BRONCHIECTASIS AND CYSTIC FIBROSIS

DR MALLESWARI

28.04.22

BRONCHIECTASIS

DEFINITION

- Abnormal permanent dilatation of
- bronchi or subsegmental bronchi with
- inflammatory destruction of peribronchial tissue [muscle, cartilage] with
- accumulation of secretions in the dependent bronchi

- Age group 5-15 Yrs.

- Causes

– Congenital / Hereditary

– Acquired

ETIOLOGY

- In developing countries

Frequently sequelae of acute infection.

- In developed world

cystic fibrosis

immune deficiencies (including HIV)

primary ciliary dyskinesia

recurrent aspiration syndromes.

CONGENITAL CAUSES

CYSTIC FIBROSIS	ABNORMAL IMMUNE FUNCTION	CILIARY DYSFUNCTION
	• **Agammaglobulinemia,** • combined immunodeficiency, • neutrophil function abnormalities	• **Kartageners syndrome, Immotile** • cilia syndrome

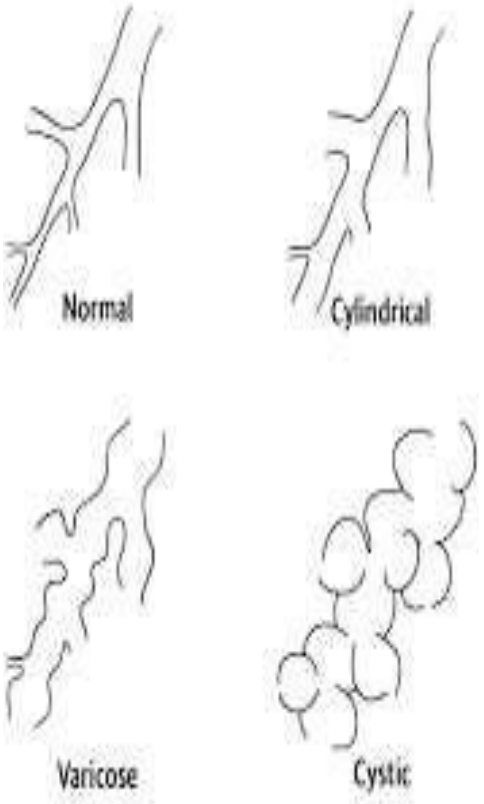
OTHERS: Structural defects, Alpha 1 Anti Trypsin Defeciency

