RECURRENT CHEST INFECTIONS

Mosab Abushqeer
Mohammed Salameh
Sae'd Al-Hamawi

Definition

- Recurrent pneumonia is defined as 2 or more episodes in a single year, or 3 or more episodes ever, with radiographic clearing between occurrences.
- Children with recurrent pulmonary infiltrate may be febrile or afebrile and may display a wide range of respiratory symptoms and signs.
- An underlying disorder should be considered if a child experiences recurrent pneumonia.

Differential diagnosis

- Foreign body
- Aspiration syndromm
- Asthma
- Immunodeficiency
- Cystic fibrosis
- Congenital anomalies
- Tuberculosis

Table 400-4	Differential Diagnosis of Recurrent Pneumonia
HEREDITARY DISORDERS	
Cystic fibrosis	
Sickle cell disease	
DISORDERS OF IMMUNITY	
HIV/AIDS	
Bruton agammaglobulinemia	
Selective immunoglobulin G subclass deficiencies	
Common variable immunodeficiency syndrome	
Severe combined immunodeficiency syndrome	
Chronic granulomatous disease	
Hyperimmunoglobulin E syndromes	
Leukocyte adhesion defect	
DISORDERS OF CILIA	
Immotile cilia syndrome	
Kartagener syndrome	
ANATOMIC DISORDERS	
Pulmonary sequestration	
Lobar emphysema	
Gastroesophageal reflux	
Foreign body	
Tracheoesophageal fistula (H type)	
Bronchiectasis	
Aspiration (oropharyngeal incoordination)	
Aberrant bronchus	

From Kliegman RM, Marcdante KJ, Jenson HJ, et al: Nelson essentials of pediatrics, *ed 5, Philadelphia, 2006, Elsevier, p. 507.*

Foreign body

Epidemiology and Etiology

- foreign body aspiration (FBA) is a potentially life-threatening event.
- The majority of children who aspirate foreign bodies are under 4 years of age especially older infants and toddlers.
- The most common objects on which children choke are food items (Nuts, seeds, hard candy, gum, bones)
- Common inorganic objects on which children choke include coins, latex balloons, jewellery, magnets and toys.
- Globular, compressible, or round objects are particularly hazardous due to their ability to completely occlude the airway.

Clinical Manifestations

- Foreign bodies of the airway have variable presentations and complications, depending on the characteristics, duration, and location of the foreign body. The clinical manifestations range from an asymptomatic state to severe respiratory distress.
- The most serious complication of foreign body aspiration is complete obstruction of the airway.

Three stages of symptoms

- **Initial event:** coughing, choking, gagging and possibly airway obstruction occur immediately when the foreign body is aspirated.
- **Asymptomatic interval:** The foreign body becomes lodged , reflexes fatigue, and the immediate irritating symptoms subside.
- **Complications:** Obstruction, erosion, or infection (complications include fever, cough, hemoptysis , pneumonia, and atelectasis)

Recurrent infection

- foreign bodies for long periods of time and may come to medical attention because of persistent cough, sputum production, or fever and recurrent unilateral pneumonia.
- persistent wheezing unresponsive to bronchodilator therapy, persistent atelectasis.
- One reason for delay in diagnosis is that children with lower airway FBs may present with subtle or nonspecific symptoms.

Diagnostic Studies

- History
- Physical findings (cough , localized wheezing, unilateral absence of breath sounds, stridor, and, rarely, bloody sputum)
- Radiology (X-ray and CT scan)
- Bronchoscopy

X-RAY

Posteroanterior (expiratory posteroanterior) and lateral chest radiographs are the standard.

Findings:

- Unilateral hyperinflation (focal air trapping).
- Obstructive emphysema.
- Lobar or segmental atelectasis.
- Mediastinal shift toward the opposite side.

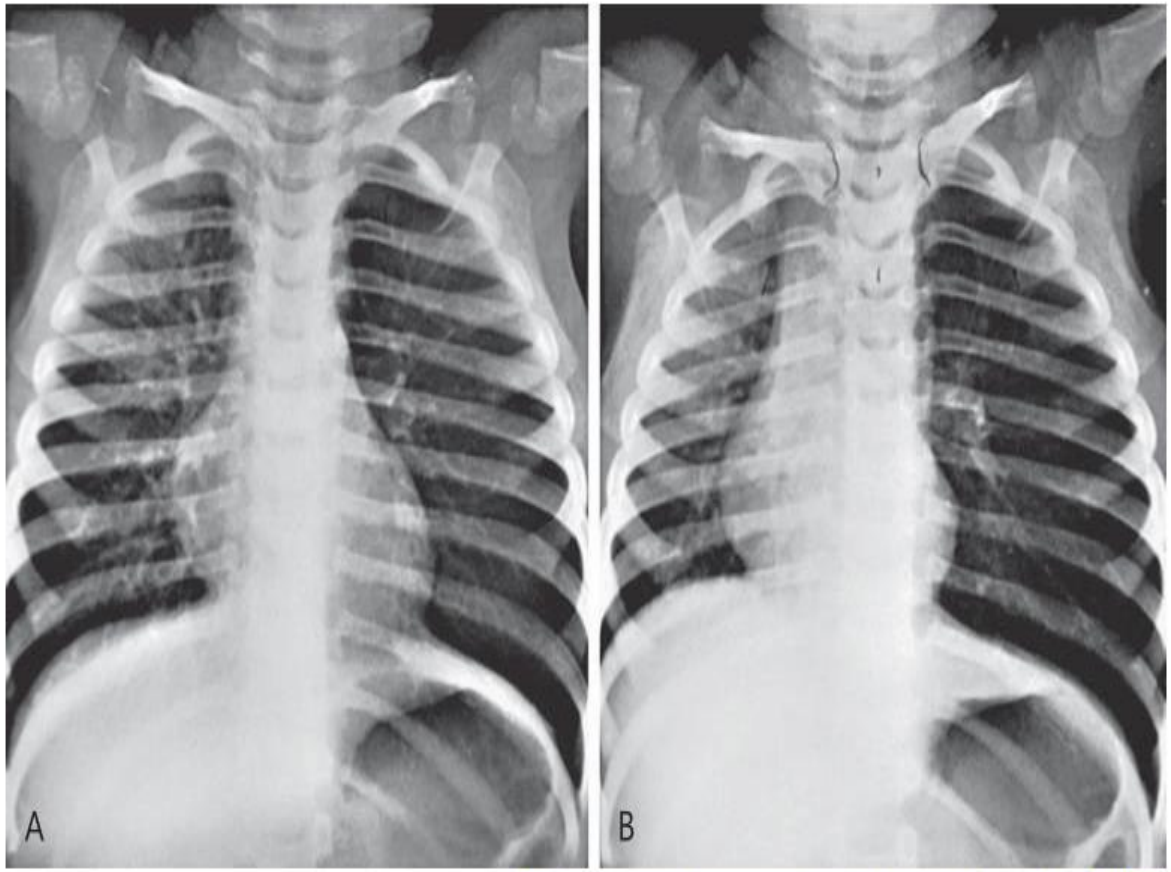


Figure 387-2 **A,** Normal inspiratory chest radiograph in a toddler with a peanut fragment in the left main bronchus. **B,** Expiratory radiograph of the same child showing the classic obstructive emphysema (air trapping) on the involved (left) side. Air leaves the normal right side allowing the lung to deflate. The medium shifts toward the unobstructed side.

Management

The treatment of choice for airway foreign bodies (rigid bronchoscopy)

Chronic Recurrent Aspiration

Aspiration syndromes :

- aspiration of materials or chemicals foreign to the tracheobronchial tree from above (e.g., aspiration of colonized oropharyngeal materials) or from below (e.g., aspiration of gastroesophageal contents).
- Most commonly, associated with gastroesophageal reflux (GER), swallowing dysfunction, neurological disorders, and structural abnormalities.
- Can cause Inflammation of the bronchial tubes and lungs and bacterial infection.
- Repeated aspiration of even small quantities of gastric, nasal, or oral contents can lead to recurrent bronchitis or bronchiolitis; recurrent pneumonia; atelectasis; wheezing; cough; apnea; and/or laryngospasm.

The extent of injury is related to :

1- the amount and quality of the aspirate .

2. the frequency of episodes.

3. the effectiveness of protective lung-clearance mechanisms.

History

The 4 syndromes that may be associated with chronic lung aspiration are:

- apnea,
- chronic cough,
- recurrent pneumonia.
- Recurrent wheezing and asthma symptoms can be related to aspiration of gastric contents.

Investigation

- **Chest radiography**
- hyperinflation
- marked diffuse interstitial or perihilar infiltrates,
- lobar or segmental infiltrates,
- bronchial wall thickening
- pleural effusion
- atelectasis

- barium esophagram .

- modified barium swallow study with video-fluoroscopy .
- 24 Hours Esophageal pH monitoring to evaluate acid reflux.
- Video fluoroscopic swallow study (VFSS).

Management

- Treat **underlying causes**
- **Conservative therapy** is the initial treatment of choice to prevent aspiration syndromes and often results in significant improvement in respiratory symptoms.
- **Diet modifications**
- **Medications** to enhance gastric emptying (prokinetic agents) / H2 receptor antagonists/ PPIs.
- **Surgical** intervention in complicated unresponsive cases

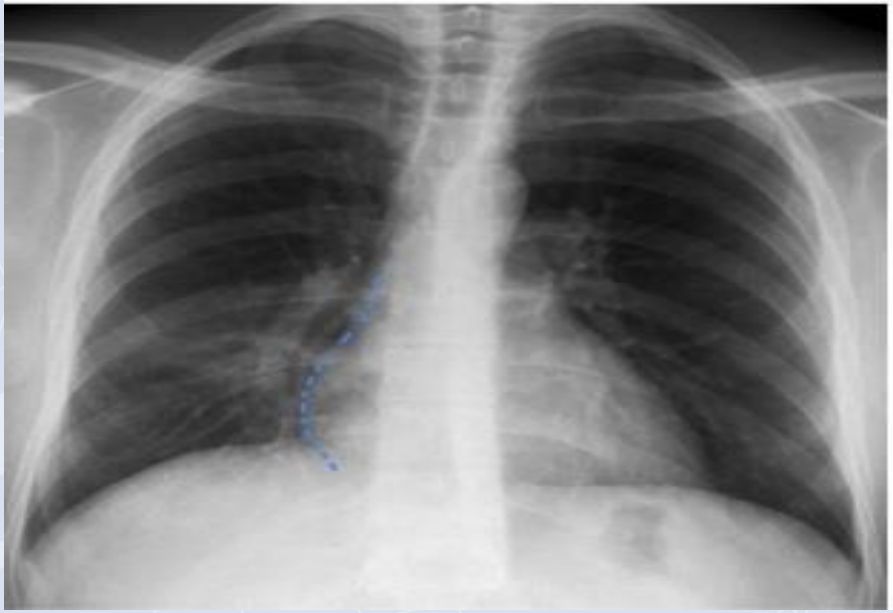
Asthma

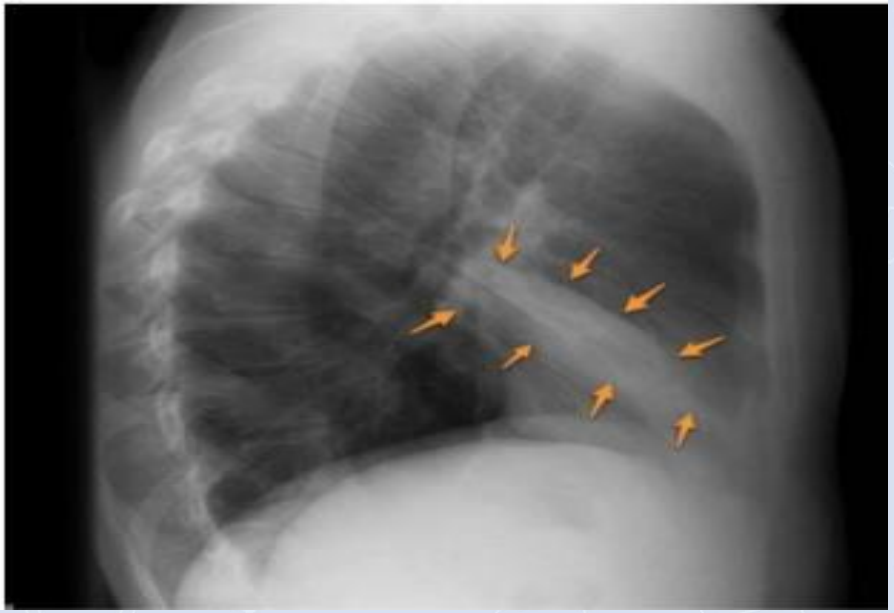
Right middle lobe syndrome

- Persistent or recurrent atelectasis of right middle lobe caused most commonly by acute asthma exacerbation.

- If area of atelectasis is small, patient may be asymptomatic
- Most common symptoms include: persistent of recurrent cough, wheezing, Hx o recurrent pneumonia, Hemoptysis and low grade fever.

- Treated with anti inflammatory therapy such as inhaled steroids. Consider systemic steroids





It is characterized by a wedge-shaped density that extends anteriorly and inferiorly from the hilum of the lung,

Immunodeficiency

Immunodeficiency

- Definition:
  Immunodeficiency represents a diverse group of abnormalities of the immune system resulting from defects in one or more components of innate or adaptive immunity, leading to recurrent, opportunistic, or life-threatening infections. .

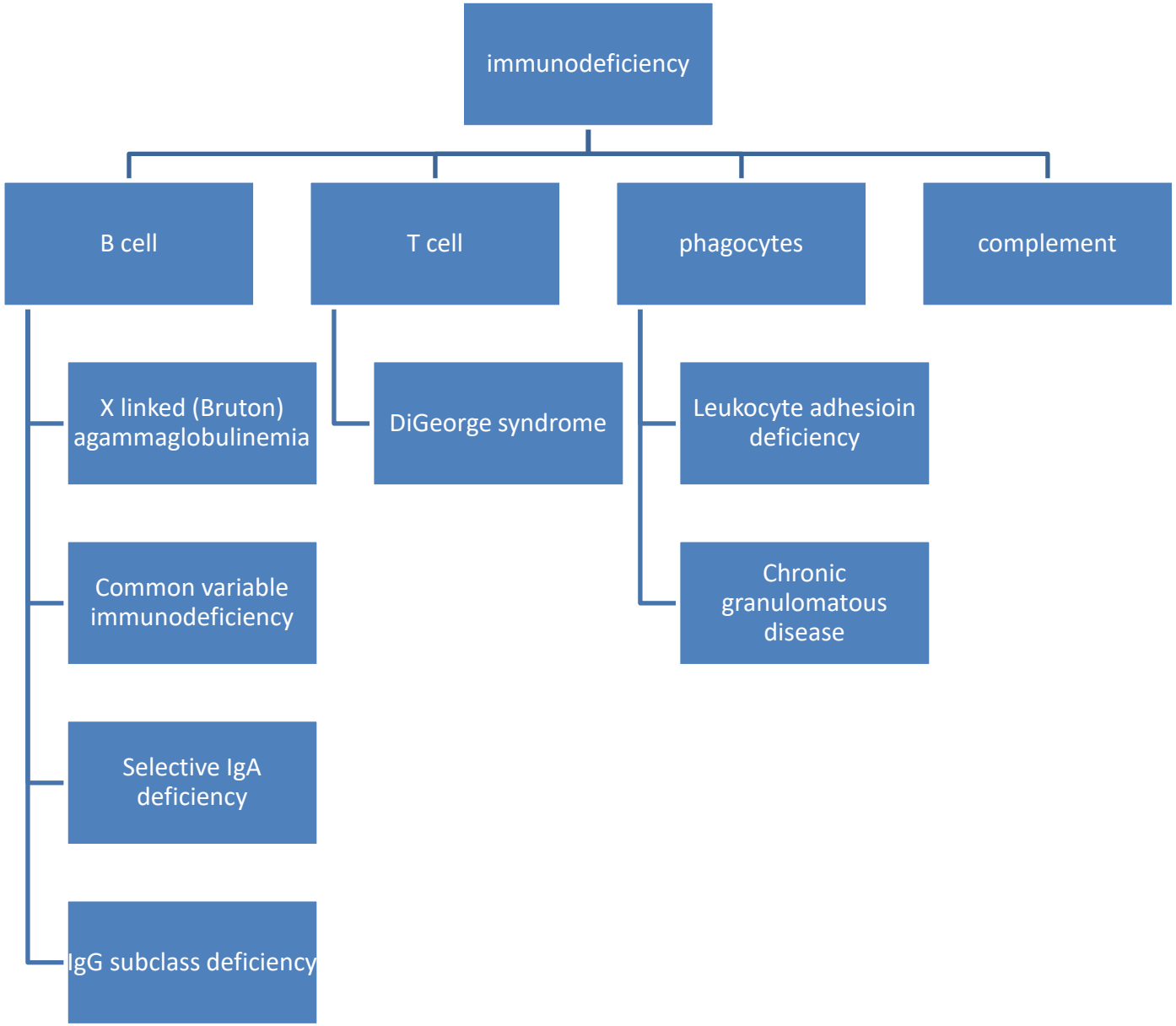
- Primary Immunodeficiency:
  Congenital (inherited).

