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7

Congenital Malformations of the Bronchi and Lung

Benjamin H. Landing

BRONCHI

Bronchial and Pulmonary Isomerism Syndromes

There are several anomaly syndromes with pulmonary and lobar bronchial isomerism.[1] The right lung isomerism syndromes include Ivemark asplenia syndrome and M-anisosplenia. Ivemark asplenia syndrome features include absence of the spleen, bilateral right lung bronchial pattern, single ventricle, functionally single atrium, anomalous pulmonary venous return, great artery position seen in transposition of great vessels, pulmonic stenosis or atresia, bilateral atrial entry of superior venae cavae, hemiazygos connection of the inferior vena cava, malrotation of the intestine, symmetric liver, and a high frequency of right aortic arch. There is a high incidence of sepsis in early life. The syndrome predominantly affects boys. The M-anisosplenia features are similar to those of Ivemark syndrome, but cardiac lesions are less severe, and two or more small spleens develop. M-anisoplenia predominantly affects boys.

The left lung isomerism syndromes include polysplenia, O-anisosplenia, and f-anisosplenia. The polysplenia features include a bilateral left lung bronchial pattern with absence of an eparterial bronchus, malrotation of the intestine, symmetric liver, atrial or ventricular septal defect, symmetric entry of pulmonary veins into the atria, hemiazygos connection of the inferior vena cava, and multiple small spleens. The gender ratio is equal. The O-anisosplenia features include bilateral left lung bronchial pattern, congenital heart disease with double-outlet right ventricle, atrioventricular septal defect, and a lesser degree of visceral situs abnormality and number of small spleens than in polysplenia. Entry of the pulmonary veins to the atria is not symmetric. The genders are equally affected. The f-anisosplenia features are like those of O-anisosplenia, with congenital heart disease other than double-outlet right ventricle or atrioventricular septal defect. The atrial entry of the pulmonary veins is not symmetric. This pattern predominantly affects girls.

Situs inversus, short-pancreas syndrome, and other malformations must be differentiated from the pulmonary isomerism syndromes. Situs inversus describes reversed pulmonary lobar bronchial and vascular patterns and is associated with a high incidence of pulmonic stenosis or atresia. Situs inversus occurs often (≈50%) in the Kartagener syndrome of abnormal ciliary motility, usually with absence of ciliary dynein arms, increased frequency of respiratory tract infections, and immobile sperm in males. A ciliary abnormality has also been associated with left lung isomerism, but the pulmonary isomerism syndromes have not been adequately studied in this regard.

The short-pancreas syndrome describes an externally bilobed right lung caused by the absence of the fissure between the right upper and middle lobes. Other features are normal lobar bronchial and pulmonary artery patterns, congenital heart disease, malrotation of the intestine, and a relatively large number of small spleens.

The possible association of right lung isomerism with Ellis-van Creveld chondroectodermal dysplasia has been mentioned, and lobar emphysema of the left upper lobe due to compression of a left eparterial bronchus by the aortic arch has been reported in patients with Ellis-van Creveld syndrome.

The isomerism syndromes must be differentiated from hypoplasia or absence of the right upper lobe bronchus in scimitar syndrome and from sling left pulmonary artery (SLPA) with bridging bronchus type 2B. Unlike the left lung isomerism syndromes, the right lung is typically smaller than normal in these disorders.

High-kilovoltage chest radiographs can provide evaluable air bronchograms, or computed tomography (CT) scans can be used to demonstrate bronchial situs in patients with these syndromes.[2] The right pulmonary artery branch to the right upper lobe, the truncus anterior of the right pulmonary artery (TARPA), is easily demonstrable by angiocardiography or by dissection at autopsy.

This demonstration can be useful in the diagnosis or later confirmation of the pulmonary isomerism syndromes.

Exceptions to the usual relations of the bronchi and pulmonary arteries occur.[3] Wells and colleagues studied three patients, two with situs solitus and one with situs inversus, who had a single ventricle and absence of the right or left upper lobe bronchus but normal TARPA. Isolated left bronchial isomerism in the apparent absence of the other features of left lung isomerism syndromes has also been described.[4,5]

Single Mediastinal Unilobar Lung

Only a single instance of single mediastinal unilobar lung has been reported.[6] The pulmonary artery was small, but the bronchial and pulmonary artery branch patterns were not described.