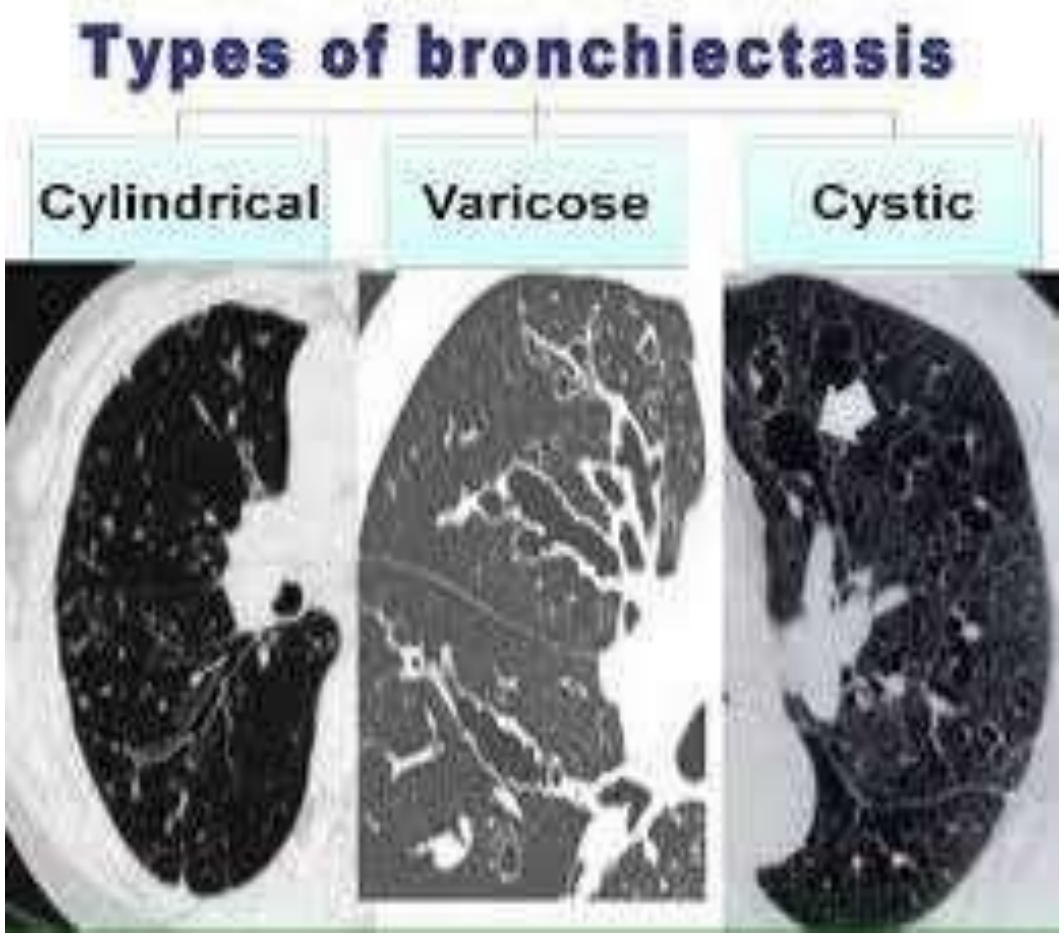
ACQUIRED CAUSES

INFECTION	OBSTRUCTION	ASPIRATION SYNDROME
• BACTERIAL • Tb ,Pertussis, • Mycoplasma • VIRAL • Measles, Viral • Pneumonia, HIV • Chronic lung allograft rejection	• Foreign body • Lymph nodes • Cardiomegaly • Tumors • Endobronchial TB • Inspissated mucus- • [cystic fibrosis, • Immotile cilia syndrome]	• Near drowning • Oropharyngeal • surgery • General anesthesia • Dental extraction • GERD • Hydrocarbon • poisoning

