

Congenital anomalies of the lung

Developmental defects, pulmonary morphology, and clinical presentation

Pulmonary hypoplasia · Foregut cysts · Pulmonary sequestration

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Learning objectives

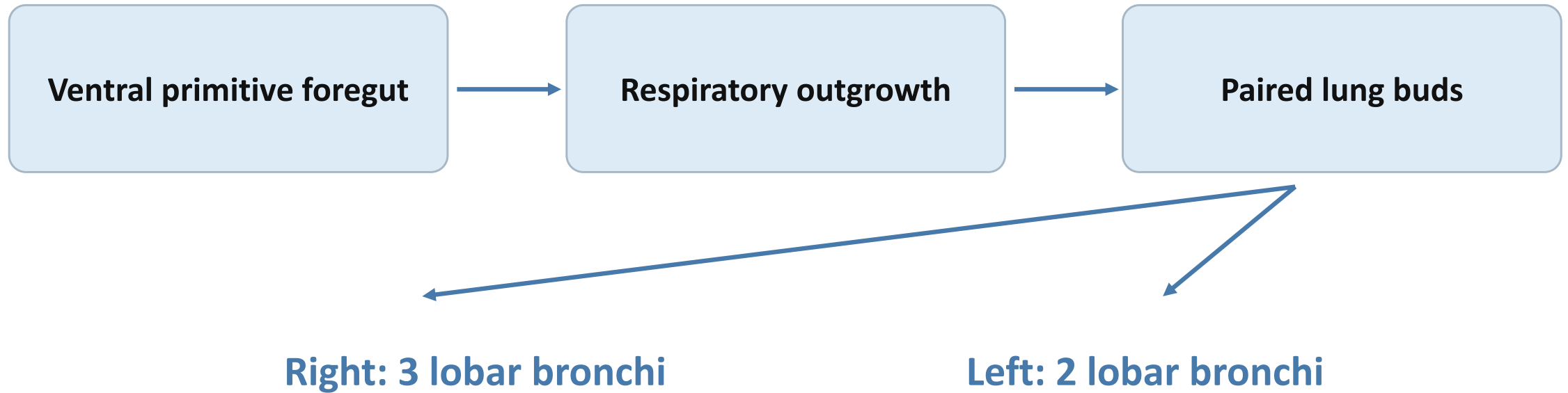
- Relate normal foregut-derived lung development to congenital pulmonary anomalies.
- Explain the mechanisms and neonatal implications of pulmonary hypoplasia.
- Recognize the locations and airway-type histology of foregut cysts.
- Distinguish intralobar from extralobar pulmonary sequestration.

Interpret anatomy, developmental mechanism, pathology, and typical presentation as one connected problem.

Organize congenital lung anomalies by the defect

- Pulmonary hypoplasia: Inadequate lung development with reduced lung size.
- Foregut cyst: Abnormal developmental detachment producing a cystic structure.
- Pulmonary sequestration: Noncommunicating lung tissue with systemic arterial supply.
- Other lesions include airway, vascular, cystic, and lobar inflation anomalies.

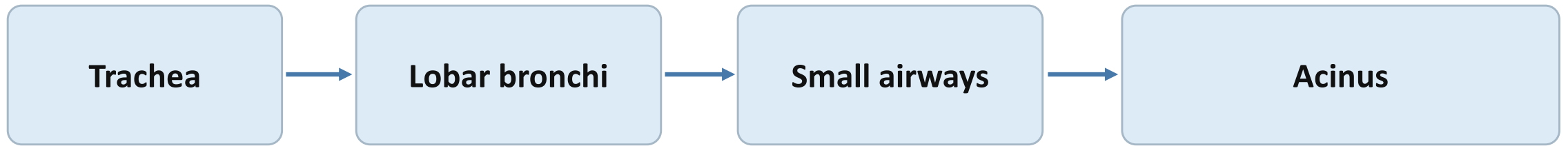
Embryologic origin: The respiratory diverticulum



Foregut origin explains shared airway phenotypes

- The respiratory system develops as an outgrowth of the ventral primitive foregut.
- The tracheal outgrowth produces paired lung buds and a branching bronchial tree.
- Abnormal separation or detachment can leave foregut-derived cystic structures.
- Embryologic origin helps interpret a bronchogenic cyst with mature airway-wall elements.

