A Systematic Classification of the Congenital Bronchopulmonary Vascular Malformations: Dysmorphogeneses of the Primitive Foregut System and the Primitive Aortic Arch System

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INTRODUCTION

Congenital bronchopulmonary vascular malformations (BPVM) encompass a broad spectrum of diseases involving abnormal communication or anastomosis of one or more components of the lung.1-3 Despite early definition of the sequestration spectrum,4-7 and scimitar syndrome,8,9 reports of incomplete-, atypical-, and pseudo-sequestration/scimitar syndrome continued to prevail in the literature, which implies that a universal understanding of the sequestration spectrum and scimitar syndrome is still lacking. It is believed that malformations of the pulmonary airways, arteries, and veins may occur independently and coincidentally.10-14 Clements et al. proposed a "wheel theory" to explain the serial formation of congenital BPVM, and coined "pulmonary malinosculation" (PM) to classify these malformations. We propose a "haphazard theory" to substantiate this concept of PM.15 However, there is no category in which to classify patients with isolated scimitar vein anomalies and meandering right pulmonary veins.17 Nevertheless, isolated scimitar vein anomalies have been observed in our clinical
practice\textsuperscript{15} and in that of others.\textsuperscript{16} In this article, we will make a review and classification from an embryologic perspective of our 33 patients and 142 patients reported in the literature who had scimitar syndrome, bronchopulmonary sequestration, sequestration spectrum, congenital horseshoe lung, crossover lung segment, and other similar conditions.

**MATERIALS AND METHODS**

Between January 1990 and January 2007, 33 consecutive patients (17 men or boys and 16 women or girls), aged 1 day to 24 years (median, 2.5 months) with congenital BPVM were included in this retrospective analysis. Patients with total or partial anomalous pulmonary venous connections were excluded from this study. Study modalities included clinical features and plain chest radiographs (n = 33) plus at least two of the following laboratory profiles: two-dimensional echocardiography with Doppler (Acuson 128XP/10c, Mountain View, CA, USA) (n = 22), cardiac catheterization with cineangiography (n = 19), cardiothoracic surgery (n = 12), bronchography (n = 10), electron-beam CT of the chest (Imatron, South San Francisco, CA, USA) (n = 11), MRI (n = 11), bronchoscopy (n = 5), barium esophagography (n = 2), chromosomal study (n = 2), autopsy (n = 2), high resolution CT of the chest (n = 1), and MRA (n = 1).

**Definitions of malinosculation, sequestration, syndrome, and scimitar syndrome**

1. *Stedman’s Medical Dictionary* (2000) defines *malinosculation* (Latin: *mal-ill*, bad, abnormal; *in-in*; *osculum*-mouth) as "the establishment of abnormal communications by means of small openings or anastomoses".

2. *The Concise Oxford Dictionary* (2001) defines *sequestration* (from the Latin *sequestrare*, to separate) as something in a state of being secluded, isolated, or separated from other things.

3. *The ENCARTA World English Dictionary* (1999) defines *syndrome* (from the Latin and Greek *sundrome*, literally, running together) as a group of symptoms that consistently occur together.

4. Scimitar syndrome was originally defined as a syndrome having a triad of 1) an anomalous pulmonary venous connection to the inferior vena cava as scimitar vein, 2) a systemic arterial supply to the right lung, and 3) bronchial anomalies of the hypoplastic right lung.\textsuperscript{8,9}

The lung is mainly composed of six basic bronchovascular trees attributed to three different systems, including: 1) airways: bronchial trees, 2) artery: systemic and pulmonary arterial trees, and 3) vein: systemic and pulmonary venous trees, and lymphatic trees. Thus, these six basic bronchovascular trees can be classified into three different systems (airways, arteries, and veins). We regard anomalies of the venous drainage of the lungs (systemic, pulmonary, and lymphatic) along with anomalies of the pulmonary airways and pulmonary arterial supply simultaneously, rather than consider them as the aftermath of malinosculations of the pulmonary airways and the pulmonary artery.

