UNIVERSITATEA DE STAT DE MEDICINĂ ȘI FARMACIE
„NICOLAE TESTEMIȚANU” DIN REPUBLICA MOLDOVA

Semiology of the Respiratory Diseases in Children

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Learning objectives

To understand:

- anatomical and physiological features of the respiratory tract in children vs adults
- aspects of history and physical examination of the respiratory tract in children
- most frequent sign and symptoms of respiratory diseases in children
- methods of paraclinical examination (imaging, functional) of the respiratory tract in children

Peculiarities of the Respiratory System in Children

Respiratory system

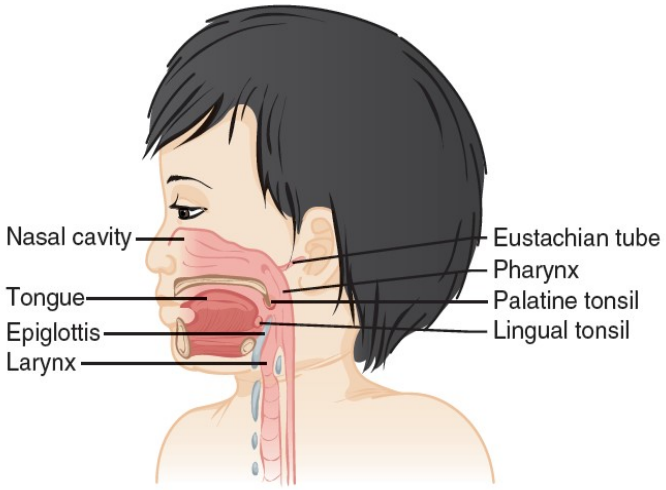


Fig 16.1 Upper airway.

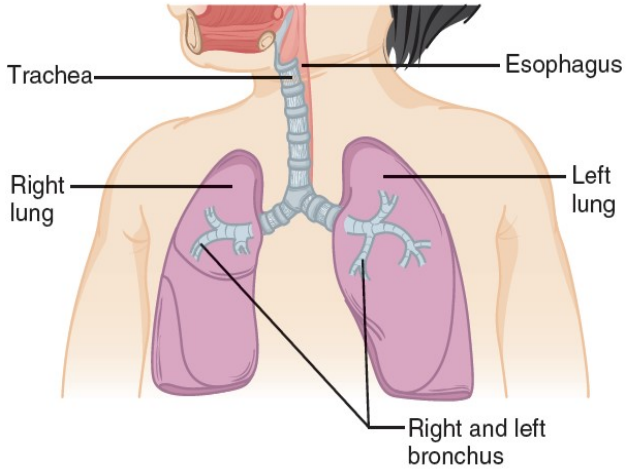


Fig 16.2 Middle and lower airways.

- The **upper airway**, containing the nasopharynx and oropharynx, serves as the pathway for gases exchanged during ventilation, the movement of oxygen into the lungs and carbon dioxide out of the lungs. The larynx divides the upper and lower airways.
- The **lower airways** (trachea, bronchi, and bronchioles) serve as the pathway of gases to and from the alveoli in the lungs.

Respiratory system

- The left lung is divided into two lobes, and the right lung is divided into three lobes.

- Alveolar sacs surrounded by capillaries are located at the end of the airways and are the site of gas exchange, where oxygen diffuses across the alveolocapillary membrane.

- Surfactant secreted by alveolar cells coats the inner surface of the alveolus to allow expansion during inspiration.

Respiratory system

- The lung tissue surrounding the airways keeps them from collapsing as the oxygen moves in and carbon dioxide moves out during ventilation.

- The lungs are positioned in the thoracic cavity, where the ribs and muscles protect the lungs from injury.

Respiratory system

- The intercostal muscles work with the diaphragm to perform the work of breathing.
- The diaphragm is a muscle that separates the abdominal and thoracic cavity contents.
- When the diaphragm contracts, it creates negative pressure that increases the thoracic volume and pulls air into the lungs.

Respiratory system

- The lungs and chest wall have the ability to expand during inspiration (compliance) and then to recoil or return to the resting state with expiration.

- The work of breathing is tied to the muscular effort required for ventilation, which can be increased in cases of airway obstruction or disorders that increase the stiffness of the lungs.

Respiratory system

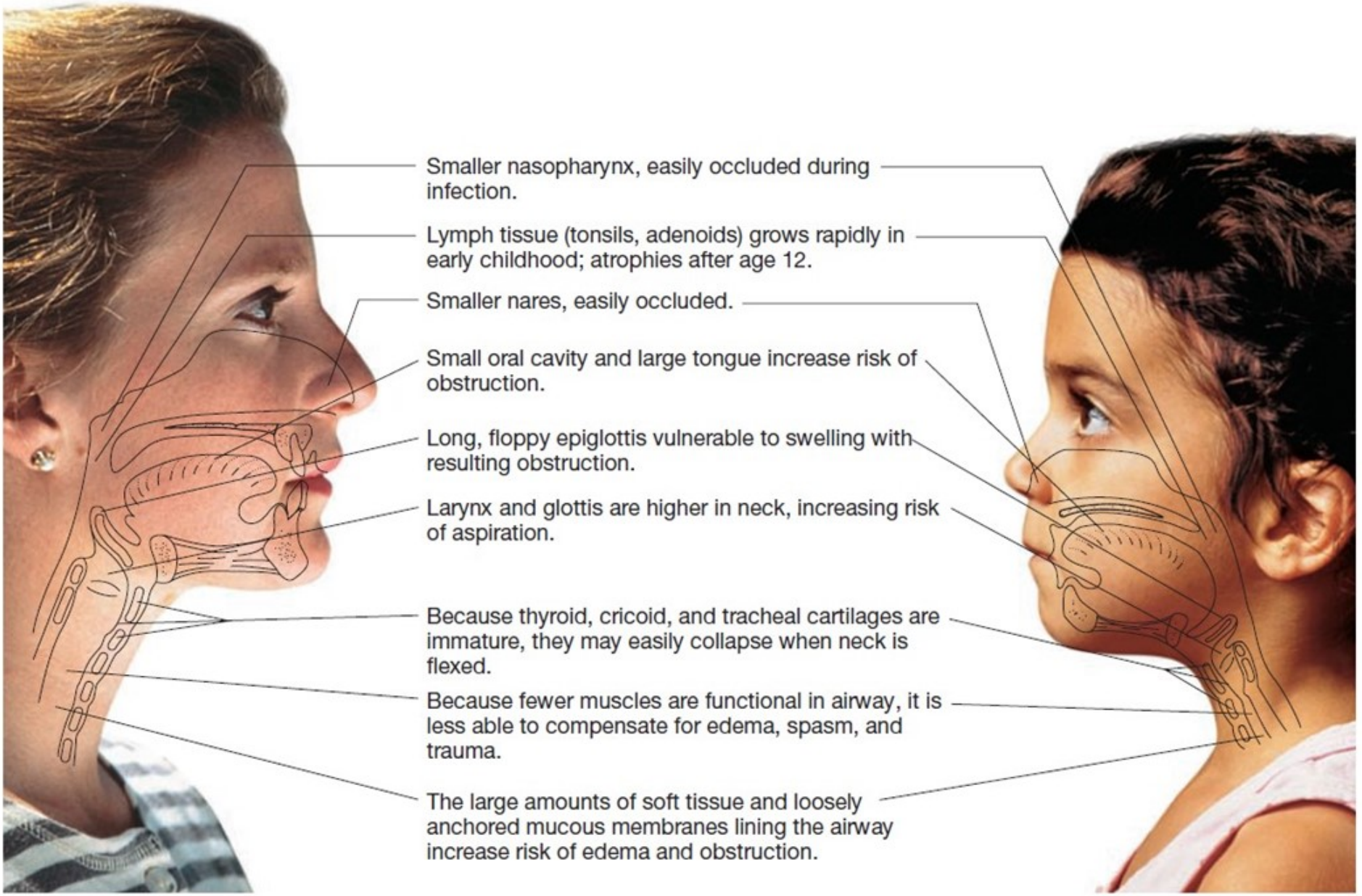
- The respiratory center in the brain controls respiration, sending impulses to the respiratory muscles to contract and relax.
- Breathing is usually automatic as the nervous system adjusts the ventilatory rate and volume to maintain normal gas exchange (Brashers, 2014).
- Chemoreceptors monitor the pH, $PaCO_2$, and PaO_2 in the arterial blood and send signals to the respiratory center to increase ventilation in cases of arterial hypoxemia.

Respiratory system

- Effective gas exchange requires a near even distribution of ventilation and perfusion (oxygenated blood flow to all portions of the lungs).
- As oxygen diffuses across the alveolocapillary membrane, it dissolves in the plasma and the resulting increased partial pressure of oxygen (PaO2) helps bind the oxygen to the hemoglobin molecules for transport to the cells for metabolism.
- Carbon dioxide produced by cellular metabolism is dissolved in the plasma (PCO2) and/or as bicarbonate and travels back to the lungs where it diffuses across the alveolocapillary membrane.