Normal airway branching: Proximal to distal



Alveolar ducts → alveolar sacs → gas-exchange alveoli

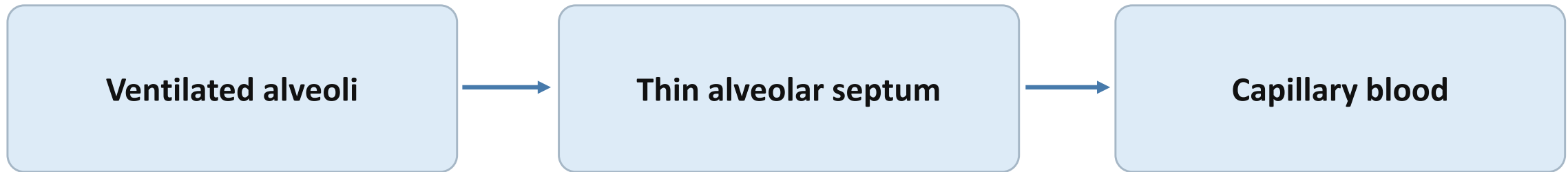
Normal lung architecture: What must develop?

- Conducting airways connect the trachea and bronchi with distal bronchioles.
- Terminal bronchioles lead to acini containing respiratory bronchioles, ducts, and alveoli.
- Alveoli are the structural site of air–blood gas exchange.
- A developmental anomaly may primarily alter lung size, airway connection, or vascular supply.

Airway-wall histology is a diagnostic reference

- Bronchi have a cartilage-supported wall and ciliated respiratory epithelium.
- Airway-associated glands produce mucus and contribute to mucosal defense.
- Smooth muscle is a normal airway-wall constituent.
- These same tissue components help identify bronchogenic cysts histologically.

Alveoli: The functional endpoint of development



Type I pneumocytes provide broad exchange surface; Type II cells make surfactant

Two circulation concepts relevant to malformations

- Normal airways are accompanied by a pulmonary and bronchial arterial supply.
- Pulmonary sequestration is defined instead by anomalous arterial supply from the aorta or its branches.
- A vessel's origin therefore helps distinguish a sequestered region from normal lung.
- Blood-supply abnormalities and airway-connection abnormalities are separate diagnostic observations.

