Congenital bronchopulmonary vascular malformations, “sequestration” and beyond

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Abstract
Congenital bronchopulmonary vascular malformations (BPVMs) include a broad spectrum of disorders that involve abnormalities in the form of disruptions of normal communication and/or presence of abnormal communication between one or more of the three main systems of the lung, namely, the airways, arteries, and veins. The establishment of abnormal communications by means of small openings or anastomoses is termed as malinosculation. The aim of this pictorial essay is to illustrate the imaging appearances of the various types of pulmonary malinosculation.

Key words: Congenital bronchopulmonary vascular malformations; malinosculation; systematic classification

Introduction
Congenital bronchopulmonary vascular malformations (BPVMs) include a broad spectrum of disorders that involve abnormalities in one or more of the three main components of the lung, namely, the airways and lung parenchyma, arteries, and veins. Malformations can involve the lung parenchyma, airways, pulmonary or systemic arteries, pulmonary or systemic veins, fistulas with the gastrointestinal tract, or defects in the diaphragm, and various combinations of these.

History and classification of BPVMs
In 1946, Pryce used the term “sequestration” to describe the abnormal lung that was “disconnected” or “secluded” from the normal bronchial tree and had anomalous systemic arterial supply (classically described as intralobar sequestration). However, he found variations and termed the variants as Pryce types I, II, and III [Table 1]. Later, as researchers encountered more and more variations which could not be grouped into Pryce’s types, different terminologies came into use. In 1974, Sade et al., coined the term “sequestration spectrum” to try and encompass all the various combinations of lung and vascular anomalies encountered. Following this, in 1987, Clements and Warner coined a new term, “malinosculation” to describe this spectrum of abnormalities where there is an anomalous communication between the different components of lung tissue, namely, the lung parenchyma, tracheobronchial tree, arteries, and veins. Malinosculation is defined as the establishment of abnormal communications by means of small openings or anastomoses. Presence of disruptions of normal communication was also encompassed in this terminology by Clements and Warne. Lee et al., further refined the concept of malinosculation and classified it as bronchopulmonary malinosculations [Table 2]. This classification is a systematic approach for the evaluation of BPVMs, taking into account isolated and concurrent abnormalities of airway, arteries, and veins [Figure 1].

Basis of classification
Bronchovascular malformations are a complex group of disease entities having variable developmental anomalies. On imaging, each disease entity can show a spectrum of variations in its pulmonary/systemic arterial and
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venous supply. For example, although systemic arterial supply is considered characteristic of sequestration, systemic arterial supply can also occur to normal lung in the absence of sequestration (pseudosequestration).\[^{6,7}\] In addition, some BPVMs like congenital pulmonary airway malformation (CPAM) may have associated abnormal venous drainage. The classically described scimitar syndrome consists of anomalous pulmonary venous drainage of the right lung to the inferior vena cava (IVC) (Scimitar sign on chest radiograph), systemic arterial supply to right lower lobe from the aorta, and hypoplasia of the right lung. Initially, scimitar sign was thought to be specific for scimitar syndrome; subsequently, however, a number of pulmonary venous anomalies like anomalous meandering pulmonary vein, anomalous unilateral single pulmonary vein (AUSPV), and scimitar variant were found to show a similar scimitar sign.\[^{8-10}\] Further, these scimitar variants can be associated with abnormalities of pulmonary airways (e.g. meandering pulmonary vein and pulmonary hypoplasia). Thus, BVPMs should not be viewed as isolated disease entities, but as complex anomalies which can show variable airway, arterial, and venous connections. Classification according to presence of bronchial, arterial, and venous abnormalities may help describing the conditions which do not meet all the criteria of a typical syndrome. For example, a scimitar vein in combination with hypoplasia lung and in the absence of systemic arterial supply would more appropriately be termed as Type E bronchovenous malnmosculation instead of scimitar syndrome. Classification also helps in the systematic study of each entity for abnormalities in airway, arterial, and venous supply. Identification of all the components can have surgical significance.

We present a pictorial essay, trying to explain the various types of pulmonary malnmosculation.

**Type A – Isolated bronchial pulmonary malnmosculation**

This group includes isolated airway abnormalities in the proximal and distal tracheobronchial tree with normal airway anatomy and normal bronchial continuity. Examples include tracheal stenosis, tracheal bronchus, tracheal cyst, congenital bronchial stenosis, bronchial atresia, bronchogenic cyst, CPAM, and congenital lobar emphysema.