**Nomenclature and definition of PM (Fig. 1)**

Type A isolated bronchial PM (area A in Fig. 1) is defined as isolated malinosculation of the pulmonary airways into the lung (with or without lung parenchymal abnormalities), but with a normal pulmonary arterial supply and a normal pulmonary venous connection. These malformations could be further subclassified into: 1) proximal lesions of tracheal airways, e.g., tracheal stenosis, tracheal bronchus, and tracheal cyst; 2) distal lesions of bronchial and smaller airways, e.g., a localized lesion of congenital bronchial stenosis, bronchial atresia, bronchogenic cyst, congenital cystic adenomatoid malformation, and congenital lobar emphysema. Type B isolated arterial PM (area B in Fig. 1) is defined as isolated malinosculation of the pulmonary artery, but with normal pulmonary arterial supply and a normal pulmonary venous connection. Patients with a systemic arterial supply (systemic arterialization) to the normal lung, without either broncho-pulmonary airways sequestration or anomalous pulmonary venous connections, have isolated arterial PM. Type C isolated venous PM (area C
in Fig. 1) is defined as isolated malinosculation of the pulmonary vein, but with normal pulmonary airways and a normal pulmonary arterial supply. Patients with isolated anomalies of the scimitar vein (to the inferior vena cava/right atrium) and meandering right pulmonary vein could be categorized into type C isolated venous PM. Type D mixed bronchoarterial PM (area D in Fig. 1) is defined as a combination of malinosculations of the pulmonary airways and the pulmonary artery but with a normal pulmonary venous connection. Type E mixed bronchovenous PM (area E in Fig. 1) is defined as a combination of malinosculations of the pulmonary airways and the pulmonary vein but with a normal pulmonary arterial supply. Tracheal stenosis, tracheal bronchus, tracheal cyst, bronchogenic cyst, bronchial stenosis, congenital cystic adenomatoid malformation, and congenital lobar emphysema could be found with anomalous pulmonary venous connections and a normal pulmonary arterial supply. Type F mixed arteriovenous PM (area F in Fig. 1) is defined as a combination of malinosculations of the pulmonary artery and the pulmonary vein, but with normal pulmonary airways. The cases of patients with a congenital fistula between a systemic or pulmonary artery and a pulmonary vein, and of patients with a scimitar vein (to the inferior vena cava) and a systemic arterial supply from the aorta (systemic arterialization) to a segment or a lobe of the lung, which is not secluded from the normal pulmonary airways (i.e., without bronchopulmonary airways sequestration), could be grouped into type F mixed arteriovenous PM. Type G mixed bronchoarteriovenous PM (area G in Fig. 1) is defined as malinosculations involving the pulmonary airways, the pulmonary artery, and the pulmonary vein. Classical scimitar syndrome with the above triads can be grouped into type G mixed bronchoarteriovenous PM. The aforementioned PM will be applied to analyze the cases of our 33 patients with congenital BPVM and those of another 142 patients from the literature with scimitar syndrome, bronchopulmonary sequestration, sequestration spectrum, congenital horseshoe lung, crossover lung segment, and other similar conditions.

RESULTS

The clinical profiles of our own 33 patients with congenital BPVM are summarized in Table 1. They were aged one day to 24 years (median, 2.5 months; range, 0.03-288 months). There were 24 infants (73%), eight children (>1 and ≤18 years old; 24%), and one adult (3%). Among these 33 patients, the salient clinical picture was recurrent pneumonia in 23 patients (70%), eight children (>1 and ≤18 years old; 24%), and one adult (3%). Among these 33 patients, the salient clinical picture was recurrent pneumonia in 23 patients (70%), respiratory distress in 21 (64%), wheezy respiration in 17 (52%), pulmonary hypertension in 14 (42%), cyanosis in 13 (39%), and congestive heart failure in 12 (36%). Situs solitus was noted in 28 patients, situs ambiguous with asplenia or heterotaxy syndrome in three, and situs inversus in two. Ellis-van Creveld
syndrome was noted in one patient, Down syndrome in one, and tibial agenesis with ectrodactyly syndrome in one. Congenital syphilis, E. coli sepsis, and Klebsiella oxytoca sepsis were documented in three individual patients. Lymphedema of the arm and shoulder was noted at the sixth day of life in a patient with congenital pulmonary lymphangiectasia. One patient had microcephaly, cleft palate, dermal sinus tract, and hyperlactic acidemia. Absence of a gallbladder was noted in one patient. Among the 33 patients with congenital BPVM, 17 patients had problems involving the bronchopulmonary system and the cardiovascular system (52%), 10 had problems of the bronchopulmonary system (30%), and six had problems of the cardiovascular system (18%). Ten patients had associated simple congenital heart disease (30%), nine of them had situs solitus (27%), and one had situs inversus (3%). In ten patients associated with simple congenital heart disease, ventricular septal defects were noted in six, patent ductus arteriosus in five, secundum atrial septal defects in five, aortic coarctation in three, sling left pulmonary artery (vascular ring) in three, and supravalvular pulmonary stenosis in one. Four patients had associated complex congenital heart disease (12%), three of them had situs ambiguous (9%), and one had situs inversus (3%). Dextroversion of the heart was found in 12 patients (36%), and dextrocardia in five (15%). Scimitar vein anomaly was found in four patients (nos. 17, 24, 32, and 33), and was silhouetted on the plain chest film as a right-sided sword in three patients (nos. 17, 24, and 32) and a left-sided sword in one (no. 33). The drainage site of the scimitar vein was the inferior vena cava in two patients (right middle and lower lobes in no. 24; right lower lobe in no. 32), the right atrium in one (left-sided scimitar in no. 33). Patient 17, who had a scimitar vein draining the whole right lung both to the left atrium and to the right atrium (a dual system of right pulmonary venous drainage), did not present with desaturation or discernible lip cyanosis. Bronchopulmonary (airways) sequestration was found in four patients (nos. 12, 20, 32, and 33), including systemic arterial supply to the left lower lobe in two patients (nos. 20 and 33), the right lower lobe in one (no. 32), and normal pulmonary arterial supply to the right lower lobe in one (no. 12). Of three patients with vascular ring (nos. 19, 21, and 30), an isolated left pulmonary artery sling was noted in one (no. 21), and a complex left pulmonary artery sling (associated with other rare aortic arch anomalies) was noted in the remaining two (nos. 19 and 30). Aortic arch anomalies included ductal sling, a right-sided patent ductus arteriosus, a left ascending aorta and aortic arch, and a right descending aorta in one (no. 19); an aberrant right subclavian artery from a left aortic arch, a retroesophageal aortic arch, and a descending aorta in one (no. 30).