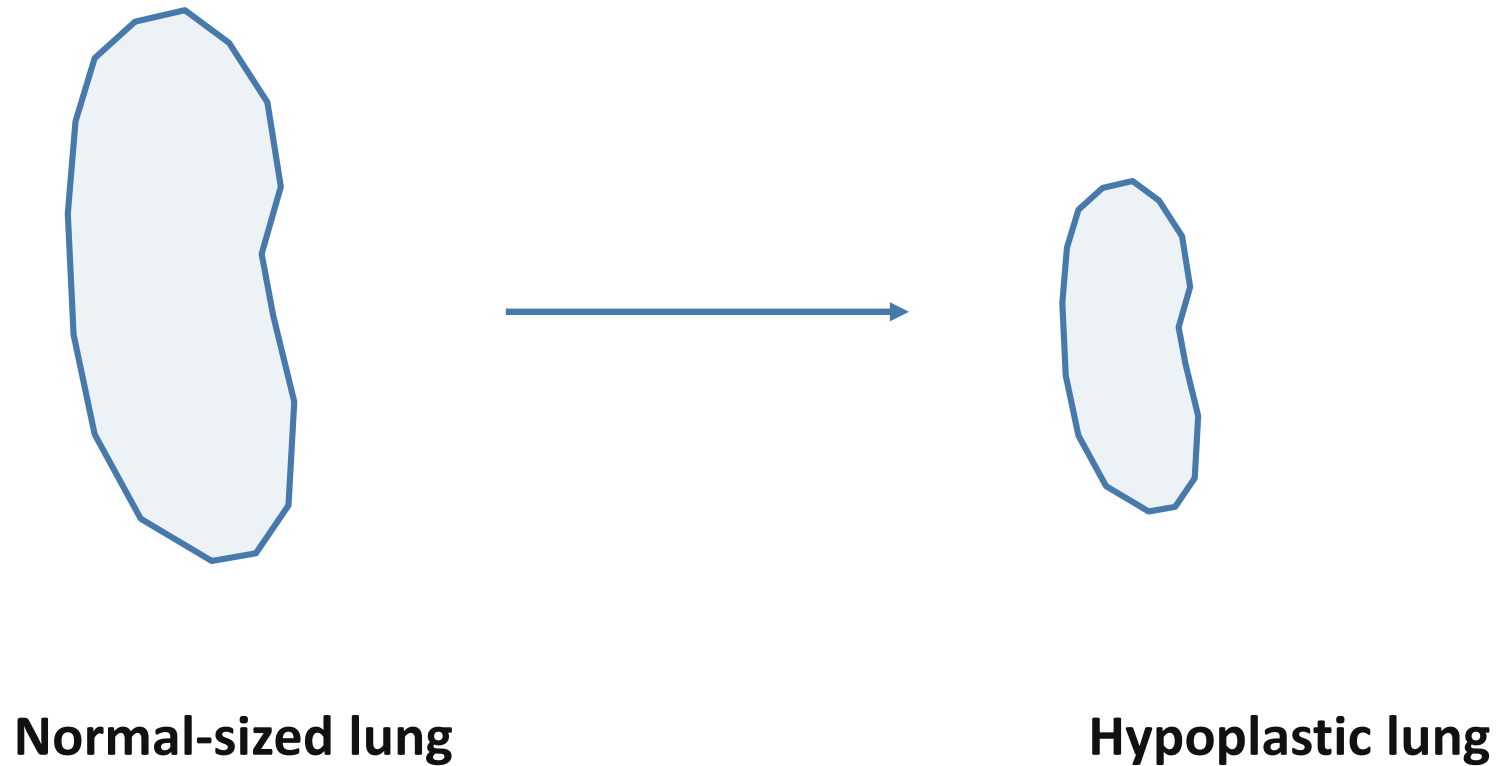
Three defining abnormalities

Anomaly	Primary defining feature	What to verify
Hypoplasia	Reduced developmental lung size	Laterality and impairment of expansion
Foregut cyst	Developmental cyst from foregut detachment	Location and epithelial / wall phenotype
Sequestration	Separate lung tissue without airway connection	Anomalous aortic arterial supply

Pulmonary hypoplasia: Definition

- Pulmonary hypoplasia is a developmental defect that reduces lung size.
- Both lungs may be affected, even when one is more severely involved.
- The core abnormality is underdevelopment rather than collapse of an originally fully developed lung.
- Severe disease has major consequences in the early neonatal period.

Reduced lung size is the morphological hallmark



Conceptual size contrast; Not a scaled anatomical depiction

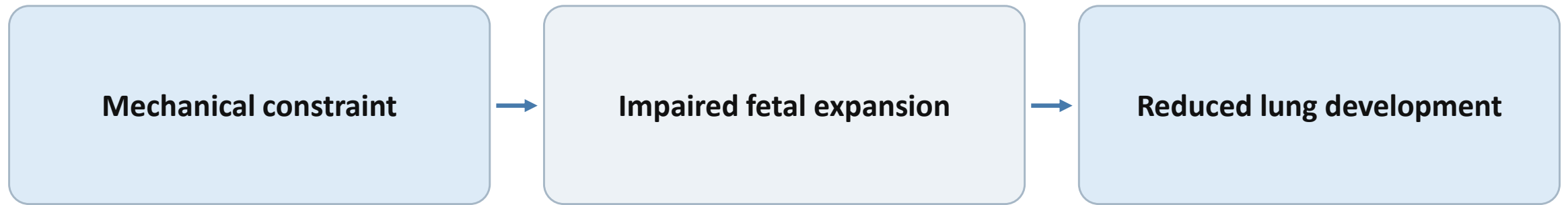
Pulmonary hypoplasia may be asymmetric

- The source describes a defect of development involving both lungs.
- Severity need not be equal: One lung may be more affected than the other.
- Asymmetry should not obscure the systemic embryonic or mechanical setting.
- Clinical interpretation begins by evaluating whether fetal lung expansion was restricted.

Restricted fetal expansion is the causal theme

- Pulmonary hypoplasia can follow abnormalities that compress the developing lung.
- A different route is impaired ability of the lung to expand in utero.
- Both mechanisms act before the transition to neonatal respiration.
- The developmental timing explains why severe deficits may be evident immediately after birth.

Pathway from in-utero constraint to small lungs



Causal schematic; Precise molecular pathway is not specified in the source.

Congenital diaphragmatic hernia and hypoplasia

- Congenital diaphragmatic hernia is a cited cause of fetal lung compression.
- The abnormal thoracic mechanical environment limits lung expansion.
- The resulting developmental deficit produces pulmonary hypoplasia.
- The lecture focuses on pulmonary consequences; Detailed hernia classification is outside this source excerpt.

Compression-associated hypoplasia

Diaphragmatic hernia



Compression of fetal lungs



Pulmonary hypoplasia

The diagram shows the textbook's compression mechanism at the organ level.

Oligohydramnios-associated hypoplasia

- Oligohydramnios is the second named setting for pulmonary hypoplasia.
- It interferes with fetal lung expansion in utero.
- The pathological endpoint is reduced lung size.
- The causal lesson is inadequate physical expansion during development, not an acquired alveolar collapse.

