# Cystic Fibrosis

- Cystic - Fluid filled Fibrotic - Fibrous tissue.
- It is a autosomal recessive disorder. with CFTR (Cystic fibrosis Transmembrane conduction regulation) is affected.

Pathology:

CFTR → Pumps chloride ions into the secretions
↓
Chloride ions draws water into the secretions
↓ × → Thickness of secretion.
Thinning of secretion.

Clinical features

1) Newborn → Meconium (first stool gets thick & sticky)
↓
Stuck in intestine
↓
Intestinal obstruction.

2) Childhood.

- Thick secretion from pancreas block the ducts.

↓ No pancreatic enzymes
↓ Protein & Fat not Absorbed.
(Weight gain, Failure to thrive)
↓ Steatorrhea (Fat containing stools)
↓ Cholilithiasis

↓ Inflammation
↓ Acute and chronic Pancreatitis
↓ Fibrosis.
↓ Endocrine damaged.
↓ Insulin dependent Diabetes Mellitus.

3) Later in childhood.

Respiratory System → Recurrent injection (Pneumonia)
↓ Thick mucus
↓ Elasticity lost
↓ Bronchiectasis
↓ Recurrent Respiratory I n.

iv) Adulthood:

- Infertility in Men (Associated with absence of vas deferens).
- Nasal Polyps.

Complication:

- ↑ Pulmonary hypertension → hypertrophy → Cor Pulmonale.
- → Intestinal obstruction, Portal hypertension, Biliary cirrhosis, Cholelithiasis.

Investigation:

↑ Sweat chloride content

Management:

Avoid ~~management~~ marriage between the relatives.