GENERAL

cystic fibrosis hesi case study

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Cystic Fibrosis HESI Case Study: A Comprehensive Guide for Nursing Students

Preparing for the Health Education Systems, Inc. (HESI) exam can feel overwhelming, especially when you're tackling complex medical conditions like cystic fibrosis (CF). A well structured case study not only tests your knowledge but also sharpens your clinical reasoning and patient centered care skills. This article presents a detailed CF HESI case study, breaking down the patient's presentation, pathophysiology, nursing interventions, and critical thinking questions. By the end, you'll be equipped to approach the exam confidently and provide top quality care to individuals with CF.

Understanding Cystic Fibrosis: A Quick Overview

What is Cystic Fibrosis?

Cystic fibrosis is an inherited, autosomal recessive disorder that primarily affects the lungs, pancreas, and other organs. It is caused by mutations in the CFTR (cystic fibrosis transmembrane conductance regulator) gene, which encodes a protein responsible for chloride and bicarbonate transport across epithelial cells. Defective CFTR function leads to thick, sticky mucus that obstructs ducts and airways, resulting in chronic infections, malabsorption, and multi system complications.

Key Clinical Features

- Respiratory: Chronic cough, recurrent pneumonia, bronchiectasis, and progressive lung function decline.
- Digestive: Pancreatic insufficiency, steatorrhea, failure to thrive, and malnutrition.
- Reproductive: Infertility in males due to congenital bilateral absence of the vas deferens.
- Other: CF-related diabetes, liver disease, and electrolyte imbalances.

Diagnostic Criteria

Diagnosis is confirmed through a combination of sweat chloride testing, genetic analysis, and clinical presentation. Newborn screening programs in many countries now enable early detection and intervention, which significantly improve outcomes.

HESI Exam: Why CF Case Studies Matter

The HESI exam evaluates a nursing student's knowledge across core domains: medical surgical, pharmacology, pathophysiology, nursing fundamentals, and patient safety. CF case studies test your ability to integrate these areas, especially:

- Understanding complex disease mechanisms.
- Applying pharmacologic treatments such as inhaled bronchodilators, mucolytics, and antibiotics.
- Implementing respiratory therapy protocols.
- Managing nutrition and pancreatic enzyme replacement.
- Addressing psychosocial and developmental aspects of chronic illness.

The Case Study: Patient Profile

Patient Information

Name: Emily R. **Age:** 12 years **Gender:** Female **Chief Complaint:**Persistent cough and shortness of breath for the past 3 weeks.

Medical History

- Diagnosed with cystic fibrosis at 4 months via newborn screening.
- History of recurrent respiratory infections; last hospitalization 2 months ago.
- Pancreatic enzyme supplementation (pancrelipase) daily.
- On inhaled dornase alfa (Pulmozyme) and albuterol nebulizations twice daily.
- Recent chest X ray shows mild bronchiectasis.

Social History

Emily lives with her parents and younger brother. She attends school and participates in adaptive sports. The family has a strong support network, but the financial burden of treatments is significant.

Presenting Symptoms

- Productive cough, green sputum.
- Shortness of breath on exertion.
- Fatigue and decreased appetite.
- Weight: 35 kg (below 5th percentile for age).

Assessment and Physical Examination

Vital Signs

- Temperature: 38.2 /°C (100.8 /°F)
- Heart rate: 110 bpm
- Respiratory rate: 28 breaths/min
- Oxygen saturation: 92% on room air, 96% with 2 /L/min nasal cannula.

General Appearance

Emily appears fatigued, with a slight nasal flaring. She is in mild respiratory distress but is alert and oriented.

Respiratory System

- Clear breath sounds at the base of both lungs.
- Coarse crackles and occasional wheezes throughout the lung fields.
- Use of accessory muscles noted during deep breaths.

Abdominal Examination

Soft, non tender, no organomegaly. Bowel sounds are normal.

Skin and Extremities

Dry skin, mild xerosis. No clubbing or cyanosis observed.

Laboratory and Diagnostic Findings

- Chest X ray: Mild bronchiectasis, increased bronchial markings.
- Complete blood count: White blood cell count 15,000/mm³ with neutrophil predominance.
- Basic metabolic panel: Mild hypokalemia (3.2 /mmol/L).
- Sweat chloride test: 85 /mmol/L (diagnostic range > 60 /mmol/L).