Restricted-expansion pathway

Oligohydramnios



Limited fetal expansion



Pulmonary hypoplasia

Two cited settings, one developmental endpoint

Context	Perturbation	Shared consequence
Diaphragmatic hernia	Fetal lung compression	Reduced lung development
Oligohydramnios	Impeded fetal expansion	Reduced lung development

Why severe hypoplasia is life-threatening

- A normally developed lung provides alveolar surface for exchange between air and blood.
- Severely underdeveloped lungs cannot provide adequate respiratory function after birth.
- Severe pulmonary hypoplasia is fatal in the early neonatal period.
- This is a severity statement; The text does not provide quantitative risk or survival estimates.

Developmental hypoplasia versus acquired atelectasis

- Hypoplasia: Defective fetal development leading to small lungs.
- Atelectasis: Incomplete neonatal expansion or collapse of previously inflated lung.
- The former reflects development; The latter describes an expansion / collapse state.
- Do not use the two terms interchangeably without analyzing the anatomical and temporal context.

Hypoplasia and atelectasis: Conceptual distinction

Feature	Pulmonary hypoplasia	Atelectasis
Primary process	Developmental undergrowth	Incomplete expansion or collapse
Core change	Reduced developed lung size	Poor aeration / reduced expansion
Timing	Established in utero	May occur in neonatal or later life
Clinical use	Analyze prenatal constraint	Analyze airway, pleura, or scarring

Case interpretation: Small neonatal lungs

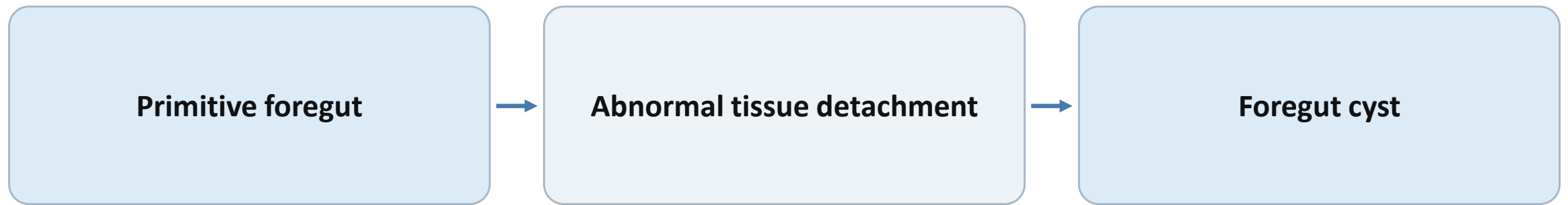
- A newborn has severe respiratory compromise and markedly small lungs.
- A history of diaphragmatic hernia would support developmental pulmonary hypoplasia.
- Intrauterine restriction of lung expansion explains the structural deficit.
- Question for learners: What additional prenatal context could produce the same endpoint?

Answer: Oligohydramnios is another cited causal context.

Foregut cysts: Developmental origin

- Foregut cysts arise when a part of the primitive foregut detaches abnormally.
- The misplaced detached tissue may retain a recognizable foregut-derived wall phenotype.
- These lesions differ conceptually from inadequate growth of the entire lung.
- Classification depends on the histological architecture of the cyst wall.

Mechanism of a foregut-derived cyst



The diagram is a developmental schematic, not a microscopic specimen.

Foregut cysts: Typical anatomic sites

Middle mediastinum

Pulmonary hilum

Frequent sites of foregut cysts

Hilum and middle mediastinum: Location matters

- The hilum and middle mediastinum as the most frequent sites.
- These regions contain major airways and other structures that may be compressed.
- The clinical consequences can reflect location rather than aggressive intrinsic growth.
- Anatomic location is supportive but cannot replace cyst-wall assessment.

Foregut cysts: Wall-based classification

Type	Classification basis	Relative frequency in excerpt
Bronchogenic	Airway-like histologic wall	Most common
Esophageal	Esophageal-type wall	Not quantified
Enteric	Enteric-type wall	Not quantified

Bronchogenic cyst: The reference phenotype

- Bronchogenic cyst is the most common type of foregut cyst in the Robbins passage.
- Its epithelial lining is ciliated pseudostratified columnar epithelium.
- The wall contains bronchial glands, cartilage, and smooth muscle.
- Together these features reproduce normal bronchial microanatomy.

A bronchogenic cyst rarely joins the airway tree

- The source states that bronchogenic cysts are rarely connected to the tracheobronchial tree.
- A cyst can therefore be lined by respiratory epithelium without being an airway segment.
- Airway connection and epithelial differentiation are distinct anatomical features.
- This distinction becomes important when comparing a cyst with sequestration.

Respiratory epithelial lining: Histologic clue

- The characteristic lining is ciliated pseudostratified columnar epithelium.
- “Pseudostratified” describes nuclei at different levels in one epithelial layer.
- Cilia support recognition of respiratory-type epithelial differentiation.
- The lining must be interpreted together with the rest of the cyst wall.