Peculiarities of the respiratory system in children

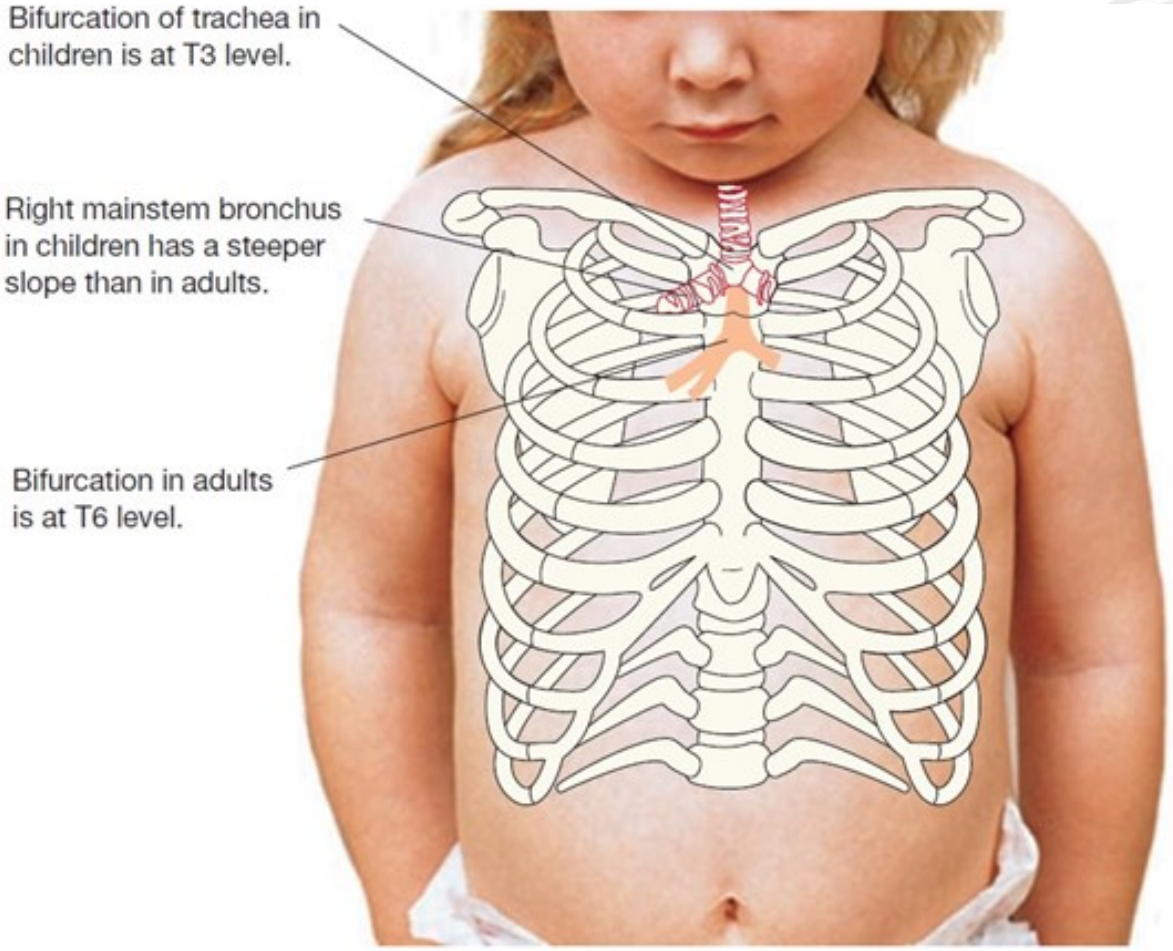
- The child's respiratory tract constantly grows and changes until about 12 years of age.
- The young child's neck is shorter than an adult's, resulting in airway structures that are closer together.
- These differences create a greater potential for obstruction


Smaller nasopharynx, easily occluded during infection.
Lymph tissue (tonsils, adenoids) grows rapidly in early childhood; atrophies after age 12.
Smaller nares, easily occluded.
Small oral cavity and large tongue increase risk of obstruction.
Long, floppy epiglottis vulnerable to swelling with resulting obstruction.
Larynx and glottis are higher in neck, increasing risk of aspiration.
Because thyroid, cricoid, and tracheal cartilages are immature, they may easily collapse when neck is flexed.
Because fewer muscles are functional in airway, it is less able to compensate for edema, spasm, and trauma.
The large amounts of soft tissue and loosely anchored mucous membranes lining the airway increase risk of edema and obstruction.

Peculiarities of the respiratory system in children

- The infant's airway diameter is approximately 4 mm (0.16 in.), about the width of a drinking straw, in contrast to the adult's airway diameter of 20 mm (0.8 in.).
- The child's little finger is a good estimate for the child's tracheal diameter and can be used for a quick assessment of airway size.
- The trachea primarily increases in length rather than diameter during the first 5 years of life.
- The tracheal division of the right and left bronchi is higher in a child's airway and at a different angle than the adult's.

Peculiarities of the respiratory system in children



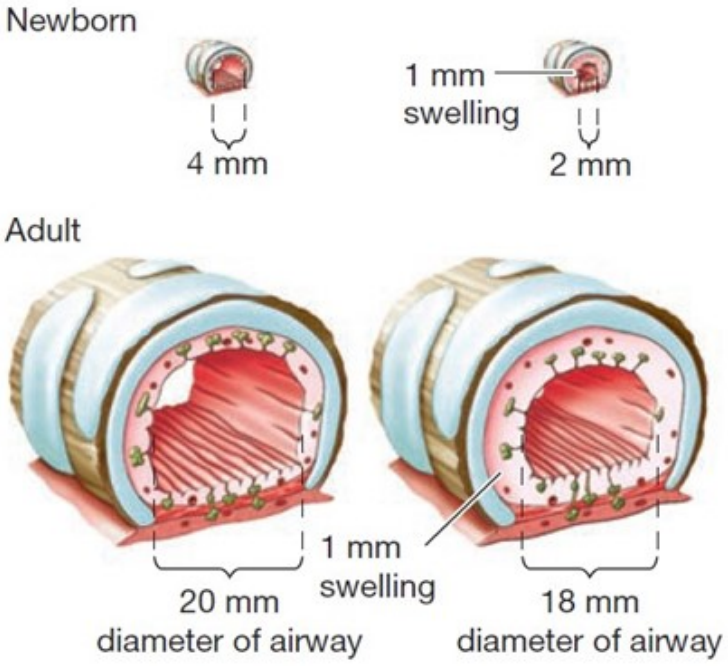
In children, the trachea is shorter and the angle of the right bronchus at bifurcation is more acute than in the adult. When you are resuscitating or suctioning, you must allow for these differences. Do you think that the angle of the right bronchus is significant in foreign-body aspiration? Why?

Peculiarities of the respiratory system in children

- The cartilage that supports the trachea is more flexible, and the airway may be compressed when the head and neck are flexed toward the chest.

- The child's narrower airway causes a greater increase in **airway resistance** (the effort or force needed to move oxygen through the trachea to the lungs) in any condition causing airway inflammation or edema

Peculiarities of the respiratory system in children



An infant's airway diameter is approximately 4 mm (0.16 in.), in contrast to the adult's 20–mm (0.8-in.) airway diameter. An inflammatory process in the airway causes swelling that narrows the airway, and airway resistance increases. Note that swelling of 1 mm (0.04 in.) reduces the infant's airway diameter to 2 mm (0.08 in.), but the adult's airway diameter is only narrowed to 18 mm (0.7 in.). Air must move more quickly in the infant's narrowed airway to get the needed amount of air into the lungs. The friction of the quickly moving air against the side of the airway increases airway resistance. The infant must use more effort to breathe and must breathe faster to get adequate oxygen.

Peculiarities of the respiratory system in children

- Newborns are obligatory nose breathers.
- The only time newborns breathe through the mouth is when they are crying.
- The coordination of mouth breathing is controlled by maturing neurologic pathways, and infants up to 2 to 3 months of age do not automatically open the mouth to breathe when the nose is obstructed.
- It is important to keep the newborn's nose patent for breathing and eating.

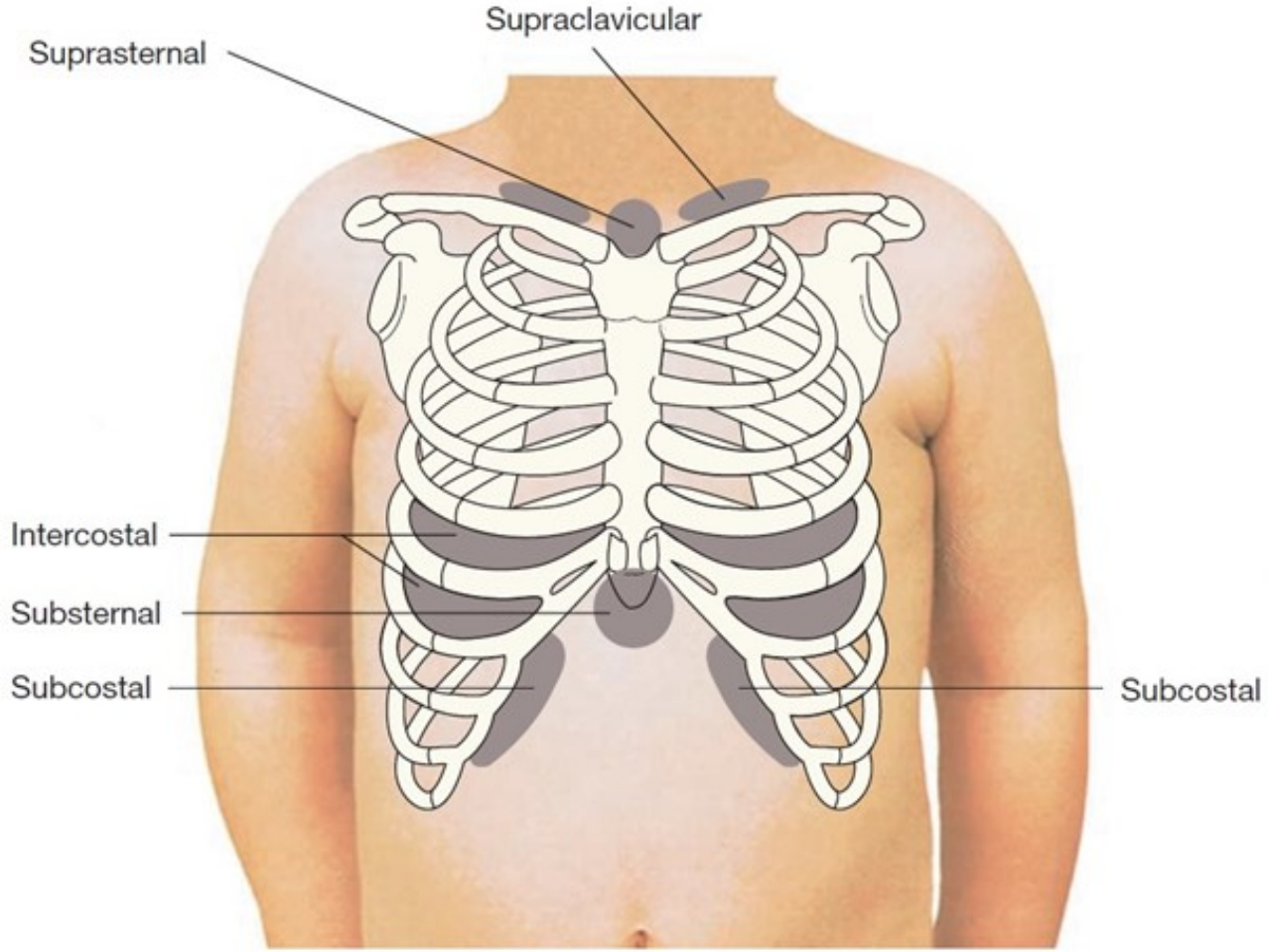
Peculiarities of the respiratory system in children

- The tracheobronchial tree is complete in the full-term newborn, but the child's lower airway is constantly growing. Beginning at 24 weeks' gestation, the lung sacs begin forming to support future gas exchange. The lung sacs begin differentiating into alveoli at 36 weeks' gestation (Rozance & Rosenberg, 2012).
- Alveoli continue developing and increasing in number for the first 5 to 8 years of age, followed by further development in size and complexity (Brashers & Huether, 2014).