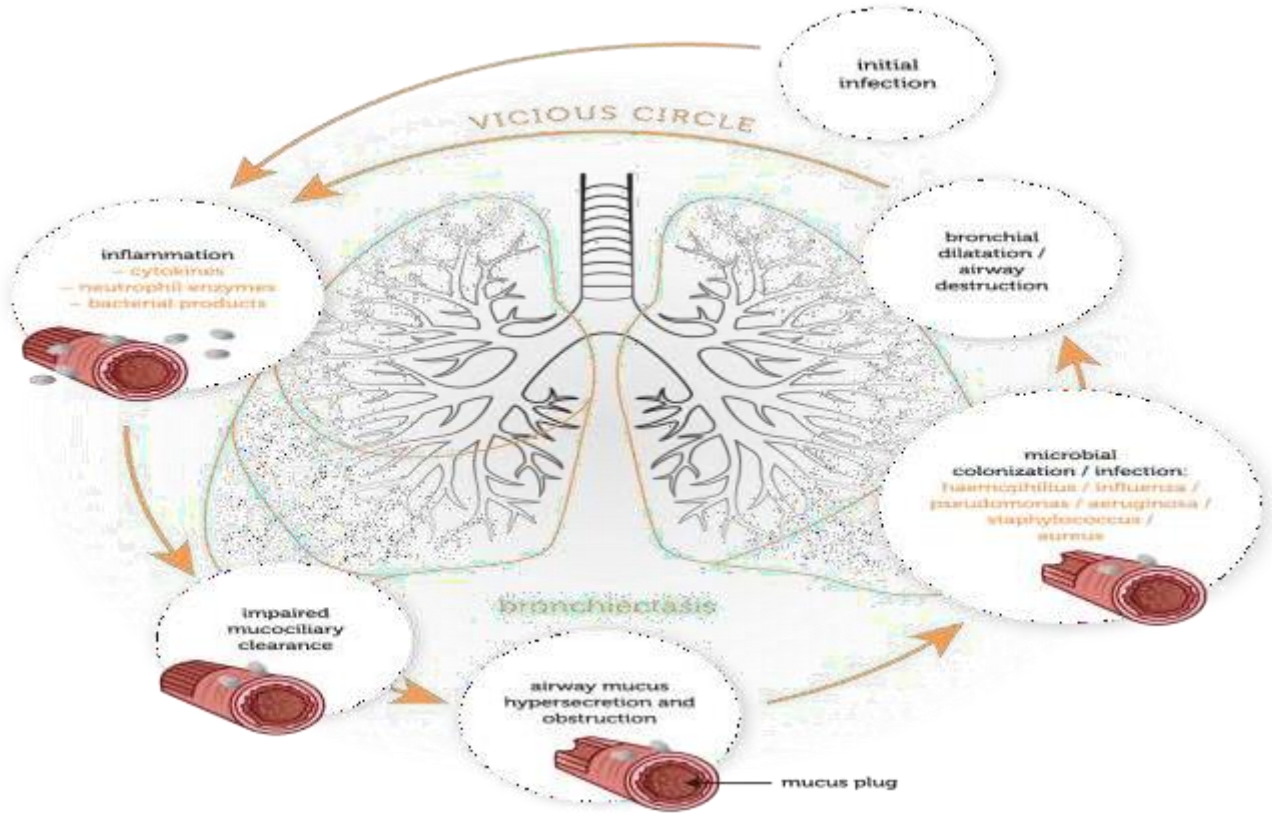
TYPES

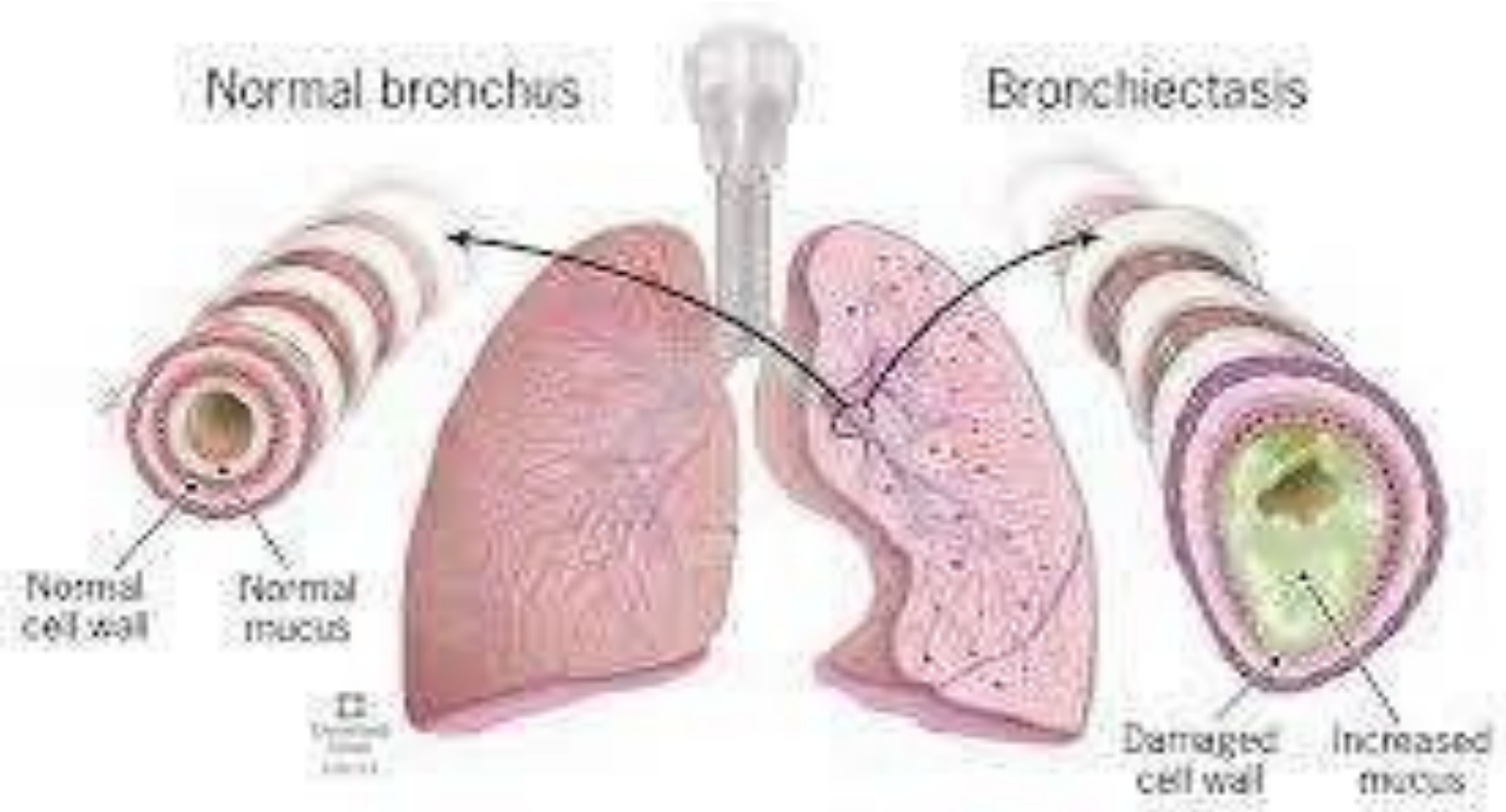
- **Classic ‘Reid’ classification (gross histological**
- appearance) - three different patterns
- **1. Cylindrical bronchiectasis - mildly uniform**
- airway dilation
- **2. Varicose bronchiectasis - focally dilated areas**
- between narrowed segments
- **3. Saccular bronchiectasis - balloon-like airway**
- dilation with more disruption of lung parenchyma


Normal
Cylindrical
Varicose
Cystic


