

# Congenital bronchopulmonary vascular malformations, “sequestration” and beyond

Aparna Irodi, Shailesh M Prabhu, Reetu Amrita John, RV Leena

Department of Radiology, Christian Medical College, Vellore, Tamil Nadu, India

**Correspondence:** Dr. Aparna Irodi, Department of Radiology, Christian Medical College, Vellore - 632 004, Tamil Nadu, India.  
E-mail: aparnashyam@gmail.com

## 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 malinosculature. The aim of this pictorial essay is to illustrate the imaging appearances of the various types of pulmonary malinosculature.

**Key words:** Congenital bronchopulmonary vascular malformations; malinosculature; 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).<sup>[1,2]</sup> However, he found variations and termed

the variants as Pryce types I, II, and III [Table 1].<sup>[1]</sup> 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.<sup>[3]</sup> Following this, in 1987, Clements and Warner coined a new term, “malinosculature” 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.<sup>[4]</sup> Malinosculature 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 Warner.<sup>[4]</sup> Lee *et al.*, further refined the concept of malinosculature and classified it as bronchopulmonary malinosculatures [Table 2].<sup>[5]</sup> 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

#### Access this article online

##### Quick Response Code:






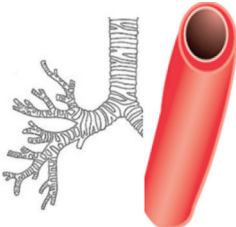

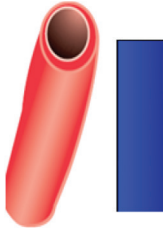
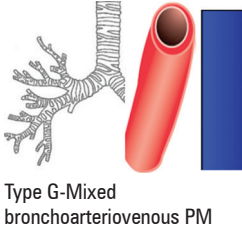
Website:  
www.ijri.org

DOI:  
10.4103/0971-3026.150138

**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

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

	Tracheal stenosis Tracheal bronchus Tracheal cyst Congenital bronchial stenosis Bronchial atresia Bronchogenic cyst CPAM Congenital lobar emphysema
	Interrupted pulmonary artery Isolated systemic arterial supply to normal lung Dual arterial supply to normal lung
	PAPVC TAPVC Isolated scimitar vein Meandering pulmonary vein Scimitar variant Anomalous unilateral single pulmonary vein
	Typical intralobar sequestration
	Combination of bronchial anomalies (Type A) and anomalous pulmonary venous connections (Type C)
	Fistula between the following: a) Systemic/pulmonary artery and pulmonary vein b) Scimitar vein and systemic arterial supply
	a) Classical scimitar syndrome b) Extralobar sequestration c) Variations of bronchial, arterial, and venous anomalies

**Table 2: Contd....**

Contd...

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

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.<sup>[8-10]</sup> 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 malinosculature 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 malinosculature.

**Type A – Isolated bronchial pulmonary malinosculature**

This group includes isolated airway abnormalities in the proximal and distal tracheobronchial tree with normal

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).<sup>[6,7]</sup> 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.

Contd...

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.<sup>[11-13]</sup>

#### 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].<sup>[14]</sup>

#### 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].<sup>[12]</sup> Cyst wall may or may not enhance.

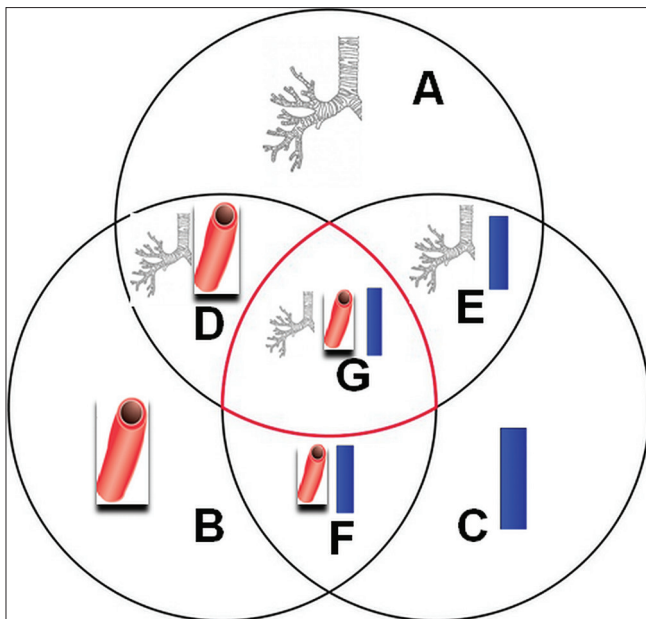
#### Congenital pulmonary airway malformation

