The anomalous vein occasionally drains to the portal vein, right atrium or coronary sinus [4, 11, 27, 61].

More variable components of scimitar syndrome include: hypoplasia or absence of the right pulmonary artery, anomalous systemic arterial supply to the lung from the lower thoracic or upper abdominal aorta with or without sequestered lung, bronchogenic cyst, horseshoe or crossover lung (a portion of the right lower lobe herniates posteriorly behind the heart and fuses with or abuts the left lower lobe), and anomalies of the diaphragm (hernia, eventration, duplication) [4, 11, 17, 27, 30, 61, 62] (Figs. 10 and 11). There are often abnormal right-side bronchial branching and lung lobation. Frequently associated cardiovascular anomalies include atrial septal defect, patent ductus arteriosus, tetralogy of Fallot, aortic coarctation, and left pulmonary artery sling (Fig. 11).

Skeletal abnormalities such as hemivertebrae and genitourinary malformations might also occur. Occasional left-side lesions are described [63].

Many cases are discovered incidentally in asymptomatic individuals. Whether clinical symptoms occur early in life depends on the degree of systemic vascularization with left-to-right shunting as well as the severity of associated cardiovascular anomalies and, particularly, the presence of pulmonary hypertension [64–66]. Multiple factors might contribute to pulmonary hypertension, including large left-to-right shunting caused by systemic arterial supply, anomalous venous drainage and intracardiac defects, as well as pulmonary vein stenosis and right lung hypoplasia [66] (Fig. 10). Symptoms in infancy include congestive failure, dyspnea and recurrent infection. Pulmonary hypertension and hemoptysis might develop in older patients [63–65].

On radiographs, scimitar syndrome is recognized by the presence of a small-volume right lung and cardiac dextroposition along with a curvilinear vertical density of the anomalous pulmonary vein (scimitar vein) in the lower right lung (Figs. 10 and 11). With the advent of angiographic MR and CT techniques and sophisticated reconstruction, excellent detail of the vascular and other anomalies associated with scimitar syndrome can be depicted [11, 28, 64, 65] (Figs. 10 and 11).

Similar issues exist in the approach to treatment of the anomalous systemic arterial supply that is associated with scimitar syndrome and other sequestration lesions. A concern is whether to repair the anomalous venous drainage. The approach depends on whether clinical symptoms are present. Surgical techniques of pulmonary venous reimplantation are still very difficult, but there is now an increased likelihood of operative success [66, 67]. Postoperative venous stenosis and thrombosis is common, and pneumonectomy is sometimes necessary [63].

Conclusion

When evaluating congenital bronchopulmonary/foregut malformations, wherever one anomaly is recognized others should be sought. Combination lesions occur frequently. Airway, esophagus, pulmonary parenchymal, arterial and venous structures should be systematically analyzed.

Many of these anomalies, especially cystic or hyperlucent lesions, appear to fit into an overlapping spectrum of an airway obstruction malformation sequence. Vascular abnormalities are frequently associated. Primary vascular anomalies are a major feature in some lesions, particularly unilateral right-side pulmonary hypoplasia. Thin-section and oblique multiplanar MR images and MR and CT angiographic techniques with multiplanar reconstructions provide excellent definition of these malformations [11, 16, 18, 30, 35] (Figs. 1, 2, 3, 4, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15 and 16). A wide spectrum of other organ system anomalies are frequently associated and should be evaluated.

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