Types of bronchiectasis
Cylindrical
Varicose
Cystic

PATHOGENESIS




Normal bronchus
Bronchiectasis
Normal cell wall
Normal mucus
Damaged cell wall
Increased mucus

CLINICAL FEATURES

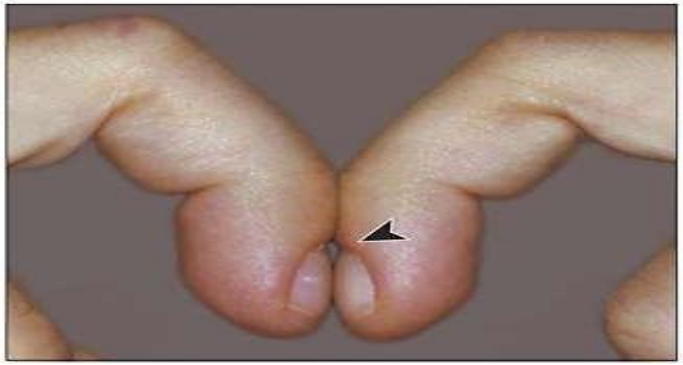
- persistent cough,typically "wet" or productive- most common symptoms, may be absent in young children
- Hemoptysis - uncommon in children, due to erosion of inflamed airway tissue adjacent to pulmonary vessels.
- Dyspnea and exercise intolerance
- Cyanosis- severe disease

