

Cystic Fibrosis

Pathophysiology

- **Autosomal recessive** predominantly in white patients.
- **Defect in chloride channel protein** causes impaired chloride and water transport, which leads to **excessively thick, vicious secretions** in the respiratory tract, exocrine pancreas, sweat glands, GI, and GU tracts.
- Prognosis has improved with the median age of death now >30.

Treatment

- Pancreatic enzyme replacement, fat-soluble vitamin supplements, chest PT, vaccinations (influenza and pneumococcal), antibiotics for infections, inhaled recombinant human deoxyribonuclease (rhDNAse) breaks down the DNA in respiratory mucus that clogs airways.

Symptoms

- Results in **obstructive lung disease** with **chronic infections** (frequently *Pseudomonas*), pancreatic insufficiency, and other GI complications.

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By [xkissmekatex](#) (kissmekate) cheatography.com/kissmekate/

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