- Pulmonary function tests (PFTs): FEV1 60% predicted, FVC 70% predicted.

Pathophysiology Recap

In cystic fibrosis, defective CFTR proteins lead to impaired chloride transport and increased sodium reabsorption. This results in dehydrated, viscous secretions in the airways. The thick mucus promotes bacterial colonization, especially by Pseudomonas aeruginosa and Staphylococcus aureus. Chronic infection and inflammation cause structural lung damage, bronchiectasis, and decreased pulmonary function.

In the gastrointestinal tract, pancreatic ducts become obstructed, leading to exocrine pancreatic insufficiency. The lack of digestive enzymes causes malabsorption of fats and fat soluble vitamins, contributing to malnutrition and growth delays.

Nursing Diagnosis and Priorities

Primary Nursing Diagnoses

1. Impaired airway clearance related to excess mucus production and bronchial inflammation.
2. Risk for infection related to chronic colonization with Pseudomonas aeruginosa.
3. Imbalanced nutrition: less than body requirements related to malabsorption.
4. Anxiety related to chronic illness and school absenteeism.
5. Risk for deficient fluid volume related to sweat chloride loss and dehydration.

Priority Interventions

- Implement airway clearance techniques and monitor effectiveness.
- Administer prescribed antibiotics and monitor for side effects.
- Ensure adequate caloric intake and pancreatic enzyme dosing.
- Provide emotional support and involve a multidisciplinary team.
- Monitor hydration status and electrolytes closely.

Comprehensive Nursing Care Plan

1. Airway Clearance

- Perform chest physiotherapy (postural drainage, percussion) before inhaled mucolytics.
- Administer nebulized dornase alfa (Pulmozyme) as prescribed to reduce sputum viscosity.
- Encourage deep breathing exercises and incentive spirometry twice daily.
- Assess sputum color, volume, and consistency; report changes promptly.

2. Respiratory Management

- Administer albuterol nebulizations for bronchodilation.
- Use high flow oxygen therapy if SpO , drops below 92%.
- Monitor peak expiratory flow rate (PEFR) and record trends.
- Educate Emily on proper inhaler technique and adherence to medication schedule.

3. Infection Control

- Initiate intravenous antibiotics (e.g., tobramycin) based on culture and sensitivity.
- Maintain strict hand hygiene and isolate if necessary to prevent cross infection.
- Encourage handwashing before meals and after bathroom use.
- Educate family on preventing transmission of Pseudomonas to others.

4. Nutrition and Pancreatic Enzyme Replacement

- Collaborate with dietitian to design high calorie, high fat meals.
- Ensure pancreatelipase dosing is appropriate (e.g., 2500–5000 units per meal).
- Supplement fat soluble vitamins (A, D, E, K) as needed.
- Monitor weight gain and adjust caloric intake accordingly.

5. Fluid and Electrolyte Management

- Encourage oral fluid intake ("e 2 /L/day) unless contraindicated.
- Administer potassium supplements if hypokalemia persists.
- Monitor daily weights and intake/output charts.
- Educate family on signs of dehydration (dry mucous membranes, decreased urine output).

6. Psychosocial Support

- Facilitate support group meetings for patients and families.
- Refer to counseling services for anxiety and coping strategies.
- Coordinate with school nurse to develop an individualized health plan.
- Encourage participation in adaptive sports to maintain physical activity.

Pharmacologic Management: Key Medications

Inhaled Therapies

- Dornase Alfa (Pulmozyme): Degrades DNA in sputum, decreasing viscosity.
- Albuterol: Short acting ;" " vöæ—7B `or bronchodilation.
- Azithromycin: Long term anti inflammatory effect against Pseudomonas.

Oral Therapies

- Pancrelipase: Enzyme replacement to aid digestion.
- High dose vitamin supplementation: Addresses malabsorption.
- CFTR modulators (e.g., ivacaftor, lumacaftor): For specific mutations to improve chloride transport.

Antibiotics

- For acute exacerbations: tobramycin, ceftazidime, or piperacillin/tazobactam based on culture.
- Long term suppression: azithromycin or inhaled colistin.

Critical Thinking Questions for HESI Preparation

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