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].<sup>[14,15]</sup> They usually communicate with the bronchi, although the communications may be abnormal.<sup>[15]</sup> On CT, CPAM appears as a multicystic air-filled mass which may show air

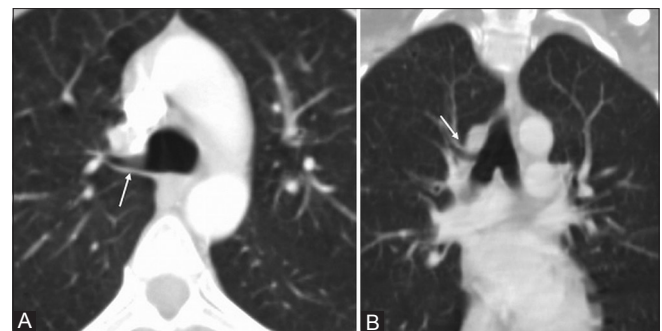
**Table 3: Types of congenital pulmonary airway malformation**

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

CAPM: Congenital pulmonary airway malformation



**Figure 1(A-G):** Graphical representation of various types of bronchopulmonary vascular malformations. Type A, B, and C represent isolated bronchial, arterial, and venous malformations, respectively. Various combinations of Type A, B, and C result in bronchoarterial (D), bronchovenous (E), arteriovenous (F), and bronchoarteriovenous (G) malformations. (Image adapted from Lee et al.<sup>[5]</sup> and used after modification with authors permission)



**Figure 2 (A and B):** Isolated bronchial malformation – 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

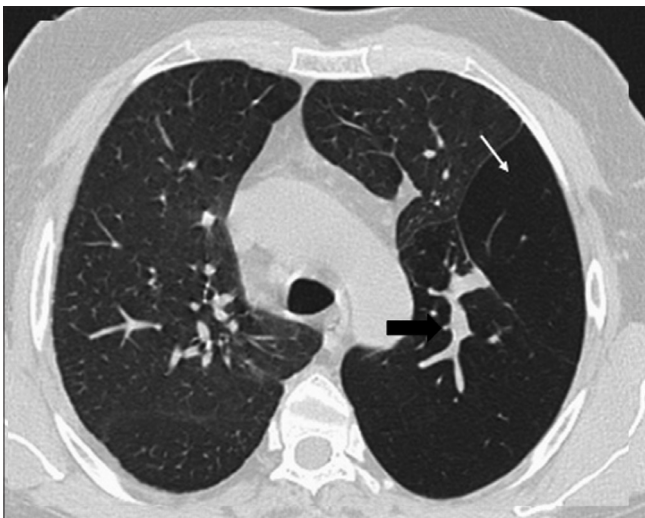
vascular structures and mass effect on the adjacent lung and mediastinal structures [Figure 6].<sup>[11-13]</sup>