- Secondary Immunodeficiency:
  Acquired. Could be transient or
  permanent.

10 Warning Signs Of Primary Immunodeficiency For Children Up To Age 18

If your child has two or more of these signs, ask your doctor about Primary Immunodeficiency. It could save your child's life

1. Four or more new ear infections within one year.
2. Two or more serious sinus infections within one year.
3. Two or more months on antibiotics with little effect.
4. Two or more pneumonias within one year.
5. Failure of an infant to gain weight or grow normally.
6. Recurrent, deep skin or organ abscesses.
7. Persistent thrush in mouth or fungal infection on skin.
8. Need for intravenous antibiotics to clear infections.
9. Two or more deep-seated infections including septicemia.
10. A family history of Primary Immunodeficiency.


immunodeficiency
B cell
T cell
phagocytes
complement
X linked (Bruton) agammaglobulinemia
Common variable immunodeficiency
Selective IgA deficiency
IgG subclass deficiency
DiGeorge syndrome
Leukocyte adhesioin deficiency
Chronic granulomatous disease

X-linked agammaglobulinemia

Caused by defect in b-cell development, leading to absence of b cells, and severe hypogammaglobulinemia

Genetics: mutation in BTK gene (bruton tyrosine kinase)

Clinical findings: recurrent sinopulmonary infections, chronic enteroviral meningoencephalitis and vaccine-associated poliomyelitis and giardiasis

presentation: first 6-12 months when maternally transferred IgG disappears

Investigations: low serum immunoglobulins, absence of b cells in flow cytometry

Therapy

1. Regular (usually every 2-4 weeks) IVIG or subcutaneous Ig (SCIG)
2. Antibiotics: prolonged therapy for suspected infections
3. Avoidance of live vaccines

Management of pneumonia in immunodeficiency:

- Follow pulmonary function in patients with recurrent pneumonia.
- Use chest physiotherapy and postural drainage in patients with recurrent pneumonia.
- vaccination with protein-conjugate vaccines to H.influenzae type b and S.pneumoniae, assaying post vaccination titers at least 1 month later.

Common variable immunodeficiency

characterized hypogammaglobulinemia developing after an initial period of normal immune function, most commonly in the second and third decades of life

Genetics: gene defect is unknown

Clinical findings:respiratory tract infections (S. pneumoniae, H. influenzae type b, and Mycoplasma), Gastrointestinal infections (Giardia, Salmonella,enteroviruses).
Autoimmune hemolytic anemia and lymphoma

Labs: low serum IgG, IgM and IgA, phenotypically normal b cell

Therapy
1. IVIG infusions
2.Bone marrow transplant before age 3 months improves survival

Selective IgA deficiency

isolated serum IgA deficiency in patient age >4 years without other immunodeficiencies

Genetics: defect is unknown, but familial pattern suggests autosomal inheritance

clinical findings: **Most patients (85%-96) are asymptomatic**
recurrent sinopulmonary infections, IgG2 subclass deficiency, food allergy, autoimmune disease(SLE, JIA) and celiac disease. Giardiasis is common

Testing: quantitative immunoglobulins (demonstrates low IgA)

Treatment: IVIG is not indicated (anaphylactic reaction)
Treat the infections

Combined Immunodeficiency Diseases

Hyper IgM syndrome

caused by failure of immunoglobulin isotype switching from IgM to IgG, IgA, or IgE, and a lack of memory responses, deficient t-cell function

Genetics: the commonest type is x-linkes,defects in CD40 ligand

Clinical findings: sinopulmonary infections, opportunistic infections (P. jiroveci and cryptosporidium parvum.)

Investigations: normal or elevated serum levels of IgM with low or absent levels of IgG, IgA, and IgE

Therapy:
IVIG replacement, trimethoprim-sulfamethoxazole to prevent PCP.
Stem cell transplantation

Severe combined immunodeficiency

profound lack of T-cell numbers or function, and B-cell dysfunction resulting from the absence of B cells from the gene defect itself, or secondary to lack of T-cell function

Genetics: x-linked is the most common form.

Clinical findings: failure to thrive, severe bacterial infections, chronic candidiasis and other fungal infections, infection with opportunistic organisms, and intractable diarrhea and eczema.

Labs: absence of T cells or NK cells in the peripheral blood, normal numbers of B cells. Immunoglobulin levels are low or undetectable because there are no CD4 T cells to stimulate B cells

Therapy: stem cell transplant
high risk of graft versus host disease

Cystic Fibrosis

-Inherited multisystem Autosomal recessive disorder.

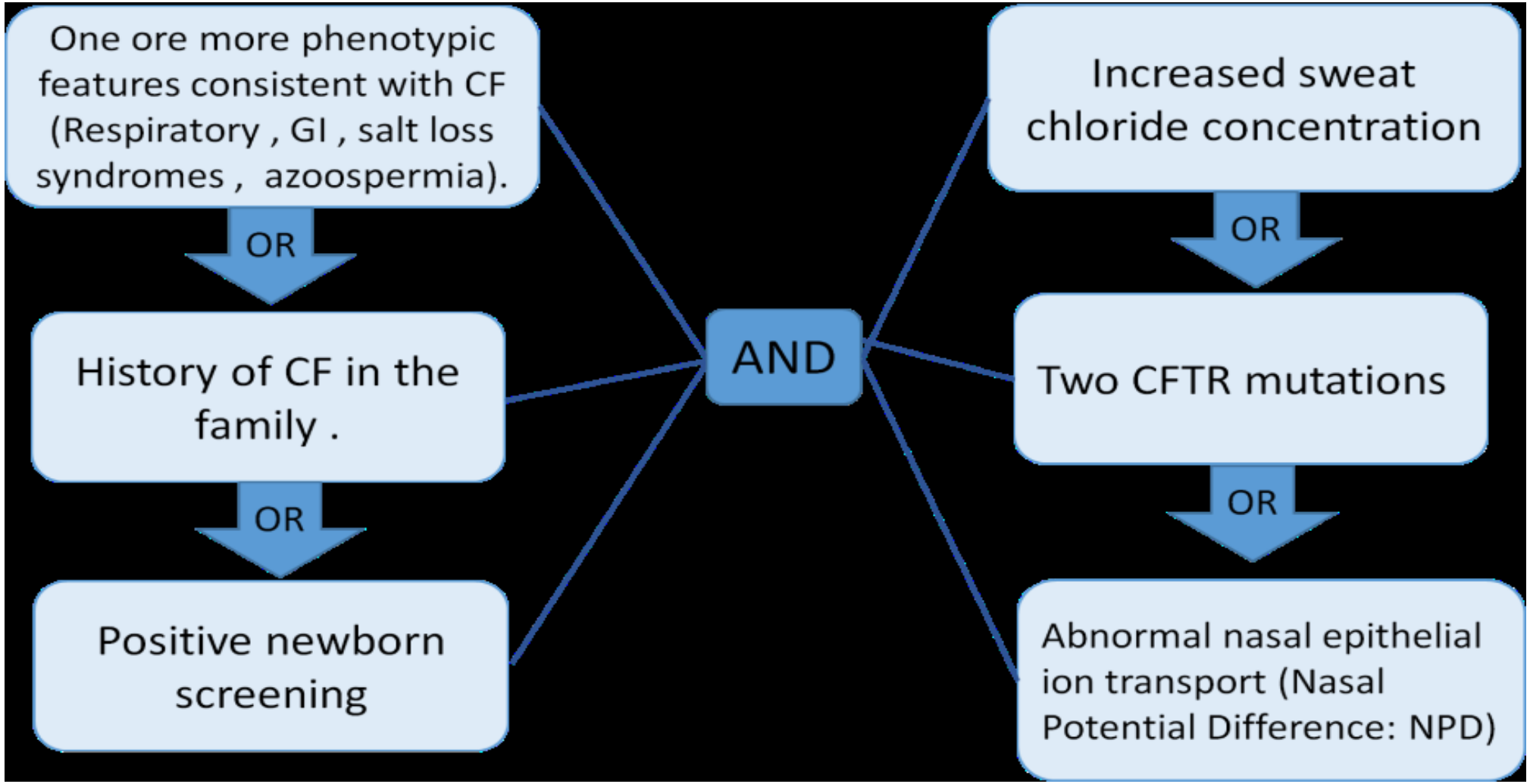
-Defect in the transmembrane conductance regulator protein(CFTR protein) which is responsible for chloride secretion in the lung and GI , reabsorbs cl in sweat glands

—increase intracellular cl – increase intracellular Na and H2o– thick mucous outside .

-Leads to pancreatic insuffiency and chronic lung disease.

-presents mostly as failure to thrive.

Diagnosis Criteria



Recurrent infection

-A likely explanation for recurrent infection is a sequence of events starting with **failure to clear inhaled bacteria** promptly and then proceeding to persistent colonization and an inflammatory response in airway walls

- Common organisms : **Staphylococcus aureus and Pseudomonas aeruginosa, hemophilus influenza**

Management

Mild acute pulmonary exacerbations of CF can be treated successfully at home with the following measures:

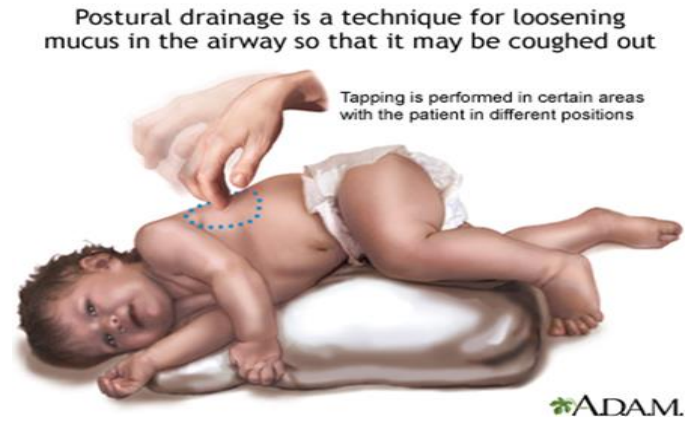
-Increasing the frequency of airway clearance

-Inhaled bronchodilator treatment

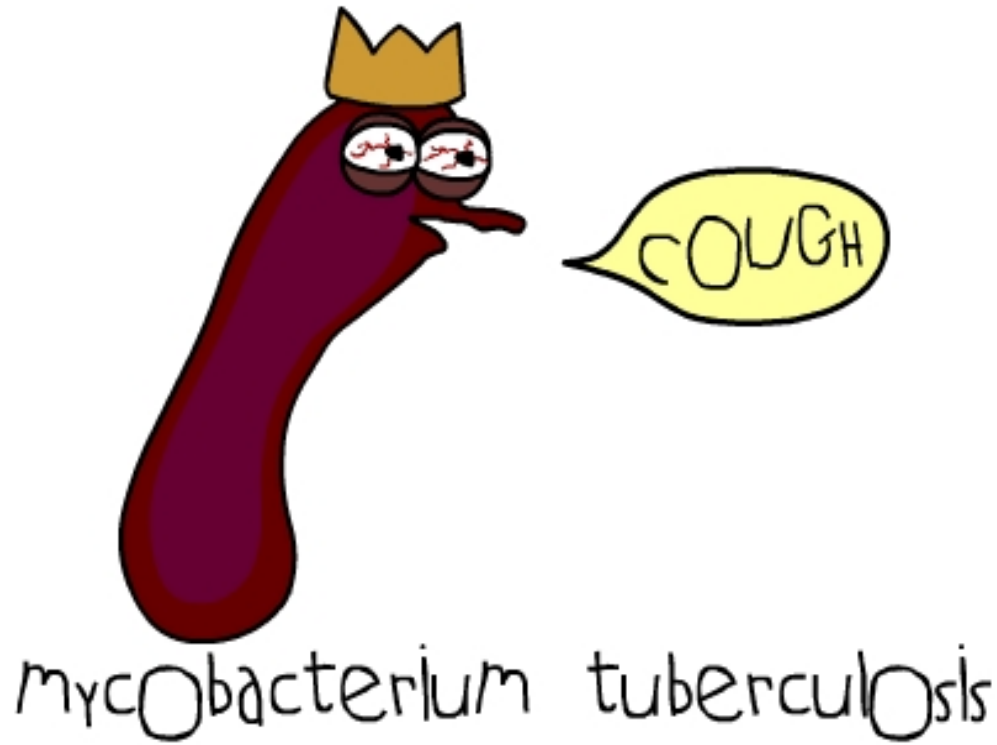
-Chest physical therapy and postural drainage

-Increasing the dose of the mucolytic agent dornase alfa

-Use of oral antibiotics (eg, fluoroquinolones)



TUBERCULOSIS

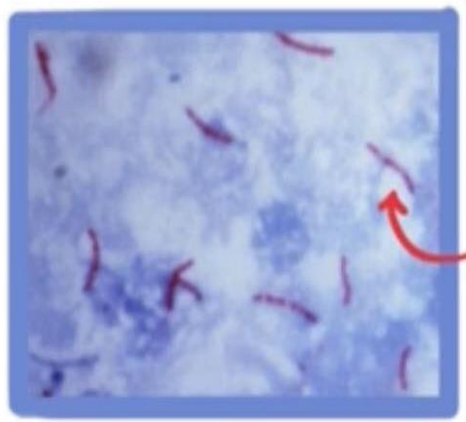


MYCOBACTERIUM TUBERCULOSIS (TB)

WAXY cell wall
- from **MYCOLIC ACID**
- "ACID-FAST"

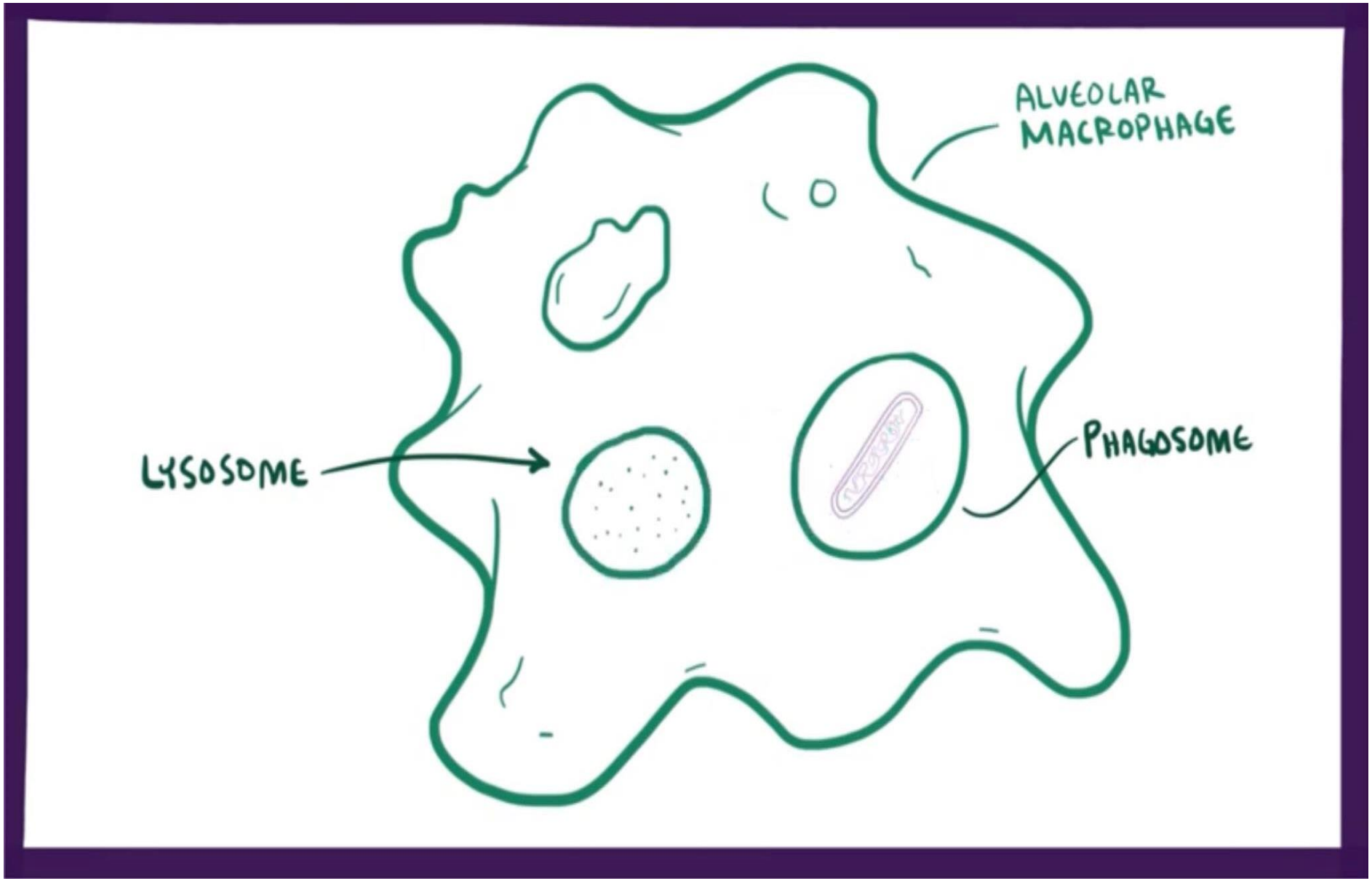
HARDY
- resists weak disinfectants
- survives on dry surfaces

