# CYSTIC FIBROSIS

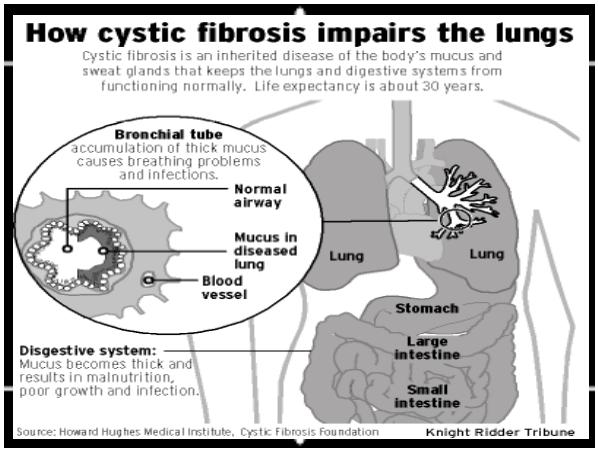
**Cystic fibrosis is an inherited disease of the mucus glands that affects many body systems. In particular, this disorder causes progressive damage to the respiratory system and chronic digestive system problems.**

## CYSTIC FIBROSIS

Cystic fibrosis is a hereditary disorder of the exocrine glands, with a high sodium chloride content in sweat and pancreatic insufficiency resulting in mal-absorption. There is hypertrophy and hyperplasia of mucus secreting glands resulting in excessive mucus production in the lining of the bronchi, which predisposes the patient to chronic bronchopulmonary infection.

## Cystic fibrosis (CF)

- Most common hereditary multisystemic disease
- Autosomal recessive disorder
- Cause: mutations in the single gene on the long arm of chromosome 7 that encodes cystic fibrosis transmembrane conductance regulator (CFTR)

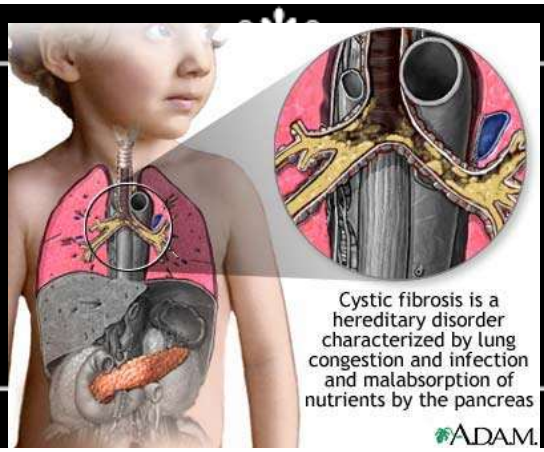



**CF is a disease of exocrine gland function, involving multiple organ systems and chiefly resulting in chronic respiratory infections, pancreatic enzyme insufficiency, and associated complications in untreated patients. Pulmonary involvement occurs in 90% of patients surviving the neonatal period. End-stage lung disease is the principal cause of death.**

**Mucus is a slippery substance that lubricates and protects the linings of the airways, digestive system, reproductive system, and other organs and tissues. In people with cystic fibrosis, the body produces mucus that is abnormally thick and sticky**

This abnormal mucus can obstruct the airways, leading to severe problems with breathing and bacterial infections in the lungs.

**These infections cause chronic coughing, wheezing, and inflammation. Over time, mucus buildup and infections result in permanent lung damage, including the formation of scar tissue (fibrosis) and cysts in the lungs.**




## Clinical features of Cystic Fibrosis

- Chronic Sino-Pulmonary Disease
- Nutritional deficiency
- Electrolyte abnormality – high level of sodium in blood
- Nasal polyps – benign lesions in nasal airways
- Frequent fowl- smelling stools
- Intestinal abnormality
- Hepatobiliary disease
- Pancreatic endocrine dysfunction
- Infertility

## Chronic Sino-Pulmonary Disease

- Chronic infection with CF pathogens
- Endobronchial disease
  - Cough producing copius, often purulent sputum production
  - Air obstruction--- dyspnoea, wheezing; evidence of obstruction on PFTs
  - Digital clubbing - Bulbous swelling at end of fingers
  - Chest x-ray anomalies
- Sinus disease
  - CT or x-ray findings of sinus disease

## Diagnosis of cystic fibrosis

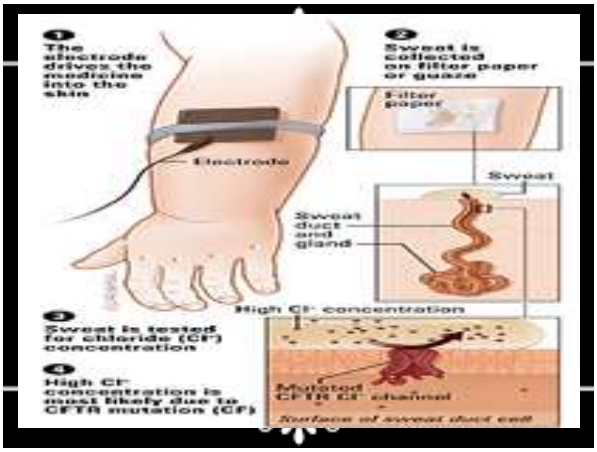
- One or more clinical features of CF

**PLUS**

- Two CF mutations

**OR**

- Two positive quantative pilocarpine iontophoresis sweat chloride values




## Cystic fibrosis---Treatment Multidisciplinary

- Airway Clearance
- Infection
- Nutrition
- Gastrointestinal
- Inflammation
- Infertility
- Social Issues

## Medical Management (Respiratory)

- Antibiotics
- Bronchodilators
- Oxygen therapy
- Mucolytic agents

## Physical therapy Management

Aims:

- To reduce bronchospasm and to clear the lung fields
- To encourage activities for maintaining physical fitness/increase exercise tolerance
- To train postural awareness and relaxation
- To educate the patient in self-management.

## Physical therapy Management

Clearing lung fields:

- Nebulization
- Positioning
- Postural drainage
- Percussion, shaking and vibration

## Physical therapy Management

Increasing exercise tolerance

- Aerobic exercise training
- Physical activities
- Participation in home and school activities

## Physical therapy Management

Postural awareness

Relaxation techniques

- Diaphragmatic breathing and breathing control
- Generalised relaxation techniques