#### **Type B – Isolated arterial pulmonary malinosculation**

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*

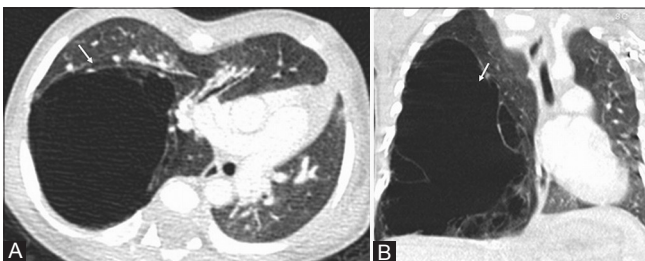
In this condition, a portion of the proximal part of pulmonary artery is absent with intact distal vascular network within the lungs. The intrapulmonary arteries receive oxygenated blood from systemic artery collaterals. Usually, the interruption occurs on the side opposite to the aortic arch. The chest radiograph findings described include reduced volume of the affected hemithorax, attenuated hilum, reduced vascularity, pleural thickening



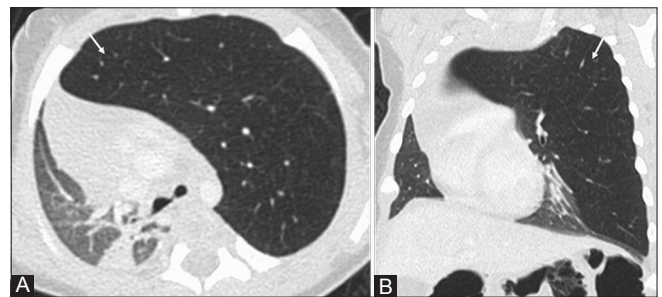
**Figure 3:** Isolated bronchial malinosculation – Bronchial atresia. Axial CT chest section in lung window shows a mucoid impacted bronchus (black arrow) supplying the apicoposterior segment of the left upper lobe with hyperinflation (white arrow) of the involved segment



**Figure 4:** Isolated bronchial malinosculation – Bronchogenic cyst. Axial CT chest section in mediastinal window shows a well-defined non-enhancing cystic lesion (white arrow) of fluid attenuation in the anterior mediastinum



**Figure 5 (A and B):** Isolated bronchial malinosculation – Congenital pulmonary airway malformation. Axial (A) and coronal (B) CT chest sections in lung window show a large multiseptated cystic lesion (arrows) with a single dominant cyst in the left lower lobe with associated mediastinal shift



**Figure 6 (A and B):** Isolated bronchial malinosculation – Congenital lobar emphysema. Axial (A) and coronal (B) CT chest sections in lung window show hyperinflated left upper lobe (arrows) with attenuated lung markings and herniation across the midline

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].<sup>[12]</sup>

#### Isolated systemic supply to normal lung

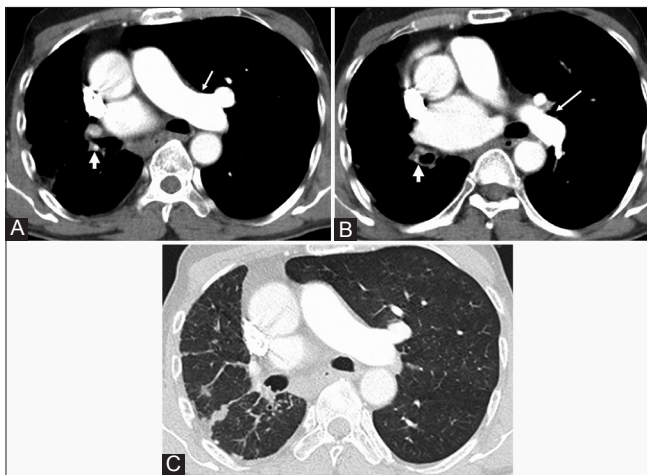
In isolated systemic supply to normal lung (ISSNL), there is aberrant supply to a lobar segment of lung (right/left lower lobe) by a systemic artery in the absence of normal pulmonary arterial supply. This entity is also known as pseudosequestration as the lung shows normal communication with tracheobronchial 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.<sup>[6,10]</sup>

#### 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.<sup>[10]</sup>

#### Type C – Isolated venous pulmonary malinosculation

Type C includes isolated malinosculation of the pulmonary vein with normal bronchial and arterial system.