ROD-SHAPED



BRIGHT RED on Ziehl-Neelsen Stain

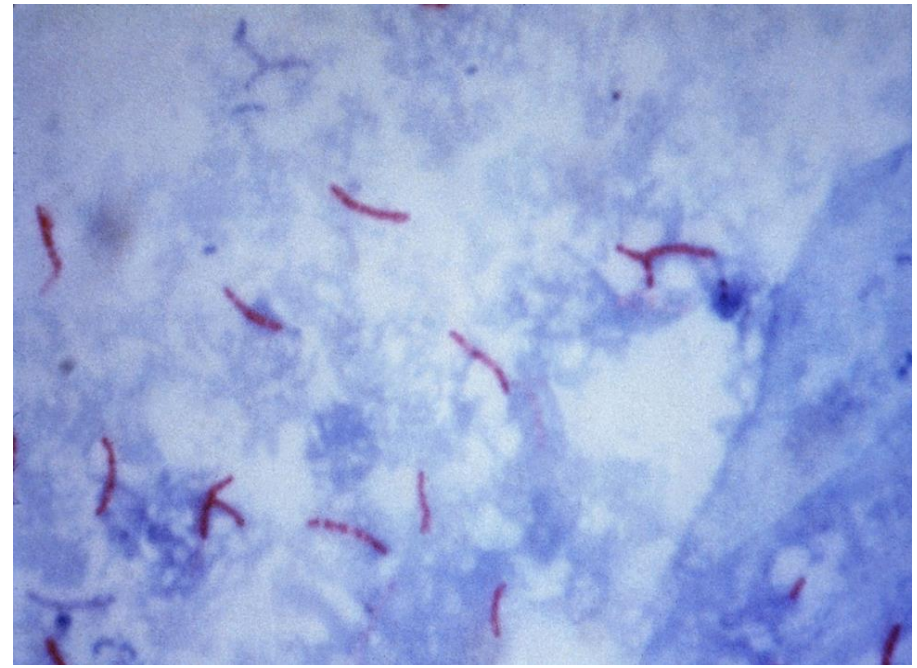
NEEDS **OXYGEN**
STRICT AEROBES


ALVEOLAR
MACROPHAGE
LYSOSOME
PHAGOSOME

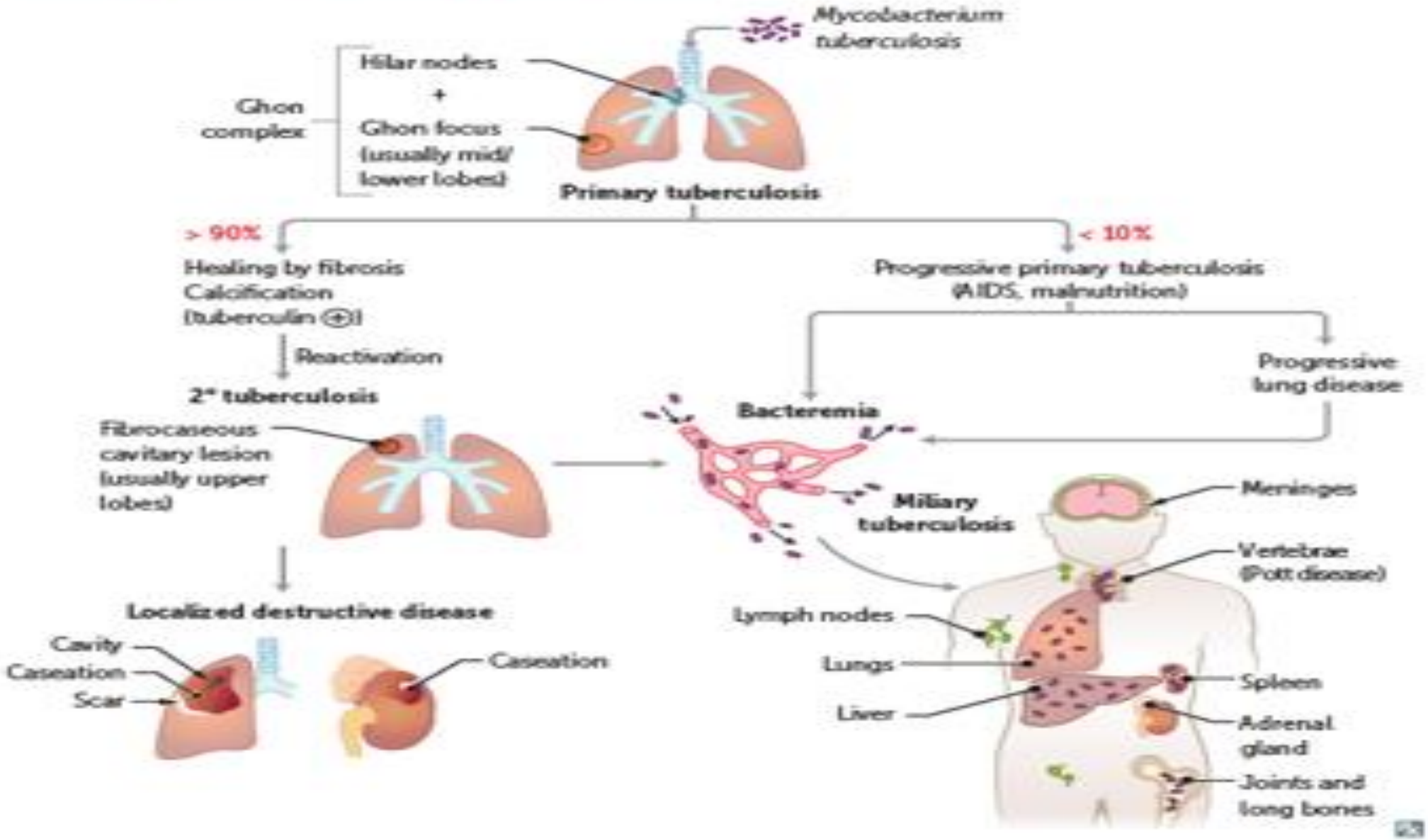
Mycobacterium Tuberculosis

- Aerobes
- Acid fast bacilli
- Transmition> inhalation
- TB in childhood has 2 peaks:

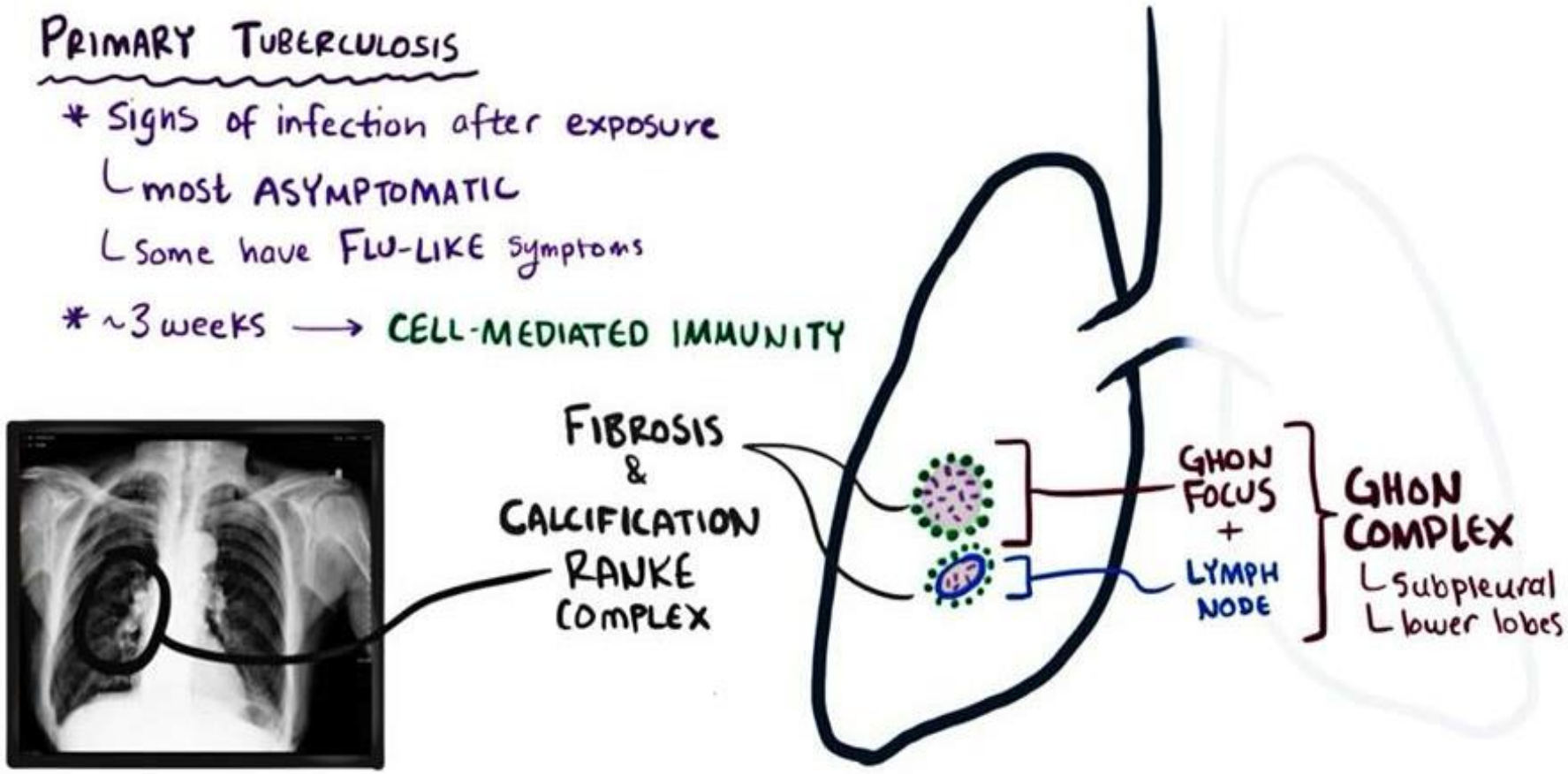
age <5 years and age > 14 years

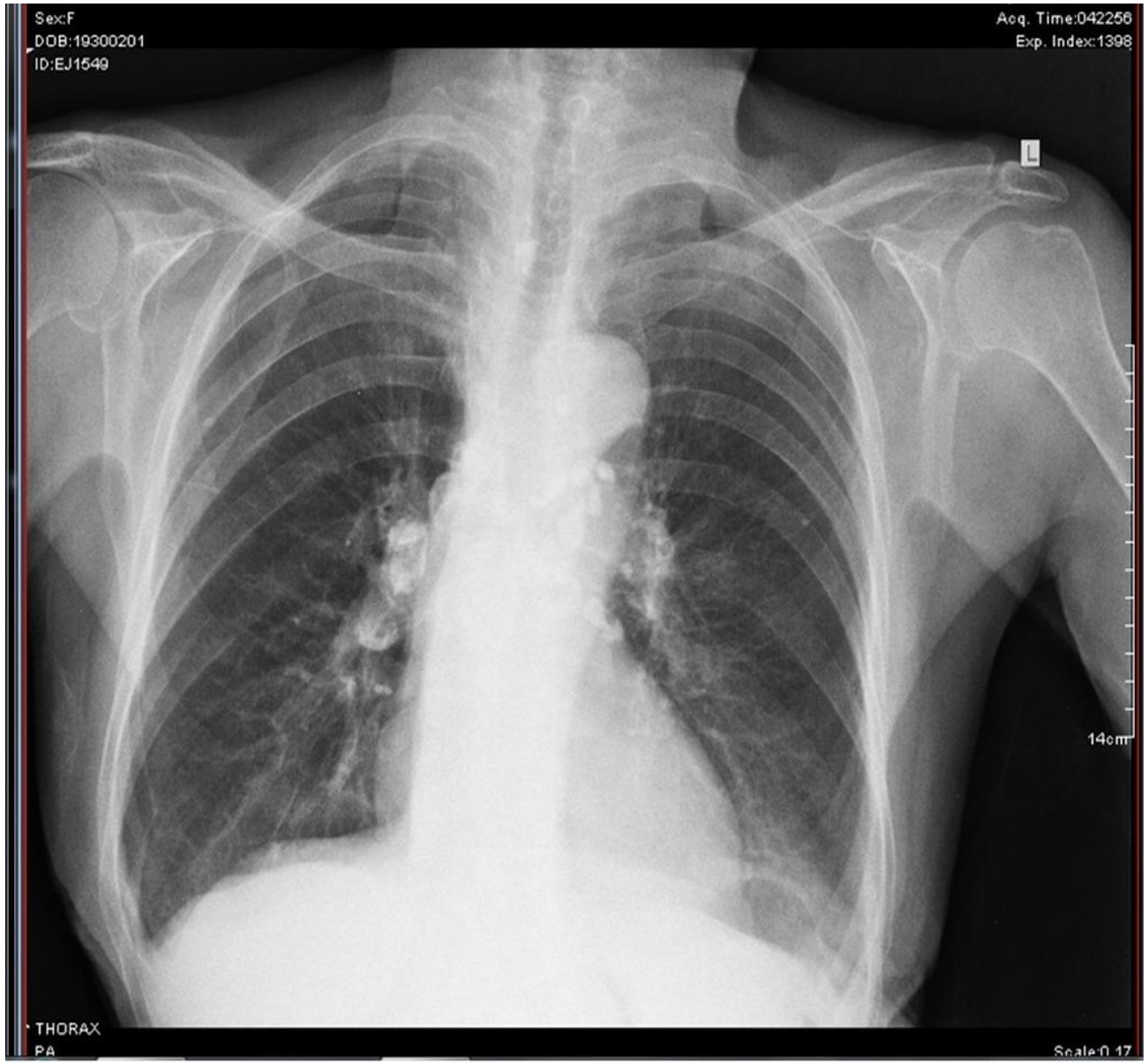


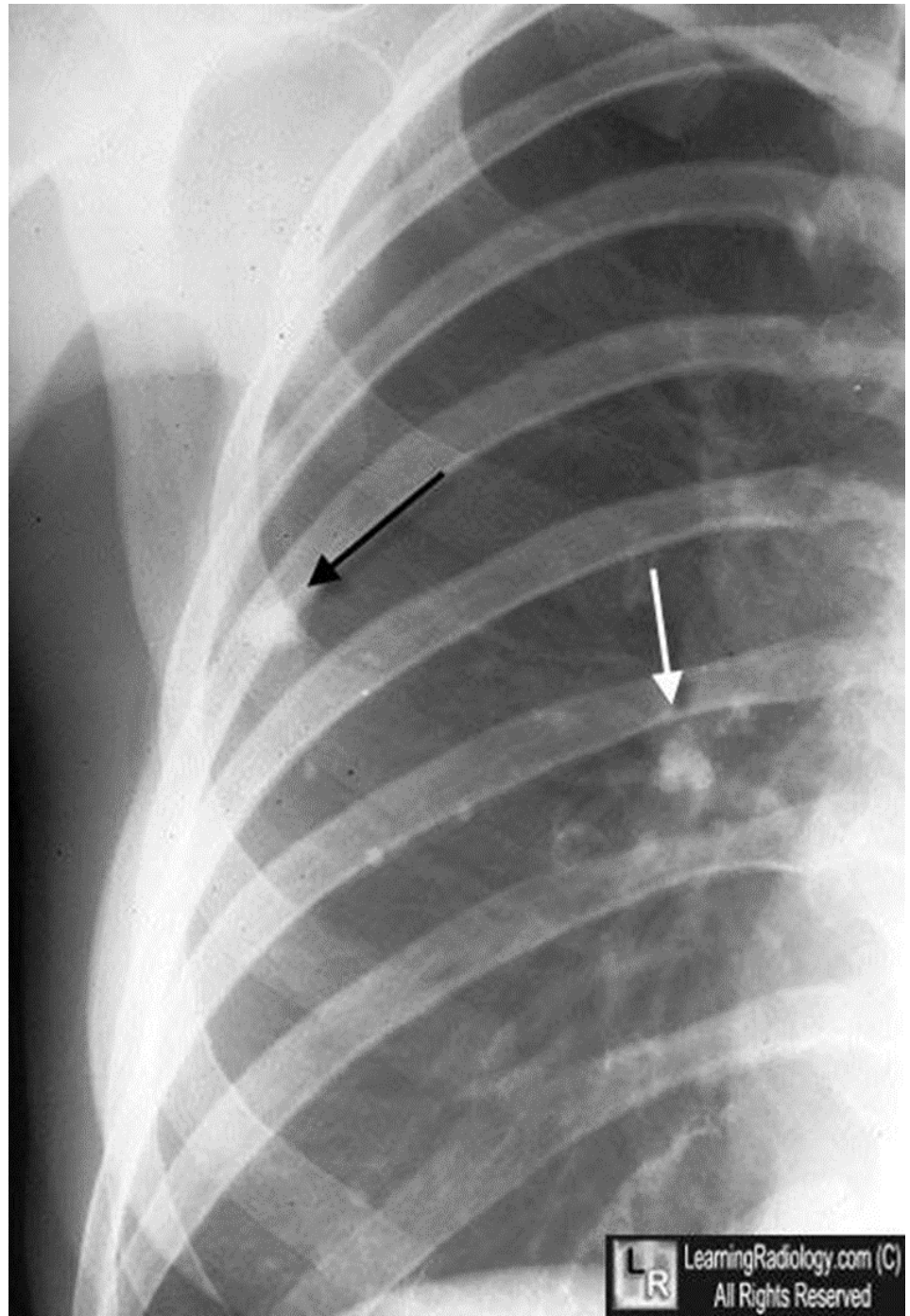
Primary and secondary tuberculosis



Primary TB (latent)