Peculiarities of the respiratory system in children

- The bronchi and bronchioles are lined with smooth muscle that develops after birth.
- Children under 6 years of age use the diaphragm to breathe because the intercostal muscles are immature. By 6 years of age the child uses the intercostal muscles more effectively.
- The ribs are primarily cartilage and very flexible. In cases of respiratory distress, the negative pressure caused by the diaphragm movement causes the chest wall to be drawn inward, causing retractions.

Peculiarities of the respiratory system in children



Infants and young children have immature chest muscles and ribs of cartilage, making the chest wall very flexible. The negative pressure created by the downward movement of the diaphragm is increased in cases of respiratory distress, and the chest wall is pulled inward, causing retractions. Intercostal retractions are seen in mild respiratory distress. As respiratory distress severity increases, substernal and subcostal retractions are seen. In cases of severe distress, supraclavicular and suprasternal retractions occur as the accessory muscles (sternocleidomastoid and trapezius muscles) are used.

Activate W

Peculiarities of the respiratory system in children

- Children consume more oxygen than adults because of their higher metabolic rate.
- This rate of oxygen consumption increases when the child is in respiratory distress.
- The child also has fewer muscle glycogen reserves, leading to more rapid muscle fatigue when accessory muscles must be used for breathing (Brashers & Huether, 2014).

Peculiarities of the respiratory system in children

- Pediatric respiratory conditions may occur as a primary problem or as a complication of nonrespiratory conditions.

- Respiratory problems may result from structural problems, functional problems, or a combination of both.

- Structural problems involve alterations in the size and shape of parts of the respiratory tract.

Peculiarities of the respiratory system in children

- Functional problems involve alterations in gas exchange and threats to the process of ventilation due to irritation by large particles and chemicals or infection.

- Alterations in other organ systems, especially the immune and neurologic systems, may also threaten respiratory function.

History and physical examination of the respiratory tract in children

Introduction

- Respiratory disease is one of the most common reasons that pediatric patients seek medical attention.
- Signs and symptoms can be subtle, and a careful history and physical examination are always useful in assessment of pediatric patients with respiratory complaints.
- Diseases of the chest can be divided into two major categories: **acquired** and **congenital**.

Introduction

- **Congenital** chest diseases are often **symptomatic at all times** rather than episodically.
  - a child who has chronic noisy breathing from a congenital vascular ring, for example, is not as likely as the patient with asthma to have intermittent periods of wheezing with long intervals of normal breathing.

- The spectrum of diseases involving the pediatric respiratory system is primarily dependent on the age of the patient; therefore, **age must be a primary consideration** in the differential diagnosis.

History

- Each pediatric history should include the **perinatal history**:
  - ex. respiratory distress at birth or intubation
- **Prematurity** with prolonged need for supplemental oxygen may **suggest bronchopulmonary dysplasia** with associated structural lung abnormalities.
- **Noisy breathing starting early in life** suggests **congenital airway obstruction** and should be evaluated.

History

- Regardless of cause, **failure to thrive** is a worrisome finding, whereas excellent weight gain in a child with noisy breathing is reassuring.
- **Distinguishing between constant and intermittent symptoms** can be one of the most important means of diagnosing diseases of the pediatric chest.
- A good "**cough history**" and "**wheeze history**" are important and have similar elements.

History

- The clinician should inquire about the
  - chronicity of the symptoms
  - association with feeding
  - upper respiratory infections
  - exposures (pets, dust, and especially cigarette smoking are important), and
  - fevers

History

The nature of the cough is important:

- ***wet*** or ***dry***
- ***paroxysmal*** or ***continuous***, and
- ***staccato*** (as seen in neonatal chlamydial pneumonia)
- **Posttussive emesis** is a “red flag” to the clinician.
- The **cough that awakens the child at night** or keeps the child up much of the night is another worrisome historical finding.

History

- A **persistent cough that disappears in sleep** strongly suggests the diagnosis of **habit** (psychogenic) **cough**.

- In pursuing a history of ***wheeze,*** it is important to ask the parents or historians what they mean by the term; it may mean "noisy breathing," and it may even be applied to stridor.

History

- In evaluating the infant with frequent episodes of cough and/or wheeze, the clinician should inquire about symptoms and signs of gastroesophageal reflux (GER):
  - food refusal
  - arching
  - pain behaviors
  - frequent spitting
  - milk or formula found on the bed next to the infant's head in the morning
  - recurrent croup
  - hoarseness
  - laryngomalacia

Because reflux is worse when the patient is lying down, **symptoms** tend to be more **prominent at night and during naps.**

History

- A **family history of atopy** including eczema and environmental allergies should be investigated.
- In inquiring about **cystic fibrosis**, an autosomal recessive trait, an **extended family medical history** including grandparents and cousins should be taken.
- **Frequent infections** in parents or siblings, particularly those requiring hospitalization, suggest **possible immunodeficiency** in the family.
- **Immunization history** is essential in identifying patients at risk for pertussis.

History

- **Exercise intolerance** is one of the primary symptoms of respiratory disease:
  - the neonate's main output of energy is in feeding, and thus difficulties with feedings should be monitored;
  - toddlers are expected to keep up with peers and/or siblings in play;
  - the school-age child's gym performance should be scrutinized.
  - wheezing or coughing fits following vigorous exercise can occur in asthma.

Physical examination

- The infant or toddler is best examined with his or her shirt off while being held upright in the arms of a parent.

- The patient should face the parent; this maximizes contact with the parent and allows the patient to feel safe.

- The room should be at a comfortable temperature.

Physical examination

- The stethoscope head should be warmed in the clinician's hand or pocket for several minutes before use.

- The **classic four steps in the physical examination** – inspection, palpation, percussion, and auscultation—are well applied to the examination of the pediatric chest.

Inspection

- **Decreased subcutaneous adipose tissue** as seen in a cystic fibrosis patient should be noted.
- The **pattern of breathing** should always be evaluated with the child disrobed.

Any use of expiratory musculature is abnormal!

- **Suprasternal** and **intercostal retractions** reflect excessive negative pleural pressure and can be seen in normal children with thin chest walls after vigorous exercise.

Inspection

- **Subcostal retractions** are **always pathologic** and are the result of hyperinflated lungs and a flattened diaphragm pulling inward on the chest wall.

- In advanced lung disease, the use of accessory muscles of inspiration can be noted:
  - the sternocleidomastoid muscle, for example, helps lift the chest (in a "bucket handle" fashion) and increase its anteroposterior diameter, thereby increasing intrathoracic volume.

Inspection

- In **respiratory muscle fatigue**, a pattern of breathing can be observed in which the diaphragm alternates with the intercostal muscles to inflate the lungs – ***respiratory alternans*** and is seen as alternating abdominal and chest expansion instead of the usual pattern of simultaneous chest and abdominal expansion.

- Chest wall deformities such as **pectus excavatum** or **pectus carinatum** should be noted.

Palpation

Palpation of the chest can reveal significant findings:

- The examiner places the hands on either side of the chest as the patient takes a deep breath.

- The chest should expand symmetrically; **asymmetry** can be seen in unilateral pulmonary hypoplasia, mainstem bronchial obstruction, and diaphragmatic paresis.

Palpation

Palpation of the chest can reveal significant findings:

- Placing fingertips on the upper abdomen just over the insertion of the rectus muscles into the lower rib cage can reveal subtle use of expiratory muscles in children with peripheral (lower) airway obstruction.

- Similarly, the anterior lower ribs should be assessed with the fingertips.

Palpation

Palpation of the chest can reveal significant findings:

- In infants with **obstructive lung disease**, the **lower ribs** can be felt to **pull inwards on inspiration.** This is the palpable aspect of a subcostal retraction.

- With the patient's head in the midline position, the trachea should be palpated at the sternal notch to evaluate for tracheal deviation, as is seen with mediastinal shift.

Palpation

Palpation of the chest can reveal significant findings:

- Vocal fremitus should be assessed in patients with suspected pleural fluid accumulation
  - the vibrations transmitted from the larynx as the child says “99” are diminished when there is an accumulation of air or fluid in the pleural space.
- Infants and children with tracheomalacia and bronchomalacia often have a **palpable vibration in the back**.
  - palpable vibrations in only one hemithorax suggest a partial obstruction of the mainstem bronchus in that hemithorax as seen in bronchomalacia.

Percussion

- Percussion of the chest can reveal much more than hyperresonance and dullness over an area of consolidation.
- **Air trapping** is the hallmark of small airway disease and results in a **depressed position of the diaphragm**:
  - ordinarily the diaphragm can be found just at or slightly below the tip of the scapula when the patient's arm is at his or her side in children 5 years old and younger.
  - In the patient with hyperinflation, the diaphragm is found several fingerbreadths below the scapular tips. This finding, even in the absence of wheezing on auscultation, suggests a lesion of the small airways.

Percussion

- An **area of consolidation** or **pleural effusion** results in **dullness to percussion**.

- Another disorder causing **asymmetry of percussion** of the two hemithoraces is **diaphragmatic eventration**, which is a congenital lesion of the diaphragm in which the diaphragm is replaced with a thin fibrous membrane without contractile properties.

Auscultation

- Auscultation of the pediatric chest requires patience.
- One often must wait a minute or two for a deep breath in order to appreciate abnormal breath sounds that are not apparent on shallow breathing.
- Augmenting the expiratory phase with a gentle squeeze of the thorax while listening with the stethoscope may bring out expiratory wheezes.

Auscultation

- Abnormal ("adventitial") breath sounds include **crackles** and **wheezes**.

- Wheezes are *continuous* sounds, whereas crackles (formerly referred to as *rales*) are *discontinuous*.

- Wheezes and crackles can be inspiratory or expiratory, although crackles are more commonly heard on inspiration and wheezes are more commonly heard on expiration.

Wheezes

- Wheezes probably arise from the vibration within the walls of narrowed large and medium-sized airways.

- In a patient experiencing an acute exacerbation of asthma, the lungs have wheezes in a range of pitches (described as *polyphonic*) with substantial regional differences in auscultation.

Wheezes

- Patients with central airway obstruction such as tracheomalacia, on the other hand, have a single pitch of wheeze that sounds the same in all lung fields *(monophonic)* and is heard loudest over the central airway that is obstructed.

- Foreign bodies can cause a monophonic wheeze that can vary in pitch depending on the degree of obstruction.