Preeparterial and Other Tracheal Bronchi

The tracheal origin of a bronchus supplying the apical segment of the right upper lobe (*i.e.*, preeparterial bronchus) is a relatively frequent anomaly (0.1%–0.2% of live births) that usually has no clinical significance.[7,8] Tracheal origin of the right upper lobe bronchus or of an accessory right upper lobe bronchus other than a preeparterial bronchus also occurs. Such bronchi can be associated with recurrent or chronic infection and are rarely identified except in this situation.[9–12] They can be identified by imaging studies, including air bronchograms, CT scans, and lung scans.[11,13] Whether reported associations with other anomalies or syndromes, such as asplenia, Down syndrome, or left upper lobe bronchial atresia, are more than coincidence is uncertain.[14,15] Lobar emphysema of the right upper lobe from compression of a right upper lobe tracheal bronchus by a right aortic arch has been observed.[16]

Congenital Bronchobiliary Fistula

Congenital bronchobiliary fistula is preferable to the term tracheobiliary fistula because the fistulous connection between the airway and biliary tree usually joins the right main bronchus. The condition may be a duplication of the foregut that is patent at both ends. The clinical picture is that of cough, emphysema or atelectasis, yellow or greenish sputum, and pneumonitis. Other associated anomalies include right lung isomerism, esophageal atresia with tracheoesophageal fistula, and high origin of the right upper lobe bronchus. The clinical features can suggest those of cystic fibrosis, but that disease rarely causes respiratory symptoms before about 3 months of age.[17–19]

High Origin of the Right Upper Lobe Bronchus

The high origin of the right upper lobe bronchus (*i.e.*, carinal trifurcation), which is usually of no clinical significance, occurs frequently in patients with esophageal atresia and tracheoesophageal fistula (Fig. 7-1) and in patients with transposition of the great arteries.[20]

FIGURE 7-1. A dissected, stained, and cleared specimen of the trachea and bronchial tree of a neonate with esophageal atresia and tracheoesophageal fistula shows the anomaly of high origin of the right upper lobe bronchus, or carinal trifurcation.

Sling Left Pulmonary Artery, Tracheal Stenosis as a Result of Absence of the Tracheal Pars Membranacea, and Bridging Bronchus

The syndrome of SLPA (*i.e.*, the origin of the left pulmonary artery from the right) with an anteesophageal course of the left pulmonary artery and tracheal stenosis due to ring tracheal cartilages is highly associated with bridging bronchus, an anomaly in which the bronchus to the right middle and lower lobes arises from the left main bronchus and crosses (*i.e.*, bridges) the mediastinum to the right lung. Wells and colleagues proposed two main forms of SLPA. In one form, the left pulmonary artery is supracarinal and occurs either with or without a tracheal accessory (*i.e.*, preeparterial) bronchus. In the other form, the right lower lung is supplied by a bridging bronchus, with a right upper lobe bronchus in SLPA type 2A, or without a right upper lobe bronchus in SLPA type 2B. The latter pattern is the most common form of SLPA.[21]

Tracheal stenosis with ring tracheal cartilages (*i.e.*, absence of the tracheal pars membranacea) is common in SLPA type 2. The tracheal cartilages do not actually form rings but helical springlike structures. Unlike tracheal splint (see Chap. 6), in this type of tracheal stenosis, the central airway can lengthen to some extent, but it cannot dilate on inspiration. Radiologic features of SLPA type 2 include demonstration of the left pulmonary artery between the esophagus and airway in lateral radiographs and a low position of the apparent carina, which is actually the junction of the bridging and left main bronchi, in air bronchograms.[22] The right upper lobe bronchus can be misinterpreted as a tracheal bronchus, and the condition under discussion could explain the respiratory symptoms reported with tracheal bronchus in some patients.[5,23] Serial section reconstruction of the central respiratory tract pattern can be achieved with serial CT scan images.[24] Patients with SLPA type 2 have an increased incidence of imperforate anus and possibly absence of the gallbladder; one patient with right upper lobe bronchus arising from the esophagus who had bridging bronchus was reported.[25]

