Table of Contents:

Cystic Fibrosis

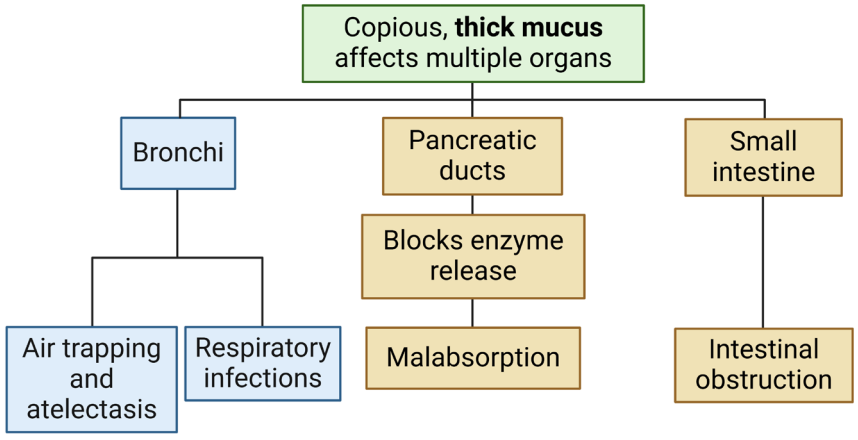
1. Pathophysiology

Cystic fibrosis (CF):

- CF = a **progressive**, **incurable** genetic disorder significantly affecting the respiratory and digestive systems, **shortening life expectancy**.
- CF is an **inherited autosomal recessive** mutation of ion channels that alters chloride and water transport in exocrine (mucus-producing) glands.
  - ✪ The child must inherit **two copies** of the gene (one from **each parent**) to **have CF**.
- Dysfunctional exocrine glands throughout the body block secretion of chloride and water → **Copious, thick, sticky mucus** builds up **in multiple organs** → **Obstructs** small passageways in **organs** (**FIGURE 1**)

✪ Diagnosed with family history, genetic testing (**newborn screening**), and an elevated **sweat chloride test**

FIGURE 1. CYSTIC FIBROSIS PATHOPHYSIOLOGY



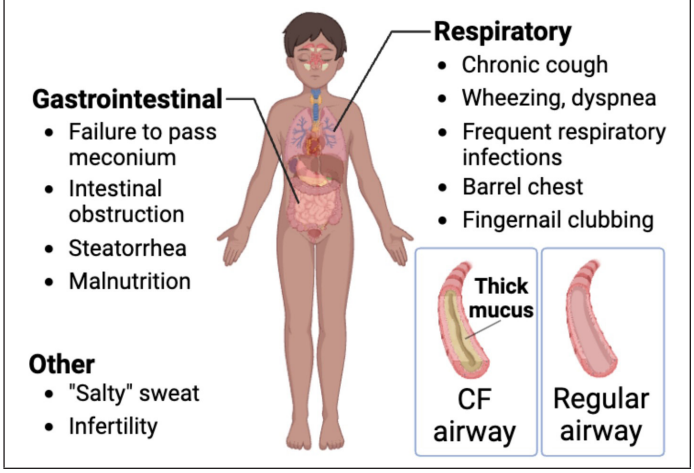
2. Assessment

CF findings are caused by **copious, thick mucus affecting multiple organ systems** (**FIGURE 2**).

✪ **Respiratory:** Thick mucus impairs ventilation and promotes bacterial growth, causing:
  - ✪ Chronic **cough**
  - Wheezing, **dyspnea**
  - ✪ **Frequent respiratory infections** (bronchitis, pneumonia)
  - Signs of **chronic** air trapping and hypoxemia:
    - Barrel chest
    - Fingernail clubbing

- **Gastrointestinal:** Thick mucus obstructs the small intestines and blocks pancreatic enzyme release, causing:
  - ✪ **Failure to pass meconium** (meconium ileus) in newborns
  - **Intestinal obstruction** in older children
  - ✪ **Malnutrition** and growth failure (failure to thrive) from malabsorption and ↑ work of breathing
  - ✪ **Bulky, frothy, foul-smelling stool** (steatorrhea)
- **Other:**
  - **"Salty"** sweat (↑ Na^+ and Cl^-)
  - **Infertility** in both males and females

FIGURE 2. CYSTIC FIBROSIS ASSESSMENT FINDINGS



3. Interventions

Nursing care focuses on:

1. **Mobilizing respiratory secretions** and preventing **respiratory complications**
2. Supporting **nutrition** and monitoring for **GI complications**
3. Providing **psychosocial support**

1. **Mobilize respiratory secretions** and prevent **respiratory complications**:

↑ **risk for respiratory complications** (infection, pneumothorax) due to thick, sticky mucus

- Administer **bronchodilators** and **mucolytics** as prescribed (**TABLE 1**).
- ✪ Perform **airway clearance therapies (ACTs)** daily to **mobilize thick secretions**:
  - **Chest physiotherapy** (CPT) includes (**FIGURE 3**):
    - Chest wall **percussion** or **vibration** (high-frequency chest compression vest)
    - **Postural drainage** (positioning client to drain secretions by gravity)
  - **Positive expiratory pressure** (PEP) devices: ↑ airway pressure to **open the airway** (flutter valve)
  - **Huff coughing**: Coughing technique (forcefully exhale, say "huff") to **expel mucus**
  - ✪ **Avoid** performing ACTs immediately **before** or **after meals** to prevent vomiting.
- ✪ To **prevent respiratory infections**:
  - Encourage **routine immunizations**, including an annual flu vaccine.
  - Promote **meticulous hand hygiene**.
  - ✪ **Avoid close contact** with other hospitalized clients **who also have CF**.
- Immediately notify the HCP for signs of **respiratory complications**:
  - ✪ **Respiratory infection**
    - May display subtle signs like ↓ **activity** and **appetite** (anorexia)
  - **Pneumothorax** (frequent infections damage the lungs)
- To manage **respiratory complications**:
  - Monitor **pulse oximetry**.
  - Give **oxygen** for respiratory distress.
  - Give **antibiotics** to treat infection.