Crackles

- Crackles are believed to arise from the popping of fluid menisci within airways.
- Coarse crackles are often audible at the mouth and are a late finding in cystic fibrosis patients with advanced bronchiectasis.

- ***Rhonchi*** refers to the sound made by pooled secretions in the central airways, which can be categorized as harsh, low-pitched central wheezes or coarse, central crackles (depending on the nature of the sounds heard).

Friction rubs

- Friction rubs are creaking sounds heard during both phases of respiration as inflamed pleural surfaces rub over one another.

- One of the most important abnormal findings in children is the **absence of breath sounds** over an area of **collapse** or **consolidation**.

Signs and symptoms of respiratory diseases in children

Cough

- Persistent or recurrent cough represents one of the most common and vexing problems in pediatrics.

- In most circumstances, the tracheobronchial tree is kept clean by airway macrophages and the mucociliary escalator, but cough becomes an important component of airway clearance when excessive or abnormal materials are present, or when mucociliary clearance is reduced, such as during a viral respiratory illness.

Cough

- A cough clears airway secretions and inhaled particulate matter through a combination of the high airflow velocities generated during the expiratory phase of the cough and compression of smaller airways, which “milks” the secretions into larger bronchi where they can be eliminated by a subsequent cough.
- Cough is generally produced by a reflex response arising from irritant receptors located in ciliated epithelia in the lower respiratory tract, but it can be suppressed or initiated at higher cortical centers.

Cough

- One of the most common causes of cough in pediatric patients is the self-limited cough of an acute viral lower respiratory illness or bronchitis that lasts 1 to 2 weeks.
- The cough that persists longer than 2 weeks is potentially more worrisome.
- A diagnostic approach to chronic cough is best served by considering the age of the child (*next slide*).

Causes of Cough According to Age

Infancy (Younger Than 1 Year Old)

Congenital and Neonatal Infections

Chlamydia
Viral (e.g., RSV, CMV, rubella)
Bacterial (e.g., pertussis)
Pneumocystis jiroveci

Congenital Malformations

Tracheoesophageal fistula
Vascular ring
Airway malformations (e.g., laryngeal cleft)
Pulmonary sequestration

Other

Cystic fibrosis
Asthma
Aspiration
Recurrent viral bronchiolitis/bronchitis
GER
Interstitial pneumonitides
Lymphoid interstitial pneumonitis
Diffuse interstitial pneumonitis

Causes of Cough According to Age

Preschool

Inhaled foreign body
Asthma
Suppurative lung disease
    Cystic fibrosis
    Bronchiectasis
Right middle lobe syndrome
Ciliary dyskinesia syndromes
Upper respiratory tract disease
Recurrent viral infection/bronchitis
Passive smoke inhalation
GER
Interstitial pneumonitides
Pulmonary hemosiderosis

Causes of Cough According to Age

School Age to Adolescence

Asthma
Cystic fibrosis
Mycoplasma pneumoniae infection
Psychogenic or habit cough
Cigarette smoking
Pulmonary hemosiderosis
Interstitial pneumonitides
Ciliary dyskinesia syndromes

Causes of Cough According to Age

All Ages

Recurrent viral illness
Asthma
Cystic fibrosis
Granulomatous lung disease
Foreign body aspiration
Pertussis infection

CMV, Cytomegalovirus; *GER,* gastroesophageal reflux; *RSV,* respiratory syncytial virus.

Causes of persistent cough

Several causes of persistent cough are **common to all pediatric age groups**:

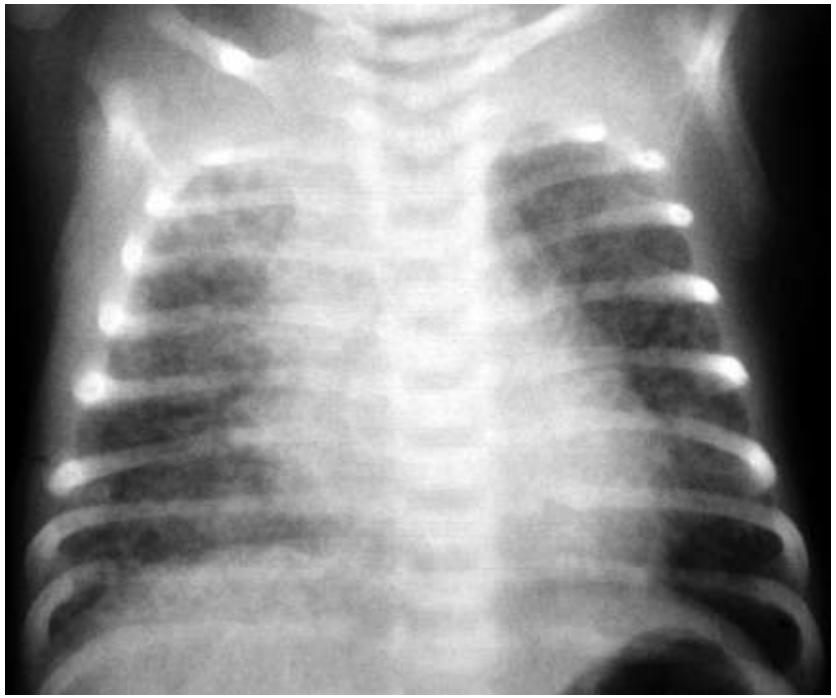
- second-hand cigarette smoke exposure
- recurrent viral bronchitis
- Asthma
- GERD
- cystic fibrosis
- granulomatous lung disease (e.g., tuberculosis)
- foreign body aspiration
- and pertussis

Cough in Infancy (< 1 y.o.)

- Cough starting at birth or shortly afterward may be a sign of serious respiratory disease and must be evaluated assiduously.
- Cough beginning at this time raises the possibility of **congenital infections** (cytomegalovirus or rubella, which are often associated with other findings, such as
  - ✓ hepatosplenomegaly
  - ✓ thrombocytopenia
  - ✓ central nervous system disease

Cough in Infancy (< 1 y.o.)

- Pneumonia due to *Chlamydia trachomatis* generally develops after the first month of life and presents with
  - ✓ afebrile pneumonitis with congestion
  - ✓ wheezing
  - ✓ fine, diffuse crackles
  - ✓ a paroxysmal cough
  - ✓ in approximately 50% of cases, a prior or concomitant inclusion conjunctivitis



Pneumonia caused by *Chlamydia trachomatis* in a 3-month-old infant with inclusion conjunctivitis

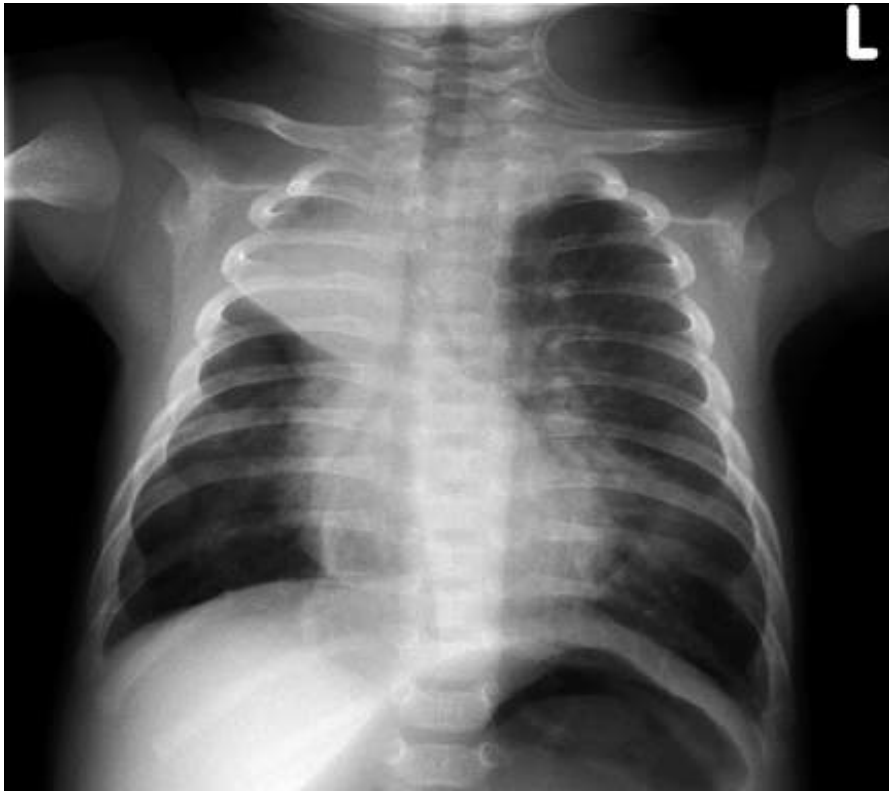
Cough in Infancy (< 1 y.o.)

- Pneumonia caused by *Bordetella pertussis* is a potentially life-threatening illness characterized by severe paroxysmal coughing episodes followed by cyanosis and apnea and is often associated with an inspiratory "whoop."
- The latter finding may be missing in young infants or those weakened by the recurrent coughing spasms.
- Newborns and young infants may have apnea as the primary sign of a *B. pertussis* infection.

Cough in Infancy (< 1 y.o.)

- The chest radiograph is nondiagnostic and can be normal or show

  - ✓ perihilar infiltrates
  - ✓ atelectasis
  - ✓ hyperinflation
  - ✓ interstitial or subcutaneous emphysema in some cases



Pertussis in a 6-week-old infant demonstrates the typical radiographic pattern of perihilar involvement. This child also has right upper lobe atelectasis

Cough in Infancy (< 1 y.o.)

- *Ureaplasma urealyticum* and *Pneumocystis jiroveci* (formerly known as *Pneumocystis carinii*) have been recognized as causes of pneumonia and **persistent cough** in this age group.

Cough in Infancy (< 1 y.o.)

- **Chronic aspiration** is a cause of respiratory symptoms that should be considered, particularly in medically complex infants.

- Common symptoms of chronic aspiration include chronic cough, wheezing, congestion, choking or gagging with feeding, failure to thrive, apnea, intermittent fever, and recurrent pneumonia.

Cough in Infancy (< 1 y.o.)

- Aspiration may be due to neurologic, anatomic, or functional disorders, often with a combination of factors present.

- Infants with neurologic disorders may have incoordination of swallowing and sucking reflexes that lead to aspiration of milk or gastric contents, as well as saliva into the lung.

Cough in Infancy (< 1 y.o.)