**Figure 7 (A-C):** Isolated arterial malinosculation – Congenital interruption of pulmonary artery. Axial CT chest mediastinal (A, B) and lung window (C) sections show absent right pulmonary artery with normal left pulmonary artery (thin white arrow) and prominent bronchial arteries (thick white arrow) at the right hilum supplying the right lung. Inflammatory changes are noted in the right lung with volume loss

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.

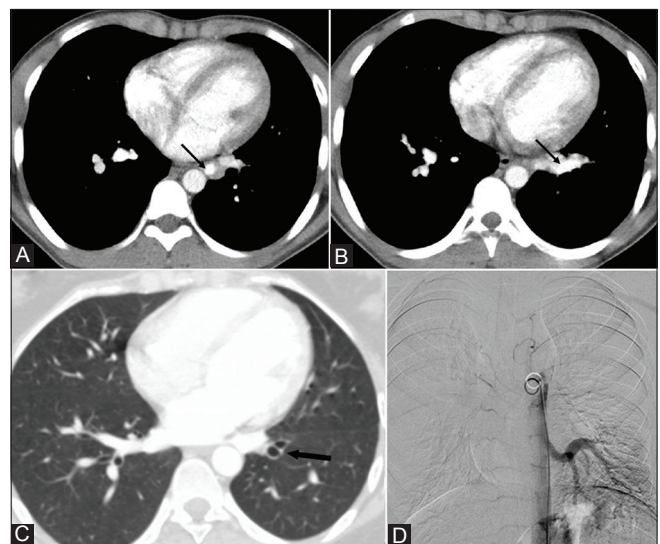
#### Partial anomalous pulmonary venous drainage

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].<sup>[16]</sup> 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.<sup>[16]</sup> 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



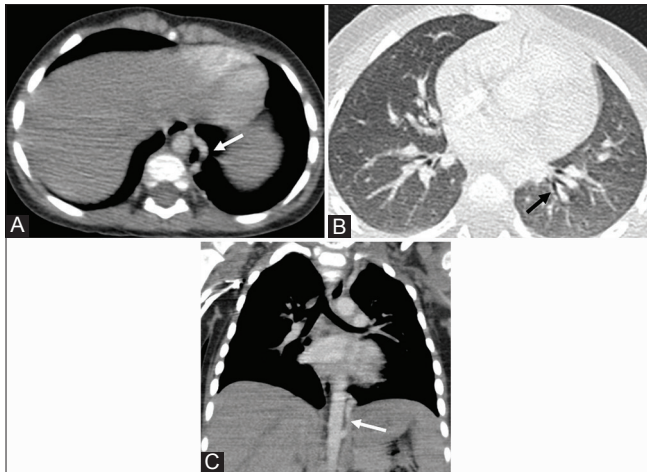
**Figure 8 (A-D):** Isolated arterial malinosculation – Isolated systemic arterial supply to normal lung. Axial chest CT sections mediastinal (A, B) and lung window (C) show prominent artery (black arrow) arising from the thoracic aorta supplying the left lower lobe. Note the absence of normal pulmonary blood vessels (black arrow) accompanying the left lower lobe bronchi. Angiogram (D) shows prominent systemic artery supplying the left lower lobe. Patient underwent coiling of the systemic artery for complaint of hemoptysis