CLINICAL SIGNS

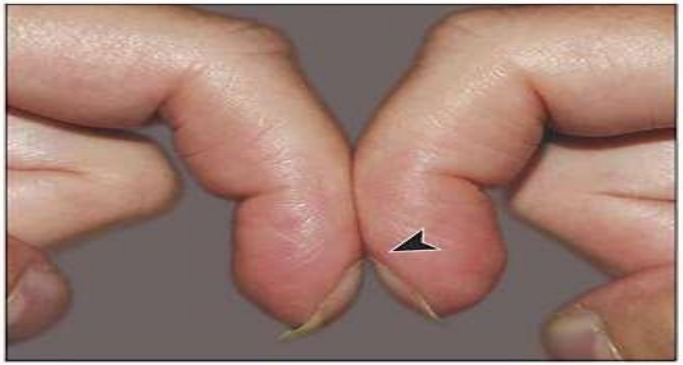
- Failure to thrive
- Sinus and ear infections
- Presence of congenital anomalies
- Clubbing
- Coarse leathery crackle over areas of bronchiectasis

A Schamroth sign

Normal

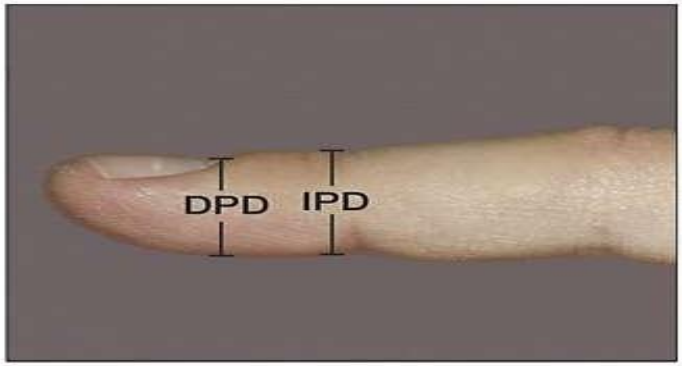


Clubbed

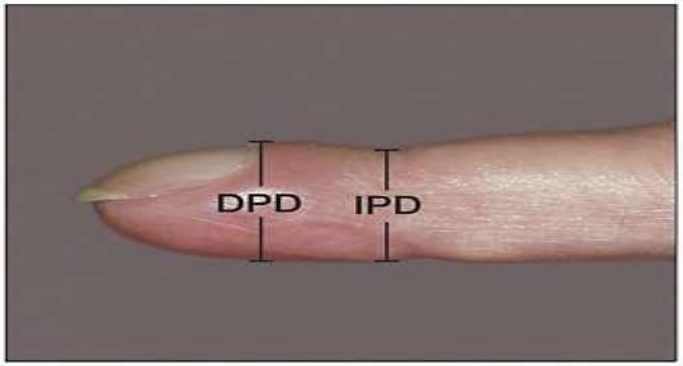


B Phalangeal depth ratio

Normal



Clubbed

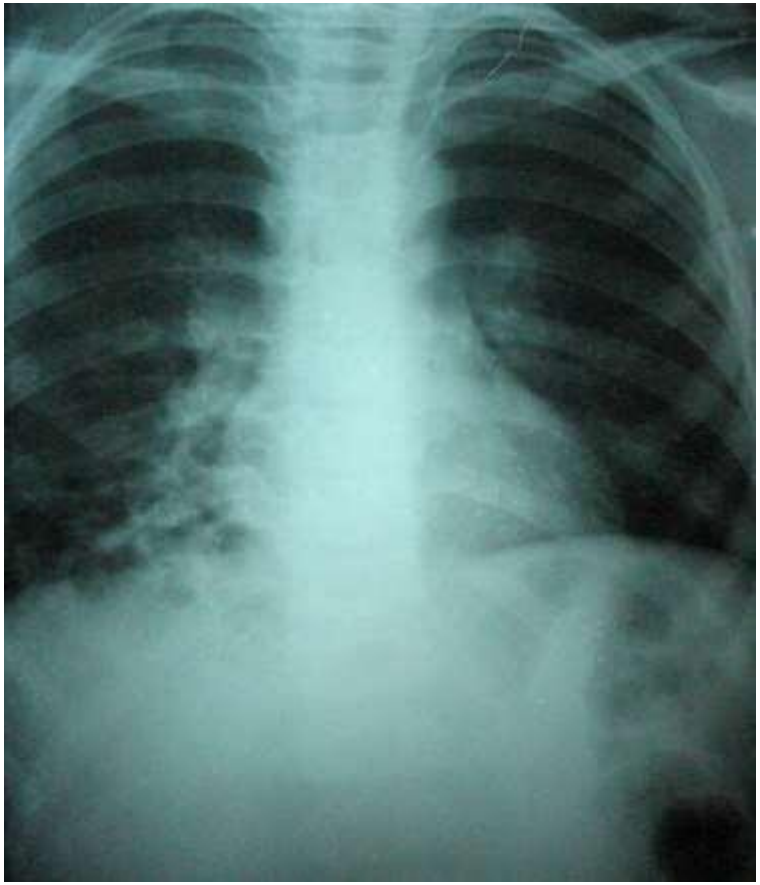


Clinical clues for aetiology

- **Failure to thrive-** cystic fibrosis (CF), immunodeficiency disorders.
- **Chronic sinusitis –** cystic fibrosis, ciliary dysfunction disorders, immunodeficiencies
- **Chronic ear infection with or without otorrhea -**ciliary dysfunction
- **Steatorrhea** — suggests cystic fibrosis.
- **Choking history**— foreign body aspiration or
- swallowing disorder
- **Dextrocardia-** primary ciliary dyskinesia