Interventional cardiac catheterization was performed in four patients (nos. 16, 22, 23, and 30), including balloon dilatation for aortic coarctation in three (nos. 16, 22, and 23) and supravalvular pulmonary stenosis in one (no. 30), and transarterial coil embolization for the systemic pulmonary artery in one (no. 16). Surgical intervention was performed in 12 patients, including thoracic surgery in six patients, cardiovascular surgery in five, and thoracic-cardiovascular surgery in five. The overall mortality rate was 8 out of 33 (24%).

A systematic classification for the congenital BPVM was: type A isolated bronchial PM in 13 patients, type B isolated arterial PM in three, type C isolated venous PM in two, type D mixed bronchoarterial PM in five, type F mixed arteriovenous PM in one, and type G mixed bronchoarteriovenous PM in nine (Table 1).

We also reappraised the cases of 142 patients from the literature reported to have had scimitar syndrome, bronchopulmonary sequestration, sequestration spectrum, congenital horseshoe lung, crossover lung segment, congenital pulmonary lymphangiectasia, meandering right pulmonary vein, and isolated scimitar vein anomaly, and applied this systematic PM to assess and classify them (Table 2).

DISCUSSION

Congenital BPVM can manifest with various symptoms. One disease entity, for example, congenital cystic adenomatoid malformation, may have different pathology and protean features. On
Table 1. A Systematic Approach to Our 33 Patients with Congenital BPVM and Their Classification