Cyst-wall glands: Recapitulation of a bronchus

- Bronchial glands are part of the bronchogenic cyst wall.
- Normal bronchi contain mucus-producing subepithelial glands.
- Glandular structures provide another correspondence between cyst and airway wall.
- The combination of features is more informative than any one wall element alone.

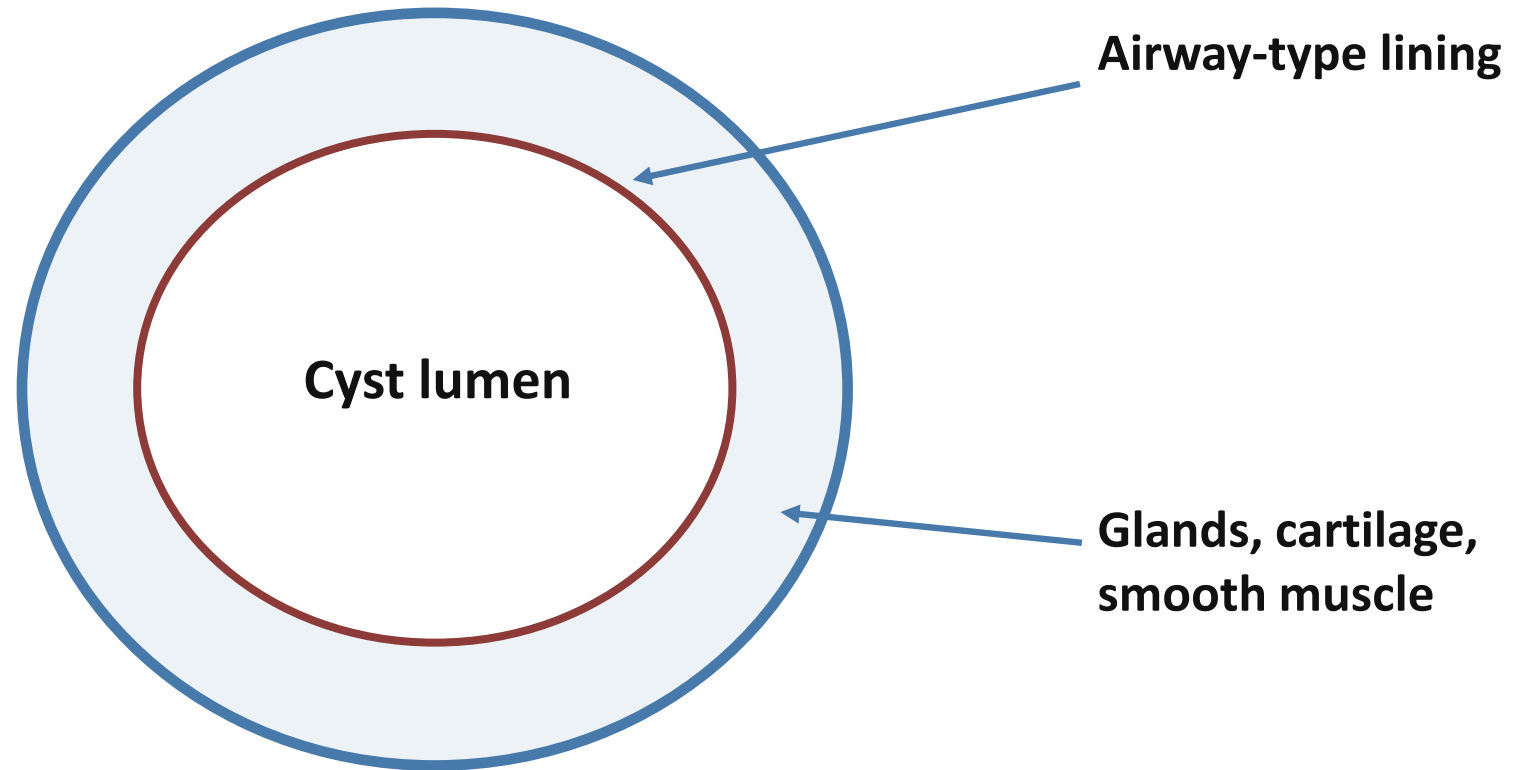
Cartilage is a critical wall component

- Bronchogenic cysts can contain cartilage within their walls.
- Cartilage is also part of the normal bronchial supporting framework.
- Its presence helps explain why the lesion resembles bronchial rather than alveolar tissue.
- Cartilage must be assessed in its histologic context with epithelium and glands.

Smooth muscle completes the airway-wall pattern

- The wall of a bronchogenic cyst may include smooth muscle.
- Smooth muscle is normally found in conducting airway structures.
- Glands, cartilage, and smooth muscle collectively mirror bronchial anatomy.
- The cyst by its wall structure, not by symptoms alone.