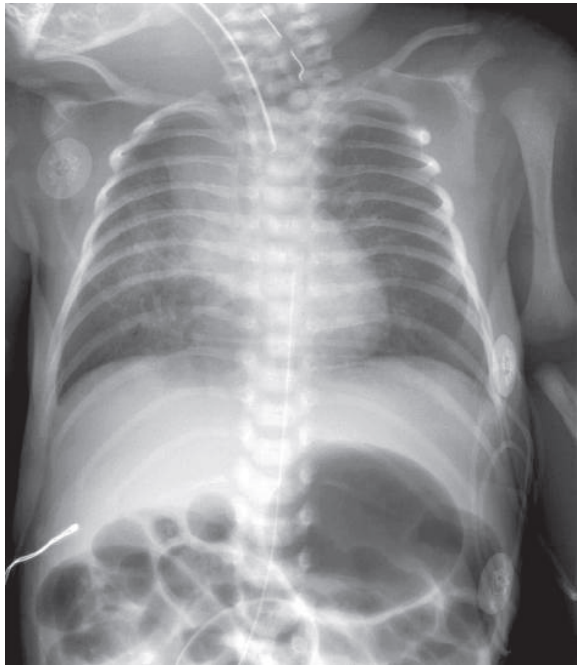
- **Anatomic malformations** (such as, a tracheoesophageal fistula, laryngeal cleft, or web) can produce cough via chronic aspiration.

- These anomalies are associated with **feeding-related coughing**, **choking**, and occasionally **cyanosis or persistent hypoxemia**.

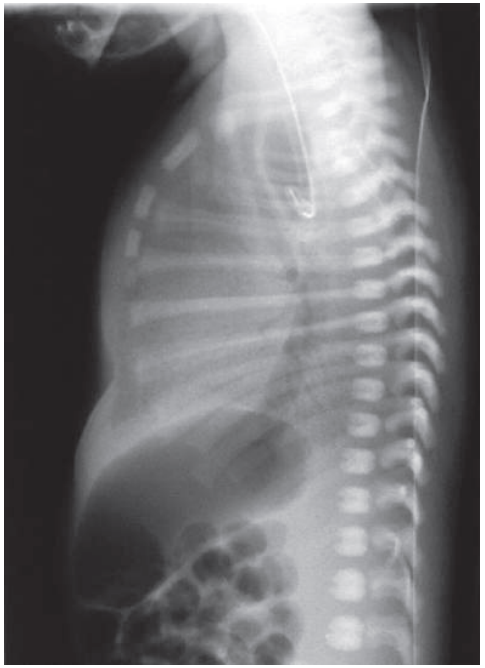
- Dysfunctional swallow may be present with or without anatomic abnormality.

Cough in Infancy (< 1 y.o.)

Tracheoesophageal fistula



A, Anteroposterior chest radiograph shows feeding tube passing no farther than proximal esophagus; there is an aspiration pneumonitis present.



B, Lateral view showing the feeding tube in the proximal esophageal pouch with air in the airway, distal esophagus, and intestine.

Cough in Infancy (< 1 y.o.)

- **Congenital thoracic malformations** may also present as persistent or recurrent cough, wheeze, or pneumonias.

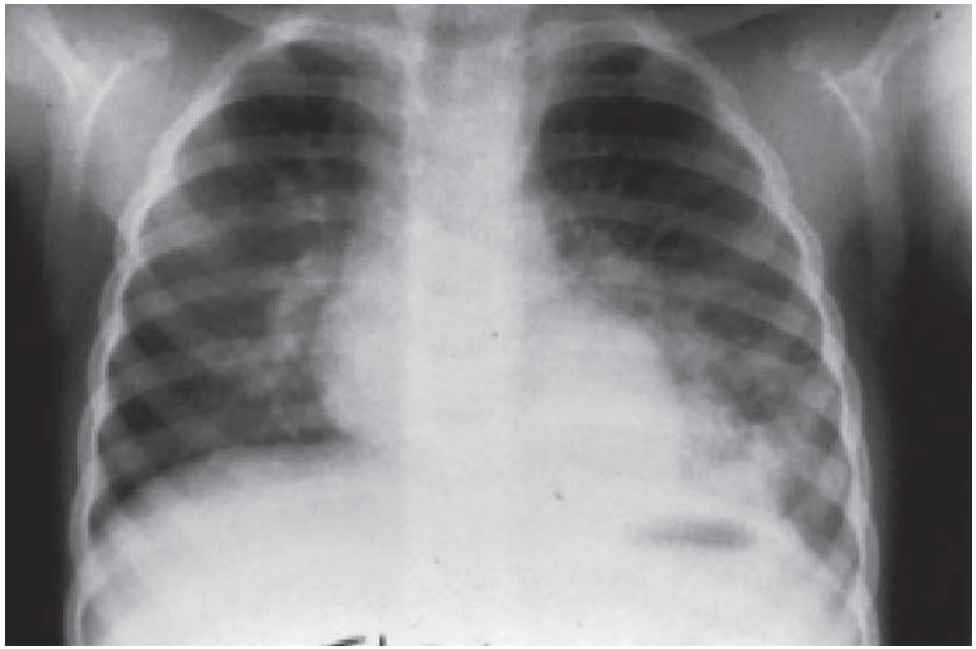
- Diagnoses within this category include:
  - ✓ bronchial atresias or hypoplasia
  - ✓ bronchogenic cysts
  - ✓ congenital pulmonary adenomatoid malformations
  - ✓ sequestrations

Cough in Infancy (< 1 y.o.)

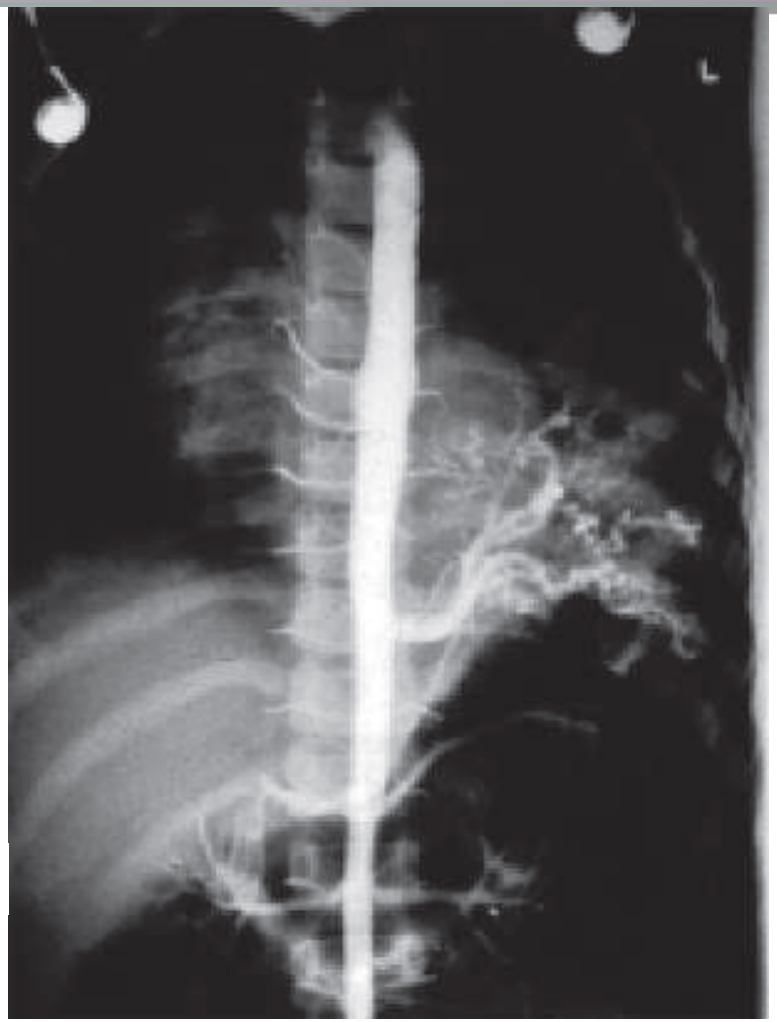
- **Pulmonary sequestration** (in which a portion of the lung is perfused by systemic, not pulmonary, arteries) and **bronchogenic cysts** (cystic structures arising from the pulmonary epithelium) are rare congenital anomalies that may compress the pulmonary tree or become infected, thereby producing a cough.

Cough in Infancy (< 1 y.o.)

Pulmonary sequestration



A, Anteroposterior film shows left lower lobe infiltrate.



B, Aortic angiogram demonstrates anomalous origin of pulmonary blood supply from abdominal aorta to the left lower lobe in a 7-year-old girl with extralobar sequestration.

Cough in Infancy (< 1 y.o.)

- A further structural consideration is **vascular anomalies** such as:
  - ✓ right-sided or double aortic arch
  - ✓ innominate artery compression
  - ✓ pulmonary artery slings

- Aberrant major blood vessels generally cause ***inspiratory stridor*** and ***expiratory wheezing*** from tracheal compression, but a brassy cough may also be observed, as may ***dysphagia*** from the associated esophageal compression.

Cough in Infancy (< 1 y.o.)

- The triad of poor weight gain, steatorrhea, and chronic cough at this age makes cystic fibrosis a strong consideration, and a sweat test at an accredited cystic fibrosis center is mandatory.
- Asthma (formerly called *reactive airway disease*) or bronchial hyperresponsiveness is a common and probably underdiagnosed cause of cough in infancy.
  - Cough or persistent wheezing can be found in these infants, who may have a history of a previous viral lower respiratory illness with or without a family history of wheezing and/or asthma.

Cough in Infancy (< 1 y.o.)

- Babies with **Gastro Esophageal Reflux** may have a combination of effortless vomiting; nocturnal cough/wheeze; pain behaviors/arching; hoarseness; laryngomalacia; and, in some cases, poor weight gain.

- The absence of a history of vomiting ("spitting up") does not eliminate GER as a diagnostic consideration in infants with persistent coughing, because occult reflux or microaspiration may induce bronchospasm.

Cough in Infancy (< 1 y.o.)

- **Childhood interstitial lung diseases** (chILDs) are a complex and rare group of pulmonary disorders.
- These disorders usually involve the pulmonary interstitium but can involve other aspects of lung parenchyma.
  - Causes of chILD are extremely variable and include infections, inhalation injury, chemotherapeutic agents, post–bone marrow transplant lung disease, systemic inflammatory diseases, pulmonary hemorrhage syndromes, structural and growth anomalies, metabolic diseases, and congenital disorders of host defense and of surfactant production.

Cough in Infancy (< 1 y.o.)

- Examples of chILD that present in infancy include
  - alveolar capillary dysplasia
  - surfactant B and C deficiencies
  - ILD associated with *ABCA3* mutations
  - pulmonary interstitial glycogenosis
  - neuroendocrine cell hyperplasia of infancy
  - follicular bronchitis of infancy

Cough in Infancy (< 1 y.o.)

