

Inhaling salt mist helps cystic fibrosis

Gene Emery
Reuters

Thursday, 19 January 2006

Inhaling the mists of salt water can reduce the pus and infection that fills the airways of cystic fibrosis sufferers, although side effects include a nasty coughing fit and a harsh taste.

That's the conclusion of two studies published in this week's issue of *The New England Journal of Medicine*.

They found that inhaling a mist with a salt content of 7 or 9% improved lung function and, in some cases, produced less absenteeism from school or work.

Cystic fibrosis, a progressive and frequently fatal genetic disease that affects about 30,000 young adults and children in the US alone, is marked by a thickening of the mucus which makes it harder to clear the lungs of debris and bacteria.

The salt water solution "really opens up a new avenue for approaching patients with cystic fibrosis and how to treat them," says Dr Gail Weinmann, of the US [National Heart, Lung, and Blood Institute](#), which sponsored one of the studies.

Mark Elkins of the [Royal Prince Alfred Hospital](#) in Sydney, Australia and colleagues authored one of the new published studies.

The team found that the 83 volunteers who regularly inhaled a 7% mist of salty water had fewer breathing problems and less absenteeism from school or work than those who inhaled a solution with a salt content of under 1%.

"Adding salt [and water] to the airway surfaces of patients with cystic fibrosis is beneficial" for both children and adults, they conclude.

All of the patients first inhaled a chemical to try to open their lung passages as much as possible.

In the second study, US-based Assistant Professor Scott Donaldson of the [University of North Carolina at Chapel Hill](#) and his colleagues found that a 7% salt mist "produced a sustained acceleration of mucus clearance and improved lung function" because it helped hydrate the lungs.

In an accompanying editorial, Dr Felix Ratjen of the [Hospital for Sick Children](#) in Toronto, Canada, cited several unpleasant side effects of the salt mist treatment including a bad taste, coughing fits and the lengthy 30 minutes it can take to administer.

He added that in the study by Elkins and team, patients may not have received the best long-term antibiotic treatment.

That would make the inhaled salt water mist appear more effective than it would have been if people were getting a better drug, says Ratjen.

Weinmann says limitations inherent to the treatment mean a salt water mist "may be just a first step" in treating cystic fibrosis.

Salt water treatment for cystic fibrosis both effective and cheap

Two teams of scientists working half a world apart, have identified what they believe is a simple, effective and inexpensive treatment to reduce lung problems associated with cystic fibrosis.

Cystic fibrosis is the leading fatal genetic illness among whites, and victims suffer from a mucus build-up in the lungs that makes breathing difficult.

Cystic fibrosis appears on average in one of four children of parents who both carry a defective copy of a gene known as CFTR.

Children born with the disease soon develop chronic lung damage, since their lungs cannot clear the excessively sticky mucus by sweeping it to the mouth, where it is swallowed and eliminated.

Respiratory failure is the leading cause of death in CF patients.

The scientists say that salt water can ease the condition, and by inhaling a saltwater aerosol solution for between 10 and 15 minutes at least twice a day, young patients should be able to avoid a significant part of the damage the disease causes to their lungs.

The new therapy also appears to be safe and easy to take.

According to the researchers the aerosolized saltwater restores the thin lubricant layer of water that normally coats airway surfaces.

Apparently the water layer promotes the clearance of the naturally occurring mucus the body uses to trap harmful bacteria, viruses and other foreign particles.

The teams from the [University of North Carolina at Chapel Hill School of Medicine](#) and [UNC Hospitals](#), worked with the [Royal Prince Alfred Hospital](#), the [University of Sydney](#) and the [Woolcock Institute of Medical Research](#), all in Sydney.

The scientists say they are excited that such a simple and inexpensive therapy appears to be so effective and well-tolerated in patients with CF.

They are optimistic that the therapy will reduce how often patients are ill, and will slow the decline of lung function over time and extend their life span.

The UNC study involved 24 CF patients who each inhaled the salt solution with or without pretreatment with a compound known as amiloride over two-week periods. Analysis of mucus clearance and lung function showed that the high-salt aerosol alone worked best, which surprised the medical scientists.