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**Table 1: Pryce’s classification of pulmonary sequestration**

| Variant type | Anomalies |
|--------------|-----------|
| Pryce type I | Normal lung with anomalous systemic arterial supply |
| Pryce type II | Anomalous artery supplying “disconnected” lung and adjacent normal lung |
| Pryce type III | Anomalous artery to “disconnected” lung |

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**Table 2: Systematic classification of congenital bronchopulmonary vascular malformations**

| Type A-Isolated bronchial PM | Tracheal stenosis | Tracheal bronchus | Tracheal cyst | Congenital bronchial stenosis | Bronchial atresia | Bronchogenic cyst | CPAM | Congenital lobar emphysema |
| Type B-Isolated arterial PM | Interrupted pulmonary artery | Isolated systemic arterial supply to normal lung | Dual arterial supply to normal lung |
| Type C-Isolated venous PM | PAPVC | TAPVC | Isolated scimitar vein | Meandering pulmonary vein | Scimitar variant | Anomalous unilateral single pulmonary vein |
| Type D-Mixed bronchoarterial PM | Typical intralobar sequestration |

| Type E-Mixed bronchovenous PM | Combination of bronchial anomalies (Type A) and anomalous pulmonary venous connections (Type C) |
| Type F-Mixed arteriovenous PM | Fistula between the following: a) Systemic/pulmonary artery and pulmonary vein b) Scimitar vein and systemic arterial supply |
| Type G-Mixed bronchocartionovenous PM | a) Classical scimitar syndrome b) Extralobar sequestration c) Variations of bronchial, arterial, and venous anomalies |

CAPM: Congenital pulmonary airway malformation, PAPVC: Partial anomalous pulmonary venous drainage, TAPVC: Total anomalous pulmonary venous drainage

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arterial and venous systems. Proximal lesions include tracheal stenosis, tracheal cyst, tracheal diverticulum, and tracheal bronchus. Distal anomalies include bronchial branching anomalies, bronchogenic cyst, bronchial atresia, and CPAM.

**Bronchial anomalies**
Presence of anomalous origin of one of the normal bronchi away from its expected origin is referred to as “displaced bronchus” and presence of extra anomalous bronchus is referred to as “supernumerary bronchus.” Most common anomalies include the accessory cardiac bronchus and tracheal bronchus. Accessory cardiac bronchus is a supernumerary bronchus arising from the medial wall of the right main bronchus or bronchus intermedius and directed medially, ending blindly, or dividing into bronchioles, and may end in rudimentary lung tissue or a cyst. Tracheal bronchus [Figure 2] can be an accessory or displaced bronchus arising from the trachea or main bronchus and directed to the upper lobe.[11-13]

**Bronchial atresia**
In this condition, there is focal atresia or interruption of a lobar, segmental, or subsegmental bronchus. Distal bronchi are formed normally, but are dilated with mucoid impaction (mucocele) and there is associated air trapping in the part of the lung supplied by the atretic bronchus due to collateral air drift through the pores of Kohn and canals of Lambert. Apicoposterior segment of the left upper lobe is most commonly affected. On chest radiographs, tubular or branching opacity with surrounding hyperlucency may be seen. Computed tomography (CT) demonstrates the abnormality better, with mucocele seen as a hypodense nonenhancing tubular and branching structure, and also shows the air trapping, in the surrounding lung [Figure 3].[14]

**Bronchogenic cyst**
Bronchogenic cysts develop from abnormal budding from the ventral foregut, which then develops into a cyst. Cysts can contain water or proteinaceous and mucoid material. Cyst wall calcification and milk of calcium within the cyst can also be seen. Mediastinal location of bronchogenic cyst is more common than location within the lung parenchyma. On CT, cysts appear as well-defined lesions with smooth or lobulated margins with fluid or soft tissue attenuation, and are situated more commonly around the carina [Figure 4].[12] Cyst wall may or may not enhance.

**Congenital pulmonary airway malformation**
Congenital pulmonary airway malformations (CPAMs) represent developmental anomalies of the lower respiratory tract which result from abnormalities in the bronchial branching. These are hamartomatous lesions with cystic and adenomatoid components arising from the tracheobronchial tree. Five types are described – Type 0-4 [Table 3].[14,15] They usually communicate with the bronchi, although the communications may be abnormal. On CT, CPAM appears as a multicystic air-filled mass which may show air

