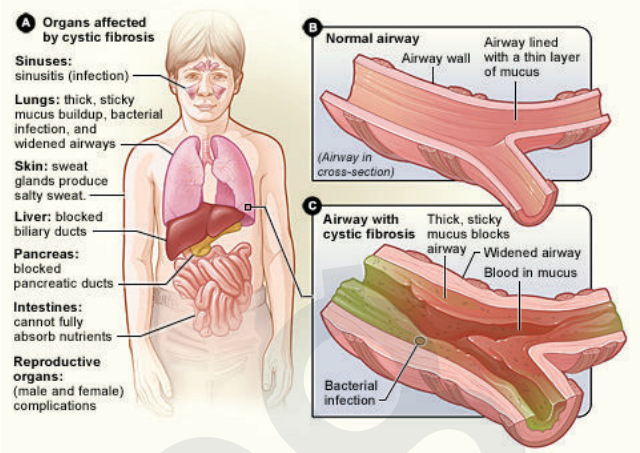
CYSTIC FIBROSIS PATHOCHART

PATHOPHYSIOLOGY

Cystic fibrosis (CF) is an autosomal recessive genetic disorder which causes abnormalities in the secretory glands that produce mucus and sweat and mostly affects the lungs, pancreas, liver, intestines and sex organs. The mucus that is produced in the body becomes thick and sticky. Instead of lubricating the lungs and other organs, it clogs the airways in the lungs and the ducts, most frequently in the pancreas and liver.



ASSESSMENT FINDINGS

- Coughing, non-productive or with thick sputum
- Meconium ileus
- Failure to thrive
- Salty taste to skin
- Oily stools or constipation
- Abdominal distention/pain
- Exercise intolerance
- Respiratory congestion

DIAGNOSTICS

- Newborn screening
- Sweat testing
- Genetic testing

NURSING PRIORITIES

- Ensure Adequate Oxygenation
- Optimize Gastrointestinal Function/Elimination
- Promote Effective Gas Exchange

THERAPEUTIC MANAGEMENT

- Chest physiotherapy
- Monitor for respiratory infections
- Supplemental oxygen
- Mucus clearance devices
- High calorie, high protein diet
- Fat soluble vitamin replacement
- Increase fluid intake
- Assess for intestinal obstruction

MEDICATION THERAPY

- Bronchodilators
- Antibiotics
- Pancreatic enzymes