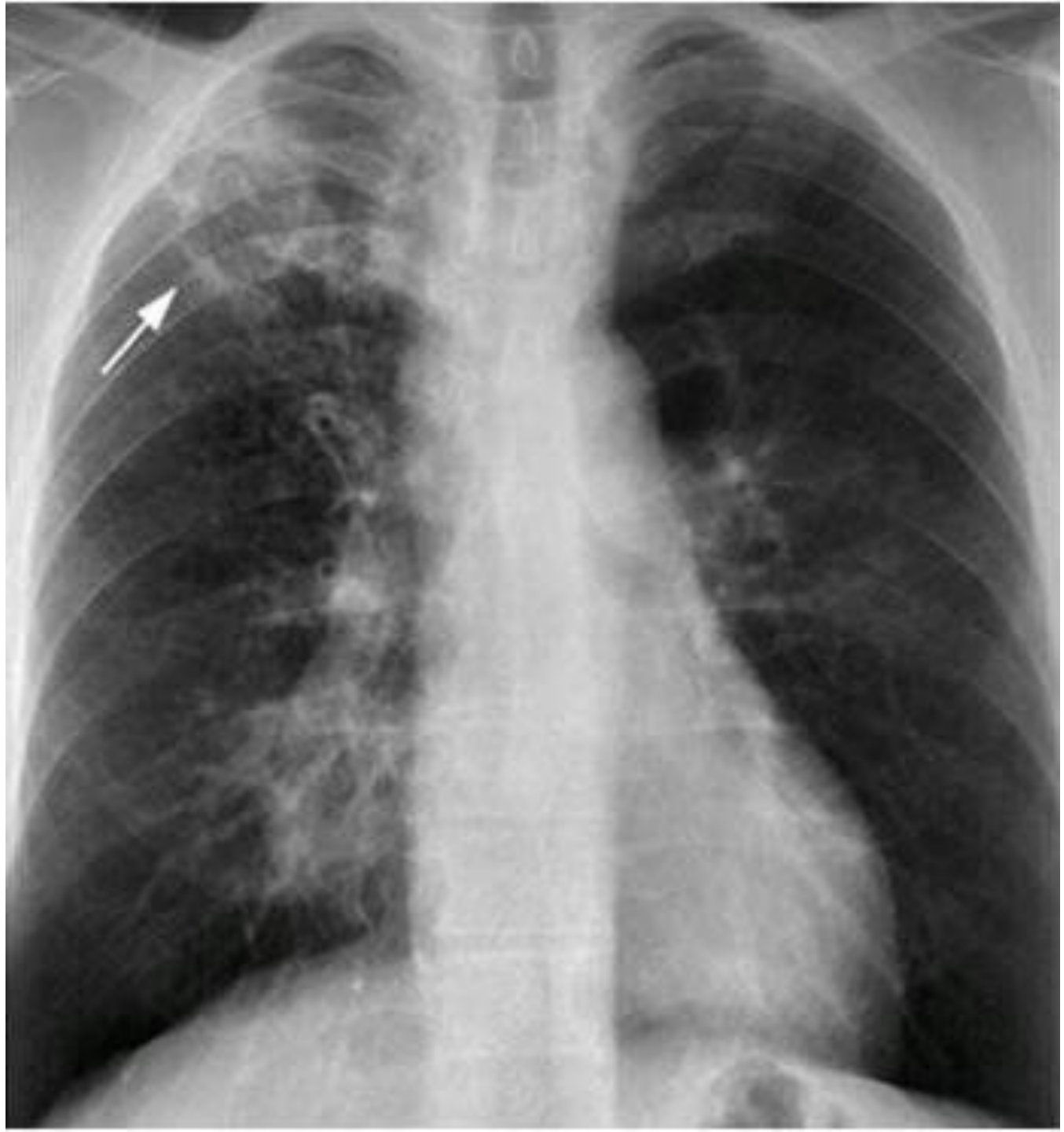

Sex:F
DOB:19300201
ID:EJ1549
Acq. Time:042256
Exp. Index:1398
L
14cm
THORAX
PA
Scale:0.17

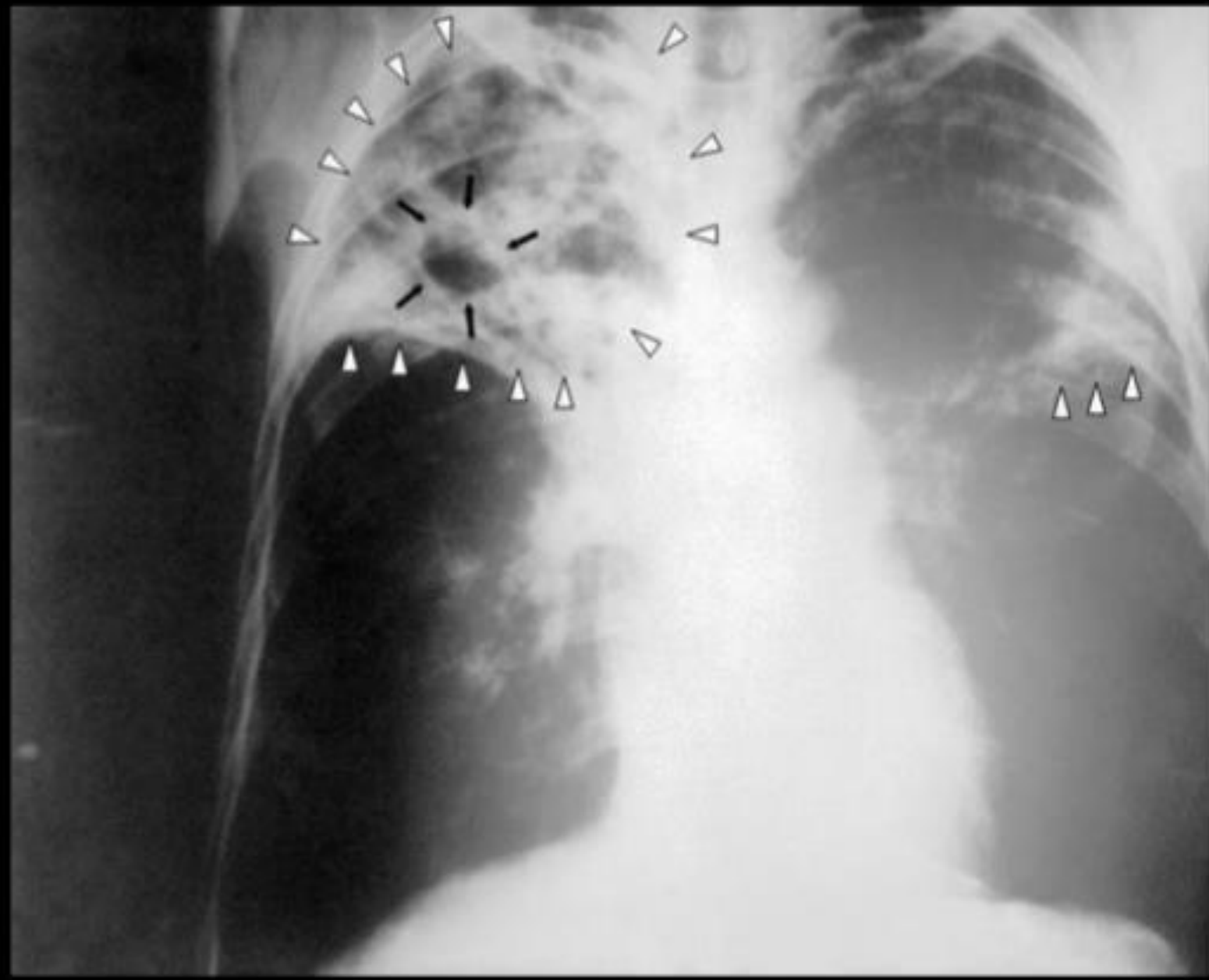

LearningRadiology.com (C)
All Rights Reserved

Secondery TB (active)

- Reactivation (HIV, malignancy, immunosuppresants, substance abuse, malnutrition)
- Most oxygenated segments
- Spreads through lymphatics & blood stream
- **Clinically**:

- Constitutional symptoms: fever, night sweats, weight loss, malaise
- Dry → purulent cough→ hemoptysis
- Apical rales on auscultation

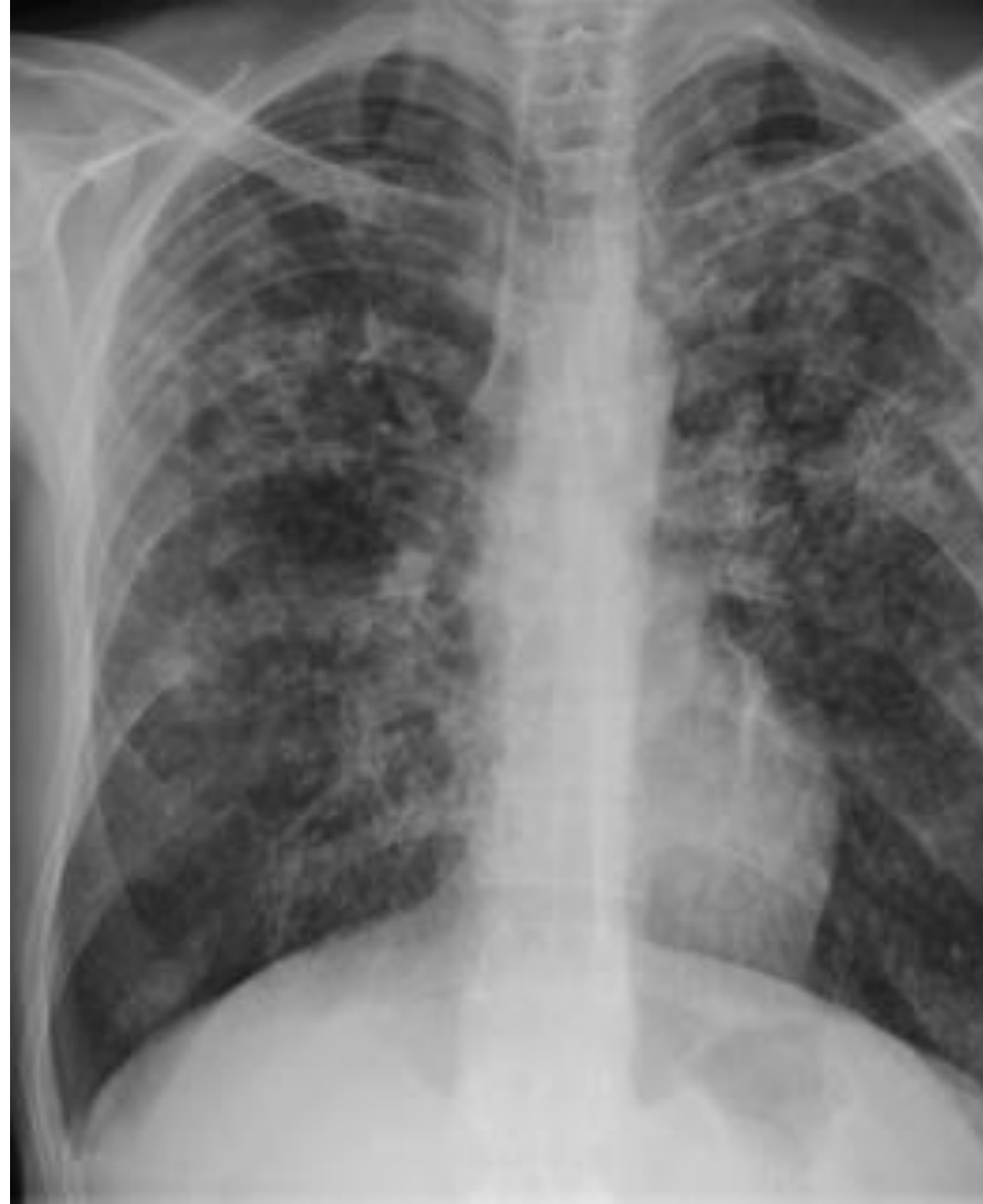




Miliary TB

- Pulmonary
- extrapulmonary (meninges, spine, Renal, Lymph nodes)
- Infants are more predisposed
- **Clinically:**

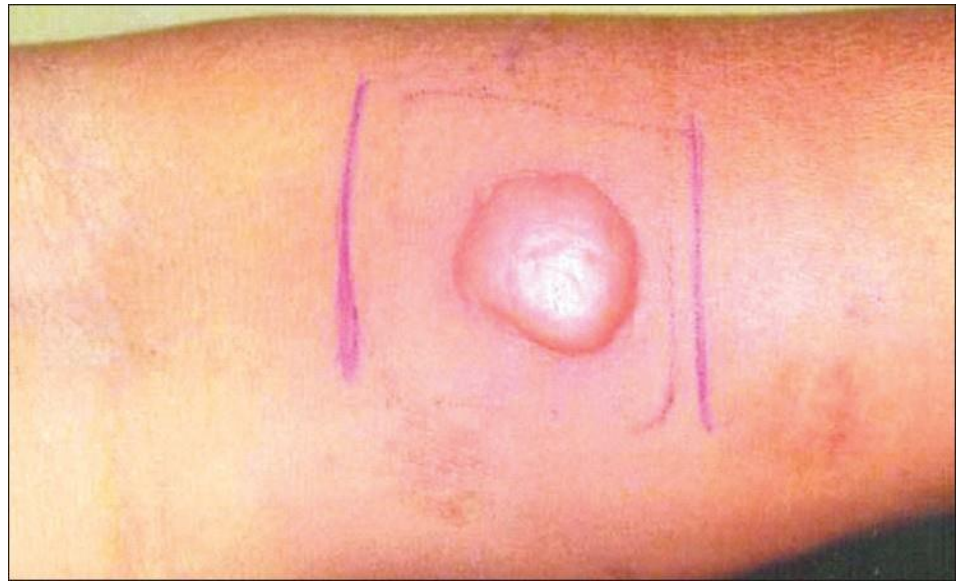
- More sever respiratory symptoms
- Lymphadenopathy
- Organomegaly



Tests

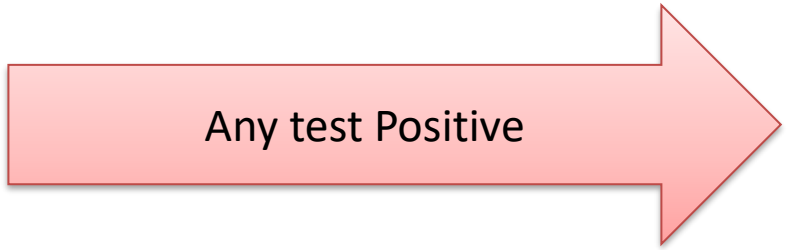
➢ Purified protein derivative (PPD)

- Intradermal injection (5 tuberculin units).. Induration in 48-72 hours →Positive
- Positive= at some point the patient was exposed to TB
- Negative doesn't exclude the exposure
- No differentiation between active and latent



➢ Interferon gamma release assay (IGRA)

- Blood test measure person's immune reactivity to M. tuberculosis (White blood cells from most persons that have been infected with M. tuberculosis will release interferon-gamma (IFN-g) when mixed with antigen)
- 1 office visit
- More specific
- Unlikely to be positive from past BCG vaccine



CHEST X-RAY

TREATMENT

- **Latent TB:** 9 months of **isoniazid** (daily or twice weekly under directly observed therapy)
- **Pulmonary** (and extra-pulmonary except meningitis):

- Start with **RIPE**: Rifampin, Isoniazid, Pyrazinamide, Ethambutol (2 months)
- Followed by **isoniazid** and **rifampin** (another 4 months)
- Once a day or twice weekly under DOT

- **Meningitis:**

- Start with **RIP** (2 months)
- Followed by **isoniazid** and **rifampin** (another 7-10 months)

RECURRENT CHEST INFECTIONS

CONGENITAL ANOMALIES OF THE RESPIRATORY TRACT

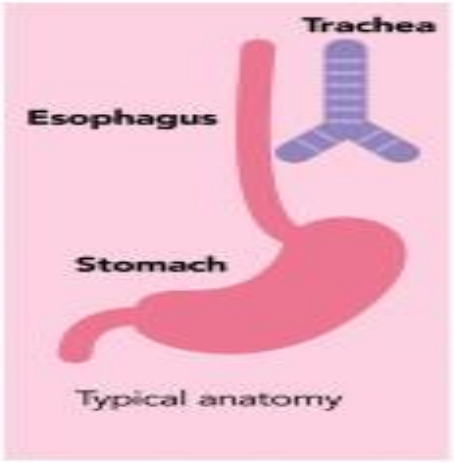
TRACHEAOESOPHAGEAL FISTULA

A Tracheoesophageal fistula is an congenital disease. Its an abnormal communication between the trachea and esophagus. Most of the patient with TEF are diagnosed immediately following after birth .

