

Case Study - What is the Relationship Between the Cell Membrane and Cystic Fibrosis?

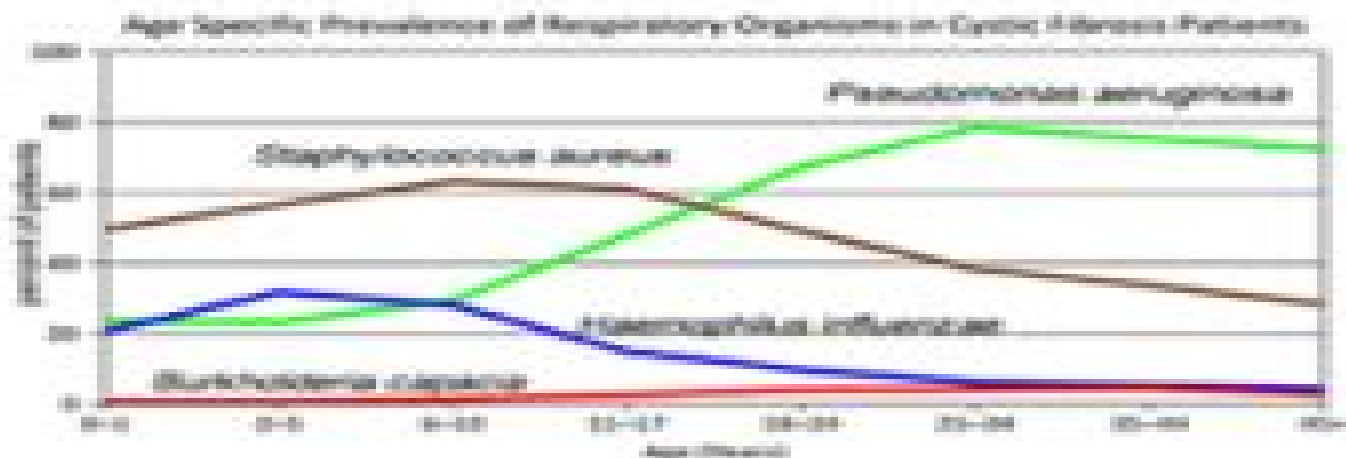
Dr. Heyland examined a six-month-old infant that had been admitted to university hospital earlier in the day. The baby's parents had brought along Joey to the emergency room because the baby had been suffering from a chronic cough. In addition, they said that Joey sometimes would "sweat" and more than they thought was normal for a child with a cold. Upon arriving at the emergency room, the attending pediatrician noted that ear crystals were present on Joey's skin and called Dr. Heyland, a pediatric pulmonologist. Dr. Heyland suspects that baby Joey may be suffering from cystic fibrosis.



CF affects more than 30,000 kids and young adults in the United States. It disrupts the normal function of epithelial cells – cells that line up the sweat glands in the skin and that line the passageways inside the lungs, pancreas, and digestive and reproductive systems.

The inherited CF gene directs the body's epithelial cells to produce a defective form of a protein called CFTR (or cystic fibrosis transmembrane conductance regulator) found in cells that line the lungs, digestive tract, sweat glands, and reproductive system.

When the CFTR protein is defective, epithelial cells can't regulate the way that chloride ions pass across cell membranes. This disrupts the balance of salt and water needed to maintain a normal thin coating of mucus inside the lungs and other passageways. The mucus becomes thick, sticky, and hard to move, and can result in infections from bacterial colonization.



1. "Given to that child which when based on the findings tested baby, He is devastated and soon will die"

This is an old saying from the eighteenth century and describes one of the symptoms of CF (salty sweat). Why do you think babies in the modern age have a better chance of survival than babies in the 18th century?

2. What symptoms lead Dr. Heyland to his initial diagnosis?

3. Consider the graph of infections, what organisms show relatively constant or numbers over a lifetime.

What organism is most likely affecting baby Joey?

4. Before 1950, what was the life expectancy for a child born with cystic fibrosis? What technology caused an improvement in life expectancy after that time period?

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