Bronchogenic cyst: Layered wall structure



Schematic cross-section—not a micrograph

Four recognizable bronchogenic cyst components

Ciliated airway epithelium

Subepithelial glands

Cartilage + smooth muscle

A bronchogenic cyst reproduces these airway-wall elements

Foregut cysts may be incidental

- Many foregut cysts are found without causing symptoms.
- Incidental presentation does not change their developmental origin.
- In an asymptomatic lesion, location and wall architecture remain central to classification.
- The source does not give a numerical probability of incidental detection.

Compression produces symptomatic foregut cysts

- Cysts can become symptomatic through pressure on neighboring structures.
- The hilum and middle mediastinum place them near major airway-related structures.
- The symptom mechanism is mechanical rather than necessarily infectious.
- Clinical features alone do not uniquely establish a bronchogenic cyst phenotype.

Superimposed infection: A second symptom mechanism

- Foregut cysts can also present following superimposed infection.
- Infection is a complication of a pre-existing developmental structure.
- Compression and infection are separate mechanisms of symptomatic presentation.
- The textbook passage does not specify organisms, laboratory thresholds, or treatment regimens.

Foregut cyst presentation: Two mechanisms

Finding	Mechanical pathway	Infective pathway
Pre-existing lesion	Congenital foregut cyst	Congenital foregut cyst
Added event	Compression of nearby structures	Superimposed infection
Interpretation	Symptoms depend on site / pressure	Symptoms follow secondary infection

Case interpretation: Mediastinal cyst

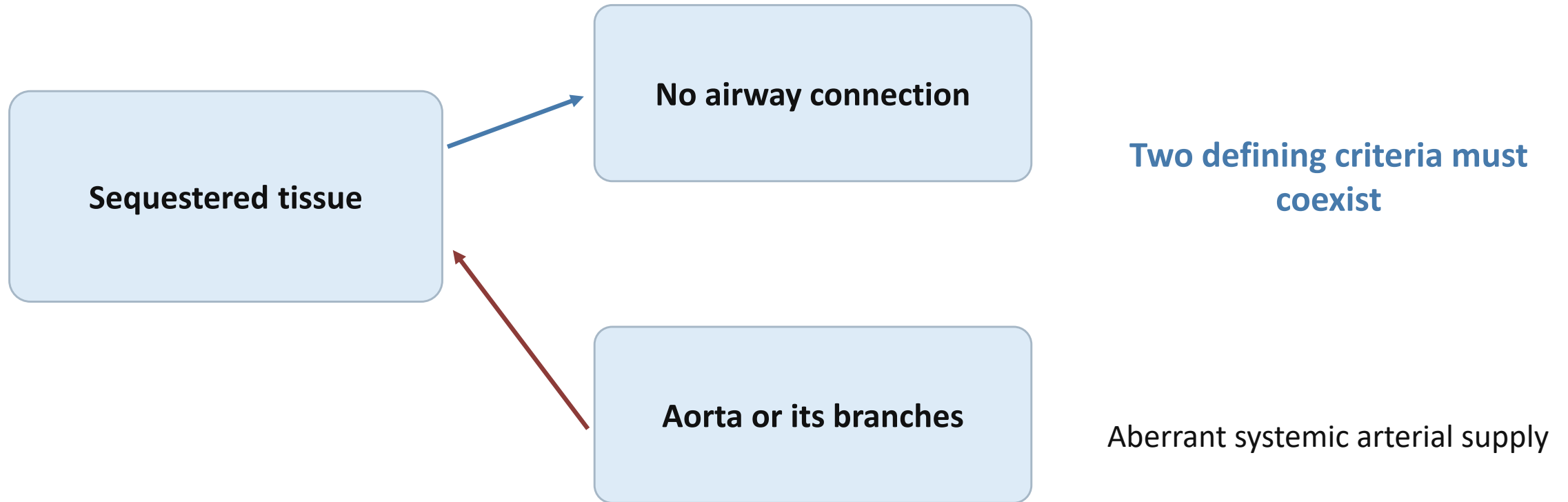
- A cystic lesion near the middle mediastinum is discovered incidentally.
- Its wall contains ciliated pseudostratified epithelium, cartilage, glands, and smooth muscle.
- The combined findings support a bronchogenic cyst.
- Question: Would the lack of airway communication exclude the diagnosis?

Answer: No. Bronchogenic cysts are rarely connected to the airway tree.

Sequestration: Two-part definition

- Pulmonary sequestration is a discrete mass or area of lung tissue.
- There is no connection to the normal airway tree.
- Its arterial supply arises anomalously from the aorta or its branches.
- The two diagnostic components are airway isolation and systemic arterial inflow.