The Australian research team then used a comparable protocol to study another 164 patients for a longer period of almost a year.

During that extended period the Sydney researchers also found fewer lung problems with the concentrated saline than with normal saline, less need for antibiotics to treat lung

infections over time, and improved attendance by patients at school, work and other activities during the 48 weeks.

The researchers say that as salt solutions are so cheap to make, the new therapy will produce clinical benefits at a minimal treatment expense, in contrast to many other available therapies.

The studies were supported by the [Cystic Fibrosis Foundation](#) and the [National Heart, Lung and Blood Institute](#).

The final reports on both studies, were collaborative and complementary, and appear in the Jan. 19 issue of the [New England Journal of Medicine](#).

Inexpensive Breakthrough Treatment for Cystic Fibrosis Involving Salt Water

Scientists may have discovered a breakthrough treatment for cystic fibrosis (CF). The treatment is inexpensive, safe and easy to do. According to researchers at the University of North Carolina, by inhaling a saltwater aerosol solution at least twice a day for between 10 and 15 minutes, patients should be able to avoid a "significant part of the damage the disease causes to their lungs".

The new saline water treatments appear to promote the "clearance of the naturally occurring mucus the body uses to trap harmful bacteria, viruses and other foreign particles." The solution may produce a "water layer" in the lungs promoting the removal.

It now appears the surfaces of the lungs of CF patients are dehydrated. Healthy people have a 5 to 10 micron thick, coating of water in the lungs that helps lubricate the open areas of the lungs. By restoring hydration with the hypertonic saline solution, you are treating the "basic cause of this disease".

Since respiratory failure is the leading cause of death in CF patients, this finding is very important. According to Dr. Scott H Donaldson, assistant professor of Medicine, "We are very excited that this simple and inexpensive therapy turned out to be so effective and well-tolerated in patients with CF."

Cystic fibrosis is the leading fatal genetic illness among whites, and affects 1 in 2,500 children born. About one in five babies with CF are diagnosed at birth. This happens when their gut becomes blocked by extra thick meconium (a black tar-like bowel contents that all babies pass soon after birth). Some children with CF don't put weight on because their pancreas is not producing enzymes that help digest fat. Typically the stools contain the extra fat and are oily and very smelly.

Patients with CF will also symptoms involving the lungs. In a healthy person there is a constant flow of mucus over the air passages in the lungs. This is an important process to remove debris and bacteria. Cystic fibrosis will cause the mucus to become excessively sticky. Bacteria can then grow in this environment.

Inhaled Hypertonic Saline

Thinning Secretions by Osmosis

By [Lori Alma](#), About.com

Updated: August 5, 2008

Inhaled hypertonic saline is one of the newer treatment options for cystic fibrosis (CF). A group of Australian researchers discovered the benefits of hypertonic saline when they noticed that surfers with CF had better outcomes than people with CF who did not surf. The researchers believed that inhaling the salty mist of ocean water during surfing was the reason that surfers had less respiratory flare-ups than their nonsurfing counterparts had, and a study confirmed those findings. The researchers published the results of their study in 2006, and since then, hypertonic saline has been available as a treatment option to many people with CF.

What is Hypertonic Saline?

Normal saline is a salt-water solution that contains the same concentration of salt as our own body fluids. Hypertonic saline is any salt-water solution that contains a greater concentration of salt than normal saline, and therefore a greater concentration of salt than our body fluids. Normal saline solution is 0.9% salt. The hypertonic saline used in CF treatment is 7% salt.

How Hypertonic Saline Works

Fluids transfer in and out of cells by a process called "osmosis," where fluids with lower salt concentrations flow across the cell membrane to mix with fluids with higher salt concentration. Simply stated, osmosis means that water follows salt.

In CF treatment, a machine called a "nebulizer" aerosolizes the hypertonic saline solution. The saline mist is inhaled and settles in the airways. Since hypertonic saline has a much greater concentration of salt than normal body fluids, it draws water from the cells in to the airway. The water mixes with the thick mucus in the airway, making it thinner and easier to cough out.