Table 3: Types of congenital pulmonary airway malformation

| Type   | Description                                                                                     |
|--------|-----------------------------------------------------------------------------------------------|
| Type 0 | Rarest, arises from trachea or bronchi and involves whole lung; commonly fatal                  |
| Type 1 | Commonest (60-70%), arises from distal bronchi or proximal bronchioles; single or multiloculated 2-10 cm sized cyst. Reported association with malignancy |
| Type 2 | 15-20% of CPAMs; multiple cysts 0.5-2 cm in diameter with intervening solid-appearing areas     |
| Type 3 | 5-10% of CPAMs. Alveolar origin; can have small cystic areas (>0.5 cm) with solid tissue or are mostly solid appearing |
| Type 4 | 10-15% of CPAMs. Acinar origin. Large air-filled or fluid-filled cysts up to 10 cm; strongly associated with pneumothorax, indistinguishable from cystic pleuropulmonary blastoma |

**Figure 1(A-G):** Graphical representation of various types of bronchopulmonary vascular malnasculation. Type A, B, and C represent isolated bronchial, arterial, and venous malnasculations, respectively. Various combinations of Type A, B, and C result in bronchoarterial (D), bronchovenous (E), arteriovenous (F), and bronchoarteriovenous (G) malnasculations. (Image adapted from Lee et al.[16] and used after modification with authors permission)

**Figure 2 (A and B):** Isolated bronchial malnasculation – Tracheal bronchus. Axial (A) and coronal (B) chest sections in lung window show the origin of tracheal bronchus (white arrow) supplying the apical segment of the right upper lobe above the level of carina
fluid levels. Mass effect can cause contralateral mediastinal shift and atelectasis of the surrounding lung [Figure 5].

**Congenital lobar emphysema**
Congenital lobar emphysema is an anomaly of the lower respiratory tract, where there is hyperinflation of one or more lobes of the lungs. Intrinsic or extrinsic bronchial obstruction can be identified in around 25% of cases. Extrinsic obstruction can be caused by vascular anomalies, mediastinal masses, or foregut cysts which can compress the bronchus. Developmental anomaly in the bronchial wall cartilage leading to collapse of the airway during expiration or intraluminal lesions like meconium plugs or mucosal folds could also cause bronchial obstruction. Chest radiograph can demonstrate hyperinflation of the affected lobe with compression of the adjacent lobes, mediastinal shift to the opposite side, and inversion of the diaphragm. Left upper lobe is most commonly involved, followed by right middle lobe and right upper lobe. CT helps in confirming the chest radiograph findings of hyperinflated lung with reduced number and caliber of vascular structures and mass effect on the adjacent lung and mediastinal structures [Figure 6].

**Type B – Isolated arterial pulmonary malformation**
This group includes isolated arterial abnormalities with normal airways and normal venous system. Abnormal development of the pulmonary artery (interrupted pulmonary artery) or anomalous systemic arterial supply to normal lung are included in this group. Anomalous systemic arterial supply to the lung can be either an isolated systemic supply to the normal lung or dual arterial supply (combination of pulmonary and systemic arterial supply).

**Interrupted pulmonary artery**
In this condition, a portion of the proximal part of pulmonary artery is absent with intact distal vascular network within the lungs. The intrapulmonary arteries receive oxygenated blood from systemic artery collaterals. Usually, the interruption occurs on the side opposite to the aortic arch. The chest radiograph findings described include reduced volume of the affected hemithorax, attenuated hilum, reduced vascularity, pleural thickening.
and peripheral reticular opacities due to transpleural collaterals, and rib notching due to intercostal collaterals. Contrast-enhanced CT scans depict either complete absence of the mediastinal portion of the pulmonary artery or presence of a short stump. Systemic collaterals including subpleural collaterals are also well seen on CT as hypertrophied intercostal arteries and subpleural reticular lines parallel to the pleura [Figure 7].

Isolated systemic supply to normal lung
In isolated systemic supply to normal lung (ISSNL), there is aberrant supply to a lobar segment of lung (right/ left lower lobe) by a systemic artery in the absence of normal pulmonary arterial supply. This entity is also known as pseudosequestration as the lung shows normal communication with tracheobronchial tree. It is more commonly described on the left side where the left lower lobe is supplied by an artery arising from descending thoracic aorta [Figure 8]. When the right lower lobe is involved, it is usually supplied by an artery arising from celiac trunk or abdominal aorta. Patients with ISSNL can be asymptomatic or present with hemoptysis and exertional dyspnea due to left side volume overload. Surgery or coiling of aberrant systemic artery may be required in patients with hemoptysis.

Dual arterial supply to normal lung
This is a rare congenital entity in which a lobar segment of lung, typically lower lobes, receives dual arterial supply from both systemic artery (arising from aorta or branches) and pulmonary artery [Figure 9]. Patients can be asymptomatic or present with hemoptysis.

Type C – Isolated venous pulmonary malmosculation
Type C includes isolated malmosculation of the pulmonary vein with normal bronchial and arterial system.