- Patients with chILD may present insidiously with some combination of
  - cough
  - tachypnea
  - retractions
  - exercise intolerance
  - resting hypoxemia or hypercarbia
  - desaturation with exercise
  - diffuse abnormalities on chest imaging
  - crackles and retractions on examination
  - growth failure

Cough in Infancy (< 1 y.o.)

- The diagnosis of a specific chILD is usually made following **lung biopsy**, but this is usually preceded by a variety of less invasive tests, such as
  - high resolution **computed tomography** (CT) scan of the chest
  - infant **lung function testing**
  - **flexible fiberoptic bronchoscopy with bronchoalveolar lavage**
- Surfactant disorders (surfactant protein B or C deficiencies, or *ABCA3* mutations) can often be diagnosed by **mutation analysis**.

Cough in Infancy (< 1 y.o.)

- The prognosis of chILD can be quite variable:
  - universally fatal (alveolar-capillary dysplasia)
  - very severe and treatable only by lung transplantation (surfactant protein B deficiency)
  - gradual improvement over months or years (neuroendocrine cell hyperplasia of infancy).

Cough in Preschool

- **Most common** reasons for a persistent cough in this age group are **recurrent viral infections** and **asthma**.

- The child with **asthma** may not manifest audible wheezing or dyspnea but rather may have persistent cough, especially with viral respiratory infections, following exposure to noxious inhalants, such as cigarette smoke, or following vigorous activity.

Cough in Preschool

- **Upper respiratory tract disease and sinusitis** have been implicated in the pathogenesis of chronic cough, presumably through the stimulation of pharyngeal cough receptors by upper airway secretions.
- **Parental smoking** (passive smoking) itself is a common cause of cough in preschool children.
- **GER** more commonly causes cough at a younger age but may appear at any age.

Cough in Preschool

- An **inhaled foreign body** in either the tracheobronchial tree or esophagus is an important cause of chronic cough, especially in toddlers.

- A **history of gagging** or **choking** may be absent at this age, physical examination may be unrevealing, and the plain chest radiograph may be normal.

Cough in Preschool

Inhaled foreign body

- Cough is present in more than 90% of cases.

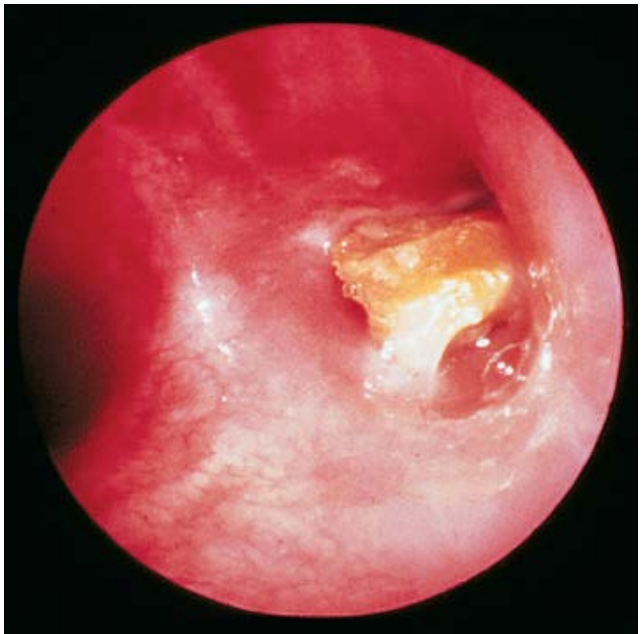
- Cough is usually of abrupt onset, but a quiescent period may occur after inhalation and cough may disappear as irritant receptors adjust to the object's presence.

- A mobile foreign body may result in the recurrence of cough as new receptors are stimulated by the object.

Cough in Preschool

Inhaled foreign body

- Although inspiratory and expiratory radiography and fluoroscopy are useful in the evaluation of a child with a possible bronchial foreign body, they may be normal and **rigid bronchoscopy** may be necessary to confirm or disprove the presence of a foreign object.

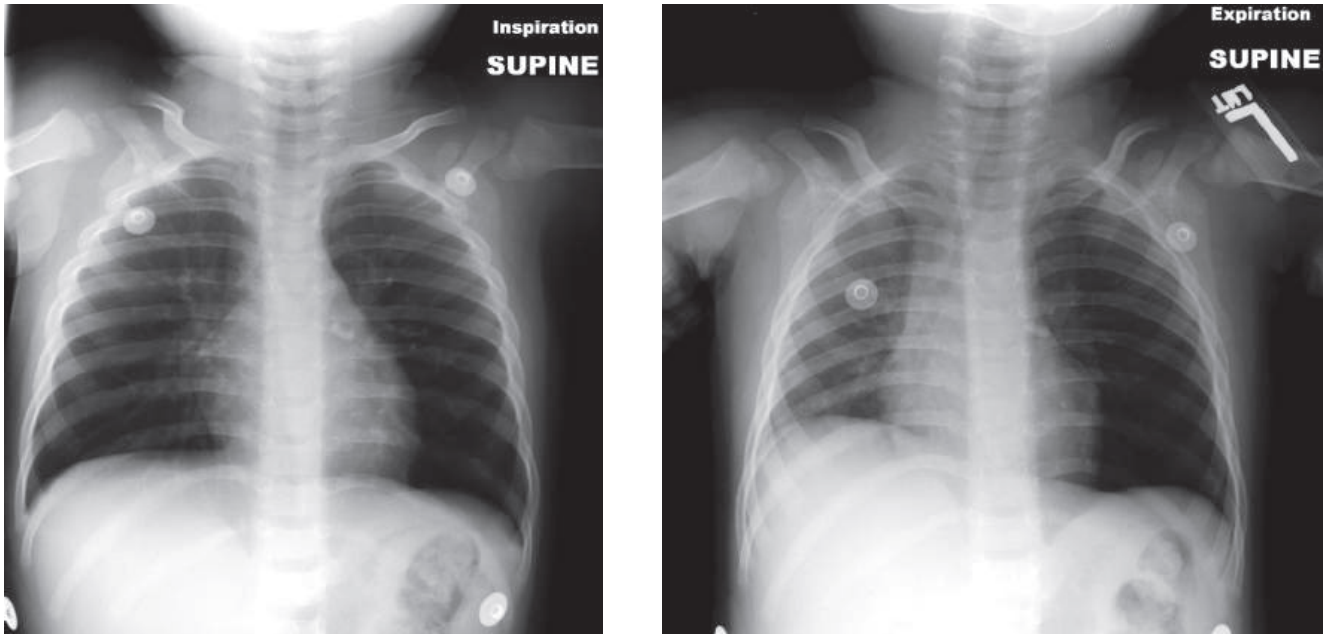


Foreign body. Portion of a carrot lodged in the right mainstem bronchus, as seen through a rigid bronchoscope

Cough in Preschool

Inhaled foreign body

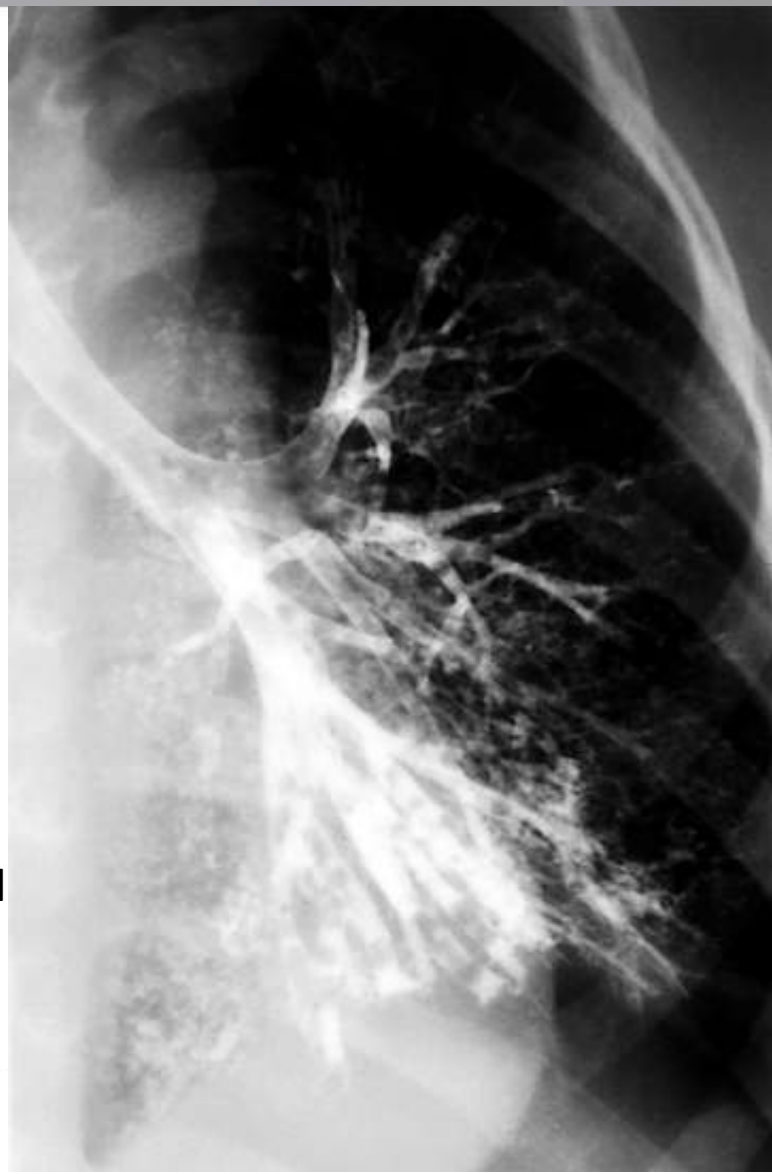
- Unilateral air trapping demonstrated by **inspiratory and expiratory radiographs** (or **left and right lateral decubitus films in younger children**) strongly suggests an inhaled foreign body.



Foreign body. Inspiratory **(A)** and expiratory **(B)** radiographs in a child with an inhaled foreign body lodged in the left mainstem bronchus reveals hyperlucency of the left hemithorax and compensatory shift of the mediastinal structures to the right on expiration.

Cough in Preschool

- Suppurative lung diseases, such as **cystic fibrosis** or **bronchiectasis** from any other causes (e.g., tuberculosis), characteristically result in a chronic cough producing purulent sputum.



Bronchiectasis. Bronchogram shows cylindrical bronchiectasis of the left lower lobe in a 5-year-old girl with recurrent pneumonia and chronic cough.