Typically , occur with (esophageal atresia) which is congenital discontinuity of esophageal lumen from pharynx to stomach during embryonic development .

Congenital and acquired TEFs are associated with multiple complications, including recurrent pneumonia, acute lung injury, acute respiratory distress syndrome, lung abscess, poor nutrition, bronchiectasis from recurrent aspiration, respiratory failure, and death.

- Approximate 17-70% of children with TEFs have associated development anomalies . These anomalies include Down syndrome, duodenal Artesia and VACTERL association
- **Etiological factors may include :-**
- Maternal alcohol and smoking
- Exogenous sex hormones
- Exposure to methimazole
- Prolonged mechanical ventilation via Endotracheal or tracheotomy tube
- In first trimester exposure to Diabetes mellitus



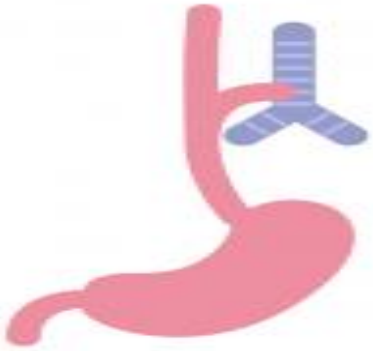
Typical anatomy

Type A: isolated esophageal atresia

Type B: atresia with proximal fistula

Type C: atresia with distal fistula

Type D: atresia with double fistula



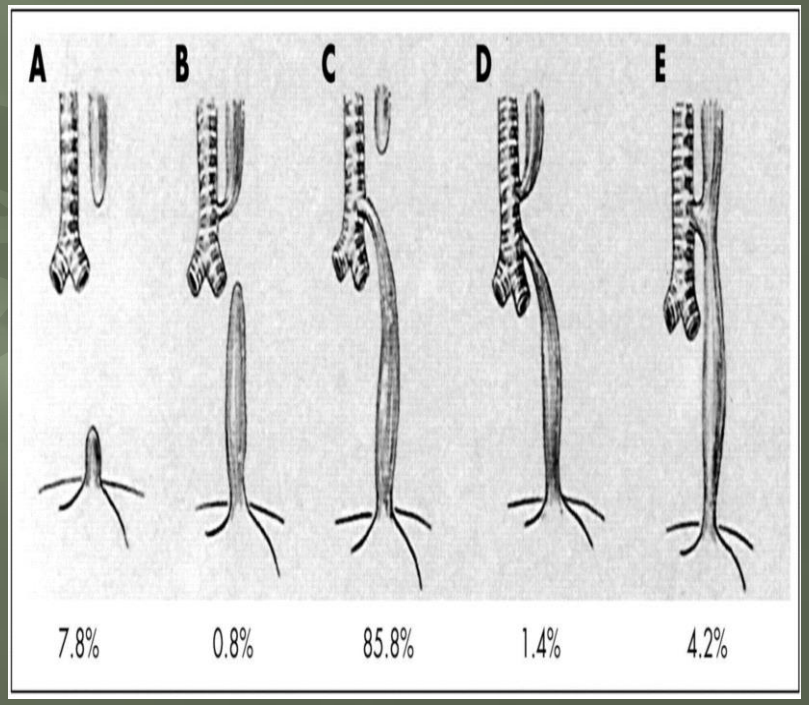
Type E: isolated fistula (H type)

TEF TYPES :-

Type c is the most common type .

Types b , c , d , e can present with recurrent
chest infections

in Type e , the most common mode of presentation is recurrent chest infections and it usually presents later on .

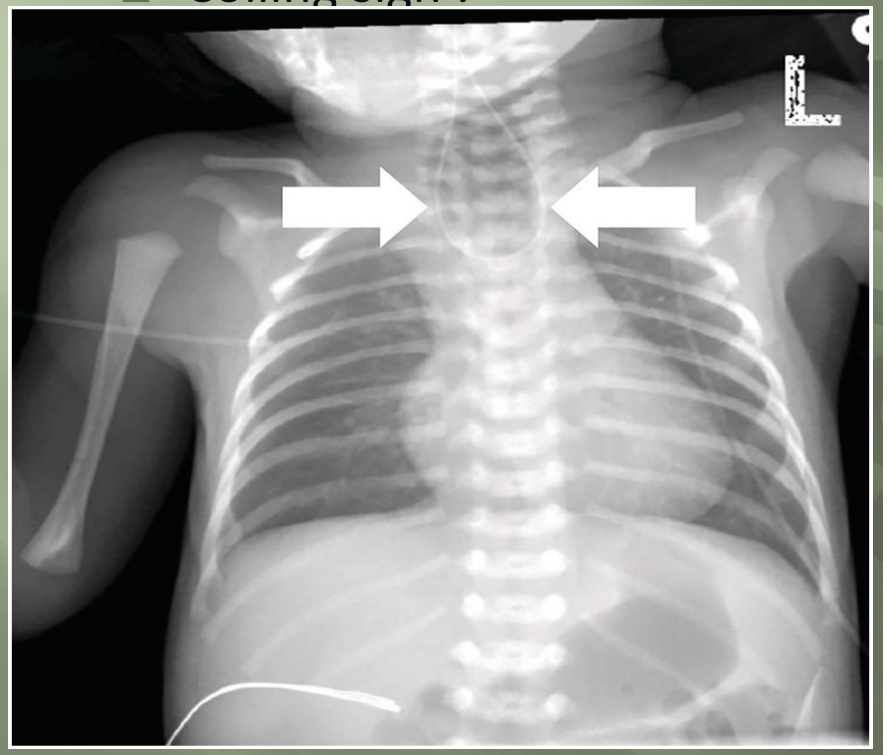


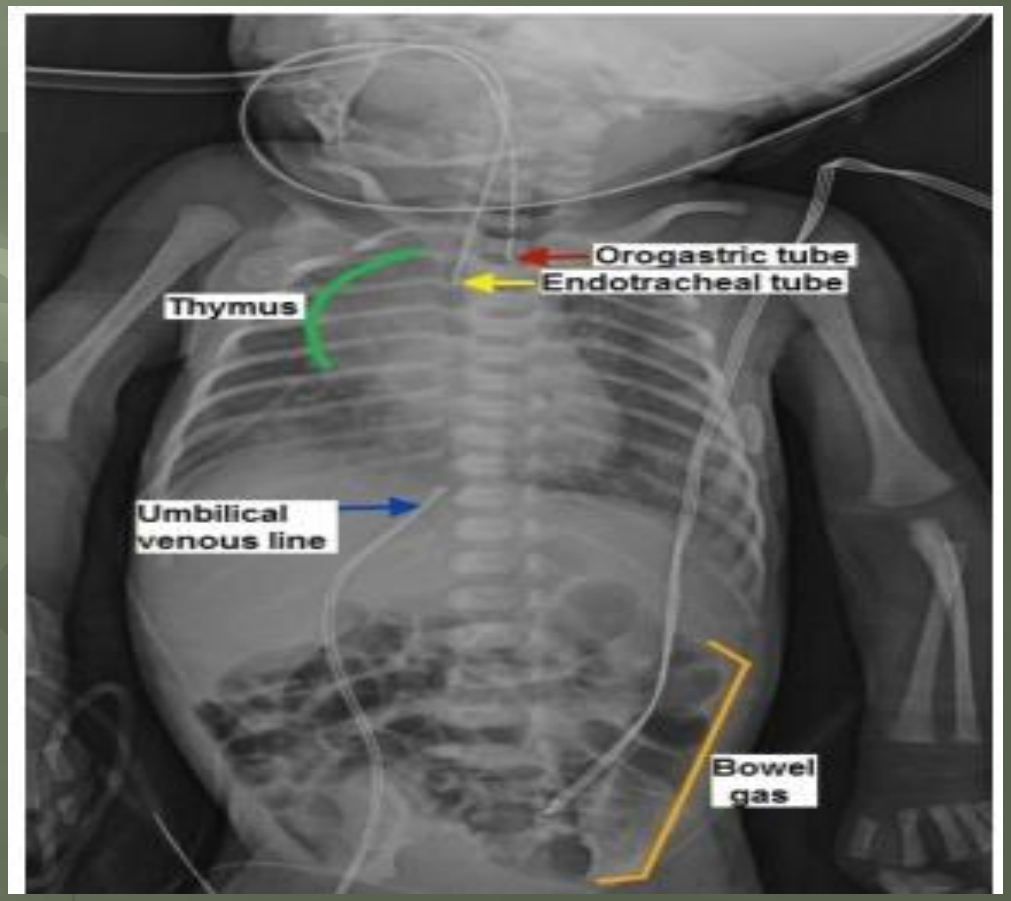
- Tracheoesophageal fistula usually present within the first few hours of life by:
- “vomiting with first feeding”
  Excessive salivation
- Respiratory distress
- Cyanosis , Choking , Coughing .
  Recurrent aspiration pneumonia
  - There will be a history of possible polyhydramnios as the affected fetus cannot swallow amniotic fluid.

Feeding exacerbates these symptoms and can precipitate aspiration

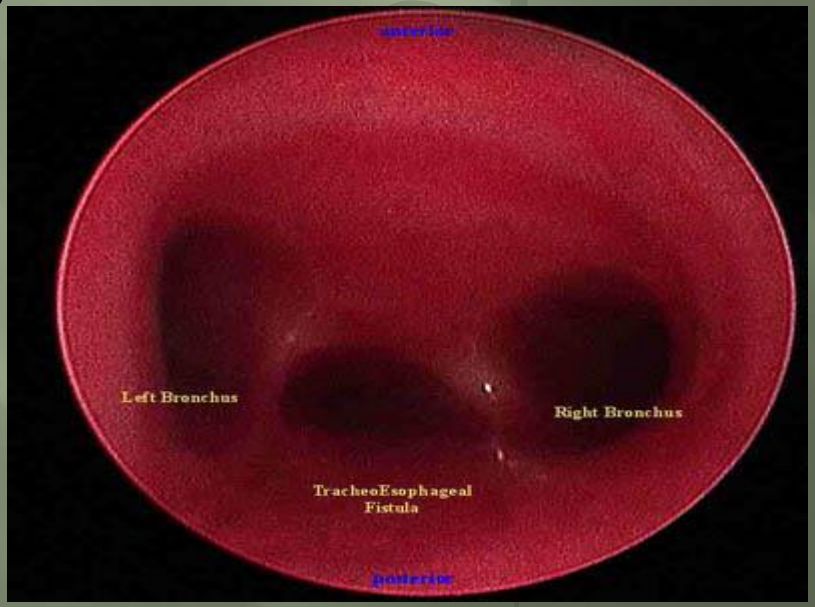
Inability to pass a nasogastric tube is pathognomonic for TEF and useful for diagnosis “ Coiling sign “ , also A gastric air bubble and esophageal air bubble can be seen on chest X-ray (CXR)

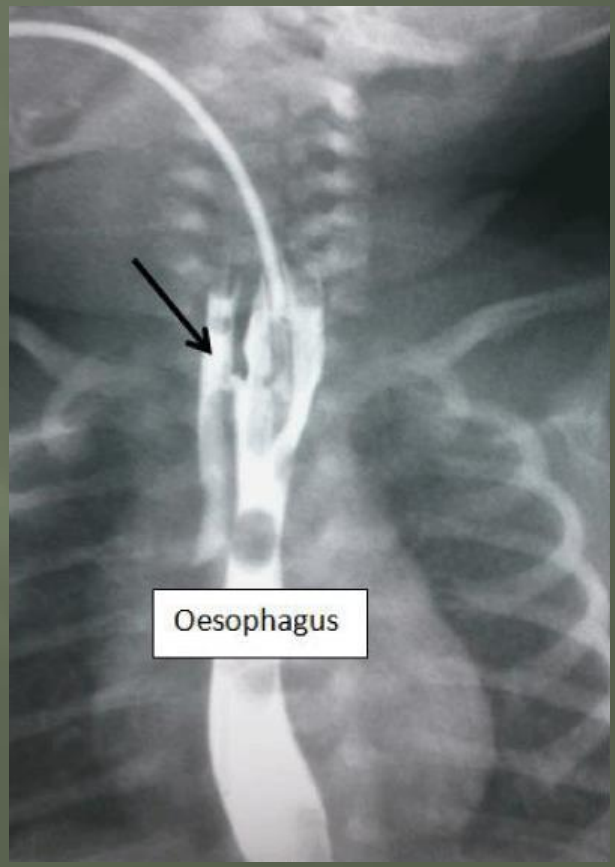
- Coiling sign :





- For H type : Esophagogram with contrast medium
- We can visualize the level of TEF by using Bronchoscopy .





PHARMOLOGIC MANAGEMENT

- Propping infant at 30 degree angle
- Nasogastric tube remains in the esophagus and it is aspirated frequently
- Nothing by mouth
- Supportive therapy include meeting nutritional requirements IV fluid, antibiotics
- respiratory support and maintaining neutral environment

SURGICAL MANAGMENT

Division and closure of the fistula and anastomosis of the two esophageal segments.