Bronchial Atresia

Bronchial atresia is usually applied to a situation of atresia of a bronchus, predominantly that to the left upper lobe, with a radiologically demonstrable mucus plug in the bronchial lumen distal to the site of atresia and with the affected area of lung aerated.[26] The condition predominantly affects boys. That the area of lung supplied by the atretic bronchus is aerated has been explained by airflow through interbronchiolar channels, the bronchoalveolar channels of Lambert, or the pores of Kohn, but why the explanation applies to this condition when essentially all other forms of bronchial occlusion lead to atelectasis of the affected region of lung is unclear. Association of this condition with extrapulmonary bronchogenic cysts may imply the cause is a problem in an early stage of lung budding during the fourth through sixth fetal weeks.[27] Hyperinflation of the affected region has been described with atresia of the apical bronchus of the left lower lobe.[28] CT and magnetic resonance imaging can be used for diagnosis.[29]

Communicating Bronchopulmonary-Foregut Malformations

This category of bronchial communication with the esophagus differs from that of tracheal or bronchial origin from the esophagus seen with tracheal agenesis. Srikanth and associates proposed a classification based on a review of 57 patients; this scheme is presented in Table 7-1.[30] The significant association with esophageal atresia and tracheoesophageal fistula with communicating bronchopulmonary-foregut malformations (CBPFM) implies a causative field defect, but more than one possible mechanism of such airway-foregut connections may apply. Why the connection of a main bronchus to the esophagus should be overwhelmingly on the right, unlike the more equally sided arrangement for foregut connections to lobar or more peripheral bronchi, is unclear. The similarity to proposals of pathogenesis of pulmonary sequestrations (see Chap. 8) are obvious, with the operational distinction being the presumed involution of the original foregut connection for pulmonary sequestration. These investigators have proposed that instances of communication of an area of lung with the airway and the esophagus result from secondary attachment of lung bud to the foregut as the lung buds curve posteriorly around the foregut during days 32 through 35 of fetal development. CBPFM has been demonstrated antenatally by ultrasonography. The differential diagnosis of a lung mass with increased echogenicity in this situation includes congenital cystic adenomatoid malformation and sequestration.[31]

TABLE 7-1
Congenital Bronchopulmonary-Foregut Malformations

Type	Number	Description
SIDE OF COMMUNICATING BRONCHOPULMONARY-FOREGUT MALFORMATIONS		
Right	40	
Left	13	
Bilateral	4	
Total	57	
GROUPS OF COMMUNICATING BRONCHOPULMONARY-FOREGUT MALFORMATIONS		
Group 1A*	7	EA/TEF plus right lung from esophagus or stomach
Group 1B	2	EA/TEF plus part of either lung from esophagus
Group 2	19	Lung (18 right from esophagus)
Group 3	26	Part of lung (right = left) from esophagus or stomach
Group 4	3	Intact airway also connects to esophagus
Group total	57	

** If whole lung connects to the gastrointestinal tract, 27/29 on right (groups 1A, 2, 4); if part of lung connects to the gastrointestinal tract, 17 right/14 left (groups 1B, 3)*

EA, esophageal atresia; TEF, tracheoesophageal fistula.

From Srikanth MS, Ford EG, Stanley P, Mahour GK. Communicating bronchopulmonary foregut malformations: classification and embryogenesis. J Pediatr Surg 1991;27:732.

Bronchial Strings and Webs

The term bronchial strings has been applied to a stringlike fibrous structure crossing the lumen of the right bronchus intermedius.[32] The lesion may be a version of the rarely reported condition of bronchial web.

Other anomalous bronchial patterns, such as a common origin of the right upper and middle lobe bronchi, are rarely of clinical significance unless pulmonary resection is done, but some patterns can be associated with recurrent pneumonitis.[33,34]

Williams-Campbell Syndrome

The Williams-Campbell syndrome of diffuse bronchiectasis with bronchial cartilage deficiency was originally thought to result from maldevelopment, but the explanation is suspect because symptomatic onset usually occurs after respiratory tract infection beyond the neonatal period.[35]

LUNGS

Useful reviews of respiratory tract and pulmonary malformations, their radiologic diagnostic features, and surgical management can be consulted for additional information on the topics covered in this section.[36–47]

A timetable of respiratory tract development (see Chap. 2) helps in understanding malformations of the lungs.[41,48] The ventral pharyngeal groove forms by 20 days of gestation. The laryngotracheal groove and respiratory tract bud are formed by 22 to 26 days, and lung bud bifurcation is achieved by 26 days of gestation. Between days 35 and 40, the lobar bronchial buds form. The pseudoglandular stage of lung development encompasses weeks 6 through 16 of gestation, and all conducting airways are formed by 16 weeks. The canalicular stage of lung development encompasses weeks 16 through 32. Initial alveolar formation occurs by 24 weeks of gestation. The terminal sac period of alveolar development encompasses weeks 26 through 32, and the alveolar stage of lung development usually occurs by 32 weeks of gestation.