Who Can Use Hypertonic Saline?

So far, hypertonic saline use has only been studied in people 6 years old and older with mild-moderate forms of CF.

Are There Any Side Effects?

Hypertonic saline is not a drug, but it does cause changes in the body that can have some unpleasant effects. Some people using hypertonic saline have experienced an increase in coughing, sore throats or chest pain caused by tissue irritation.

Isn't Saline Just Salt Water? Can it Be Made at Home?

No. The concentration of salt must be precise, and this would be difficult to accomplish in a homemade solution. More importantly, the solution must be sterile to prevent introducing potentially life-threatening bacteria in to the airways.

Source:

Donaldson, S.H., et al. "Mucus Clearance and Lung Function in Cystic Fibrosis with Hypertonic Saline". 2006. *New England Journal of Medicine* 354(3): 1848-1851. 4 August, 2008.

Saltwater therapy helps ease cystic fibrosis

By Liz Szabo, USA TODAY

Australian surfers have helped inspire a new way to treat a deadly genetic disease.

Patients with cystic fibrosis, a rare disorder that damages the lungs, have reported feeling better after surfing, says Mark Elkins, a researcher at Sydney's Royal Prince Alfred Hospital. Surfers said their chests and sinuses felt clear, and they coughed up much of the thick mucus that clogs their lungs.

Doctors wondered why, Elkins says. Was it the exercise? Or was it the saltwater?

Scientists have known for years that salt plays a key role in cystic fibrosis, Elkins says. Cystic fibrosis, which afflicts about 30,000 Americans, is caused by a defect in a gene that controls the amount of salt and water that line the airways of the lungs. Without sufficient lubrication, mucus builds up and blocks the airways, providing a fertile home for bacteria.

Doctors in Australia and the USA decided to test whether saltwater might replace that missing lubrication. They hoped the extra salt would draw water out of lung tissue onto the airway, providing a thin layer of liquid to ease mucus out of the lung, says Richard Boucher, who directs the cystic fibrosis center at the University of North Carolina-Chapel Hill and led the American study.

Researchers found that inhaling an intensely salty solution — almost twice as salty as the Atlantic Ocean — improved patients' lung function and slowed the progression of the disease, according to articles published in Thursday's *New England Journal of Medicine*. Australian doctors found that 41% of those who received the treatment avoided serious complications — such as weight loss, coughing up blood or a dangerous infection — compared with 16% of the other patients. The solutions helped remove mucus from the lung for at least eight hours, according to the UNC study of 24 patients, also published in the journal.

Doctors probably will begin using the treatments right away, says Peter Mogayzel, director of the Johns Hopkins Cystic Fibrosis Center, who was not involved in the study. His center began offering the treatment last year after seeing preliminary results at a conference.

The treatments don't cure cystic fibrosis and won't replace current therapies, Elkins says. Some patients already spend hours a day treating their disease, so adding another 30-minute therapy could be a burden. To make the treatments easier to use, researchers are testing a device that works four times more quickly than current systems.

Felix Ratjen of the University of Toronto, who wrote an editorial accompanying the studies, notes that the saltwater treatment may have a limited benefit, because it may not reach the most clogged airways. But Boucher hopes the treatments could prevent damage, especially in babies whose lungs are still unscarred. He plans to begin tests soon in infants as young as 2 months.

The new therapy is also relatively cheap. The Cystic Fibrosis Foundation estimates the cost to be about \$110 a month, less than one-tenth the cost of other drugs. Scott Donaldson, an assistant professor at UNC and co-author of that study, says, "Something simple has turned out to be very good."

Saline Solution Helps Cystic Fibrosis Patients Breathe Easier

By Ed Edelson
HealthDay Reporter

WEDNESDAY, Jan. 18 (HealthDay News) -- Researchers believe they've found a way to clear the thick mucus from the lungs of people with cystic fibrosis so they will be less vulnerable to life-threatening infections.

The idea is to induce a slick, watery surface to the airways so that this mucus can slide away, explained Dr. Richard C. Boucher, director of the Cystic Fibrosis Pulmonary Treatment and Research Center at the University of North Carolina, Chapel Hill.