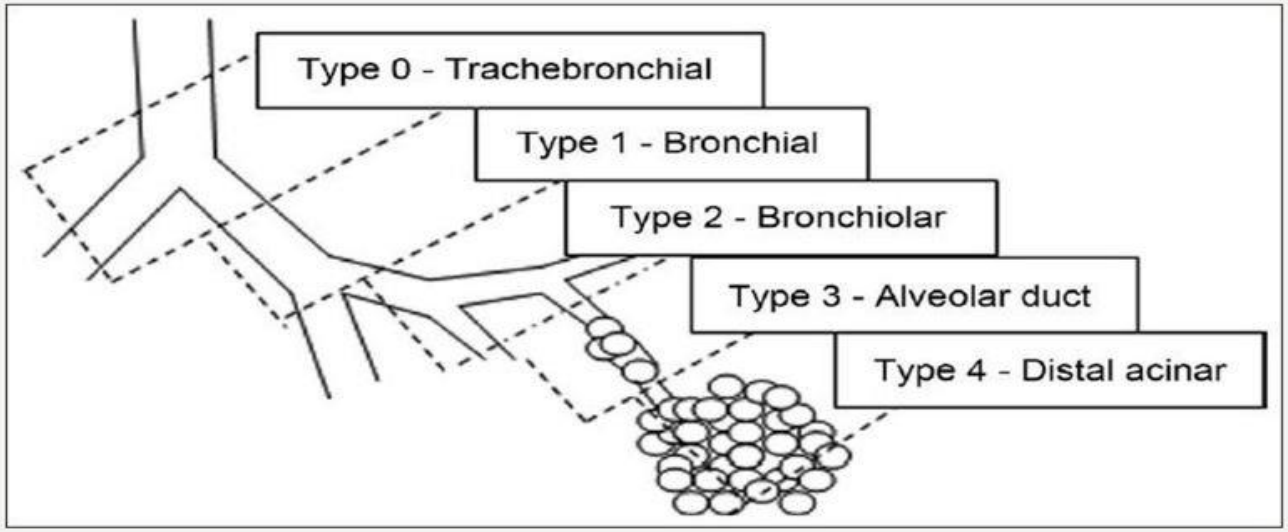
Congenital Pulmonary Airway Malformation
congenital cystic adenomatoid malformation (CCAM)

- This is the first CLM that as diagnosed antenatally in 1975 in Australia
- **Its non functioning cystic lesion**
- **They account for 25% of congenital lung lesions.**
  With prevalence 1 : 3800 live births
- **M > F**
- **Mostly Sporadically**

- **<u>Lesions are hamartomatous :</u>**
- Contain **cystic and adenomatous elements**
- Connected to **tracheobronchial tree (vs bronchogenic cysts)**
- Have blood supply **from pulmonary circulation**
- Usually **unilateral** and **limited to one lobe**

CPAM-CLASSIFICATION

- **There are 5 types of CPAM (types 0-4)**. (Stoker classification)
- Classified **pathologically**, according to the **level of the insult to the airways** and the **different stages of lung development**.

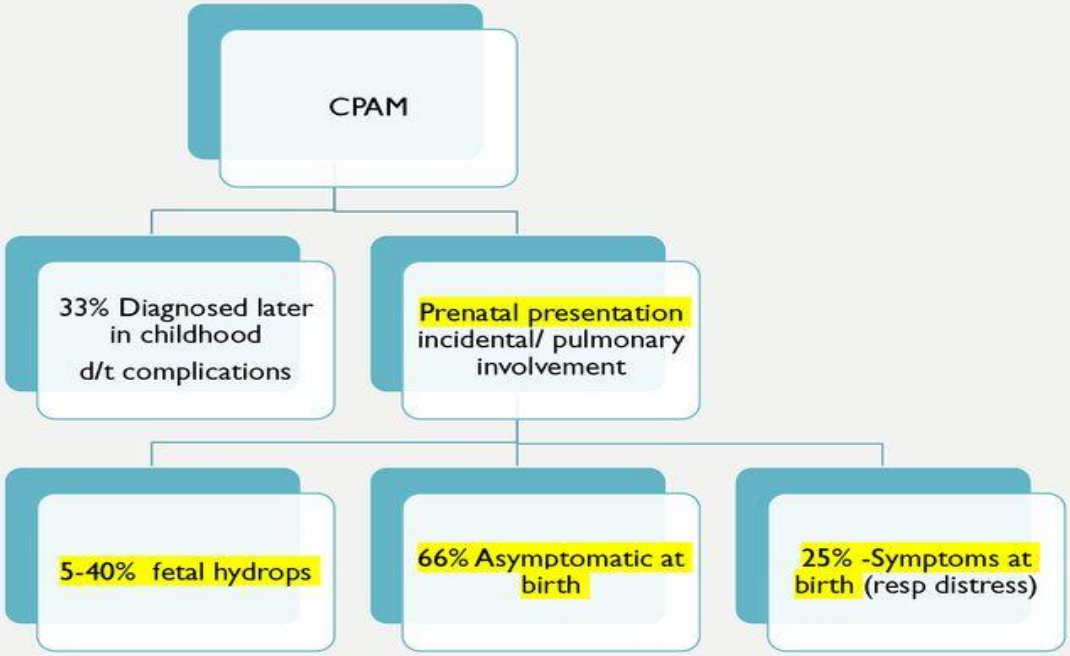


CPAM- 5 TYPES PATHOLOGICALLY

Types	Percentage	Location in the airways	Lesion description	Prognosis	Comments
Type 0	1-3%	From the trachea/bronchi and **involve the whole lung**	Solid, lungs are small and firm	**Fatal-** incompatible with life.	
Type 1	**60-70%**	From distal bronchi or proximal bronchioles	**One Large cyst** 2-10cm, presentation may be late	Most common type **–good prognosis**	**Associated with malignancy BAC (rare)**
Type 2	**15-20%**	Bronchiolar origin	**Multiple small cysts** 0.5-2cm Neonatal period	**Poor prognosis**	**Associated with other anomalies**
Type 3	5-10%	Alveolar origin	Small cystic area with solid tissue. **Solid appearance**	Respiratory distress in neonatal period/death	**Severe**
Type 4	10-15%	Acinar origin	Thin walled Large fluid filled/ air filled cysts up to 10cm.	**Good prognosis**	**Associated with pneumothorax and PPB**

- **Presentation:**
- Some maybe asymptomatic!
- In utero compression of normal fetal lung can cause Pulmonary hypoplasia → **respiratory distress**
- Recurrent URTI
- Pneumothorax
- Malignancy Sx.

CPAM- CLINICAL PRESENTATION



- **Diagnosis:**

- Prenatal ultrasound (appear as solid or cystic intrathoracic mass, also there can be a mass effect where the heart may appear displaced to the opposite side) → follow up echocardiogram + fetal MRI
- CT allows accurate diagnosis and sizing of the lesion and is indicated even in asymptomatic neonates

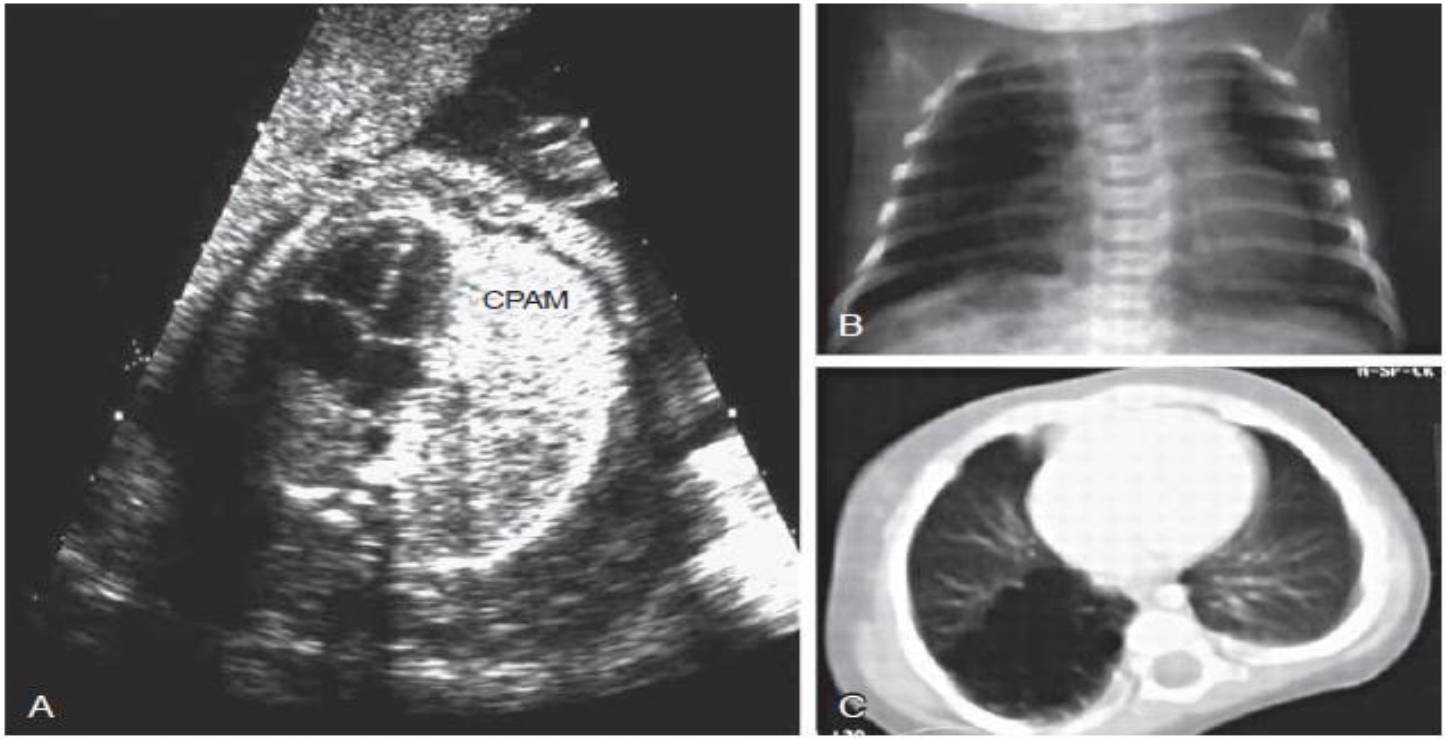
Definitive diagnosis by **Histological examination**

- It may grow, stay, regress … FOLLOW UP

CPAM DIAGNOSIS-RECOMMENDATIONS

- **Family history** of cancers/ cystic lesions (type 4-genetic predisposition).
- **Prenatal US+/- prenatal MRI.**
- **CXR** (all patients)
  - single lesions of large air filled cysts (type 1,4),
  - small numerous air filled cysts (type 2),
  - large solid mass (type 3)

  Plain CXR **often fail to detect CPAM in asymptomatic patients**.



re 395-1 Imaging of congenital pulmonary airway malformation of the lung (CPAM) on the same patient with prenatal u
hest radiograph **(B),** and CT scan **(C).** Note that the lesion is not visible on the chest radiograph. *(From Lakhoo K: Managemer*
adenomatous malformations of the lung, Arch Dis Child Fetal Neonatal Ed *94:F73–F76, 2009.)*

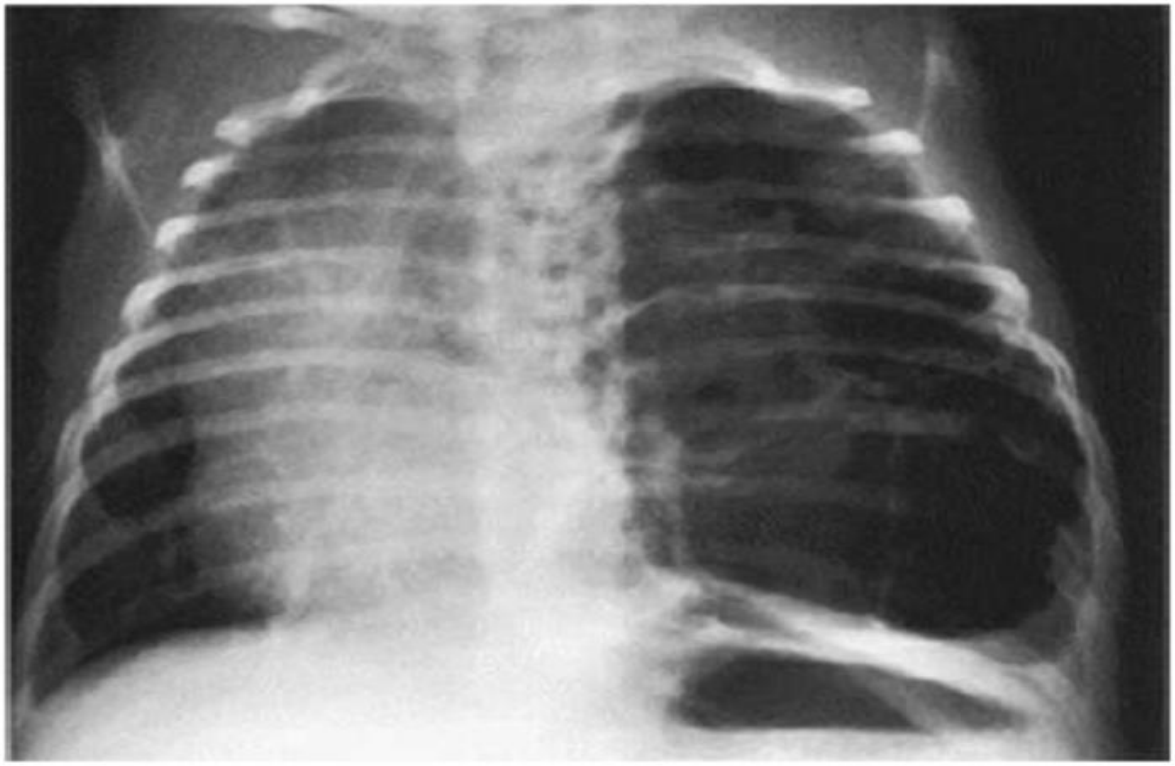


Figure 395-2 Neonatal chest x-ray showing large multicystic mass in the left hemithorax with mediastinal shift as a result of congenital pulmonary airway malformation (CPAM). *(From Williams HJ, Johnson KJ: Imaging of congenital cystic lung lesions,* Paediatr Respir Rev *3:120–127, 2002.)*

- **Management**:
  - Antenatal intervention in severely affected infants is controversial but can include:

1. excision of the affected lobe for microcystic lesions
2. aspiration of macrocystic lesions, and
3. rarely, open fetal surgery

- In the postnatal period→ surgery for symptomatic patients.
- Although surgery may be delayed for asymptomatic infants because postnatal resolution has been reported, true resolution appears to be very rare in that abnormalities usually remain detectable on CT or MRI.
- Sarcomatous and carcinomatous degeneration have been described in patients with CPAM, so surgical resection by 1 year of age is recommended to limit malignant potential. The mortality rate is <10%.
- Another indication for surgery is to rule out pleuropulmonary blastoma, a malignancy that can appear radiographically similar to type I CPAM.

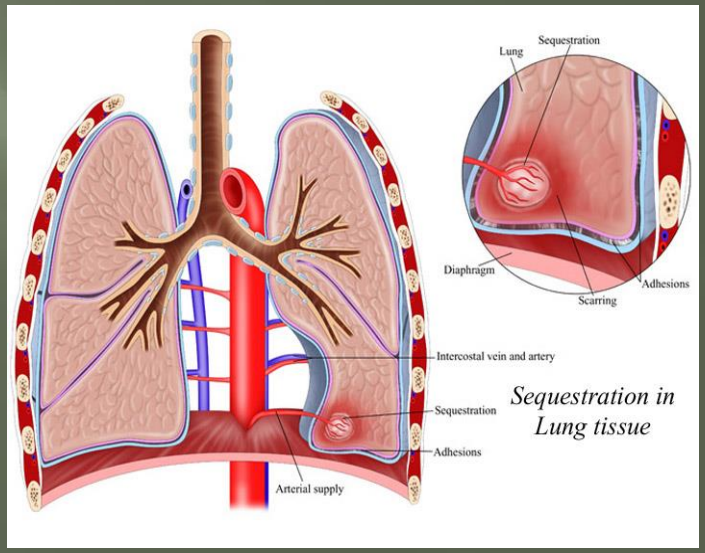
Pulmonary Sequestration (accessory lung)

- Cystic-solid mass of nonfunctioning lung tissue
- functions as a space-occupying lesion
- completely separated from the airways (does not connect to a bronchus) and receives its arterial supply from the systemic arteries.
- Usually in the left lower chest
- Gastric or pancreatic tissue may be found

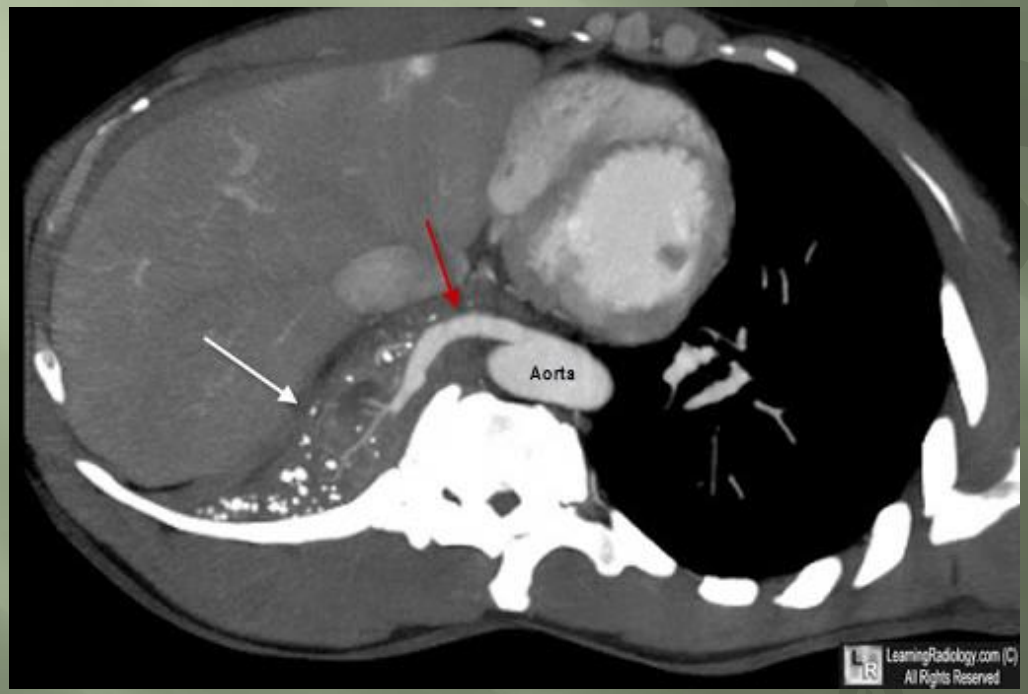
within the sequestration.

