

PATHOPHYSIOLOGY

A genetic disease that affects many organs and lethally impairs lung function. The underlying problem with CF is blocked chloride transport in the cell membranes. Poor chloride transport causes the formation of mucus that has little water content and is thick. The thick sticky mucus causes problems in lungs, pancreas, liver, salivary glands and tests. The mucus plugs up the airways in the lungs and the granular tissues in non-pulmonary organs, causing atrophy and organ dysfunction.

CF is most common among whites, and about 4% are carriers. It is rare among African Americans and Asians. Males and females are affected equally. Family history is another *risk factor*.

CFTR Gene Mutation
Autosomal Recessive

SIGNS & SYMPTOMS

Salt tasting skin
Persistent coughing
Dyspnea
Wheezing
Failure to pass Meconium
Weight loss
Frequent resp. infection
clubbing of the fingers.

DIAGNOSTICS

Chloride Sweat Test (>60)
Chest X-Ray
Genetic Screen (CFTR)

ASSESSMENT : NURSING

Nonpulmonary	Pulmonary
Abdominal distention	Chest congestion
Gastroesophageal reflux	Limited exercise tolerance
Rectal prolapse	Cough & Sputum production
Foul-smelling stools	Use of accessory muscles
Steatorrhea (excessive fat in stools)	Decreased pulmonary function

COMPLICATIONS

Infection
Infertility
Diabetes (secondary)
Intestinal obstruction
Kidney/Liver
ABG studies show acidosis (low pH), greatly reduced aerial oxygen (PaO2) levels, increased arterial carbon dioxide (PaCo2) levels, and increase bicarbonate levels.
With infection the patient has fever, an elevated white blood cell count, and decreased oxygen saturation.

TREATMENT

PHARM	NON-PHARM
Digestive enzymes (vitamins A,D,E, & K)	Aerobic exercise
Lung transplant	Chest physiotherapy
Mucolytics	Exacerbation therapy
Bronchodilators/Steroids	
Antibiotics	

EDUCATION

Yearly FLU Vaccine
High Calorie & Protein Diet
Fluids
Genetic counseling prior to having a baby
Cystic Fibrosis Fondation (Resource)

Carrier Testing

