CYSTIC FIBROSIS

PEDIATRIC NURSING • NGN NCLEX HIGH-YIELD REVIEW

★ 2026 NCLEX TEST PLAN STANDARD ★

AUTOSOMAL RECESSIVE disorder caused by a mutation in the CFTR gene (chromosome 7) → defective chloride/sodium transport → thick, sticky mucus clogs lungs, pancreas, GI tract & reproductive organs. Most common lethal genetic disease in Caucasian children.

🎯 MULTI-SYSTEM INVOLVEMENT

RESPIRATORY
Chronic cough, wheezing, recurrent infection

GI / PANCREAS
Steatorrhea, malabsorption, FTT

SKIN
Salty taste when kissed (hallmark)

SKELETAL
Clubbing, barrel chest, delayed growth

REPRODUCTIVE
Male infertility, delayed puberty

PATHOPHYSIOLOGY

Both parents = carriers → 25% child affected, 50% carrier, 25% unaffected

- **CFTR gene** mutation → defective chloride channel
- ↓ Cl^- and H_2O secretion → mucus becomes thick/dehydrated
- Mucus obstructs airways, pancreatic ducts, bile ducts
- Leads to chronic infection, fibrosis, organ failure

CLINICAL SIGNS

- **Salty-tasting skin** (classic/hallmark sign) 🧂
- **Meconium ileus** in newborn (first sign!)
- Steatorrhea: bulky, foul, fatty, frothy stools
- Failure to thrive despite big appetite
- Chronic productive cough, wheezing, crackles
- Recurrent pneumonia (*Pseudomonas, S. aureus*)
- Clubbing of fingers, barrel chest
- Rectal prolapse, nasal polyps

SWEAT CHLORIDE TEST — GOLD STANDARD DX

Pilocarpine iontophoresis stimulates sweat → chloride measured. Done on **2 separate occasions**.

Normal: < 40 mEq/L | **Borderline: 40-59** | **POSITIVE: ≥ 60 mEq/L**

DIAGNOSTICS

- **Newborn screen:** ↑ IRT (immunoreactive trypsinogen)
- **Sweat chloride ≥ 60 mEq/L** = diagnostic
- Genetic testing: CFTR mutation (ΔF508 most common)
- Chest X-ray: hyperinflation, atelectasis
- Pulmonary function tests (PFTs)
- 72-hr fecal fat: elevated
- Sputum culture for *Pseudomonas*

PRIORITY NURSING CARE

- **CPT / Vest therapy** 2-4x/day — **BEFORE meals** (prevents vomiting)
- Postural drainage + percussion after bronchodilator
- Encourage coughing, deep breathing, exercise
- ↑ Fluids to thin secretions
- Monitor O_2 sat, resp status, sputum color
- Strict infection control — avoid sick contacts
- Cluster care — conserve energy
- Emotional support: lifelong chronic illness

MEDICATIONS

- **Pancreatic enzymes** (pancrelipase/Creon) — **WITH every meal & snack**
- Do NOT crush/chew enzymes; sprinkle on applesauce (NOT milk/hot food)
- **Dornase alfa (Pulmozyme)** — thins DNA in mucus
- Hypertonic saline (3-7%) nebulizer
- Bronchodilators: albuterol BEFORE CPT
- Inhaled/IV antibiotics (tobramycin)
- **CFTR modulators:** ivacaftor, Trikafta (elexacaftor/tezacaftor/ivacaftor)

NUTRITION

- **HIGH calorie** (150% RDA) — growth priority
- **HIGH protein, HIGH fat** diet
- **Fat-soluble vitamins: A, D, E, K** (water-miscible)
- Extra **salt** added to food — especially in heat/exercise
- ↑ Fluids — prevent dehydration & thick mucus
- Small, frequent meals
- G-tube feedings may be needed for FTT

MNEMONIC: REMEMBER "CF PATIENT"

C Chronic cough	**F** Failure to thrive	**P** Pancreatic insufficiency	**A** Alkalosis / salt loss
T Thick mucus	**I** Infections (Pseudomonas)	**E** Enzymes w/ meals	**N** Nutrition — ↑cal/fat
T Transplant (end-stage)	Salty sweat + steatorrhea + chronic lung dz = think CF!		

COMPLICATIONS TO MONITOR

- **Respiratory failure** (#1 cause of death)
- Pneumothorax, hemoptysis
- CF-Related Diabetes (CFRD)
- Cor pulmonale
- DIOS (distal intestinal obstruction)
- Liver disease, cirrhosis
- Osteoporosis, delayed puberty
- Infertility (males: absent vas deferens)

★ NGN NCLEX HIGH-YIELD PEARLS ★

- **Enzymes WITH meals**, not after. If dose missed → do NOT double-up.
- **CPT BEFORE meals** or 1 hr after — prevents vomiting.
- Add **extra SALT** to diet — especially in hot weather/fever/exercise.
- **Sweat Cl^- ≥ 60 mEq/L** on 2 tests = diagnostic.
- **Pseudomonas aeruginosa** = most common chronic lung pathogen.
- **Meconium ileus** in newborn → suspect CF immediately!
- Two carrier parents → **25% chance** child has CF.
- Diet: **HIGH calorie, HIGH protein, HIGH fat** + ADEK vitamins.
- AVOID cough suppressants — need to clear mucus!
- Strict **cohort isolation** — CF pts should NOT mingle (cross-infection).