# Cystic Fibrosis Cheat Sheet

by RoseCher via cheatography.com/87410/cs/20230/

## 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 muscus 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 & SYMTOMS

- 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
- Infetility
- 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.

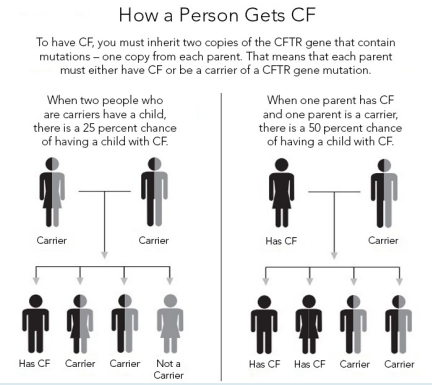
## TREATMENT

| PHARM | NON-PHARM |
|---|---|
| Digestive enzymes (vitamins A,D,E, & K) | Aerobic exercise |
| Lung transplant | Chest physio-therapy |
| 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





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