Cough in Preschool

- "**Right middle lobe syndrome**," which is commonly associated with enlargement of lymph nodes surrounding the right middle lobe bronchus in tuberculosis, has also been described in asthma and a number of other illnesses and may be associated with chronic cough.

- Recurrent infection of the middle lobe can ultimately lead to the development of bronchiectasis or fibrosis.

Cough in Preschool

- **Disorders of ciliary motility** (primary ciliary dyskinesia and acquired ciliary dyskinesia) may produce insidious symptoms of chronic productive cough, nasal drainage, recurrent middle ear infections, and fever.
  - Primary ciliary dyskinesia is most often transmitted in an autosomal recessive inheritance pattern.
  - Genetic characteristics are quite heterogenous, with 31 genes presently identified as disease causing.

Cough in Preschool

Disorders of ciliary motility

- Clinical findings include **basilar crackles** (which can be expiratory) and, later, **radiographic changes** of recurrent lower lobe infections and bronchiectasis.
  - Repetitive infections occur unless measures (such as, chest physical therapy, postural drainage, and liberal use of antibiotics) are employed.
  - It is now recognized that the classic triad described by Kartagener of situs inversus, sinusitis, and bronchiectasis fits only a limited number of patients, because situs inversus occurs in only about half of all patients with primary cilia dyskinesia.

Cough in Preschool

Disorders of ciliary motility

- Far more common is an acquired ciliary dyskinesia that can follow certain lower respiratory infections (including adenovirus, *Mycoplasma,* respiratory syncytial virus, and influenza).
  - Diagnosis can be made via **biopsy of the respiratory epithelium**, either from curettage of the nasal turbinate in the office or forceps biopsy of the bronchus via rigid bronchoscope under anesthesia.
  - It may also be suggested by a **reduced fraction of nitric oxide in exhalate from the nose**.

Cough in School Age to Adolescence

Most frequent causes:

- recurrent viral infection
- asthma
- allergic rhinosinusitis
- cystic fibrosis
- pulmonary hemosiderosis
- interstitial pneumonitis
- primary ciliary dyskinesia

Characteristics of Chronic Cough and Associated Conditions

Characteristic	Associated Condition
Loose, productive	Cystic fibrosis, bronchiectasis, ciliary dyskinesia
Croupy	Laryngotracheobronchitis
Paroxysmal	Cystic fibrosis, pertussis syndrome, foreign body inhalation, *Mycoplasma, Chlamydia*
Brassy	Tracheitis, upper airway drainage, psychogenic cough
After feedings	Pharyngeal incoordination, pharyngeal mass, tracheoesophageal fistula, GER
Nocturnal	Upper respiratory tract disease, sinusitis, asthma, cystic fibrosis, GER
Most severe in morning	Cystic fibrosis, bronchiectasis
With exercise	Asthma (including exercise induced), cystic fibrosis, bronchiectasis
Loud, honking, or bizarre	Psychogenic cough
Disappears with sleep	Psychogenic cough

GER, Gastroesophageal reflux.

Diagnostic Approach to Cough

- Complete history and physical examination
- Chest and sinus radiographs
- CBC with differential
- Pulmonary function tests (including bronchoprovocation tests)
- Sweat test (pilocarpine iontophoresis method)
- Trial of bronchodilators
- Sputum for Gram stain, AFB (Acid-fast bacillus), bacterial, viral, and fungal cultures
- Quantitative immunoglobulins
- Tuberculin skin test/anergy panel
- Serologic tests or PCR for *Mycoplasma pneumoniae*
- Bronchoscopy with bronchoalveolar lavage
- Barium swallow

Stridor

- Stridor is characteristically a harsh inspiratory noise created by obstruction of the larynx or the extrathoracic trachea.

- With a mild degree of airway narrowing, breath sounds may be normal when the infant or child is at rest, but with any activity that increases tidal breathing (e.g., crying, feeding, agitation), inspiratory stridor may become noticeable.

Causes of Recurrent or Chronic Stridor

- Croup
  - Infectious
  - Allergic/angioneurotic edema, GER
- Laryngomalacia
- Tracheomalacia
- Subglottic stenosis
- Extrinsic airway compression
  - Vascular ring
  - Mediastinal mass
  - Lobar emphysema
  - Bronchogenic cyst
  - Foreign body in esophagus
  - Thyromegaly
- Pharyngeal or laryngeal masses
  - Papilloma
  - Hemangioma
  - Laryngocele
  - Web
  - Foreign body
- Tracheoesophageal fistula
- Vocal cord paralysis
- Psychogenic

GER, Gastroesophageal reflux.

Stridor

- The most common cause of inspiratory stridor in the pediatric population is **infectious croup** (acute laryngotracheobronchitis).

- This disease is most commonly caused by a respiratory virus (parainfluenza, respiratory syncytial, influenza, or rhinovirus), and the patient typically has coryza for 24 to 48 hours before the appearance of croupy cough, hoarseness, and stridor.

Stridor

- Occasionally the inflammatory process may spread to the smaller airways and produce wheezing in addition to these symptoms.
- The “steeple sign” is a characteristic radiographic sign on anteroposterior projections that may be accompanied by marked dilation of supraglottic structures, particularly on lateral films.
- In the majority of patients, serious airway obstruction does not occur and the disease is self-limited.

Stridor

- **Acute angioneurotic edema** is a less common cause of stridor. In most cases, it results from an allergic reaction and is potentially fatal.

- **Gastro Esophageal Reflux**: some children with anatomically normal airways suffer recurrent bouts of stridor, usually in the middle of the night, in the absence of signs of viral infection.
  - Treatment for GER is often helpful in these patients, suggesting that for many, occult GER explains these bouts of recurrent airway obstruction.

Stridor

- The stridor associated with **congenital laryngomalacia** generally begins within the first week of life, varies with activity, and is more noticeable in the supine position.
- Clinical symptoms may suggest the diagnosis; but if severe, bronchoscopic visualization of airway dynamics by flexible bronchoscopy is a safe and reliable method of excluding other causes of stridor.
- Parents can be reassured that this entity is self-limited, becomes less marked after 6 to 10 months of age, and rarely causes serious problems.

Stridor

- Narrowing of the subglottic region can be congenital or acquired, such as in **subglottic stenosis associated with endotracheal intubation**.

- **Congenital subglottic stenosis** improves as the child grows older, but narrowing associated with tracheal intubation may require a tracheostomy, particularly if the infant remains dependent on ventilatory support.

Stridor

- **Congenital laryngeal or pharyngeal masses** can also produce stridor by obstructing airflow.
- Although inspiratory stridor may be observed, hoarseness is a more common presenting feature.

- **Laryngeal webs**, **papillomas**, or **hemangiomas** may all present with ongoing stridor, as well as brassy or dry cough.
- Hemangiomas of the larynx or trachea may also produce stridor or a brassy or dry cough.

Wheezing

- Wheezing is a continuous sound that results from obstruction of airflow in intrathoracic airways.
- This obstruction can be at the lower trachea “downstream” to the small bronchi and bronchioles.
- Wheezes can be heard on expiration or, less commonly, during both phases of respiration.

Wheezing

- The pitch of the wheeze, the variation in its pitch throughout the lung fields, and an association with hyperinflation as defined by percussion (described earlier) can help differentiate wheezing resulting from obstruction in the small airways (polyphonic) from that in the large airways (monophonic).

Wheezing

- Response to bronchodilator and/or steroids is a useful way of differentiating true asthma (which should improve with these treatments) from wheezing resulting from tracheomalacia or bronchomalacia (which does not improve and may even worsen with bronchodilators).

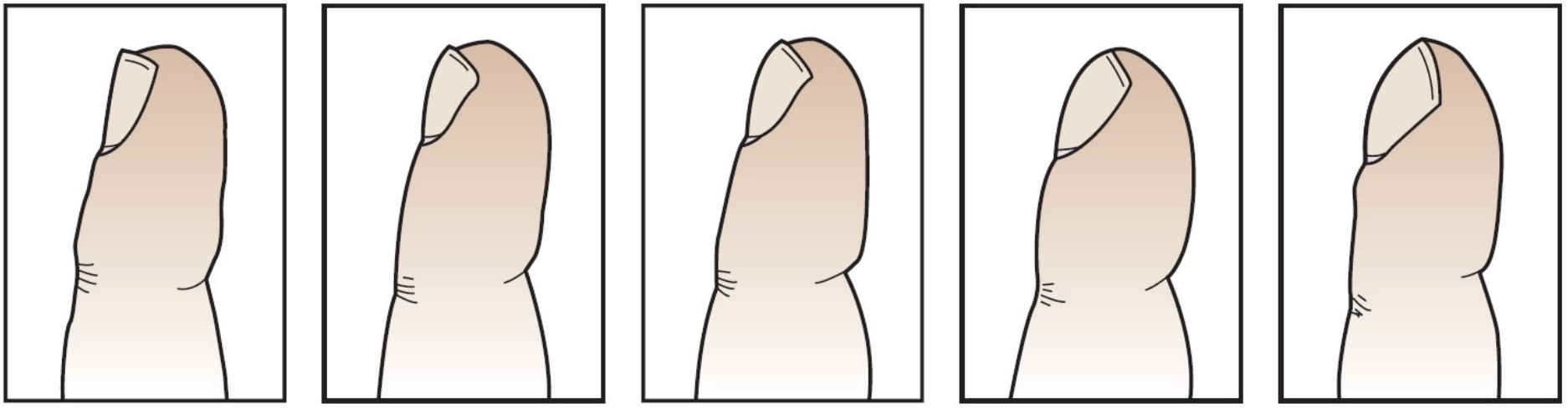
Causes of Chronic or Recurrent Wheezing

- Asthma
  - Exercise-induced asthma
- GER
- Hypersensitivity reactions (e.g., ABPA)
- Cystic fibrosis
- Aspiration
  - Tracheoesophageal fistula
  - Foreign body
  - GER
  - Laryngeal cleft
  - Pharyngeal dysmotility
- Extrinsic masses
  - Vascular ring
  - Cystic adenomatoid malformation
  - Lymph nodes
  - Tumors
- Ciliary dyskinesia syndromes
- Tracheomalacia and/or bronchomalacia
- Congestive heart failure
- Bronchopulmonary hemosiderosis or Heiner syndrome
- Endobronchial lesions including localized stenosis
- Interstitial pneumonitides
- Bronchiolitis obliterans

ABPA, Allergic bronchopulmonary aspergillosis; *GER,* gastroesophageal reflux.

Digital clubbing

- Digital clubbing may point to the presence of lung disease. There are various stages of clubbing, from mild to severe:



Digital clubbing. The 0- to 4-point scale describes the spectrum of digital clubbing as follows:
1+, very mild; 2+, mild; 3+, moderate; and 4+, severe.