His group, along with researchers in Australia, are achieving that goal by having cystic fibrosis patients inhale a concentrated salt solution.

But that's just the beginning, he said.

"We have provided a road map for future therapies designed to hydrate the airway surface," Boucher said. "It is not perfect, but it has laid the groundwork for development of better therapies."

Two reports on use of "inhaled hypertonic saline," the formal name for a salt solution, at North Carolina and in Australia, appear in the Jan. 19 issue of the *New England Journal of Medicine*. The number of patients involved in the studies is small -- 24 patients in North Carolina study, 164 in the Australian cohort -- but both groups are reporting a significant reduction in lung problems associated with mucus removal.

"They had fewer troubling symptoms such as wheezing," said Dr. Scott H. Donaldson, an associate professor of medicine and lead researcher for the North Carolina team. They also had markedly improved breathing capacity, he said.

Cystic fibrosis is an inherited disease affecting approximately 30,000 Americans. Its major symptoms include the accumulation in the airways of thick, clogging mucus that becomes a breeding ground for germs. Use of antibiotics and intense therapy has extended the once-short life span of cystic fibrosis patients, but most still won't live past the mid-30s.

Experts have long debated the cause of mucus accumulation and how to prevent it, Boucher said. However, work in the laboratory, and then with mice and humans, has shown that "dehydration is the critical factor" and that coating the airways with a thin film of water lets the mucus slide away.

The treatment now is being used routinely for adults treated for cystic fibrosis at the University of Carolina, Donaldson said. They generally inhale the salt solution twice a day, although "four times a day gets better results," he said.

But the real hope is for people newly diagnosed with the condition, Donaldson said. "Regular use of the agent at a very early stage of lung disease may prevent infections and add years of life," he said. Meanwhile, research into better ways to hydrate the airways is ongoing, Boucher said.

"The overall concept is very exciting," said Dr. Felix Ratjen, head of respiratory medicine at the Hospital for Sick Children in Toronto. However, he cautioned that there still are blanks to be filled in.

"It needs to be proven that if you do it in patients with less severe early disease, it will stop the progression of the disease," Ratjen said. "There is no data on how it will be tolerated early."

Also, it still has to be shown whether the beneficial effects reported in these trials will be sustained over time, he said.

"It also needs to be sorted out how you can make it work better," Ratjen said. "This [the inhaled salt solution] is a relatively crude approach. But, because it is a very simple concept, it is very attractive."

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Effect of increasing doses of hypertonic saline on mucociliary clearance in patients with cystic fibrosis

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BACKGROUND: Patients with cystic fibrosis are known to have decreased mucociliary clearance. It has previously been shown that inhalation of a 7.0% solution of hypertonic saline significantly improved mucociliary clearance in a group of adult patients with cystic fibrosis. The aim of this study was to measure the response to increasing concentrations of inhaled hypertonic saline. **METHODS:** Ten patients (seven men) of mean (SE) age 22 (4) years and mean forced expiratory volume in one second (FEV1) 52.0 (6.7)% predicted completed the study. Mucociliary clearance was measured using a radioaerosol technique for 90 minutes after the interventions which comprised 0.9% NaCl + voluntary cough (control), 3.0% NaCl, 7.0% NaCl, and 12% NaCl. **RESULTS:** There was a significant increase in the amount of activity cleared from the right lung with all concentrations of hypertonic saline (HS) compared with control. The amount cleared at 90 minutes on the control day was 12.7% (95% confidence interval (CI) 9.8 to 17.2) compared with 19.7% (95% CI 13.6 to 29.5) for 3% HS, 23.8% (95% CI 15.9 to 36.7) for 7% HS and 26.0% (95% CI 19.8 to 35.9) for 12% HS. The improvement in mucociliary clearance was not solely due to coughing as the number of coughs recorded on the control day exceeded that recorded on any other day. The hypertonic saline did not induce a clinically significant change in FEV1. **CONCLUSIONS:** Within the range of concentrations examined in this study, the effect of hypertonic saline appears to be dose dependent. Inhalation of hypertonic saline remains a potentially useful treatment for patients with cystic fibrosis.