Two defining findings in pulmonary sequestration



Why absent airway connection matters

- Sequestered tissue does not communicate with the tracheobronchial tree.
- The absent connection separates it anatomically from ordinarily ventilated lung.
- This is a defining criterion rather than a secondary feature of infection.
- A disconnected airway segment alone is insufficient without the abnormal arterial supply.

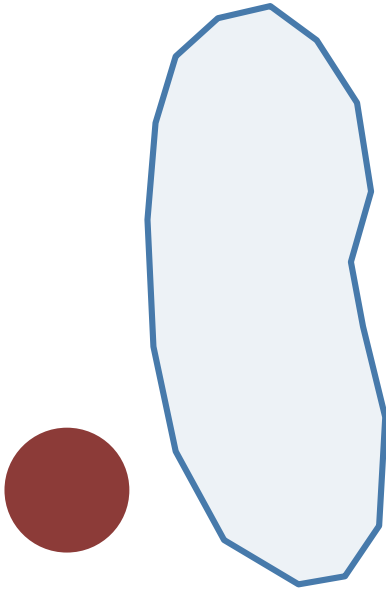
Abnormal arterial origin is indispensable

- Blood supply is derived from the aorta or one of its branches.
- This is not the ordinary pulmonary arterial arrangement of lung parenchyma.
- The artery's origin is part of the definition, not merely an associated finding.
- Anatomic evaluation should establish both blood supply and airway connection.

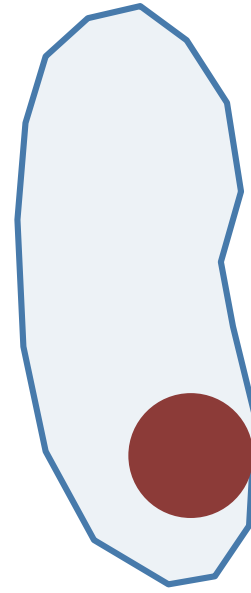
Airway and blood supply define different axes

- Airway anatomy asks whether tissue connects to a patent tracheobronchial pathway.
- Vascular anatomy asks where arterial inflow originates.
- Pulmonary sequestration is defined by an abnormality in both axes.
- Its location relative to the surrounding lung determines the intralobar / extralobar subtype.

Extralobar versus intralobar location



Extralobar: Outside the lung



Intralobar: Within the lung

Schematic location only—not anatomical imaging

Extralobar sequestration: Outside the lung

- Extralobar sequestration is located external to the lung.
- It is an anatomically separate focus of lung tissue.
- It still meets the shared criteria: No airway connection and aortic-derived arterial supply.
- The extralobar descriptor states its location, not a different embryologic cell type.

Extralobar sequestration: Infant mass lesion

- Extralobar lesions most often present in infants.
- The typical described presentation is as a mass lesion.
- An early mass presentation differs from later infectious manifestations of intralobar lesions.
- These are tendencies in the passage; Precise age limits or frequencies are not supplied.

Extralobar lesions and other anomalies

- Extralobar sequestration may coexist with other congenital abnormalities.
- A concurrent malformation should not be assumed to cause sequestration.
- The coexistence is clinically important when interpreting complex congenital presentations.
- The source does not enumerate specific associated defects or their frequencies.

Intralobar sequestration: Within lung parenchyma

- Intralobar sequestration is located within the lung.
- Unlike the extralobar form, it is not an external mass of tissue.
- It retains the basic sequestration features of airway separation and systemic artery.
- The “intra-” versus “extra-” distinction describes position relative to lung.

Intralobar sequestration tends to present later

- Presentation most often in older children.
- Repeated localized infections are a common reason for recognition.
- Bronchiectasis is another presenting association noted in the source.
- A later symptomatic presentation does not negate a congenital structural origin.

Recurrent localized infection: An anatomic clue

- Intralobar sequestration is associated with recurrent localized infection.
- Localization is a clue to a persistent focal anatomic abnormality.
- Repeated episodes can prompt consideration of a congenital malformation.
- A complete sequestration interpretation must still establish airway and vascular anatomy.

Bronchiectasis is a documented association

- Bronchiectasis is named as a presentation of intralobar sequestration.
- It signifies an airway-damage pattern rather than defining sequestration itself.
- A localized bronchiectatic process can coexist with sequestered lung tissue.
- The supplied source does not describe the detailed microscopic progression.