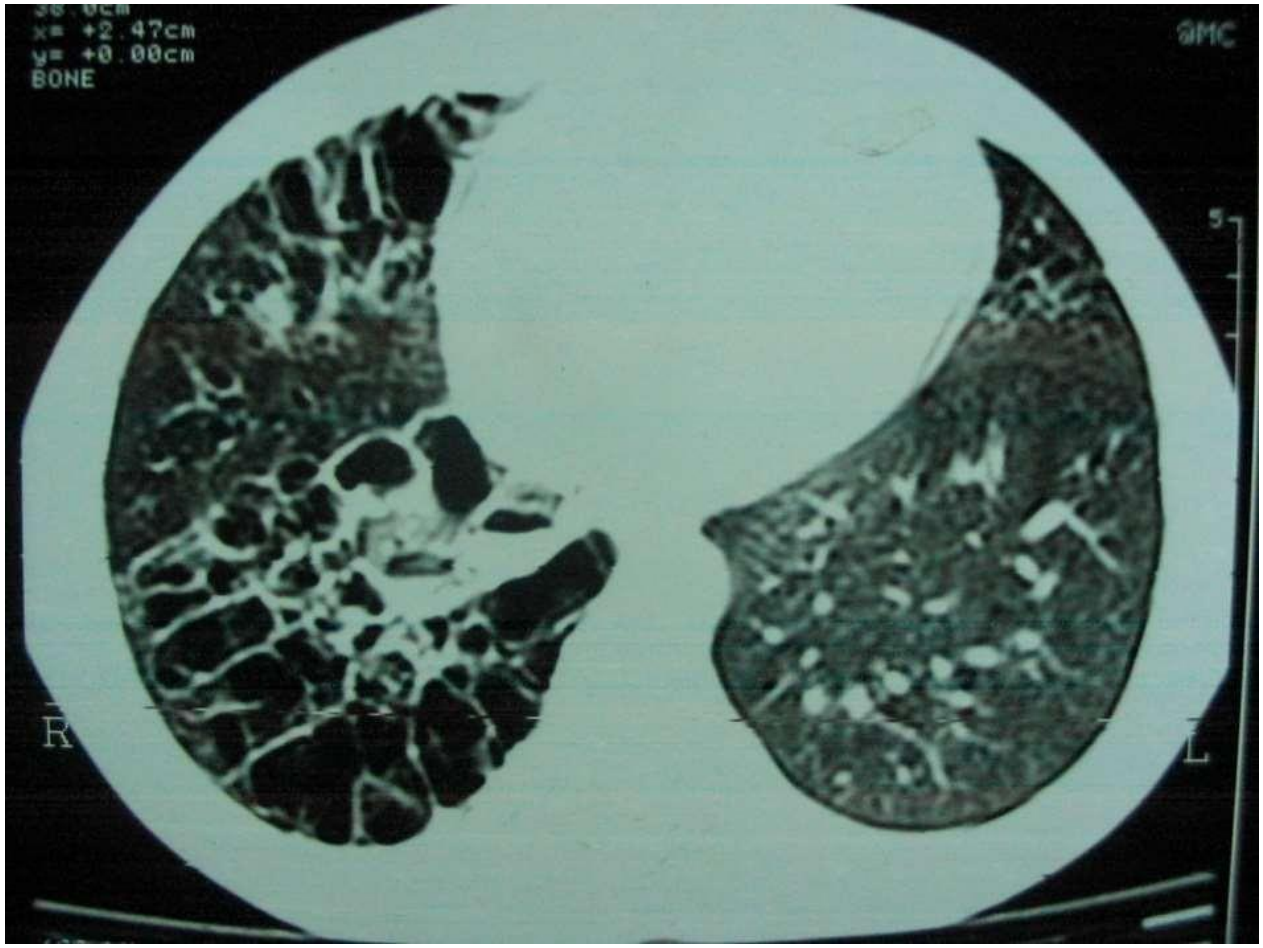
DIAGNOSIS

- Diagnosis depends on radiographically or anatomically visualizing abnormal dilatation of airways
- Diagnostic procedure of choice- **High-Resolution Computed Tomography (HRCT)scan**
- Other tests- diagnose underlying conditions.



- Dilated airways
- Tram track appearance
- Peribronchial fibrosis

Honey comb appearance and signet ring shadows



INVESTIGATIONS

- Complete blood count with differential
- Sputum exam- volume, gram stain, C & S ,
- Immunodeficiency- Total IgM, IgA and IgG
- Tests for Tuberculosis –Mantoux,Chest X-ray, Resting Gastric juice for AFB
- Test for cystic fibrosis (sweat chloride and/or DNA testing)

- Flexible bronchoscopy
- shows structural alteration of bronchial tree
- bronchoalveolar lavage - remove mucus plugs/ to obtain lower airway cultures.
- If an airway foreign body is discovered-rigid bronchoscopy for removal
- HIV screening
- Ciliary biopsy

- Test for GERD : 24 hour pH monitoring, upper gastrointestinal endoscopy and technetium milk scan scintigraphy.
- Test for aspiration due to inadequate airway protective mechanisms during swallowing or dysphagia -evaluated by video fluoroscopy
- Pulmonary function tests-obstructive lung disease
- Test for allergic bronchopulmonary aspergillosis

TREATMENT PILLARS

- Control infection
- Physiotherapy
- Nutritional support
- Identify etiology and treat accordingly

TREATMENT

- Treatment –any identified underlying disorder
- Therapy
  - reduce airway secretions
  - facilitating their removal - with chest physiotherapy & mucolytic agents
- Pharmacotherapy
  - to improve mucociliary clearance.
- Antibiotics
  - prevent & treat recurrent infections
- Surgery- if localized disease -may be considered.

COMPLICATIONS

- Broncho- pneumonia
- Empyema
- Lung abscess
- Hemoptysis
- Metastatic abscess-brain
- Osteomyelitis
- Cor pulmonale
- Amyloidosis

PREVENTION

- Childhood immunization for measles and pertussis
- Screening for tuberculosis and treatment wherever needed
- Aggressive appropriate therapy of lower respiratory tract infections
- Therapy of child with chronic or recurrent respiratory problems due to recurrent aspiration and/or gastroesophageal reflux disease