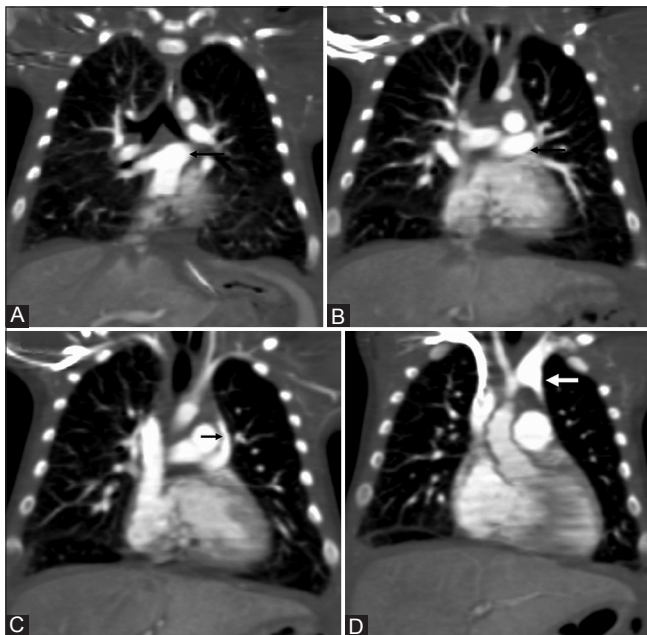
Type 4: Mixed type in which the pulmonary veins drain into different locations.

*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



**Figure 9 (A-C):** Isolated arterial malinosculature – 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 11 (A-D):** Isolated venous malinosculature – Total anomalous pulmonary venous drainage (TAPVC), supracardiac variety. Coronal CT sections (A-D) in lung window show anomalous drainage of common pulmonary vein trunk (black arrows) into the left brachiocephalic vein (white arrow at the junction)

coursing vein can be mistaken for scimitar vein which opens into IVC. CT can demonstrate the anomalous coursing vein opening into the left atrium.<sup>[9]</sup>

*Anomalous unilateral single pulmonary vein*

In this condition, a single pulmonary vein drains an entire lung and terminates into left atrium.<sup>[8]</sup>

*Scimitar variant*

In this condition, an anomalous pulmonary vein terminates into both IVC and left atrium.<sup>[9]</sup>

**Type D – Mixed bronchoarterial pulmonary malinosculature**

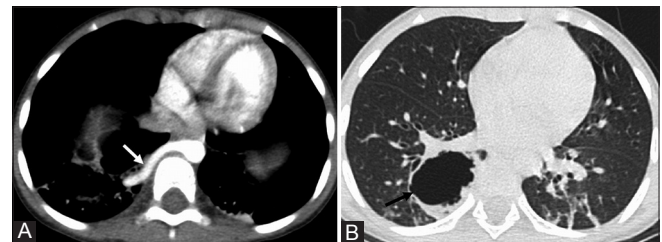
Type D or mixed bronchoarterial abnormalities include abnormality of the pulmonary artery and the airways. Conventional intralobar sequestration where abnormal lung is supplied by anomalous systemic artery with normal pulmonary venous drainage is a typical example.

*Intralobar sequestration*

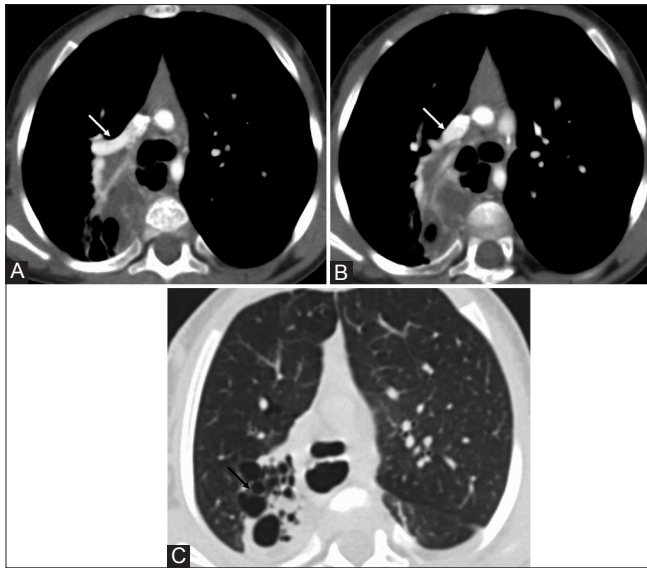
Pulmonary sequestration is defined as a segment of nonfunctioning lung that does not communicate with the tracheobronchial tree and is supplied by an anomalous systemic artery. Intralobar sequestration does not have a separate pleural covering, is separated from normal bronchial tree, and has a systemic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