Presentation pattern: Extra- versus intralobar

Extralobar presentation



Intralobar presentation

Infancy / mass lesion

Older childhood / infection

Typical clinical patterns—not obligatory ages

Sequestration subtypes: Structured comparison

Feature	Extralobar	Intralobar
Relationship to lung	External to lung	Within lung
Usual presentation	Infant mass lesion	Older child: Repeated local infection
Associated findings	May coexist with anomalies	Bronchiectasis may occur
Shared definition	Disconnected + systemic artery	Disconnected + systemic artery

Why timing cannot substitute for anatomy

- Infant mass lesions suggest the usual extralobar presentation.
- Older childhood with localized infection suggests the usual intralobar presentation.
- Age and symptoms are informative patterns, not the defining classification criteria.
- Definitive conceptual distinction remains outside-versus-within lung tissue.

Case interpretation: Recurrent focal infection

- An older child has repeated infections affecting one localized lung region.
- A congenital intralobar sequestration is a relevant explanatory possibility.
- The anatomic questions are airway communication and systemic arterial origin.
- Question: Does recurrent infection itself prove sequestration?

Answer: No. Both defining anatomic features must be established.

Other congenital abnormalities: Scope of Robbins

- Additional anomalies affect the trachea, bronchi, pulmonary vessels, and lung parenchyma.
- The text names airway atresia, stenosis, and tracheoesophageal fistula.
- It also lists vascular anomalies, congenital pulmonary airway malformation, and lobar overinflation.
- This chapter passage does not expand their subtypes, histology, or treatment.

Less common anomalies: Organ-system map

Anatomic domain	Named entities in the source	Nature of source detail
Conducting airways	Atresia; Stenosis; Tracheoesophageal fistula	Named only
Pulmonary vessels	Vascular anomalies	Named only
Lung development	Congenital pulmonary airway malformation	Named only
Lobar inflation	Congenital lobar overinflation (emphysema)	Named only

Airway anomalies: Separate the named concepts

- Atresia and stenosis are listed as tracheal or bronchial developmental anomalies.
- Tracheoesophageal fistula is also listed within that airway group.
- These entities should not be conflated with a bronchogenic cyst.
- Robbins provides no detailed causal mechanism for these lesions in this paragraph.

Vascular and parenchymal anomalies: Boundaries

- Vascular malformations are included among less common congenital pulmonary defects.
- Congenital pulmonary airway malformation is specifically named.
- Congenital lobar overinflation, also called emphysema in the excerpt, is also named.
- Specific clinical criteria and morphologic subtypes require material beyond this passage.

Do not confuse congenital entities

Entity	Distinguishing source-defined point
Pulmonary hypoplasia	Reduced developmental size of lung tissue
Bronchogenic cyst	Foregut-derived cyst with characteristic airway-type wall
Pulmonary sequestration	Noncommunicating tissue + systemic artery
Lobar overinflation	Named separately; Mechanism not elaborated here

Integrated classification: Ask three anatomical questions

Small developed lung?



Hypoplasia

**Cyst near hilum /
mediastinum?**



Foregut cyst

**Disconnected tissue +
systemic artery?**



Sequestration

A disciplined congenital-lung diagnostic description

- Start with the anatomical compartment: Lung tissue, airway, cyst, or vessel.
- Record the developmental feature: Small lung, cystic separation, or isolated tissue.
- If sequestration is considered, establish airway connection and arterial origin.
- For cysts, interpret wall histology together with the mediastinal / hilar location.

Case set: Distinguish three mechanisms

- A newborn with a history of oligohydramnios has unusually small lungs.
- An incidental hilar cyst has respiratory epithelium, glands, cartilage, and smooth muscle.
- An older child has localized recurrent infection and an isolated region of lung tissue.
- Match each pattern to the most defensible congenital anomaly.

Case set: Reasoned answers

Pattern	Most consistent interpretation	Decisive source feature
Small lungs + oligohydramnios	Pulmonary hypoplasia	Restricted in-utero expansion
Hilar airway-type cyst	Bronchogenic cyst	Wall recapitulates bronchus
Focal infection + isolated tissue	Possible intralobar sequestration	Must also show systemic artery

Five defensible take-home principles

- Pulmonary hypoplasia represents reduced development, often after restricted fetal expansion.
- Foregut cysts arise through abnormal detachment; Bronchogenic cysts reproduce an airway wall.
- Sequestration requires both absent airway connection and abnormal aortic arterial supply.
- Extralobar and intralobar forms differ in position and usual clinical presentation.
- Other congenital anomalies are named but not detailed in this Robbins section.

Reference and scope of this lecture

- Robbins, Cotran & Kumar. Pathologic Basis of Disease. 11th ed. Chapter 15: The Lung.
- Primary congenital anomalies section: printed page 613.
- Normal lung development and histology: introductory material on page 613.
- Acquired atelectasis contrasted only where it clarifies the developmental distinction.
- Schematic figures are original editable teaching illustrations; no clinical specimen image is reproduced.