



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 	Gastrointestinal 	Other 
<ul style="list-style-type: none"> Progressive disease with intermittent acute exacerbations Mucous plugging → chronic airway inflammation & infections → bronchiectasis Common pathogens: S. aureus, H. influenza, P. aeruginosa, B. cepacia  	<ul style="list-style-type: none"> Intestinal obstruction Steatorrhea Pancreatic insufficiency, initially exocrine → eventually also endocrine (CF related diabetes) Fat soluble vitamin deficiencies (ADEK) CF related liver disease 	<ul style="list-style-type: none"> 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 
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MANAGEMENT

<ul style="list-style-type: none"> Airway clearance techniques: chest physiotherapy, nebulized hypertonic saline, dornase alfa  Regular pulmonary function tests Lung transplantation 	<ul style="list-style-type: none"> Nutrition to maintain BMI >50th percentile Pancreatic enzyme supplementation  Fat soluble vitamin supplementation (ADEK) Increased salt intake Oral glucose tolerance tests 	<ul style="list-style-type: none"> CFTR modulator therapies Routine immunizations Exposure avoidance (no contact with other individuals with CF; no hot tubs or humidifiers)  Mental health screening (patient & family)
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**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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