



CF is an autosomal recessive genetic condition characterized by mutations of the **cystic fibrosis transmembrane conductance regulator (CFTR)** gene found on chromosome 7. The most common mutation is delta F508 deletion.

PATHOPHYSIOLOGY

Dysfunction of the CFTR protein found in epithelial cells of respiratory, gastrointestinal, reproductive system & sweat glands → dysregulation of chloride, bicarbonate, & sodium channel transport → ↓ H₂O concentration of secretions → impaired clearance

EPIDEMIOLOGY

Most prevalent in Caucasians → ~ 1 in every 3000 – 3500 live births

CLINICAL MANIFESTATIONS

Respiratory



- Progressive disease with intermittent acute exacerbations
- Mucous plugging → chronic airway inflammation & infections → bronchiectasis
- Common pathogens: *S. aureus*, *H. influenza*, *P. aeruginosa*, *B. cepacia*

Gastrointestinal



- Intestinal obstruction
- Steatorrhoea
- Pancreatic insufficiency, initially exocrine → eventually also endocrine (CF related diabetes)
- Fat soluble vitamin deficiencies (ADEK)
- CF related liver disease

Other



- Failure to thrive
- Delayed puberty & infertility
- Dehydration & sodium depletion
- Stress incontinence
- Low bone mineral density
- Clubbing

INVESTIGATIONS

Newborn screening test



Sweat chloride test



CF genetic testing



MANAGEMENT

- Airway clearance techniques: chest physiotherapy, nebulized hypertonic saline, dornase alfa
- Regular pulmonary function tests
- Lung transplantation



- Nutrition to maintain BMI >50th percentile
- Pancreatic enzyme supplementation
- Fat soluble vitamin supplementation (ADEK)
- Increased salt intake
- Oral glucose tolerance tests



- CFTR modulator therapies
- Routine immunizations
- Exposure avoidance (no contact with other individuals with CF; no hot tubs or humidifiers)
- Mental health screening (patient & family)



Pediatric Case Study Cystic Fibrosis Evolve

**American Association of Critical-Care
Nurses**

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Cystic Fibrosis Club Abstracts Cystic Fibrosis Club. Meeting, 1982 **Huether and McCance's Understanding Pathophysiology, Canadian Edition - E-Book** Kelly Power-Kean, Stephanie Zettel, Mohamed Toufic El-Hussein, Sue E. Huether, Kathryn L. McCance, 2022-01-08 Textbook and Academic Authors Association TAA Textbook Excellence Award Winner 2024 Prepare for Canadian nursing practice with a solid understanding of pathophysiology and disease Huether and McCance's Understanding Pathophysiology 2nd Canadian Edition covers the basic concepts of pathophysiology and disease processes from a Canadian perspective Clear descriptions and vibrant illustrations make it easier to understand body systems and the mechanisms of disease and online resources bring pathophysiology concepts to life Developed for Canadian nursing students by educators Kelly Power Kean Stephanie Zettel and Mohamed Toufic El Hussein this text prepares students for success on the Next Generation NCLEX CPNRE and REx PNTM and also in clinical practice Introduction to Pathophysiology provides an entrance to the science of pathophysiology and explains why it is important Lifespan coverage includes nine separate chapters on developmental alterations in pathophysiology and special sections with aging and pediatrics content Canadian drug and treatment guidelines familiarize you with aspects of clinical practice you will encounter Coverage of diseases includes their pathophysiology clinical manifestations and evaluation and treatment Canadian lab values provide the core fundamental information required for practice in Canada Canadian morbidity statistics provide you with the Canadian context in which you will be practising Algorithms and flowcharts of diseases and disorders make it easy to follow the sequential progression of disease processes Health Promotion boxes emphasize evidence based care and align with the Canadian curriculum Risk Factors boxes highlight important safety considerations associated with specific diseases Quick Check boxes test your understanding of important chapter concepts End of chapter Did You Understand summaries make it easy to review the chapter's major concepts Key Terms are set in blue boldface type and listed at the end of each chapter Glossary of approximately 1 000 terms is included on the Evolve website with definitions of important terminology

BOPOD - Wong's Nursing Care of Infants and Children Marilyn J. Hockenberry, David Wilson, 2014-01-30 The most trusted authority in pediatric nursing Wong's Nursing Care of Infants and Children provides unmatched comprehensive coverage of pediatric growth development and conditions Its unique age and stage approach covers child development and health promotion as well as specific health problems organized by age groups and body systems Leading pediatric experts Dr. Marilyn Hockenberry and David Wilson provide an evidence based clinical perspective based on nearly 30 years of hands on experience Easy to read and extensively illustrated this edition focuses on patient centered outcomes and includes updates on topics such as the late preterm infant immunizations the H1N1 virus and childhood obesity A clear straightforward writing style makes content easy to understand Unique Evidence Based Practice boxes help you apply both research and critical thought processes to support and guide the outcomes of nursing care Unique Atraumatic Care boxes contain

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Advances in Pediatrics V47 Lewis A.

