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 does 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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