| No./Age/Sex | Systems | Study modalities | Clinical diagnosis and annotations | Malformations | PM |
|-------------|---------|------------------|-----------------------------------|---------------|----|
| 1/2 d/F     | BP/CV   | Bg, Cr, E        | CTS; CTB; CHD; SI; dextrocardia    | Yes           | No | A  |
| 2/32 m/M    | BP      | Bg, CT, CR, E, Es| CTBS; CBB                        | Yes           | No | A  |
| 3/2 d/M     | BP      | Bg, CR           | CTS                              | Yes           | No | A  |
| 4/4 m/F     | BP      | Bg, Cr           | CTS                              | Yes           | No | A  |
| 5/4 m/F     | BP      | Bg, Cr           | CTS                              | Yes           | No | A  |
| 6/5 y/F     | BP      | CR, MRI, Su      | CCAM; dextroversion               | Yes           | No | A  |
| 7/1 m/M     | BP/CV   | An, Bg, Ch, CT, CR, E | CTBS; CHD                      | Yes           | No | A  |
| 8/10 m/M    | BP      | Bg, CR, MRI, Su  | CBC                              | Yes           | No | A  |
| 9/20 m/M    | BP      | CR, MRI, Su      | CBC                              | Yes           | No | A  |
| 10/12 d/F   | BP      | CT, CR, Su       | CLE, LUL; dextroversion           | Yes           | No | A  |
| 11/15 d/M   | BP      | CT, CR, Su       | CLE, RUL                         | Yes           | No | A  |
| 12/1.5 m/F  | BP/CV   | An, Bg, CR, E    | BPS with normal RPA, right; CHD   | Yes           | No | A  |
| 13/13 y/M   | BP      | An, CR, CT, E    | HRLS with normal RPA/RPV; dextroversion | Yes           | No | A  |
| 14/1 m/M    | CV      | Au, CR, E        | S-art, right; complex CHD; SI; dextrocardia | No           | Yes | No | B  |
| 15/1.5 m/F  | CV      | An, Bg, CR, E    | S-art, both; complex CHD; SA; RAI; dextrocardia; asplenia | No           | Yes | No | B  |
| 16/2.5 m/F  | CV      | An, CR, E, Su    | S-art, left; CHD                  | No            | Yes | C  |
| 17/3.5 y/F  | CV      | An, CT, CR, E    | isolated SV anomaly, right        | No            | Yes | C  |
| 18/6 d/M    | CV      | CT, CR, MRI      | CPL                              | No            | No | C  |
| 19/4 d/M    | BP/CV   | An, Bg, CT, CR, E| Es, Su                          | CBB; CTBS; SLPA; CHD | Yes | No | D  |
| 20/14 y/M   | BP/CV   | CR, MRI, Su      | BPS with S-art, left; dextroversion | Yes           | No | D  |
| 21/20 d/M   | BP/CV   | CT, CR, E, Su    | CTBS; SLPA; CHD                  | Yes           | No | D  |
| 22/15 d/F   | BP/CV   | An, Bg, CT, CR, E| RUL hypoplasia; CBS; CHD; dextroversion | Yes           | No | D  |
| 23/2.5 m/M  | BP/CV   | An, CT, CR, E    | CLS (a branch of RPA to LLL); CHD | Yes           | No | D  |
| 24/8 m/F    | CV      | An, CR, E, MRA, Su| S-art/SV, right; complex CHD; SA; RAI; dextrocardia; asplenia | No           | Yes | Yes | F |
| 25/4 d/M    | BP/CV   | An, CR, E        | agenesis of RL/RPA/RPVs; dextroversion | Yes           | Yes | G  |
| 26/3 m/M    | BP/CV   | An, CR, E, MRI   | agenesis of RL/RPA/RPVs; dextroversion | Yes           | Yes | G  |
| 27/11 m/F   | BP/CV   | An, Bg, CR, E, MRI| agenesis of RL/RPA/RPVs; CHD; dextroversion | Yes           | Yes | G  |
| 28/15 m/F   | BP/CV   | An, Bg, CR, E, MRI| agenesis of RL/RPA/RPVs; dextroversion | Yes           | Yes | G  |
| 29/8 y/M    | BP/CV   | An, CR, E, MRI   | agenesis of LL/LPA/LPVs           | Yes           | Yes | G  |
| 30/1 d/M    | BP/CV   | An, Bg, Ch, CR, E| agenesis of RL/RPA/RPVs; CTBS; SLPA; CHD; dextroversion | Yes           | Yes | G  |
| 31/24 y/F   | BP/CV   | An, CT, CR, E    | HRLSs right unilobulation; hyparterial RMB; RPA hypoplasia; single RPV; dextroversion | Yes           | Yes | G  |
| 32/2 m/F    | BP/CV   | An, CR, MRI, Su  | S-syn: BPS/S-art/SV/right; dextroversion | Yes           | Yes | G  |
| 33/4 d/F    | BP/CV   | An, Au, Bg, CR, E, MRI| S-syn: BPS/S-art/SV/left; complex CHD; SA; RAI; dextrocardia; asplenia | Yes           | Yes | G  |

An, angiography; Au, autopsy; Bg, bronchography; BP, bronchopulmonary; BPS, bronchopulmonary sequestration; BPVM, bronchopulmonary vascular malformations; Bs, bronchoscopy; CB, congenital bridging bronchus; CBC, congenital bronchogenic cyst; CBS, congenital bronchial stenosis; CCAM, congenital cystic adenomatoid malformation; CHD, congenital heart disease; Ch, chromosomal study; CLE, congenital lobar emphysema; CLS, crossover lung segment; CPL, congenital pulmonary lymphangectasia; CR, chest radiography; CT, computer tomography; CTR, congenital tracheal bronchus; CTBS, congenital tracheobronchial stenosis; CTS, congenital tracheal stenosis; CV, cardiovascular; E, echocardiography; Es, esophagography; HRLS, hypogenetic right lung syndrome; LL, left lung; LLL, left lower lobe; LPA, left pulmonary artery; LPVs, left pulmonary veins; LUL, left upper lobe; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging; RAI, right atrial isomerism; RL, right lung; RLL, right lower lobe; RMB, right main bronchus; RPA, right pulmonary artery; RPVs, right pulmonary veins; RUL, right upper lobe; SA, situs ambiguus; S-art, systemic arterial supply (arterialization) to the lung; SI, situs inversus; SLPA, sling left pulmonary artery; S-syn, scimitar syndrome; Su, surgery; SV, scimitar vein.
### Table 2. Retrospective appraisal of 142 Cases from the Literature with Scimitar Syndrome, Bronchopulmonary Sequestration, Sequestration Spectrum, Congenital Horseshoe Lung, and Other Similar Conditions

