

# Care of the Adult Patient with Cystic Fibrosis



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Author Talk

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# Why write the article?

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- By 2005, adults with CF > 10,000, and > 40% of total CF population
- Significant improvements in basic scientific discoveries and clinical care
- Recently published consensus statements and conference proceedings from CF Foundation
- Few comprehensive articles in nursing literature about the care of the adult with CF
- Collaboration between experienced writer and experienced nurse in a well-established adult CF center



# Format of the article

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- Target audience, target journal
- Evidence-based
- CF multi-system genetic disease
- Comprehensive, multidisciplinary care
- Diagnosis→end of life issues and transplantation
- Detailed case study of a real patient



# About CF

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- Inherited, autosomal-recessive disease
- Disrupts ion transport in epithelial-lined organs
- Protein product of CF gene identified
  - Cystic fibrosis transmembrane conductance regulator (CFTR)
  - CFTR involved in transport of Cl and Na across the epithelial lining of ducts of exocrine glands
  - Most common mutation  $\Delta F508$



# Diagnosing CF

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- Phenotypic features consistent with CF
  - Chronic sinopulmonary disease characteristics
  - Gastrointestinal/nutritional abnormalities
  - Salt loss syndromes
  - Male urogenital abnormalities



# Confirming the diagnosis

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- Elevated sweat chloride concentration (>60mmol/L)
  - Quantitative pilocarpine iontophoresis sweat test
- Identifications of CF mutation in each CFTR gene
- Demonstration of abnormal ion transport across nasal epithelium



# Genetic considerations

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- Inherited autosomal-recessive disease
- Genetic testing recommendations



# Pulmonary characteristics

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- Impaired airway mucus clearance
- Chronic inflammation and infection
- Airway wall thickening, persistent neutrophilic infiltration
- Plugging of small and large airways with mucus
- Chronic dilatation of the airways and enlarged bronchial arteries
- Pulmonary function declines ( $FEV_1$ ) exercise intolerance and exacerbations



# Pulmonary treatment

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- Identification and susceptibility patterns of pathogens (synergy testing)
- 2 IV antibiotics during exacerbations
- Aerosol administration of antibiotics (Tobramycin)
- Airway clearance techniques
- Bronchodilators, Pulmozyme, antiinflammatory drugs, steroids



# Infection Control

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- Standard precautions
- Transmission based precautions (contact, droplet or airborne)
- Private room
- Special sources of transmission
- Patient/family education



# Cystic Fibrosis-Related Diabetes (CFRD)

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- Characterized by insulin deficiency and impaired glucose metabolism
- Therapy goals
- Nutritional recommendations
- Types of glucose tolerance
  - CFRD with hyperglycemia
  - CFRD without hyperglycemia



# Gastrointestinal Problems

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- Pancreatic insufficiency
- GERD
- Distal intestinal obstruction syndrome (DIOS)
- Pancreatitis
- Liver disease



# Reproductive Issues

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- Obstructive azoospermia
- Decreased fertility with women
- Pregnancy outcomes



# End-Of-Life Issues

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- Ethical dilemmas
- How palliative care differs with CF patients
- Transplantation
  - Newer approaches
  - Survival rates



# Advantages of a Detailed Case-Study Approach

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- “Puts a face to the disease”
- Demonstrates how a standard of care is operationalized in the care of a real patient
- Makes difficult concepts understandable
- Describes how nursing care is individualized
- One of the best way to emphasize psychosocial/developmental/cultural/spiritual/ethical issues
- Describes multidisciplinary contributions to care



# Article submission process

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- Target audience/target journal
- Author guidelines
- Query letters
- Submission of article with cover letter
- Peer-review process
- Acceptance with revision (major/minor) or rejection
- Article revisions (CEU series)
- Page proofs



# The Writing Process

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- Choosing a topic of interest
- The actual writing process
- Challenges faced
- What have you learned?
- Tips to pass on to other potential authors