Cystic Fibrosis Diagnosis and Treatment

Cystic fibrosis is a hereditary disease leading to problems with Cl channels in the body. It is the most common lethal genetic disease in the Caucasion population. Patients develop recurrent pulmonary infections, bronchitis, infertility, pancreatic insufficiency, steatorrhea and malabsorption.



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Diagnosis

Sweat Chloride Test > 60 mmol/L
Sweaty-sweatgland Chlorine-dispenser tested as Greater Than 60
A diagnostic lab finding is a sweat chloride level > 60 mmol/L, as these patients cannot reabsorb chloride through their sweat glands.

Meconium Ileus
Meconium-macaroni Eels
Meconium is a black, tarry stool passed by newborns in their first 24 hours of life. Meconium ileus is something commonly seen in CF patients, as GI secretions are limited and patients form dry mucus. Thus, patients who have not passed meconium in their first 24 hours of life have meconium ileus, which is often a diagnostic tip-off to CF.

Treatment

N-acetylcysteine
N-seagull-Sistine
N-acetylcysteine is a medication given to patients with cystic fibrosis, as it loosens mucus plugs. This drug works by cleaving disulfide bonds within mucus glycoproteins.

Antibiotic Prophylaxis
ABX-guy with Purple-axes
Due to mucus stasis, these patients deal with recurrent pulmonary infections and are typically given antibiotic prophylactic medications to help prevent infections.

Pulmonary Maintenance
Lungs Maintanance-guy
Pulmonary maintenance in these patients. This includes therapy of percussive vibration vests to break and loosen mucus, BiPAP, and hypertonic saline to loosen secretions.

Lung Transplant
Lungs Train-plant
As lung function and exercise tolerance decreases, lung transplant is necessary in patients. Due to the proclivity to develop infection, both lungs must be transplanted.

Vitamin Replacement
Vikings
Due to malabsorption and pancreatic insufficiency, these patients must take Vitamin replacement, especially those which are fat-soluble (A, D, E, and K).

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