| Author | Cases* (n = 142) | Malosascular of the Airways Artery Vein | Diagnosis in the original report | Clinical annotations | PM type |
|--------|------------------|----------------------------------------|----------------------------------|---------------------|---------|
| Sade⁶   | 1 case           | No | Yes | Yes | ILS | Systemic arterial supply anomalous PVD | F |
| Blesovsky⁷ | 1 case          | Yes | No | No | ELS | Normal PVD | A |
| Halasz⁸ | Nos. 1-3         | Yes | Yes | Yes | Bronchial and arterial anomalies; Drainage of the RL into the IVC | Scimitar syndrome | G |
| Neill⁹  | No. 1            | Yes | No | Yes | Scimitar syndrome | No systemic artery | E |
| Neill⁹  | No. 2            | Yes | Yes | Yes | Scimitar syndrome | Scimitar syndrome | G |
| Juraszek¹⁰ | 1 case          | No | No | Yes | Isolated left-sided SV connecting all LPVs to the right IVC | Isolated SV anomaly, left-sided | C |
| Thilenius¹¹ | No. 1           | No | Yes | Yes | Spectrum of pulmonary sequestration | Normal bronchial airways | F |
| Thilenius¹¹ | No. 2           | Yes | Yes | Yes | Spectrum of pulmonary sequestration | Scimitar syndrome with CHL | G |
| Morgan¹² | No. 1            | Yes | Yes | No | Scimitar syndrome | Scimitar sign rather than SV | D |
| Kanemoto¹³ | No. 1           | Yes | Yes | No | Pseudo-scimitar syndrome | Scimitar sign rather than SV | A |
| Herer¹⁴  | No. 1            | Yes | No | Yes | Scimitar sign with normal PVD and Anomalous IVC | Scimitar sign rather than SV | A |
| Cukier¹⁵ | No. 1            | No | Yes | No | Scimitar syndrome or BPS | Systemic artery to the lung | B |
| Gikonyo¹⁶ | No. 1           | Yes | Yes | Yes | Scimitar syndrome | Scimitar syndrome | G |
| Gikonyo¹⁶ | Nos. 2-4        | Yes | No | Yes | Scimitar syndrome | No systemic artery | E |
| Partridge¹⁷ | Nos. 1-12      | Yes | Yes | Yes | Scimitar etcetera-DRL | Scimitar syndrome | G |
| Partridge¹⁷ | Nos. 13-15      | Yes | Yes | No | Scimitar etcetera-DRL | No SV | D |
| Partridge¹⁷ | Nos. 16-18      | Yes | No | No | Scimitar etcetera-DRL | No systemic artery; no SV | A |
| Pearl¹⁸  | No. 1            | No | No | Yes | Scimitar variant | Isolated SV anomaly only | C |
| Tumbarello¹⁹ | No. 1         | No | Yes | No | Scimitar variant | Isolated SV anomaly only | C |
| Oakley²⁰  | Nos. 1-9        | No | No | Yes | SV syndrome | Isolated SV anomaly only | C |
| Ross²¹  | No. 1            | No | No | Yes | Incomplete scimitar syndrome | Isolated SV anomaly only | C |
| Frank²²  | Nos. 1-6        | Yes | Yes | Yes | CHL | Scimitar syndrome with CHL | G |
| Dupuis²³  | Nos. 1-6        | Yes | Yes | Yes | CHL | Scimitar syndrome with CHL | G |
| Clements²⁴ | Nos. 1-3       | Yes | Yes | Yes | Crossover lung segment | Scimitar syndrome with CHL | G |
| Gao²⁵   | Nos. 1,5-7,11,13 | Yes | Yes | Yes | Scimitar syndrome | Scimitar syndrome with CHL (Nos. 5-7, 11), and BPS (Nos. 1, 13) | G |
| Gao²⁵   | Nos. 2,3,8-10,12 | No | Yes | Yes | Scimitar syndrome | Normal bronchial airways | F |
| Gao²⁵   | Nos. 4          | No | No | Yes | Scimitar syndrome | Isolated SV anomaly only | C |
| Geggel²⁶ | No. 1            | No | No | Yes | Scimitar syndrome | Isolated SV anomaly only | C |
| Dickinson²⁷ | Nos. 1,2,4     | Yes | Yes | Yes | Scimitar syndrome | Scimitar syndrome | G |
| Dickinson²⁷ | No. 3           | No | Yes | Yes | Scimitar syndrome | Normal bronchial airways | F |
| Farnsworth²⁸ | Nos. 1,2      | Yes | Yes | Yes | Scimitar syndrome | Scimitar syndrome | G |
| Schramel²⁹ | Nos. 1-4,7      | Yes | Yes | Yes | Scimitar syndrome | Scimitar syndrome | G |
| Schramel²⁹ | Nos. 5,6        | Yes | No | Yes | Scimitar syndrome | No systemic artery | E |
| Rutledge³⁰ | No. 1          | Yes | Yes | Yes | Scimitar syndrome | Scimitar syndrome with ‘left-sided’ SV | G |
| Le Rochais³¹ | No. 1         | Yes | Yes | Yes | Scimitar syndrome | Plus PAVM | G |
| Beitzke³² | No. 1            | Yes | Yes | Yes | Scimitar syndrome | Absence of RPA | G |
| Heron³³  | No. 1            | Yes | Yes | Yes | HRLS | Scimitar syndrome | G |
| Tummers³⁴ | No. 1          | Yes | Yes | Yes | Scimitar syndrome | Scimitar syndrome | G |

