THE STUDY VAULT

Cystic Fibrosis

Respiratory

DISEASE / PATHOLOGY

- Inherited CFTR disorder causes thick, sticky secretions.
- Primary process: mucus obstruction causes chronic infection and impaired absorption.
- Why it matters: affects lungs and pancreas, causing respiratory decline and malnutrition.

RISK FACTORS

- *Who is most at risk?* Children with autosomal recessive inheritance or family history.
- History / exposure: recurrent respiratory infections and pancreatic insufficiency.
- Genetics / lifestyle: poor nutrition worsens disease burden.

SIGNS AND SYMPTOMS

- *Hallmark findings*: chronic cough, thick sputum, recurrent infections, poor weight gain.
- Early: wheezing, steatorrhea, salty-tasting skin, sinus issues.
- Late / urgent: clubbing, dehydration, severe respiratory distress.

DIAGNOSTIC TESTING

- *Tests, labs, imaging, procedures*: newborn screening, sweat chloride test, genetic testing, sputum cultures, chest imaging.
- Gold standard: sweat chloride testing with supportive genetics.
- What confirms it: positive testing with characteristic symptoms.

TREATMENT

- First-line: assess airway clearance and nutrition first.
- Chest physiotherapy, bronchodilators, mucolytics, antibiotics, pancreatic enzymes, and high-calorie diet are common.
- Nursing considerations: monitor growth, stools, hydration, oxygenation, and infection.
- Nursing need-to-know: give pancreatic enzymes with meals and snacks.
- Nursing need-to-know: thick secretions require aggressive airway clearance.

PATIENT EDUCATION

- *Home care*: follow airway-clearance routines and take meds and enzymes as prescribed.
- Increase calories, protein, and fluids as recommended.
- When to call: fever, weight loss, worsening cough, or breathing changes.

ADDITIONAL NOTES:

- NCLEX must know: CF commonly affects lungs and pancreas.
- Red flag: increasing respiratory distress or dehydration.
- Common exam trap: malabsorption and poor growth are high-yield clues.
- Priority action: maintain airway clearance and nutrition.