FIGURE 3. CHEST PHYSIOTHERAPY

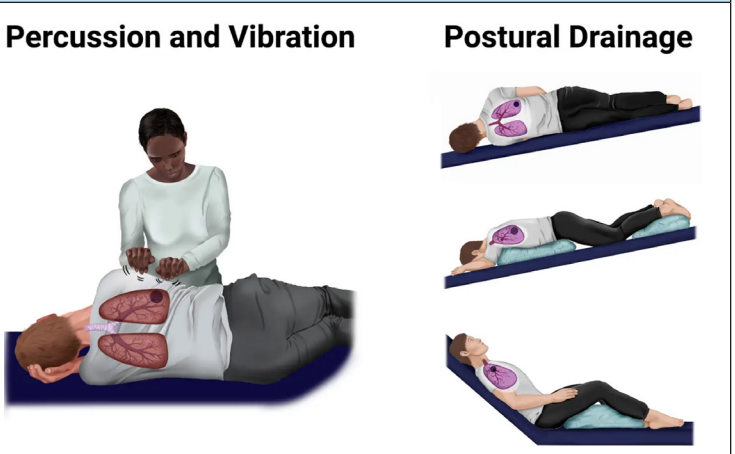


TABLE 1. CYSTIC FIBROSIS MEDICATIONS

Medications & Mechanism	Considerations
Bronchodilators open the airways. • albuterol	✪ Administer **bronchodilators before ACTs** to improve expulsion of secretions. ✪ Give **bronchodilators before mucolytics** to improve delivery of mucolytics.
Mucolytics delivered via nebulizer to thin respiratory mucus • hypertonic saline • recombinant DNase (dornase alpha)	
Pancreatic enzyme replacement therapy (PERT) replaces lipase, protease, and amylase to break down fats, proteins, and carbohydrates. • pancrelipase	✪ Give **within 30 min** of **all meals** and **snacks**. ✪ **Do not crush** or **chew** capsules (destroys enteric-coating). • For clients unable to swallow capsules, open capsules and **sprinkle contents** on **food**.

NCLEX Star Points

❶ **Mobilizing secretions**: Administering **bronchodilators before** performing **airway clearance therapies** (CPT, PEP) helps mobilize secretions.

❷ **Malnutrition**: Provide a **high-calorie**, **high-protein** diet and supplemental **fat-soluble vitamins** (A, D, E, K) to meet increased nutrient requirements.

3. Interventions, Continued

2. Support **nutrition** and monitor for **GI complications**:

   Clients with CF require **additional nutrients** and **vitamins** due to **malabsorption** of fats, proteins, and carbohydrates and ↑ metabolic demand.

   - Promote a **high-protein**, **high-calorie**, unrestricted fat diet.
     - Encourage ↑ **salt** and **fluid intake** during hot weather and exercise.
   - Administer supplemental pancreatic enzymes (**PERT**) **within 30 min** of **eating** (see **TABLE 1**).
   - Administer supplemental **fat-soluble vitamins A, D, E, and K** (↓ absorption of fat and fat-soluble vitamins).
   - Immediately **notify HCP** for signs of **GI complications**:
     - **Intestinal obstruction**
       - Abdominal **pain** and **distension**
     - **Diabetes mellitus** (pancreatic obstruction damages insulin-producing beta-cells)

3. Provide **psychosocial support**:

   CF is a **progressive**, **incurable** condition that can **cause stress** and **disrupt** normal childhood **development**.

   - Promote **goal-setting** and allow client to **take part in care** activities (ACT, PERT) to foster independence.
   - Connect clients and families with **community resources** for support (CF Foundation).
   - Encourage age-appropriate **physical activities** like **sports** (as tolerated) to improve lung function and ↑ peer interaction.
   - Address concerns about **fertility** and **childbearing**.

NCLEX Star Points

❸ **Notify HCP for complications**: **Decreased activity** and **decreased appetite** are subtle signs of **respiratory infection**. **Abdominal pain and distension** are signs of **intestinal obstruction**.

❹ **Pancreatic enzyme replacement therapy (PERT)**: Administer **PERT within 30 minutes of eating** to improve absorption of **fats**, **proteins**, and **carbohydrates**.

❺ **Psychosocial support**: Encourage participation in **physical activities** like sports as tolerated and connect the client and family with **community resources** like support groups.

NCLEX Top 5 Targets

1. To mobilize respiratory secretions, the nurse should **administer** _____ **(which type of medication?) before** performing _____ **therapies**.
2. Signs of **intestinal obstruction** include **abdominal** _____ and _____. Subtle signs of **respiratory infection** include _____ and _____.
3. Clients with CF require a **special diet** that is **high-** _____ and **high-**_____. They also require **supplemental fat-soluble vitamins** _____, _____, _____, and _____.
4. To optimize **nutrient absorption**, the client should take _____ **(which medication?) within** _____ **minutes of eating**.
5. **True or False**: The nurse should **encourage** clients with CF to **participate in sports**.

Answers: 1. bronchodilators, mucolytics, clearance 2. distention, pain, decreased activity, anorexia 3. calorie, protein, A, D, E, K
4. pancreatic enzyme replacement therapy, 30 5. True

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