*PM* = Pulmonary Mainstem

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the contrary, many different entities have similar pathology and clinical findings, for example, congenital bronchogenic cyst, cystic adenomatoid malformation, and bronchopulmonary sequestration. To add to the confusion, each disease entity may have a normal (pulmonary) or abnormal (systemic) arterial supply, as well as a normal or abnormal pulmonary venous drainage. Therefore, a reappraisal and clarification of the "terminology" of various disease entities in congenital BPVM is mandatory. Among these malformations, the sequestration spectrum and the scimitar syndrome are the most intriguing and instructive examples. However, the dysmorphogeneses of the primitive foregut system and the primitive aortic arch system that lead to the haphazard PM, and their associations with the asplenia syndrome will be discussed first.

Dysmorphogeneses of the asplenia syndrome, the primitive foregut system, and the primitive aortic arch system

During embryogenesis, the normal spleen
develops by Horizon XIII, and thus asplenia syndrome originates then or earlier. Most cardiac defects in asplenia syndrome originate at Streeter Horizon XIII, which is defined as a 26- to 28-day embryo. Embryos with extracardiac anomalies, either vascular or non-vascular, exhibited a spectrum of developmental defects involving the pulmonary, gastrointestinal, genitourinary, musculoskeletal, facial, endocrinologic, and central nervous systems. Among the pulmonary vascular developmental defects, systemic arterialization to the lung with sequestration has never been reported in asplenia syndrome, nor has right atrial isomerism been reported with pulmonary atresia in the English literature. Interestingly, we found that the congenital cardiovascular defects and bronchopulmonary anomalies in patient 33 can be temporally correlated after reviewing the developmental landmarks in cardiac morphogenesis, the development of the pulmonary artery and the bronchial arteries, and the growth of the pulmonary vascular bed in the healthy and diseased lung. In the embryo, the pulmonary artery receives blood from the primitive ventral and dorsal aorta and sends its branches into the foregut-mesodermal post-branchial pulmonary plexus. The foregut-mesodermal post-branchial pulmonary plexus, which communicates with primitive bronchial arteries arising from the aorta, normally regresses. However, these embryonic communications may persist as anomalous pulmonary arteries of systemic origin, either within the thorax or drawn down below the diaphragm by the caudal movement of vessels, such as the celiac axis. The persistence of these two early embryonic or primitive communications (at about the fourth gestational week) may lend credence to the proposed formation of the systemic pulmonary collaterals, in conjunction with or without pulmonary atresia in asplenia syndrome, at Streeter Horizon XIII, which is defined as a 26- to 28-day embryo. Such systemic pulmonary collateral arteries may have arisen directly from the thoracic aorta in patient 15 without sequestration, and the primitive bronchial artery may have taken off directly from the abdominal aorta in patient 33 with sequestration. What orchestrates this disharmonious evolution in the development of the pulmonary vascular supply and the bronchopulmonary airways and parenchyma in patients 15 and 33 with asplenia syndrome may be related to a focal developmental disturbance in laterality, which occurs primarily at Horizon XIII.

Human spleen, the pulmonary artery anlage, the primitive systemic collateral arteries, and the primitive foregut develop at about four to five weeks of life. Abnormal division of the primitive foregut by the tracheo-esophageal septum, which leaves the respiratory primordium on the enteric primordium, may be ascribed to a sequence of changes that produces various types of bronchopulmonary foregut malformations or sequestration spectrum. Normally, the respiratory mesenchyme invests the budding epithelium by producing epithelium growth factor to form the respiratory primordium. The systemic pulmonary vessels, which are supplied antegradely by the sixth aortic arch and foregut-mesodermal post-branchial pulmonary plexus and retrogradely by primitive bronchial arteries from the primitive aorta, develop after the respiratory mesenchyme and bronchial epithelium concomitantly form the respiratory primordium. Neither normal nor abnormal respiratory primordia, which are either isolated or connected, can secure a blood supply entirely from the pulmonary artery or entirely from the aorta. That is why normal lungs can be supplied by the systemic artery, and sequestration can be performed without such a vessel. Many theories concerning the etiology of pulmonary sequestration have been proposed. These are informative to the concept of the haphazard branching theory that we proposed. On the basis of the unusual combinations of extralobar pulmonary sequestration, left scimitar syndrome, asplenia syndrome, and complex congenital heart disease in patient 33, we suggest that these anomalies may share a common defect in normal embryonic organizer control, which occurs mainly during the fourth to fifth week of life. Insufficient pulmonary arterial supply may be related to the presence of systemic pulmonary collaterals. We also agree that there is no causal relationship between the nonfunctioning lung and systemic artery. Their concurrence is incidental rather than causal.