The total number of alveoli at term has been estimated at between 10 to 150 million (mean, 55 million) and at 34 ± 5

million.[48,49] Pulmonary hypoplasia can result from reduced alveolar number or size, reduced number of bronchial or bronchiolar branchings, or combinations of these, but conditions with reduced air passage number (*e.g.*, the pulmonary hypoplasia seen with oligohydramnios) must reflect developmental disturbances preceding fetal week 16.[13] Compensatory lung growth, as with congenital absence of one lung or after complete or partial pneumonectomy appears to be triggered by lung stretch, but only during the period of normal lung growth. The functional lung surface area rises from less than 1 m^2 at 28 fetal weeks to 4 m^2 at term.[49] Cooney and colleagues described increased pulmonary acinar complexity with increased acinar radial count despite pulmonary hypoplasia in lungs of fetuses with polyhydramnios, and abnormal lungs with decreased functional surface area despite increased numbers of bronchial and nonrespiratory bronchiolar branchings have been reported in leprechaunism.[50,51]

Ectopia of the Lungs

Upward herniation (*i.e.*, ectopia) of lung tissue into the lower neck may occur at least with iniencephaly, Klippel-Feil brevicollis syndrome, and cri du chat syndrome (*i.e.*, 5p−). Proposals that the cause is decreased intrathoracic space or of forceful expiration against a stenotic larynx are suspect, because the usual effects of such physiologic situations are different, and connective tissue abnormality seems more probable.[52–54] In iniencephaly and Klippel-Feil syndrome, herniation of lung through the diaphragm can occur.[52,53] This pattern must be differentiated from that of extralobar pulmonary sequestration (see Chap. 8).

Abnormal Pulmonary Fissures and Septa

Connective tissue septa are most numerous in the subpleural regions of edges and angles of the lungs and on the diaphragmatic faces, but they are infrequent on the lateral costal lung surfaces. The loci of the more common accessory fissures and lobes are similar; they include the infracardiac lobe of the right lower lobe, the dorsal lobe of Nelson of the superior segment of either or both lower lobes, a horizontal accessory fissure of the right upper lobe, an apical fissure of either lung for the subclavian artery, and the azygos and hemiazygos lobes. Failure of formation of one or more of the normal fissures between the lung lobes is not uncommon, and externally bilobate right lung or monolobate right or left lungs with normal lobar bronchial patterns occur; the most commonly absent fissure is that between the right middle and upper lobes (Fig. 7-2). Kohler described a familial syndrome of hypoplastic left heart, talipes, tetramelic postaxial polydactyly, atypical giant cells in pancreatic islets, material polyhydramnios, absent lung lobation, and other anomalies.[55]

Bilateral Pulmonary Hypoplasia

Bilateral pulmonary hypoplasia has been found in 8% to 11% of neonatal autopsies, and in 50% of neonates who have other anomalies. Diagnostic criteria have been based on the ratio of lung weight to body weight:

Normal: 0.18 ± 0.003
Definite pulmonary hypoplasia: <0.009
Probable pulmonary hypoplasia: 0.01 to 0.012
Possible pulmonary hypoplasia: 0.013 to 0.017.

Bilateral pulmonary hypoplasia from reduced intrathoracic volume in infants with several types of chondrodystrophic skeletal dysplasia is discussed in Chapter 7. Another major cause of bilateral pulmonary hypoplasia often incompatible with postnatal life is that seen with oligohydramnios (*i.e.*, Potter syndrome or oligohydramnios tetrad) as a result of renal agenesis, renal hypodysplasia, and congenital urinary tract obstruction. The pulmonary hypoplasia in this situation is considered to date from 12 to 16 weeks of gestation and presumably differs in its basic mechanism from the pulmonary hypoplasia seen with protracted (>2 weeks) amniotic fluid leak in later pregnancy.[43]

Other important causes of congenital pulmonary hypoplasia include intrathoracic masses such as cystic hygroma; pleural effusions as in erythroblastosis fetalis, nonimmune hydrops, and cystic adenomatoid malformation of the lungs; and pulmonary hypoplasia associated with fetal muscular weakness due to fetal central nervous system or skeletal muscle abnormalities and attributed to fetal compression by the uterine wall (*e.g.*, Pena-Shokeir syndrome, allied fetal akinesia deformation sequence syndromes).[56–63] However, polyhydramnios has been reported with congenital myotonic dystrophy. Pulmonary hypoplasia in the apparent absence of the causative mechanisms previously listed has been reported as primary pulmonary hypoplasia (Figs. 7-3 and 7-4).