Partial anomalous pulmonary venous drainage (PAPVC), total anomalous pulmonary venous drainage (TAPVC), meandering pulmonary vein, AUSPV, and scimitar variant can be included in this group.

Partial anomalous pulmonary venous drainage
In partial anomalous pulmonary venous connection, one of the pulmonary veins (superior or inferior) does not drain into the left atrium. It most commonly occurs on the right with the right superior pulmonary vein draining into the right atrium or the superior vena cava (SVC). The left superior pulmonary vein drains into the brachiocephalic vein or coronary sinus [Figure 10]. An isolated scimitar vein draining the whole of one lung into the IVC without associated pulmonary hypoplasia and systemic arterial supply can be included in this group.

Total anomalous pulmonary venous drainage
In total anomalous pulmonary venous connection, the four pulmonary veins do not drain into the left atrium. Depending on which structure they drain into, they are classified into four types:

Type 1: Supracardiac is the most common type. The pulmonary veins form a single vertical vein which most commonly drains into the left brachiocephalic vein [Figure 11]

Type 2: Cardiac in which the pulmonary veins drain into the coronary sinus or the right atrium

Type 3: Infracardiac type drains into an infra-diaphragmatic vessel, either in the systemic or the portal venous circulation. This type is prone for obstruction, usually at the level of the diaphragm
Type 4: Mixed type in which the pulmonary veins drain into different locations.

Meandering pulmonary vein (pseudo-scimitar)
In this entity, an anomalously coursing pulmonary vein (typically right or left inferior pulmonary vein) draining a part of one lung follows a circuitous route through the lung parenchyma before finally opening into the left atrium. On chest radiograph, this anomalously coursing vein can be mistaken for scimitar vein which opens into IVC. CT can demonstrate the anomalous coursing vein opening into the left atrium.

Anomalous unilateral single pulmonary vein
In this condition, a single pulmonary vein drains an entire lung and terminates into left atrium.

Scimitar variant
In this condition, an anomalous pulmonary vein terminates into both IVC and left atrium.

Type D – Mixed bronchoarterial pulmonary malformation
Type D or mixed bronchoarterial abnormalities include abnormality of the pulmonary artery and the airways. Conventional intralobar sequestration where abnormal lung is supplied by anomalous systemic artery with normal pulmonary venous drainage is a typical example.

Intralobar sequestration
Pulmonary sequestration is defined as a segment of nonfunctioning lung that does not communicate with the tracheobronchial tree and is supplied by an anomalous systemic artery. Intralobar sequestration does not have a separate pleural covering, is separated from normal bronchial tree, and has a systemic arterial supply and drains into the pulmonary vein.

Figure 9 (A-C): Isolated arterial malformation – Dual arterial supply to normal lung. Axial chest CT sections mediastinal (A) and lung window (B) show prominent artery (white arrow) arising from aorta supplying the left lower lobe. Note the presence of normal pulmonary artery branches (black arrow) accompanying the left lower lobe bronchi indicating dual arterial supply. Coronal chest section (C) depicts systemic arterial supply to left lower lobe.

Figure 10 (A-E): Isolated venous malformation – Partial anomalous pulmonary venous drainage (PAPVC). Axial CT sections in mediastinal window (A-E) show anomalous drainage of the left superior pulmonary vein (thin white arrow) into the left brachiocephalic vein (thick white arrow).

Figure 11 (A-D): Isolated venous malformation – Total anomalous pulmonary venous drainage (TAPVC), supracardiac variety. Coronal CT sections (A-D) in lung window show anomalous drainage of common pulmonary venous trunk (black arrows) into the left brachiocephalic vein (white arrow at the junction).

Figure 12 (A and B): Combined bronchoarterial malformation – Intralobar sequestration. Axial CT chest sections in mediastinal (A) and lung window (B) show a prominent systemic artery (white arrow) arising from the thoracic aorta supplying a cystic lesion (black arrow) in the posterior basal segment of the right lower lobe. This segment of lung did not show bronchial communication and had normal pulmonary venous drainage (not shown).
Congenital bronchopulmonary vascular malformations

Type A – Pulmonary airway malformation
Type A is defined as a combination of malformations of the pulmonary airways with or without air fluid levels seen predominantly in the lower lobes with loss of bronchial communication and showing systemic arterial supply [Figure 12].

Type B – Partial anomalous pulmonary venous drainage
Type B is defined as a combination of malformations of pulmonary venous drainage of the right superior pulmonary vein into the superior vena cava (white arrow) associated with a multicystic lesion (CPAM) in the right upper lobe (black arrow).