- **Complications:**

hemorrhage + chronic infection

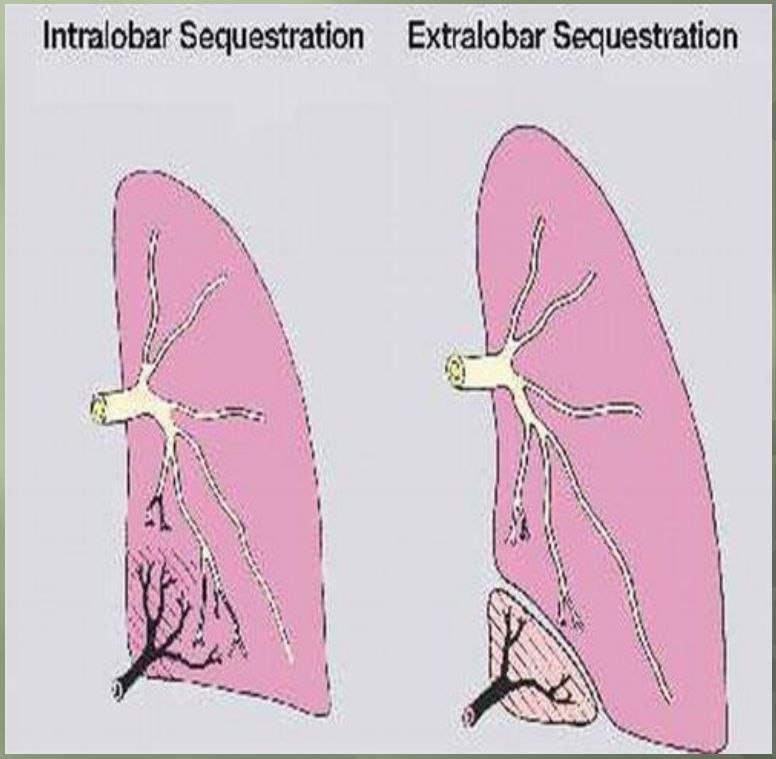


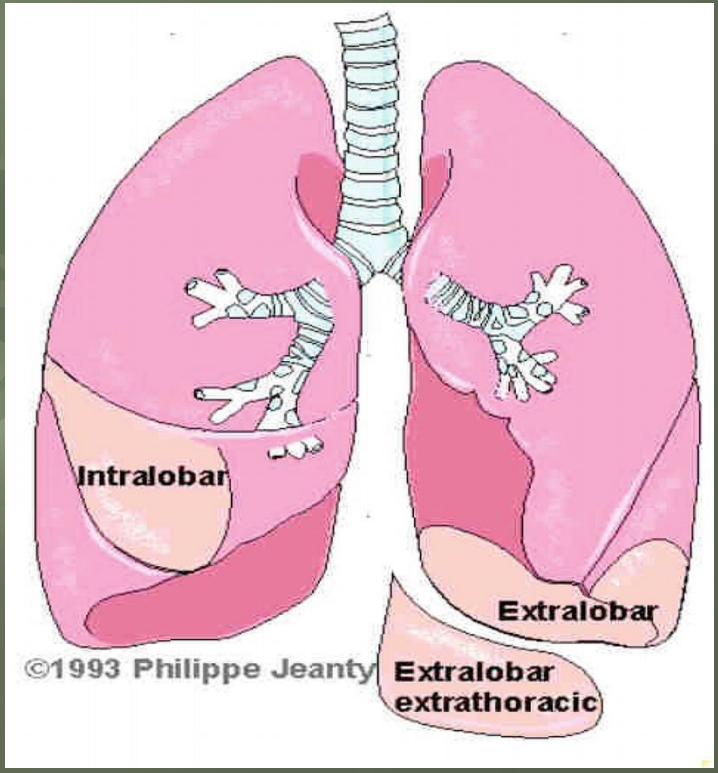
CLINICAL MANIFESTATIONS AND DIAGNOSIS



- **Physical findings in patients with sequestration include:**
  - an area of dullness to percussion
  - decreased breath sounds over the lesion.
  - If infection, crackles may also be present.
  - A continuous or purely systolic murmur may be heard over the back.

If findings on routine chest radiographs are consistent with the diagnosis → **CT with contrast or MR angiography + U\S**


Intralobar Sequestration
Extralobar Sequestration


Intralobar
Extralobar
©1993 Philippe Jeanty
Extralobar
extrathoracic

Intrapulmonary vs. Extrapulmonary

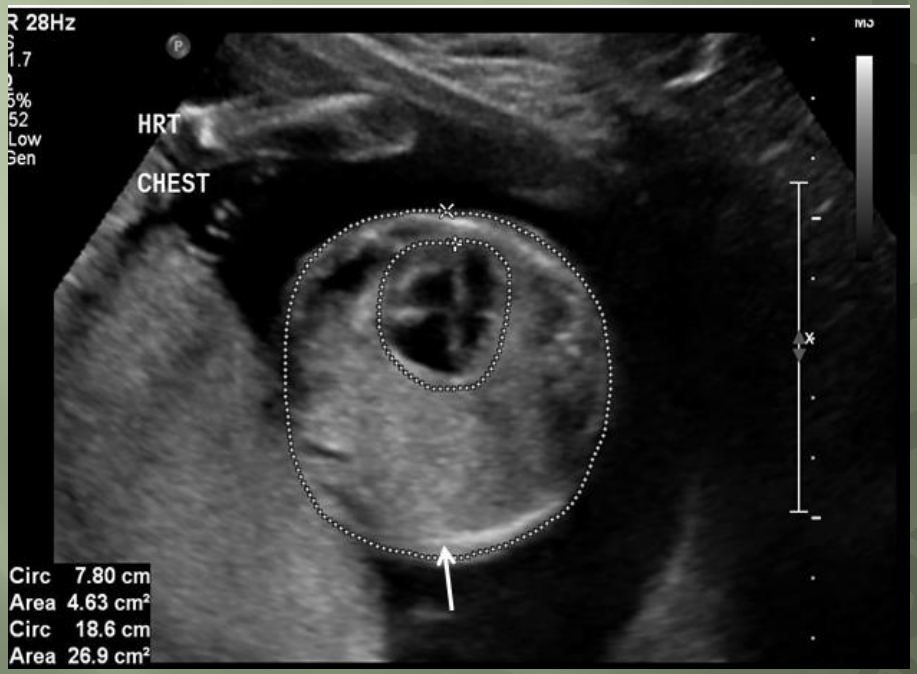
Intrapulmonary

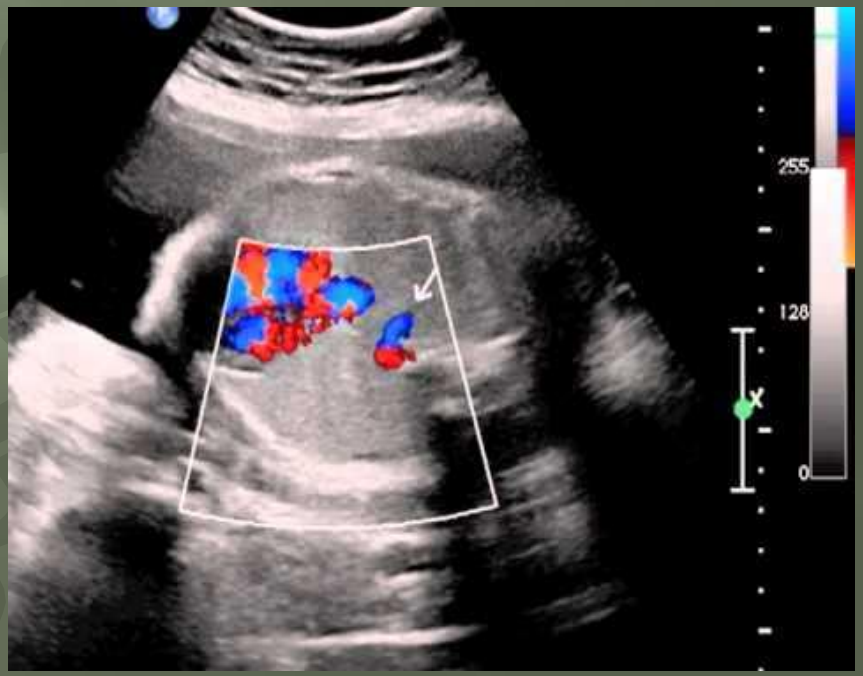
- generally found in a lower lobe
- does not have its own pleura.
- Patients usually present with infection.
- In older patients, hemoptysis is common.
- There is no difference in the incidence of this lesion in each lung.

Extrapulmonary

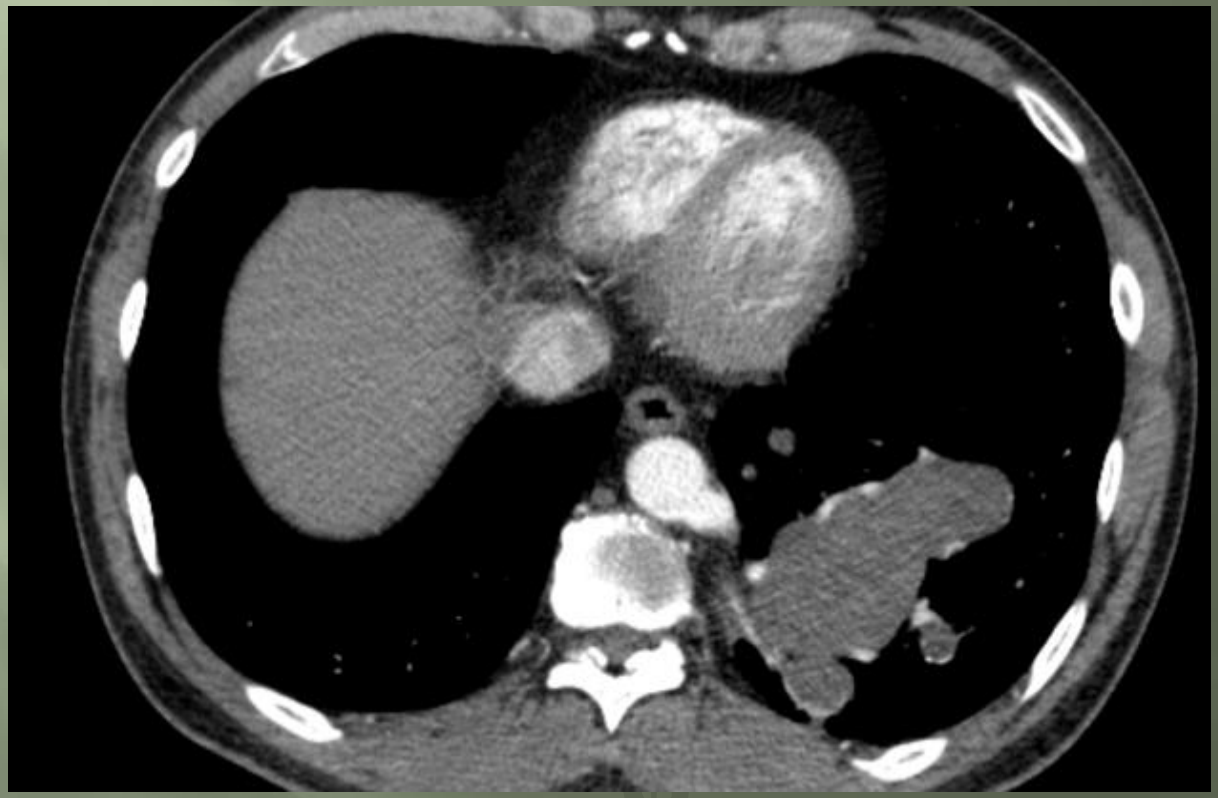
- much more common in boys
- almost always involves the left lung.
- This lesion is enveloped by a pleural covering and is associated with diaphragmatic hernia and other abnormalities such as **colonic duplication, vertebral abnormalities, and pulmonary hypoplasia.**
- Many of these patients are asymptomatic when the mass is discovered by routine chest radiography.
- Other patients present with respiratory symptoms or heart failure.
- Subdiaphragmatic extrapulmonary sequestration can manifest as an abdominal mass on prenatal ultrasonography.

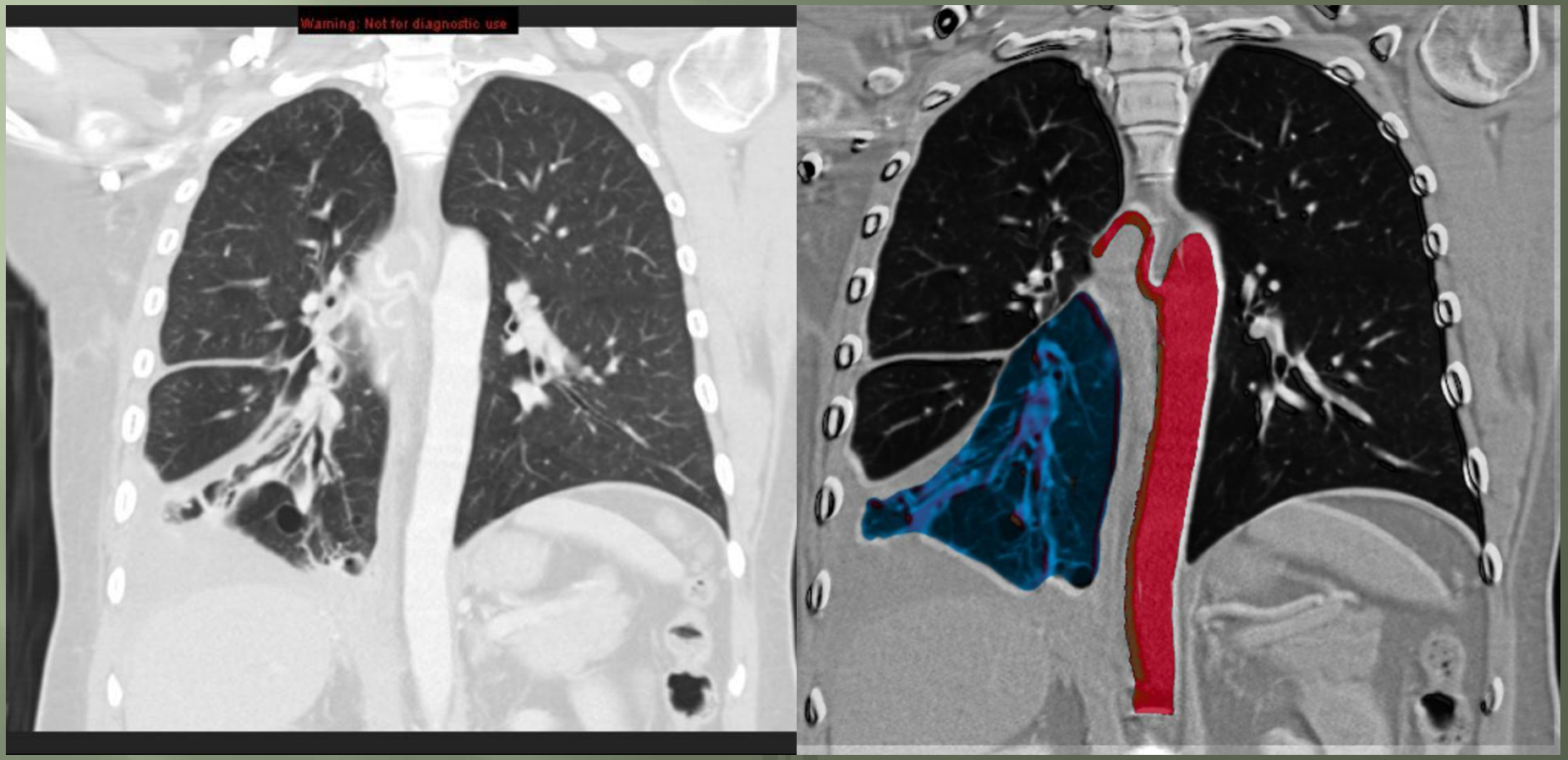
- Diagnosis is usually made prenatally and confirmed on postnatal CT scan