Many reports indicate that the pathogenesis of pulmonary hypoplasia may be more complex than has been appreciated. The

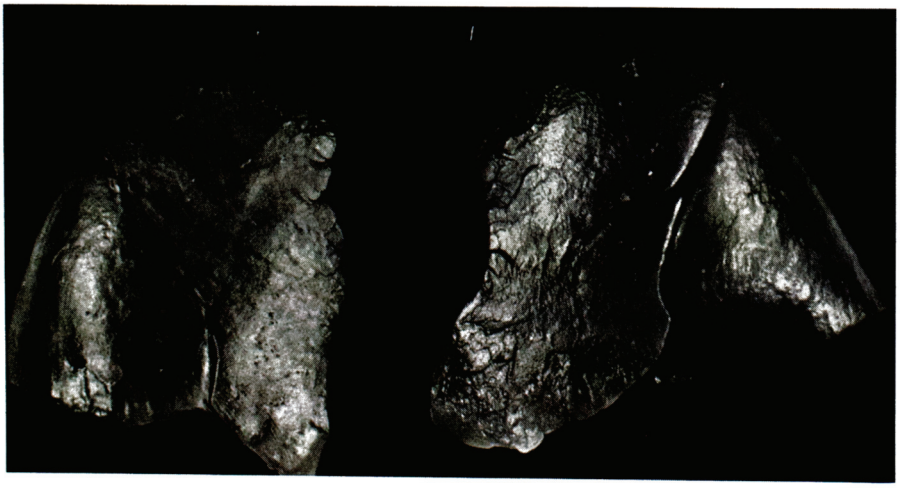

FIGURE 7-2. This newborn infant has no fissure between the right upper and middle lobes. There is an imperfect fissure between the lingular segment and the remainder of the left upper lobe.

FIGURE 7-3. In this striking example of bilateral pulmonary hypoplasia in a diprosopus conjoined twin, the larynx is larger than either lung.

pulmonary hypoplasia associated with the oligohydramnios of renal agenesis and other major urinary tract anomalies may reflect deficiency of a pulmonary growth factor produced by the kidneys, and the lungs may secrete a pulmonary-derived renotropin.[64] This area needs further study, especially in view of the lack of evidence for renal abnormality in many of the categories of pulmonary hypoplasia (see Chap. 2). The lesser degree of pulmonary hypoplasia and Potter syndrome features seen with infantile polycystic disease of liver and kidneys has been assumed to result from diaphragmatic elevation. The wide range of causes of nonimmune fetal hydrops, with which pulmonary hypoplasia is associated in about 50% of patients, appears to preclude a need for an explanation other than intrathoracic space occupation by pleural effusions and diaphragmatic elevation from ascites, although abdominal masses as a cause of pulmonary hypoplasia in the absence of oligohydramnios have been questioned.[58–61]