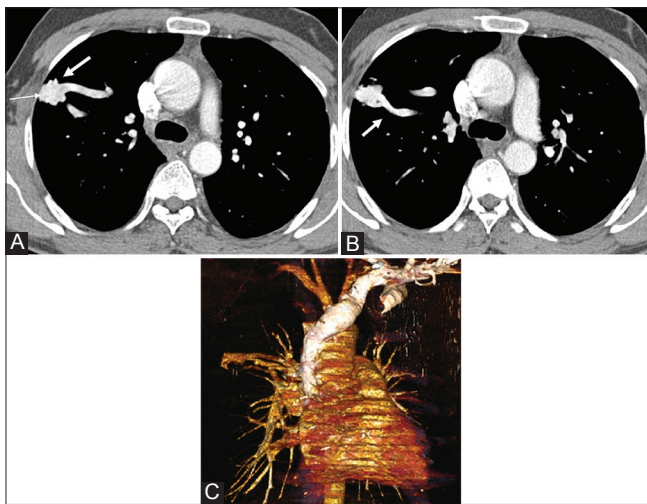
**Figure 10 (A-E):** Isolated venous malinosculature – Partial anomalous pulmonary venous drainage (PAPVC). Axial CT sections in mediastinal window (A-E) show anomalous drainage of the left superior pulmonary vein (thin white arrow) into the left brachiocephalic vein (thick white arrow)



**Figure 12 (A and B):** Combined bronchoarterial malinosculature – 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)



**Figure 13 (A-C):** Combined bronchovenous malinosculatation – Congenital pulmonary airway malformation (CPAM) with partial anomalous pulmonary venous drainage. Axial CT chest sections mediastinal (A, B) and lung window (C) show partial anomalous 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)

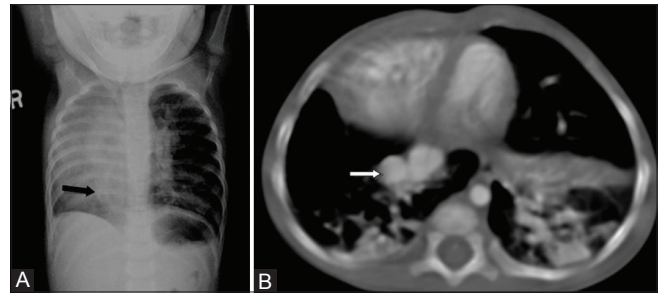


**Figure 15 (A-C):** Combined arteriovenous malinosculatation – Arteriovenous malformation (AVM). Axial CT chest mediastinal window (A, B) shows a nidus (thin white arrow) of prominent tortuous tangle of vessels in the right upper lobe supplied by a prominent pulmonary artery and drained by prominent pulmonary vein (thick white arrows). 3D volume-rendered image (C) confirms the vascular nature of the lesion with prominent supplying pulmonary artery and vein

On imaging, a homogenous well-defined mass or multicystic lesion with or without air fluid levels is seen predominantly in the lower lobes with loss of bronchial communication and showing systemic arterial supply [Figure 12].<sup>[12]</sup>

#### Type E – Mixed bronchovenous pulmonary malinosculatation

Type E is mixed bronchovenous pulmonary malinosculatation which is defined as a combination of malinosculatations of pulmonary airways and pulmonary vein, but with a normal



**Figure 14 (A and B):** Combined bronchovenous malformation – Pulmonary hypoplasia and scimitar vein. Chest radiograph (A) shows mediastinal shift to the right side with prominent scimitar vein (arrow) in the right lung base. CT axial section (B) shows scimitar vein draining the right lung into the IVC (arrow) with mediastinal shift to the right due to lung hypoplasia. No anomalous systemic artery was seen to diagnose scimitar syndrome

pulmonary arterial supply. Any of the pulmonary airway malinosculatations (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 malinosculatation