PROGNOSIS

- In absence of an underlying condition-children with isolated bronchiectasis- good prognosis
- Progressive bronchiectasis from underlying disease or ongoing pulmonary insult –causes progressive obstructive defect & ultimately, respiratory compromise

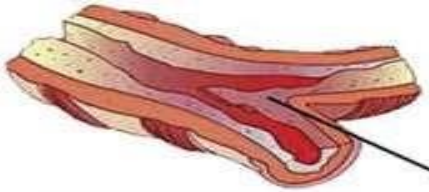
CYSTIC FIBROSIS

DEFINITION

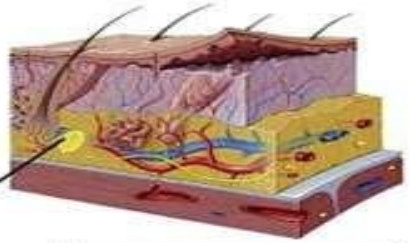
- Autosomal recessive disorder due to a mutation in the CFTR gene leading to failure of chloride conductance by epithelial cells

CLINICAL FEATURES

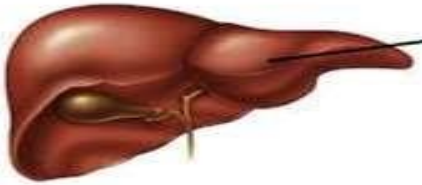
- recurrent chest infections
- malabsorption
- failure to thrive



- Small airways obstruction
- Recurrent respiratory exacerbations

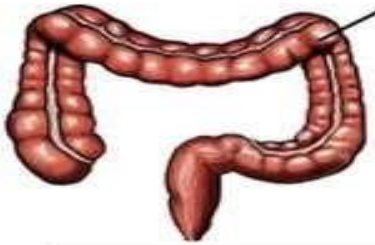


- Excessively salty sweat

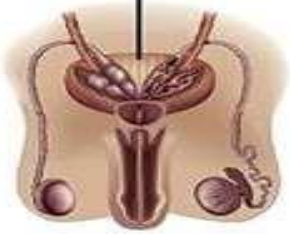


- Biliary cirrhosis
- Gallstones
- Hepatic steatosis

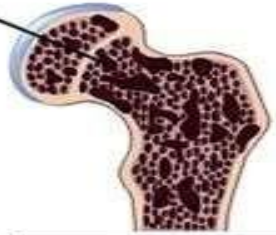
- Exocrine pancreatic insufficiency
- Fat soluble vitamin malabsorption
- Cystic fibrosis-related diabetes



- Meconium Ileus
- Distal intestinal obstruction syndrome
- Fibrosing colonopathy



- Congenital bilateral absence of the vas deferens



- Osteoporosis
- Arthritis
- Hypertrophic pulmonary osteoarthropathy

WHEN TO SUSPECT

- *child presenting with meconium ileus,*
- *recurrent pneumonia*
- *and / or malabsorption of pancreatic origin*
- *hypochloremic metabolic alkalosis,*
- *airway colonization with P. aeruginosa ,*
- *abnormal pancreatic function tests*
- *obstructive azoospermia in post pubertal males.*

DIAGNOSIS

1. demonstration of a high sweat chloride (>60 mEq/L) on at least two occasions or
2. By identifying two CF causing mutations or
3. By suggestive nasal potential difference measurements

TREATMENT

- Respiratory management
- Nutritional care
- Early diagnosis of liver disease, diabetes and other organ dysfunction.
- Airway clearance techniques include adequate hydration, chest physiotherapy and mucolytic agents.
- Antibiotics can be used via intravenous, oral or inhalational route, when needed.

- Low dose azithromycin has immunomodulatory effect.
- Supportive care

A. Increased calorie intake
B. supplementation of fat soluble vitamins
C. replacement of pancreatic enzymes.

REFERENCES

- IAP UG TEACHING SLIDES
- IJPP VOL 14 NO.3
- CLEVECLAND CLINIC, SCIENCE DIRECT (IMAGES)

THANK YOU