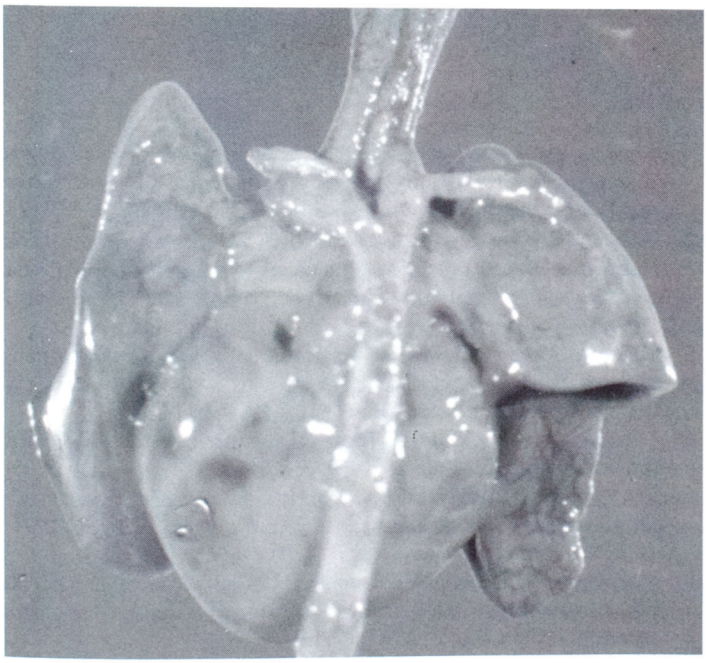

FIGURE 7-4. An example of primary pulmonary hypoplasia in a newborn allows comparison between the size of the lungs and the size of the heart in this posterior view of the thoracic contents. There was no congenital heart disease. (Courtesy of Juan Payser, M.D., Miami, FL.)

Giant abdominal wall defects may increase the risk of pulmonary hypoplasia, perhaps by chest deformity with down-slanting ribs from a low position of the diaphragm. Reported causes include gastroschisis, omphalocele (about 80% of total patients), the lower midline syndrome called cloacal exstrophy, and pentalogy of Cantrell.[62]

The pulmonary hypoplasia of the Pena-Shokeir and allied fetal akinesia deformation syndromes has been attributed to ischemic brain damage in the infant of a cocaine-using mother.[63] Further study of the timing of cocaine use would be of interest. The possible pathogenetic mechanisms of pulmonary hypoplasia in instances of expanded multiple pterygium syndrome are unclear.[65]

Skeletal Dysplasia Leading to Pulmonary Hypoplasia

The small conical or bell-shaped thorax found in several congenital skeletal dysplasias is associated with pulmonary hypoplasia and neonatal respiratory difficulty or death. These conditions can be categorized as skeletal dysplasias with short trunk, short-rib polydactyly syndromes (SRPS), other short-rib syndromes, or syndromes with platyspondyly. Skeletal dysplasias with a short trunk include achondrogenesis 1 (*i.e.*, Parenti-Fraccaro syndrome), which is autosomal recessive; achondrogenesis 2 (*i.e.*, Langer-Saldino syndrome), which is autosomal recessive; hypochrondrogenesis; atelosteogenesis; and fibrochondrogenesis, which is autosomal recessive.

SRPSs include the Saldino-Noonan syndrome (*i.e.*, SRPS I), which is autosomal recessive. The Saldino-Noonan syndrome also produces a gross deficiency of respiratory tract cartilage.[37] Other syndromes in this group are the Majewski syndrome (*i.e.*, SRPS II), which is autosomal recessive and may be the same as the condition called Mohr syndrome, and the Naumoff syndrome (*i.e.*, SRPS III), which is also autosomal recessive.

Other short-rib syndromes include Jeune syndrome type 1 (*i.e.*, thoracic asphyxiating dystrophy), Jeune syndrome type 2, and Ellis-van Creveld syndrome (*i.e.*, chondroectodermal dysplasia), all of which are autosomal recessive. This group may include those who have Beimer syndrome. Ellis-van Creveld syndrome also causes short trachea and a cross connection of the tracheal cartilages (*i.e.*, tracheal splint). Some patients also have a right lung bronchial pattern in the left lung.

Syndromes with platyspondyly include thanatophoric dysplasia and thanatophoric dysplasia with cloverleaf skull. Thanatophoric dysplasia with cloverleaf skull may be associated with tracheal cartilage sleeve (*i.e.*, fusion or failure of separation of the tracheal cartilage rings) with functional tracheal splint.

The antenatal diagnosis of these syndromes is discussed by Zimmer and associates.[66] The autosomal dominant disease thoracolaryngopelvic dysplasia (*i.e.*, Barnes syndrome) is not a cause of neonatal death.[67] In addition to the small, right, bell-shaped

thoracic cage and small chest volume, patients with this disorder also have a funnel-shaped larynx with tracheal stenosis below the cricoid cartilage and scoliosis. Rib abnormalities, including increased rib number, are associated with thoracic spine and thumb anomalies in some patients with unilateral pulmonary agenesis.[68]