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

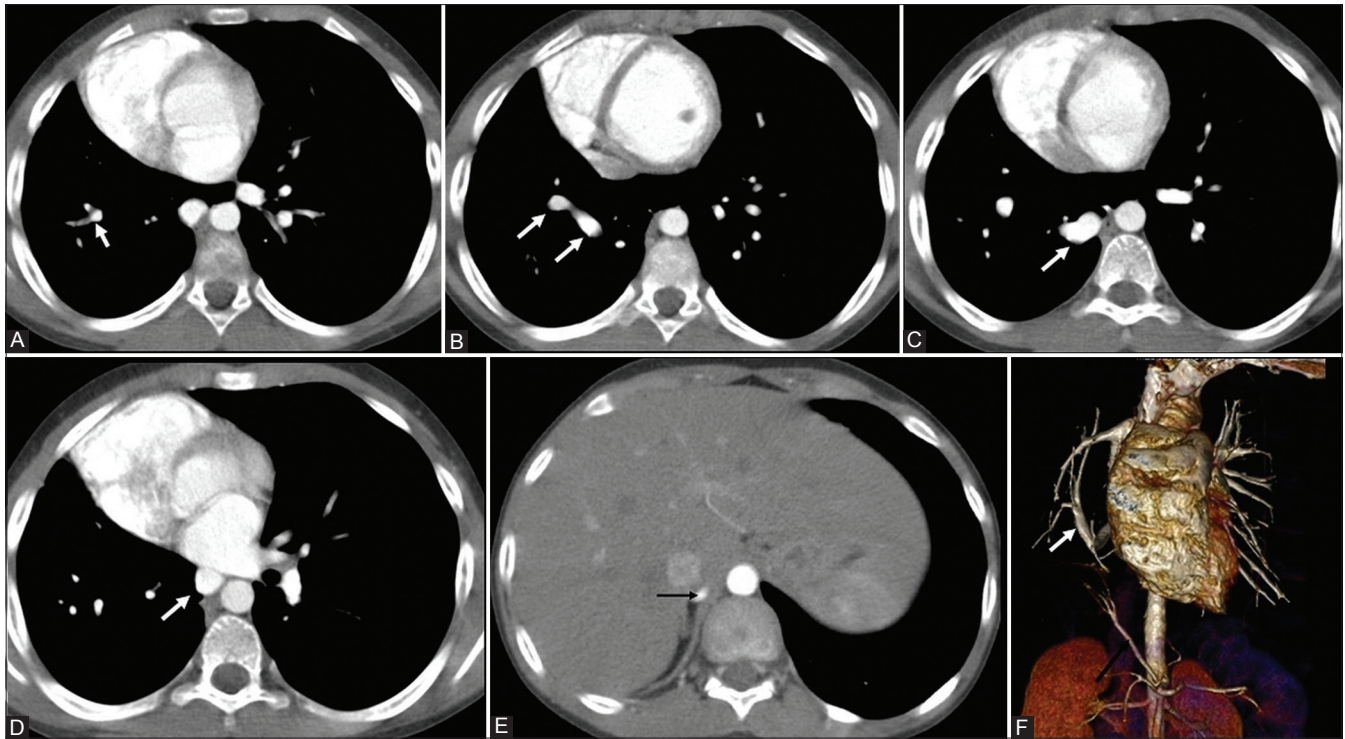
##### Arteriovenous malformation

Pulmonary AVMs or fistulas show abnormal communication between pulmonary arteries and veins.<sup>[12]</sup> 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 malinosculatation which is defined as a combination of malinosculatations 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].