In summary, dysmorphogeneses of the respira-
tory primordium (respiratory mesenchyme and budding epithelium) and the pulmonary vessels (pulmonary artery and systemic pulmonary collaterals), which are temporally related, may happen coincidentally. The concurrence of the anomalous connection (malinosculation) of either one or more of the airways, parenchyma (respiratory primordium), arteries, and veins (pulmonary vessels) occurs by chance, so that a variety of congenital BPVMs can be accordingly categorized into bronchial, arterial, and bronchoarterial malinosculations, with or without parenchymal abnormalities. The abnormal connection (malinosculation) between the respiratory primordium and the enteric primordium is correlated during embryogenesis. Thus, dysmorphogeneses of the primitive foregut system and the primitive aortic arch system can develop into many varieties of congenital BPVM.

Reappraisal and clarification of the terminology

Extralobar sequestration, rather than intralobar sequestration, is truly "secluded" from normal lungs. Thus, bronchopulmonary sequestration is clear to the letter to inform us of PM involving the pulmonary airway, without mentioning "extralobar" in character. However, we should remember that this refers to malinosculation of the pulmonary airways only, and does not imply the involvement of a systemic vascular supply, and vice versa. The notion of taking a systemic arterial supply (or systemic arterialization) for granted in bronchopulmonary sequestration should be abandoned, for bronchopulmonary sequestration may have a normal pulmonary arterial supply.

Because bronchopulmonary sequestration can derive its arterial supply from a systemic artery or from a pulmonary artery, and a systemic artery can supply a normal or a sequestered lung (bronchopulmonary sequestration) by means of coincident malinosculation of the pulmonary airways and pulmonary artery. Vascular sequestration is a nebulous term to imply arbitrarily a systemic vascular supply to bronchopulmonary sequestration and vice versa. To the letter, vascular sequestration cannot be specific because it does not refer to a vascular supply that is secluded or isolated from the normal pulmonary arterial territory. In this case, pulmonary arterial sequestration will be a more appropriate term than vascular sequestration to indicate a vascular supply that is secluded from the territory of the normal pulmonary arterial. Neither one can denote bronchopulmonary sequestration. We still recommend "systemic arterialization" to denote a systemic arterial supply to the lung without bronchopulmonary sequestration and to express a separate event of PM of the pulmonary artery in the lung, rather than "pulmonary arterial sequestration". Neither systemic arterialization, nor vascular sequestration, nor pulmonary arterial sequestration occurred in all cases of bronchopulmonary sequestration. Neither systemic arterialization, nor vascular sequestration, nor pulmonary arterial sequestration occurred in all cases of bronchopulmonary sequestration.

Since the early introduction of the concept of sequestration in 1946, variants of sequestration have inundated the literature and finally led to the concept of the sequestration spectrum in 1974. We retrospectively analyzed this sequestration spectrum, which may include: 1) an abnormal artery to a normally connected lung, 2) an abnormal artery to both a sequestered mass and adjacent normal lung, 3) an abnormal artery confined to the sequestered mass, and 4) sequestered areas of lung with normal vascularure. The first variant is normal lung with a systemic arterial supply, or systemic arterialization without bronchopulmonary (airways) sequestration. The third variant is bronchopulmonary sequestration with a systemic arterial supply and is most commonly encountered. The second variant is a combination of the first variant and the third variant. The fourth variant is bronchopulmonary sequestration with parenchymal cystic changes and a normal pulmonary arterial supply (so-called intralobar bronchopulmonary sequestration). There were reports of intralobar sequestration (with parenchymal cystic changes of bronchogenic cysts, congenital lung cysts, congenital lobar emphysema, and cystic adenomatoid malformation) with a normal pulmonary arterial supply. By definition, they are not genuine bronchopulmonary (airways) sequestrations. Nonetheless, these malformations may be considered as type A isolated bronchial PM with a normal pulmonary arterial supply and normal pulmonary venous connection. The first and fourth variants should
not be dumped into the sequestration spectrum. A liberal application of the sequestration spectrum may undermine our understanding of the pathology of congenital BPVM, let alone its classification.