Pulmonary hypoplasia with osteogenesis imperfecta, seen especially with osteogenesis imperfecta type 2, with defective synthesis of α-1(1)-procollagen has been attributed to thoracic cage deformity, but Shapiro and colleagues reported arrest of bronchial branching pattern at the 10-week stage, raising the possibility that intrinsic biochemical defects in pulmonary connective tissue can lead to pulmonary hypoplasia.[69] Study is necessary to determine whether similar considerations apply to pulmonary hypoplasia with neonatal hypophosphatasia; the lethal form of Larsen syndrome, which is characterized by flat facies, cleft soft palate, skeletal abnormalities, multiple joint dislocations, and pulmonary hypoplasia; the oromandibular limb hypogenesis syndrome, which produces abnormalities of the mandible, tongue, maxilla, and limbs; or the pulmonary hypoplasia of an autosomal recessive disease producing fetal growth retardation, hydromphalus, hypoplastic multilobed lungs, and other anomalies.[70–73] The Larsen syndrome is thought to result from a new mutation dominant gene defect and presumably reflects abnormal production of a structural protein. Older patients with infantile polycystic disease appear to have unusually friable connective tissue.

Quantitative studies of the composition of hypoplastic lungs have shown reduced alveolar number, radial alveolar count, and pulmonary elastic tissue in oligohydramnios-associated and other types of pulmonary hypoplasia, and there is reduced peribronchial cartilage in patients with Potter syndrome but not with the oligohydramnios of amniotic fluid leak.[74–77] Moessinger discusses chronic lung disease in infants with sublethal pulmonary hypoplasia.[78]

Unilateral Pulmonary Hypoplasia and Agenesis

The association of unilateral pulmonary agenesis with tracheal stenosis and esophageal atresia and tracheoesophageal fistula is discussed in Chapter 6. The association can also be with unilateral hypoplasia of a lung or of lung lobes.[79] An interestingly similar pattern of associations in an autosomal recessive condition producing agenesis of the left lung or absence of right upper and middle lobes in association with thumb abnormalities and other anomalies has been described.[80] Patients with small right lungs who have scimitar syndrome, horseshoe lung, or SLPA type 2 should be differentiated from those with absence of the right pulmonary artery or with pulmonary sequestration, although right pulmonary agenesis has been seen with SLPA.[81,82] Primary left upper lobe hypoplasia has been associated with recurrent respiratory tract infections.

Bilateral Pulmonary Agenesis

Bilateral pulmonary agenesis is a rare finding. It has been described in a fetus with nonimmune hydrops in which the pulmonary arteries and veins were absent and the pulmonary artery led only to the ductus arteriosus.[84] An infant whose respiratory tract consisted only of a trachea extending to the carinal level was suggested to have a severe degree of the hydrolethalus syndrome or the Fryns syndrome, both of which include central nervous system lesions.[85]

Congenital Acinar Dysplasia

Pathologically different from all other categories of pulmonary hypoplasia is a condition called acinar dysplasia, in which the lungs consist only of bronchiolelike passages in increased mesenchymal tissue, with no true pulmonary parenchymal development.[86,87] An apparently identical lesion has been reported as an autosomal recessive familial disorder associated with tetraamelia.[88]

Horseshoe Lung and Crossover Lung Segment

The definition of horseshoe lung specifies fusion of the lower lungs behind the heart.[89,90] Unilateral lung hypoplasia, predominantly if not invariably of the right lung, always exists. The condition is highly associated with scimitar syndrome and has been seen with SLPA, both of which are associated with right lung hypoplasia.[91] Separated from horseshoe lung by definition but also associated with scimitar syndrome is the lesion called crossover lung segment. In this anomaly, the medial inferior portion of the right lung extends behind the heart within a saccular extension of the right pleural cavity into the left lower hemithorax but is not fused with the left lung.[92,93] The two conditions can look similar angiographically.[94]

Congenital Mesenchymal Malformation and Rhabdomyomatous Dysplasia of the Lung

The condition called congenital mesenchymal malformation of the lung is rarely reported (*i.e.*, perhaps 3 cases). The patient of Warren and associates had most of the left lung replaced by a grossly solid mass of fascicles of spindle-shaped cells distributed through apparently normally developed fetal lung tissue.[95] Malformations that may be associated with rhabdomyomatous dysplasia of the lung include horseshoe lung, intralobar and extralobar sequestration, and cystic adenomatoid malformation of the lung.[96,97]

Neuroglial nodules in the lungs of infants with anencephaly are presumably caused by implantation of cells shed from the exposed neural plate into the amniotic fluid and aspirated by the fetus.[7] Ectopic adrenal tissue in the lung has been described.[7]

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