Barness,2000-07 **Wong's Nursing Care of Infants and Children - E-Book** Marilyn J. Hockenberry,David

Wilson,2014-08-28 The leading text in pediatric nursing Wong's Nursing Care of Infants and Children takes a unique easy to understand developmental approach to describe the care of children at each age and stage of development Childhood diseases and disorders are organized by age groups and body systems and described through the nursing process framework This edition includes updates on topics such as the new CPR guidelines immunizations and pain assessment and management Written by pediatric experts Dr Marilyn Hockenberry and David Wilson each with nearly 30 years of hands on experience this bestseller provides an evidence based clinical perspective that shows how the quality of nursing care can impact quality patient outcomes A good neonatal and paediatric reference and textbook Reviewed by Jo Wilson on behalf of Nursing Times March 2015 A developmental approach addresses the differences at each stage of the child's growth and enables you to individualize care at the appropriate level for each child UNIQUE Quality Patient Outcomes are discussed for major childhood diseases and disorders showing how nursing care directly impacts patient outcomes Nursing Care Plans provide models for planning patient care with rationales explaining why specific nursing interventions have been chosen and include nursing diagnoses patient family goals nursing interventions rationales expected outcomes and NIC and NOC guidelines A focus on family centered care emphasizes the role and influence of the family in health and illness with a separate chapter and Family Centered Care boxes Critical thinking exercises use case studies to help you test and develop your own analytical skills Nursing Care Guidelines provide clear step by step detailed instructions on performing specific skills or procedures Nursing Tips include helpful hints and practical clinical information and Nursing Alerts provide critical information that must be considered in providing care An emphasis on wellness includes principles of health promotion and injury prevention for

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Table of Contents Pediatric Case Study Cystic Fibrosis Evolve

1. Understanding the eBook Pediatric Case Study Cystic Fibrosis Evolve
 - The Rise of Digital Reading Pediatric Case Study Cystic Fibrosis Evolve
 - Advantages of eBooks Over Traditional Books
2. Identifying Pediatric Case Study Cystic Fibrosis Evolve
 - Exploring Different Genres
 - Considering Fiction vs. Non-Fiction
 - Determining Your Reading Goals
3. Choosing the Right eBook Platform
 - Popular eBook Platforms
 - Features to Look for in an Pediatric Case Study Cystic Fibrosis Evolve
 - User-Friendly Interface
4. Exploring eBook Recommendations from Pediatric Case Study Cystic Fibrosis Evolve

- Personalized Recommendations
- Pediatric Case Study Cystic Fibrosis Evolve User Reviews and Ratings
- Pediatric Case Study Cystic Fibrosis Evolve and Bestseller Lists

5. Accessing Pediatric Case Study Cystic Fibrosis Evolve Free and Paid eBooks

- Pediatric Case Study Cystic Fibrosis Evolve Public Domain eBooks
- Pediatric Case Study Cystic Fibrosis Evolve eBook Subscription Services
- Pediatric Case Study Cystic Fibrosis Evolve Budget-Friendly Options

6. Navigating Pediatric Case Study Cystic Fibrosis Evolve eBook Formats

- ePUB, PDF, MOBI, and More
- Pediatric Case Study Cystic Fibrosis Evolve Compatibility with Devices
- Pediatric Case Study Cystic Fibrosis Evolve Enhanced eBook Features

7. Enhancing Your Reading Experience

- Adjustable Fonts and Text Sizes of Pediatric Case Study Cystic Fibrosis Evolve
- Highlighting and Note-Taking Pediatric Case Study Cystic Fibrosis Evolve
- Interactive Elements Pediatric Case Study Cystic Fibrosis Evolve

8. Staying Engaged with Pediatric Case Study Cystic Fibrosis Evolve

- Joining Online Reading Communities
- Participating in Virtual Book Clubs
- Following Authors and Publishers Pediatric Case Study Cystic Fibrosis Evolve

9. Balancing eBooks and Physical Books Pediatric Case Study Cystic Fibrosis Evolve

- Benefits of a Digital Library
- Creating a Diverse Reading Collection Pediatric Case Study Cystic Fibrosis Evolve

10. Overcoming Reading Challenges

- Dealing with Digital Eye Strain
- Minimizing Distractions
- Managing Screen Time

11. Cultivating a Reading Routine Pediatric Case Study Cystic Fibrosis Evolve

- Setting Reading Goals Pediatric Case Study Cystic Fibrosis Evolve
- Carving Out Dedicated Reading Time

12. Sourcing Reliable Information of Pediatric Case Study Cystic Fibrosis Evolve

- Fact-Checking eBook Content of Pediatric Case Study Cystic Fibrosis Evolve
- Distinguishing Credible Sources

13. Promoting Lifelong Learning

- Utilizing eBooks for Skill Development
- Exploring Educational eBooks

14. Embracing eBook Trends

- Integration of Multimedia Elements
- Interactive and Gamified eBooks

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