- Postnatal CT scan



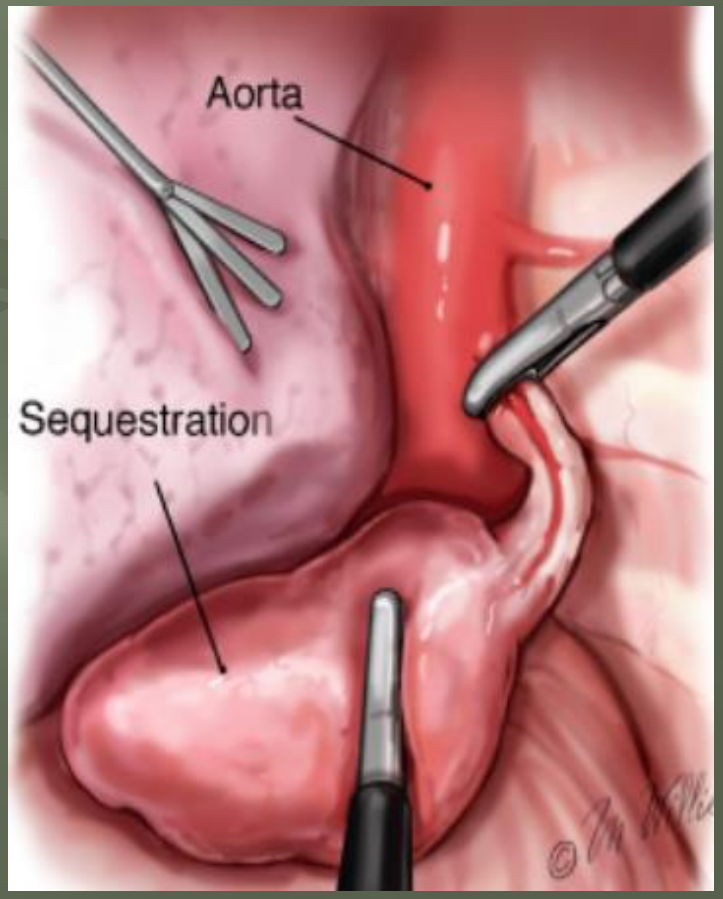

Warning: Not for diagnostic use

Managment

- intrapulmonary →is surgical removal of the lesion, a procedure that usually requires excision of the entire involved lobe, but Segmental resection occasionally suffices.

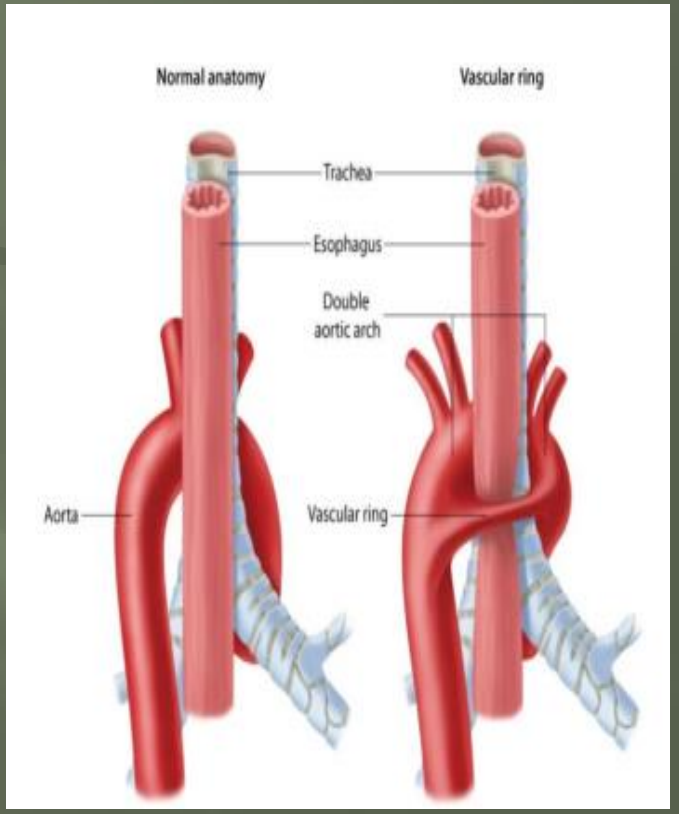
- extrapulmonary sequestration →Surgical resection of the involved area is recommended.

- Coil embolization of the feeding artery has also been successful



Vascular ring

- Vascular rings, or slings, result from abnormal development of the aortic arch, causing tracheal, bronchial, and/or esophageal compression.
  - It is a congenital problem, which means it is present at birth.
  - This happens when certain parts of the aorta that normally disappear during fetal development persist abnormally.
  - They can be either complete (circumferential around the trachea and/or esophagus), such as a double aortic arch, or incomplete (pulmonary artery sling).
  - Vascular rings present in patients age <1 with respiratory (stridor, wheezing, coughing) and/or esophageal (dysphagia, vomiting, difficulty feeding) symptoms.



Clinical Presentation

STRIDOR	**18 (90%)**
NOISY BREATHING	10 (50%)
BRASSY COUGH	12 (60%)
CHOKING EPISODES	10 (50%)
WHEEZES (misdiagnosed asthma)	15 (75%)
INTERRUPTED FEEDING	13 (65 %)
RECURRENT CHEST INFECTION	8 (40 %)
ATTACKS OF CYANOSIS	9 (45%)
RECURRENT VOMITTING	4 (20%)
APNEA	3 (15 %)

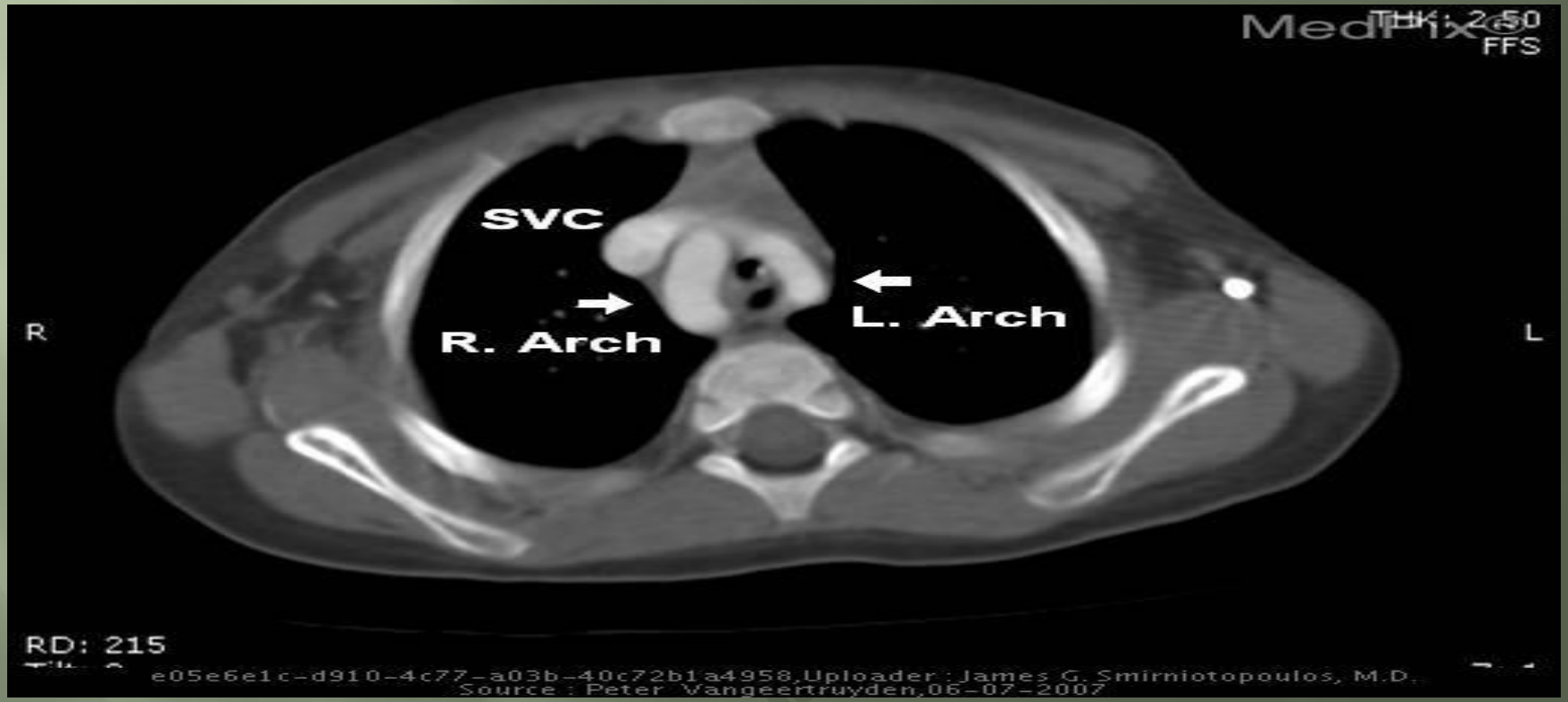
Diagnosis and treatment

- Diagnosis can be made with a CT scan to delineate the precise anatomy forming the vascular ring and evaluate any associated tracheal abnormalities.

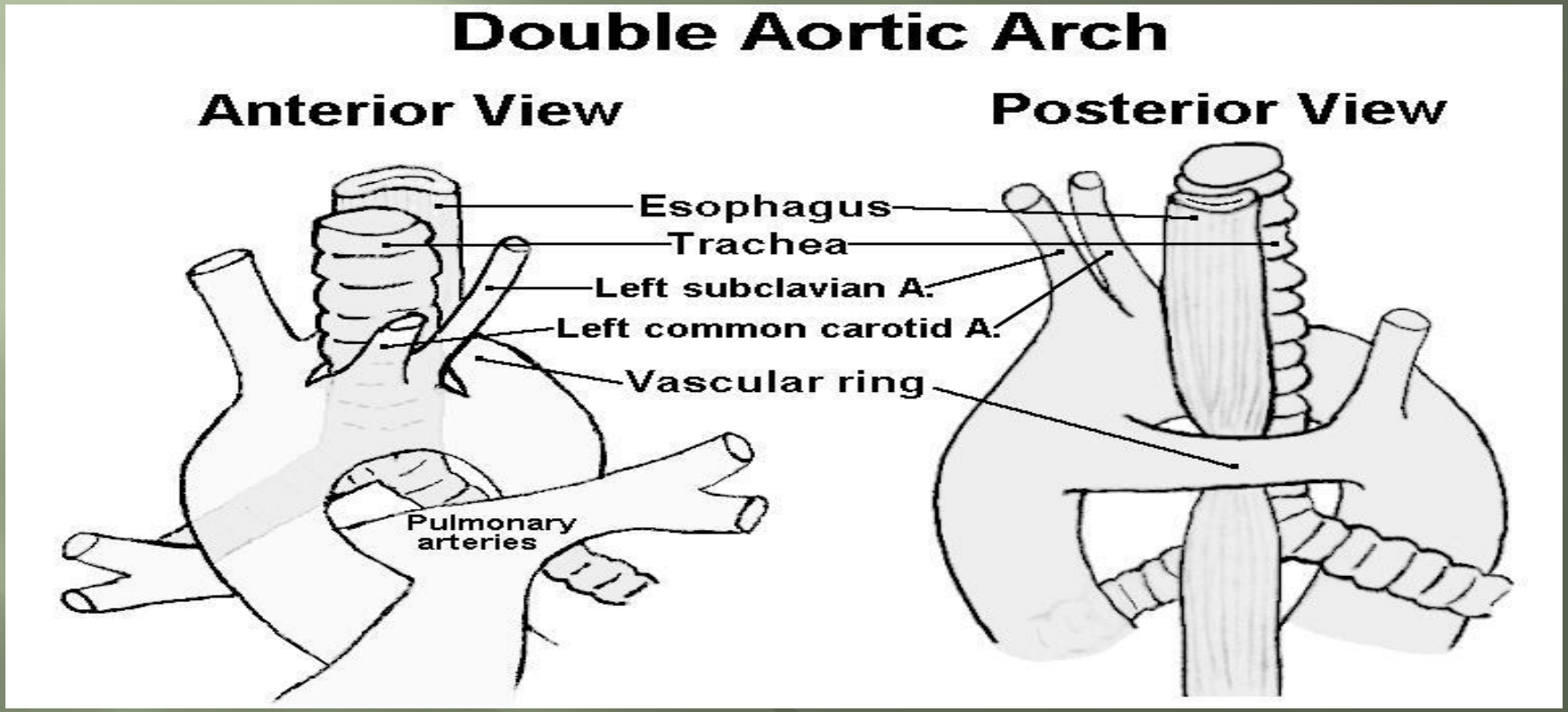
Up to 50% of patients also have a cardiac anomaly (ventricular septal defect, tetralogy of Fallot)

- Due to possible concurrent cardiac and airway abnormalities, all patients require direct laryngoscopy, bronchoscopy, and echocardiogram.

- Treatment is surgical division of the structures creating the ring


SVC
R. Arch
L. Arch
R
L
RD: 215
e05e6e1c-d910-4c77-a03b-40c72b1a4958,Uploader:James G.Smirniotopoulos, M.D.
Source:Peter Vangeertruyden,06-07-2007

Double Aortic Arch



Primary Ciliary Dyskinesia (Immotile Cilia Syndrome, Kartagener Syndrome)

- Primary ciliary dyskinesia (PCD) is an autosomal recessive genetic condition in which the microscopic cells in the respiratory system called cilia do not function normally. Ciliary dysfunction prevents the clearance of mucous from the lungs, paranasal sinuses and ears. Bacteria and irritants in the mucous lead to frequent respiratory infections .
- Kartagener syndrome is a type of Primary ciliary dyskinesia associated with a mirror-image orientation of the heart and other internal organs.

- Motile cilia, the type of cilia impaired in PCD, are found in the lungs, sinuses, ears, ventricles of the brain and the organs of reproduction.
- Vigorously beating motile cilia, working together with airway mucus, provide a critical, first-line defense against unwanted particles (debris, pathogens, etc.) in the airways.
- In PCD, inherited genetic mutations alter the structure or function of motile cilia, dramatically impairing normal ciliary clearance.
- its prevalence in children with repeated respiratory infections has been estimated to be as high as 5%.

- The functional impairment caused by PCD results in:

- Chronic lung disease leading to respiratory failure (chronic sinopulmonary disease) chronic coughing /need for transplant
- Chronic middle ear and sinus infection
- Hearing loss
- Infertility/subfertility (ectopic pregnancy)
- Increased risk of hydrocephalus and retinitis pigmentosa (inherited blindness)

- Motile cilia activity is also required for organ placement during embryonic development.
- About 50% of people with PCD have organ ‘laterality defects,’ meaning their organs are not in the ‘normal’ place.
- Sometimes the organs are in a complete mirror image arrangement called **‘situs inversus.**
- As unusual as it is to have backwards organs, **situs inversus is not generally dangerous.**
- Sometimes in PCD the organs are neither completely reversed nor completely where they should be. This condition, **called heterotaxy or ‘situs ambiguous’ is potentially life threatening.**

DIAGNOSIS

- diagnosed definitively through examination of lung or sinus tissue obtained from a biopsy. (electron microscopy visualization of ciliary abnormalities)
- Screening for levels of nasal nitric oxide is helpful to identify individuals who may have Primary ciliary dyskinesia and should proceed with a biopsy .
- Early diagnosis is important in order to provide prophylactic treatment to prevent or decrease damage to the respiratory system from recurrent infections.

MANAGEMENT

- **AIRWAY CLEARENCE THERAPY** is used to keep the lung tissue healthy for as long as possible. This therapy may include **routine washing and suctioning of the sinus cavities and ear canals. Antibiotics, bronchodilators, steroids and mucus thinners** are also used to treat Primary ciliary dyskinesia .
- **hearing evaluation** is important for young children and speech therapy and hearing aids may appropriate for children with hearing loss and speech problems.
- **Lung transplantation** is an option for severe, advanced lung disease. Surgery may be indicated if heart defects are present.

THANK YOU

- References
  - Nelson textbook
  - Nelson essentials
  - Kaplan pediatrics
  - Medscape