Scimitar signs, scimitar veins, and scimitar syndrome must be well defined before applying the aforementioned nomenclature and classification for congenital BPVM. George Cooper and Raoul Chassinat individually reported cases of rare congenital malformations involving anomalous pulmonary venous drainage below the diaphragm. The terms "scimitar" and "scimitar syndrome" appeared in the literature in 1956 and 1960, respectively. We can identify three major constant features, or triads, from these two original reports, namely, anomalous pulmonary venous connection to the inferior vena cava (scimitar vein), a systemic arterial supply to the right lung, and bronchial anomalies of the hypoplastic right lung. Two additional features that occur more sporadically, dextroversion of the heart and bronchopulmonary sequestration, also merit discussion. First of all, dextroversion of the heart is secondary to hypoplasia of the right lung. Secondly, bronchopulmonary sequestration is not the sole airway anomaly in the scimitar syndrome. Congenital horseshoe lung is another pathology to be seen in patients with the scimitar syndrome. Thus, dextroversion of the heart and bronchopulmonary sequestration are not sine qua non diagnostic criteria for scimitar syndrome. Scimitar vein is the most indispensable characteristic of the scimitar syndrome. Nonetheless, the term "isolated scimitar vein anomaly" should be reserved to describe patients with a scimitar vein anomaly only. It is invalid to regard a scimitar sign on chest radiograph as a certain sign of a scimitar vein. Scimitar signs refer only to the radiographic image that is silhouetted on the plain chest film as a curved Turkish sword (or a scimitar). It should not be taken for granted that the radiographic scimitar sign automatically indicates an anomalous pulmonary venous connection to the inferior vena cava, and vice versa. This fascinating image could be noted on the plain chest radiographs in patients with normal pulmonary venous drainage. Conversely, a scimitar vein is not always silhouetted as a scimitar sign on the plain chest radiographs in patients with the scimitar syndrome. There could be false positives and false negatives of a scimitar sign in predicting the presence of a scimitar vein. Thus, the presence of scimitar signs is not a diagnostic criterion for scimitar syndrome. It is inappropriate to regard a radiographic scimitar sign with a normal pulmonary venous connection as pseudo-scimitar syndrome. There have been reports of scimitar variants, scimitar vein syndrome, and incomplete scimitar syndrome. These reports documented neither a systemic arterial supply nor bronchial anomalies. These patients suffered from isolated scimitar vein anomalies (type C isolated venous PM), rather than from scimitar syndrome (type G mixed bronchoarteriovenous PM). We must reverse the trend toward misusing the term "scimitar syndrome," which may have stemmed from the fact that "scimitar" is a catchy name that arouses interest. Partridge et al. reported 18 patients with dysmorphic right lungs, but only 12 of them were actually patients with scimitar syndrome. Presumably, the study intended to report that the full-fledged picture of scimitar syndrome could not be found in the other six patients, and to replace the use of the term "scimitar syndrome" with "dysmorphic right lung." The term "dysmorphic right lung" cannot help us understand the essential pathology behind scimitar syndrome, and it may inevitably cause more confusion than clarification. Moreover, patients with scimitar syndrome may have malinosculation of the pulmonary airways other than the agenesis, aplasia, dysplasia, hypoplasia, or hypogenensis found in the dysmorphic right lung.

Bronchopulmonary sequestration, congenital horseshoe lung, bronchogenic cysts, congenital lobar emphysema, and congenital cystic adenomatoid malformation may present as isolated bronchial airway disease to a pediatric pulmonologist. To the pediatric cardiologist, patients with congenital malformations of the lung, anomalous systemic arterial supply, and anomalous pulmonary venous connection may present as having congestive heart failure, cyanosis, and pulmonary hypertension. Congenital horseshoe lung can be found in patients with or without the scimitar syndrome. In patients with congenital horseshoe lung, we must
go further and perform cardiac catheterization and angiography to identify whether there is a scimitar vein to the inferior vena cava, a systemic arterial supply to the lung, or an abnormal branching pattern of the pulmonary artery (type G mixed bronchoarteriovenous PM), so that the optimal interventional therapy can be chosen. Congenital horseshoe lung, without a scimitar vein, should be categorized into type D mixed bronchoarterial PM. In the reports of three patients with a crossover lung segment,\textsuperscript{2,46} all of them suffered from type G mixed bronchoarteriovenous PM.

Finally, the combination of abnormal tracheal branching and left pulmonary artery sling\textsuperscript{47-50} should be classified as type D mixed bronchoarterial PM. Boothroyd et al. reported a patient with a right-sided accessory lobe of the lung, to which a bridging bronchus (from the left main bronchus) and an accessory artery (from the right pulmonary artery) opened.\textsuperscript{51} This is an example of type D mixed bronchoarterial PM. Thus, congenital pulmonary venolobar syndrome\textsuperscript{52} can be incorporated into this system-based PM.

Clinical reappraisal of the congenital BPVM reported in the literature

We have also reviewed a total of 142 patients with scimitar syndrome, bronchopulmonary sequestration, sequestration spectrum, congenital horseshoe lung, crossover lung segment, and other similar conditions,\textsuperscript{4,5,12,15,20,26,30,30-43} and tabulated these patients in Table 2. In conclusion, the major advantages of reappraising these patients systemically in terms of PM are fourfold: 1) rapid appreciation of problems of congenital BPVM by assessing the three basic bronchovascular systems of the lung independently, 2) recognition of different disease entities sharing the same pathology, 3) clear clarification of incomplete, variant, or pseudo-scimitar syndrome, and 4) acknowledgment of isolated scimitar vein anomaly and meandering right pulmonary vein.

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