**Figure 16 (A-F):** Combined bronchoarteriovenous malinosculation – Meandering pulmonary vein with right lung hypoplasia and systemic arterial supply to lung. Chest CT axial sections (A-E) in mediastinal window show anomalous course of the right inferior pulmonary vein (white arrow) taking a circuitous course through the right lung before draining into the left atrium (meandering pulmonary vein) and a prominent systemic artery (black arrow) arising from the aorta supplying the right lower lobe. 3D volume-rendered image (F) confirms systemic supply to the right lower lobe (black arrow) and meandering right inferior pulmonary vein (white arrow)

#### *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.<sup>[5]</sup>

#### *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.<sup>[12]</sup>

#### *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.<sup>[11-13]</sup>

## 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.

## References

1. Pryce DM. Lower accessory pulmonary artery with intralobar sequestration of lung; A report of seven cases. *J Pathol Bacteriol* 1946;58:457-67.
2. McDowell C, Robb D, Indyk JS. Two cases of intralobar sequestration of the lung. *Thorax* 1955;10:73-8.
3. Sade RM, Clouse M, Ellis FH Jr. The spectrum of pulmonary



- sequestration. *Ann Thorac Surg* 1974;18:644-58.
4. Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary-vascular malformations: Nomenclature and classification based on anatomical and embryological considerations. *Thorax* 1987;42:401-8.
  5. Lee ML, Lue HC, Chiu IS, Chiu HY, Tsao LY, Cheng CY, *et al.* A Systematic classification of the congenital bronchopulmonary vascular malformations: Dymorphogeneses of the primitive foregut system and the primitive aortic arch system. *Yonsei Med J* 2008;49:90-102.
  6. Singh AS, Subbain SK, Subramanian KG, Agarwal R, Dave DJ, Kulkarni S, *et al.* Pseudosequestration of the Left Lung. *Tex Heart Inst J* 2007;34:195-8.
  7. Agarwal PP, Matzinger FR, Seely JM, Rasuli P, Shamji FM. An unusual case of systemic arterial supply to the lung with bronchial atresia. *AJR Am J Roentgenol* 2005;185:150-3.
  8. Odenthal C, Sarikwal A. Anomalous unilateral single pulmonary vein versus scimitar syndrome: Comparison of two paediatric cases and a review of the literature. *J Med Imaging Radiat Oncol* 2012;56:247-54.
  9. Rodrigues MA, Ritchie G, Murchison JT. Incidental meandering right pulmonary vein, literature review and proposed nomenclature revision. *World J Radiol* 2013;5:215-9.
  10. Irodi A, Cherian R, Keshava SN, James P. Dual arterial supply to normal lung: Within the sequestration spectrum. *Br J Radiol* 2010;83:e86-9.
  11. Berrocal T, Madrid C, Novo S, Gutiérrez J, Arjonilla A, Gómez-León N. Congenital anomalies of the tracheobronchial tree, lung, and mediastinum: embryology, radiology, and pathology. *Radiographics* 2003;24:e17-62.
  12. Lee EY, Boiselle PM, Cleveland RH. Multidetector CT evaluation of congenital lung anomalies. *Radiology* 2008;247:632-48.
  13. Paterson A. Imaging evaluation of congenital lung abnormalities in infants and children. *Radiol Clin North Am* 2005;43:303-23.
  14. Lee EY, Dorkin H, Vargas SO. Congenital pulmonary malformations in pediatric patients: Review and update on etiology, classification, and imaging findings. *Radiol Clin North Am* 2011;49:921-48.
  15. Kao SW, Zuppan CW, Young LW. AIRP best cases in radiologic-pathologic correlation: Type 2 congenital cystic adenomatoid malformation (Type 2 congenital pulmonary airway malformation). *Radiographics* 2011;31:743-8.
  16. Ferguson EC, Krishnamurthy R, Oldham SA. Classic imaging signs of congenital cardiovascular abnormalities. *Radiographics* 2007;27:1323-34.

**Cite this article as:** Irodi A, Prabhu SM, John RA, Leena RV. Congenital bronchopulmonary vascular malformations, "sequestration" and beyond. *Indian J Radiol Imaging* 2015;25:35-43.

**Source of Support:** Nil, **Conflict of Interest:** None declared.