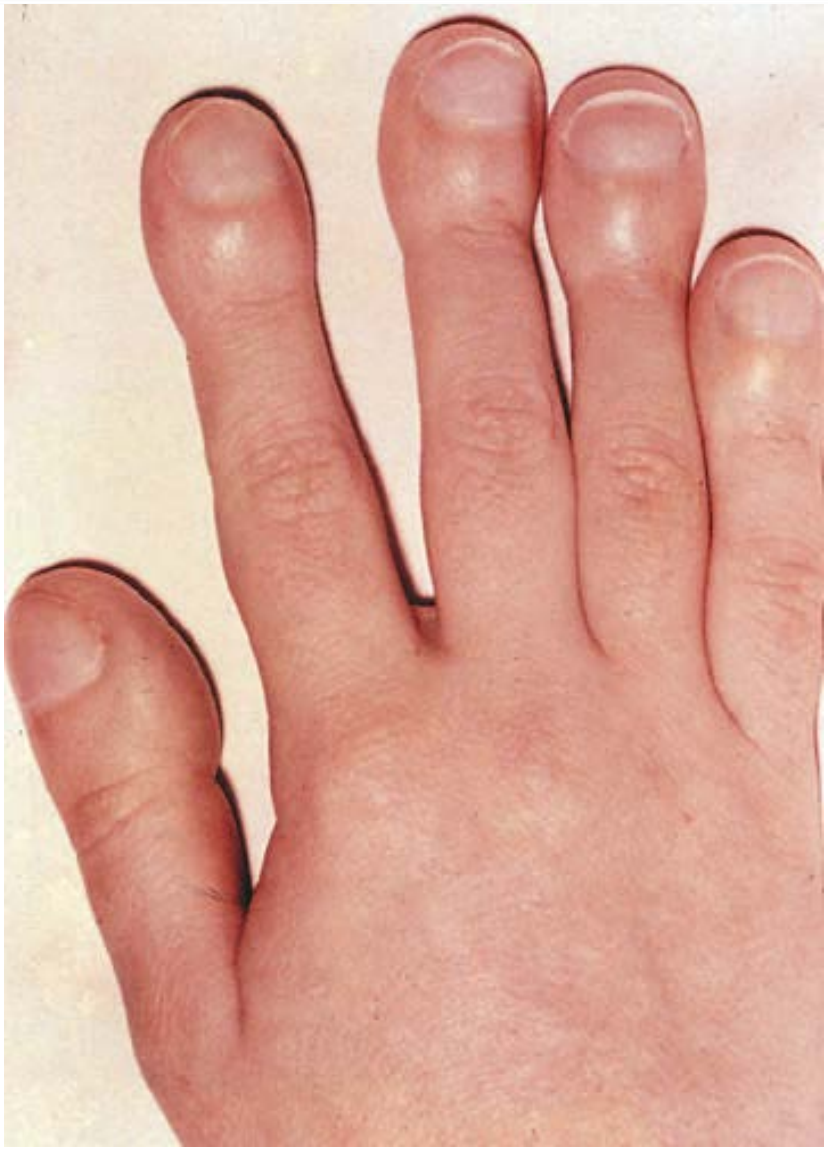
Digital clubbing

Causes of Clubbing

- Pulmonary
  - Cystic fibrosis
  - Other bronchiectasis
  - Pulmonary abscess
  - Empyema
  - Neoplasms
  - Interstitial fibrosis
  - Pulmonary alveolar proteinosis
  - Interstitial pneumonitis
  - Chronic pneumonia
- Cardiac
  - Cyanotic congenital heart disease
  - Subacute bacterial endocarditis
- Gastrointestinal or hepatic
  - Ulcerative colitis
  - Crohn disease
  - Polyposis
  - Biliary cirrhosis/atresia
- Familial
- Thyrotoxicosis

Digital clubbing

- Bronchiectasis from **cystic fibrosis** or from other chronic infectious causes is the major cause of clubbing among all pulmonary diseases.



The reminding of the physical examination

- **Nasal polyps** can be associated with cystic fibrosis, triad asthma, or significant atopy.

- An **increased second heart sound** could suggest pulmonary hypertension.

ASSESSMENT GUIDE | The Child in Respiratory Distress*

Assessment Focus	Assessment Guideline
Position of comfort	• Is the child comfortable lying down? • Does the child prefer to sit up or in the **tripod position** (sitting forward with arms on knees for support and extending the neck)?
Vital signs	• Assess the rate and depth of respirations. See Table 5–9 for age-related respiratory rates. Is **tachypnea** (abnormally rapid respiratory rate) present? • Assess the pulse for rate and rhythm. See Table 5–11 for age-related heart rates.
Lung auscultation	• Are breath sounds bilateral, diminished, or absent? • Are **adventitious sounds** (wheezes, crackles, or rhonchi) present?
Respiratory effort (work of breathing)	• Is **stridor** (audible crow-like inspiratory and expiratory breath sounds) or wheezing present? Is grunting heard on expiration? • Is breathing easy or labored? • Are retractions present or are accessory muscles used to breathe? • Is nasal flaring present? • Can the child say a full sentence or is a breath needed every few words? Is the cry strong or weak? • Do the chest and abdomen rise simultaneously with inspiration or is **paradoxical breathing** present in which the chest and abdomen do not rise simultaneously?
Color	• What is the color of the mucous membranes, nail beds, or skin (pink, pale, cyanotic, or mottled)? • Does crying improve or worsen the color?
Cough	• Is the cough dry (nonproductive), wet (productive, mucousy), brassy (noisy, musical), or croupy (barking, seal-like)? • Is the coughing effort forceful or weak?
Behavior change	• Is irritability, restlessness, or change in level of responsiveness present?

Act
Go t

Clinical Manifestations of Respiratory Failure and Imminent Respiratory Arrest

PHYSIOLOGIC CAUSE	CLINICAL MANIFESTATIONS
INITIAL SIGNS OF RESPIRATORY FAILURE	
The child is trying to compensate for oxygen deficit and airway blockage. Oxygen supply is inadequate; behavior and vital signs reflect compensation and beginning hypoxia.	Restlessness Tachypnea Tachycardia Diaphoresis
EARLY DECOMPENSATION	
The child tries to use accessory muscles to assist oxygen intake; hypoxia persists and efforts now waste more oxygen than is obtained.	Nasal flaring Retractions Grunting Wheezing Anxiety, irritability Mood changes Headache Hypertension Confusion
SEVERE HYPOXIA AND IMMINENT RESPIRATORY ARREST	
The oxygen deficit is overwhelming and beyond spontaneous recovery. Cerebral oxygenation is dramatically affected; central nervous system changes are ominous.	Dyspnea Bradycardia Cyanosis Stupor and coma

Activate
Go to Setti

Diagnostic and Laboratory Procedures/Tests for the Respiratory System

Diagnostic and Laboratory Procedures/ Tests for the Respiratory System

DIAGNOSTIC PROCEDURES	LABORATORY TESTS
Bronchoscopy	Arterial blood gas analysis
Chest radiograph	Cultures
Polysomnography (sleep study)	Neonatal screening for cystic fibrosis
Pulse oximetry	Protein-purified derivative (PPD), the Mantoux test
Spirometry (pulmonary function tests)	
Sweat chloride test	

* See Appendices D and E for information about these diagnostic procedures and tests.

Radiology

- The width of the chest on the lateral projection in the chest radiograph of a normal infant is about the same as the transverse dimension on a frontal projection, and the lungs may appear relatively radiolucent.

- Further, in contrast with the older child (>2 years old), the cardiothoracic ratio in the infant normally may be as high as 0.65.

Radiology

- The width of the superior mediastinum at this age may also be striking because the thymic shadow is particularly prominent during the first few months of life before the normal process of involution occurs.

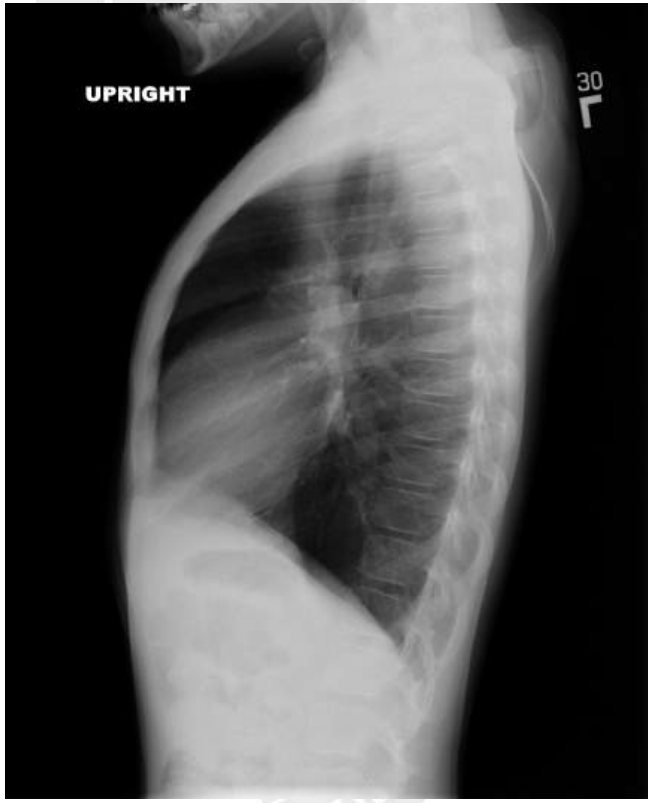
- A normal chest radiograph of an older child should show the diaphragm on an inspiratory film at the eighth or ninth rib posteriorly (sixth rib anteriorly), a cardiothoracic ratio of 0.5, and pulmonary vessels extending two-thirds of the way to the periphery.

Radiology

- In most situations, a lateral radiograph should accompany the posteroanterior (PA) view because some pathologic findings may be missed on a single projection.
  - a lateral examination yields the best information about the anterior mediastinum and the tracheal air column and may reveal a small pleural effusion that is unsuspected on the basis of a PA radiograph alone

Radiology

- In combination with the PA view, the lateral projection may help localize an abnormal finding to a particular lobe or segment or document hyperinflation with diaphragmatic flattening.



- In most situations, the chest radiograph taken at full inspiration is most helpful.

Radiology

- In the evaluation for bronchial foreign bodies, a comparison of inspiratory and expiratory views (or left and right lateral decubitus films in the younger patient) can help if one lung is unable to empty.

- In looking for a small pneumothorax, the expiratory film is more helpful because the smaller lung volume allows extrapulmonary air to expand to become more evident.