Type C – Partial anomalous pulmonary venous connection
Type C is defined as a combination of malformations with an abnormal pulmonary venous connection (Type C) is included in this type. Example includes combination of CPAM with PAPVC [Figure 13] and scimitar vein in combination with hypoplasia of lung and no systemic arterial supply [Figure 14].

Type D – Complete anomalous pulmonary venous connection
Type D is a complete anomalous pulmonary venous drainage which is defined as a combination of malformations of pulmonary airways and pulmonary venous drainage of the right superior pulmonary vein into the superior vena cava (white arrow) associated with a multicystic lesion (CPAM) in the right upper lobe (black arrow).

Type E – Mixed bronchovenous pulmonary malformation
Type E is mixed bronchovenous pulmonary malformation which is defined as a combination of malformations of pulmonary airways and pulmonary vein, but with a normal pulmonary arterial supply. Any of the pulmonary airway malformations (Type A) associated with an abnormal pulmonary venous connection (Type C) is included in this type. Example includes combination of CPAM with PAPVC [Figure 13] and scimitar vein in combination with hypoplasia of lung and no systemic arterial supply [Figure 14].

Type F – Mixed arteriovenous pulmonary malformation
Type F is a mixed arteriovenous pulmonary malformation which is defined as a combination of malformations of pulmonary arteries and vein, but with a normal pulmonary airway system. Arteriovenous malformation (AVM) can be included in this group.

Arteriovenous malformation
Pulmonary AVMs or fistulas show abnormal communication between pulmonary arteries and veins. They can be single (most common) or multiple and bilateral. They can be associated with Osler Weber Rendu syndrome or hereditary hemorrhagic telangiectasia, especially when multiple. They are more often located subpleurally or in lower lobes. They can be classified as simple AVM when only single feeding artery is seen or complex AVM when multiple feeding arteries are seen. On chest radiographs, rounded or ovoid opacities can be seen subpleurally with feeding artery and draining vein seen as “rabbit ears.” Contrast-enhanced CT shows a rounded enhancing nodule which is a tangle of tortuous vessels supplied by a prominent feeding pulmonary artery branch and draining pulmonary vein [Figure 15]. In complex AVM, more than one feeding artery may be seen.

Type G – Mixed bronchoarteriovenous malformation
Type G is a mixed bronchoarteriovenous pulmonary malformation which is defined as a combination of malformations of pulmonary airways, arteries, and vein. This group can include the classical scimitar syndrome, extralobar sequestration, pulmonary hypoplasia/aplasia/agenesis, or any combination of Type A, B, and C, for example, meandering pulmonary vein with right lung hypoplasia and systemic arterial supply to lung [Figure 16].
Scimitar syndrome
Scimitar syndrome consists of anomalous pulmonary venous drainage of the right lung to the IVC, systemic arterial supply of the right lower lobe from aorta, hypoplasia of the right lung with resultant cardiac dextroposition, and right pulmonary artery hypoplasia. CT is helpful in demonstration of the complete spectrum of abnormalities and differentiating it from incomplete forms and other variants.\[5\]

Extralobar sequestration
Extralobar sequestration, in comparison to intralobar sequestration, is a combined bronchoarteriovenous malinosculation as it shows isolation from tracheobronchial tree, has systemic arterial supply, and drains into systemic veins. On CT, a well-defined mass or multicystic lesion is seen commonly at the left lung base having its own pleural covering with loss of bronchial communication and showing systemic arterial supply from aorta and systemic venous drainage into azygous or hemiazygous veins.\[12\]

Pulmonary hypoplasia/aplasia/agenesis
Pulmonary hypoplasia, aplasia, and agenesis are associated with varying degrees of non-development of all the three systems. In pulmonary agenesis, there is complete absence of lung, bronchial, and vascular supply on one side. Pulmonary aplasia is similar to agenesis except for the presence of small rudimentary bronchus which ends as a blind pouch. In pulmonary hypoplasia, there is reduced lung volume with decrease in the number of bronchial divisions, number of alveoli, and reduced size of pulmonary vessels.\[11-13\]

Conclusions
Congenital bronchopulmonary vascular malformations include a broad spectrum of disorders that involve abnormal communication or anastomoses between one or more of the three main systems of the lung, namely, airways, arteries, and veins. Each disease entity can show a spectrum of variations on imaging in its pulmonary/systemic arterial and venous supply. Classification of BPVM according to the components involved helps provide a systematic approach for evaluation of each disease entity. Contrast-enhanced CT is very helpful in elaborating the anomalies of different components. Instead of trying to label the anomalies as different syndromes, it is more important to define in detail the different components of the complex anomalies that can be encountered.

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