DRUG INFORMATION ROUNDS

Hypertonic Saline Treatment of Cystic Fibrosis

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OBJECTIVE: To review the literature concerning the use of hypertonic saline (HS) in patients with cystic fibrosis and explain the rationale for its use.

DATA SOURCES: A MEDLINE search was conducted through February 2007. Search terms included hypertonic saline, mucociliary clearance, cystic fibrosis, and human DNASE 1 protein. Additional data were identified through subsequent bibliographic reviews.

STUDY SELECTION AND DATA EXTRACTION: All articles in English identified from the data sources were evaluated. Pertinent studies using HS in patients with cystic fibrosis were included in the analysis.

DATA SYNTHESIS: Cystic fibrosis is caused by a deficiency in the cystic fibrosis transmembrane regulator gene, resulting in reduced chloride secretion and excessive sodium absorption. The most significant changes are seen in the airway lumen of the lungs. HS has been shown to improve mucociliary clearance versus placebo. A short-term efficacy trial showed a modest and variable increase in forced expiratory volume in 1 second (FEV₁) over a 2 week period ($15.0 \pm 16.0\%$ from baseline vs $2.8 \pm 13.1\%$ with HS 6% and NaCl 0.9%, respectively; $p = 0.004$). A long-term efficacy trial of either HS 7% or NaCl 0.9% twice daily for 48 weeks has shown a modest sustained improvement in FEV₁ and a significantly increased exacerbation-free survival rate (76% vs 62% for HS 7% and NaCl 0.9%, respectively; $p = 0.03$).

CONCLUSIONS: HS preceded by a bronchodilator is an inexpensive, safe, effective additional therapy in cystic fibrosis patients with stable lung function. Its use has been associated with a modest improvement in lung function and reduced frequency of pulmonary exacerbations.

Key Words: cystic fibrosis, hypertonic saline

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Cystic fibrosis salt/fluid controversy: In the thick of it

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A new study shows that a mouse model of cystic fibrosis has lower levels of liquid on the surface of its airway epithelium which suggests that thickened airway secretions might have a role in the disease process and that rehydrating the airways might be of benefit to patients.

Cystic fibrosis (CF) is associated with impaired mucociliary clearance, abnormal mucous, inflammation and chronic respiratory infections by bacteria such as *Staphylococcus aureus* and *Pseudomonas aeruginosa*¹. Although it is recognized that lung damage caused by bacterial infection is a major component of the disease, it has been difficult to understand why the lungs of CF patients become colonized with bacteria. Normal human airways are continually exposed to bacteria but the lower airways remain sterile in healthy individuals. The normal airway has many lines of defense against bacterial colonization, including protective processes such as mucociliary clearance that traps and removes inhaled bacteria, and cell-surface barriers that limit the ability of bacteria to bind to the airway epithelium. Recently, two competing theories have been proposed to explain how bacterial colonization occurs in CF. One theory, the 'fluid' model, contends that hyper-absorption of fluid by the airway surface epithelium leads to a lower than normal airway-surface liquid (ASL) volume (Fig. 1a). This reduced volume, combined with defective fluid production by the submucosal glands, in turn leads to under-hydrated mucous and impaired mucociliary clearance. Impaired mucociliary clearance and thick airway secretions contribute to the establishment of an environment in the airway that promotes colonization of the lungs by bacteria². The second theory, the 'salt' model, states that the salt content of airway fluid in CF is too high and thus prevents salt-sensitive defensin molecules in the ASL from killing bacteria, leading to increased susceptibility to lung infections³. The two theories describe two different ways of developing therapeutic strategies. Whereas the fluid model predicts that adding more liquid to the airway surface would improve health, the salt model predicts that reducing the salt concentration of the airway fluid to reactivate the natural defenses against infection would be a better therapeutic strategy. Resolution of the salt-fluid controversy thus has important implications to the design of therapeutic strategies for CF.

Figure 1

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Cystic fibrosis salt/fluid controversy: In the thick of it

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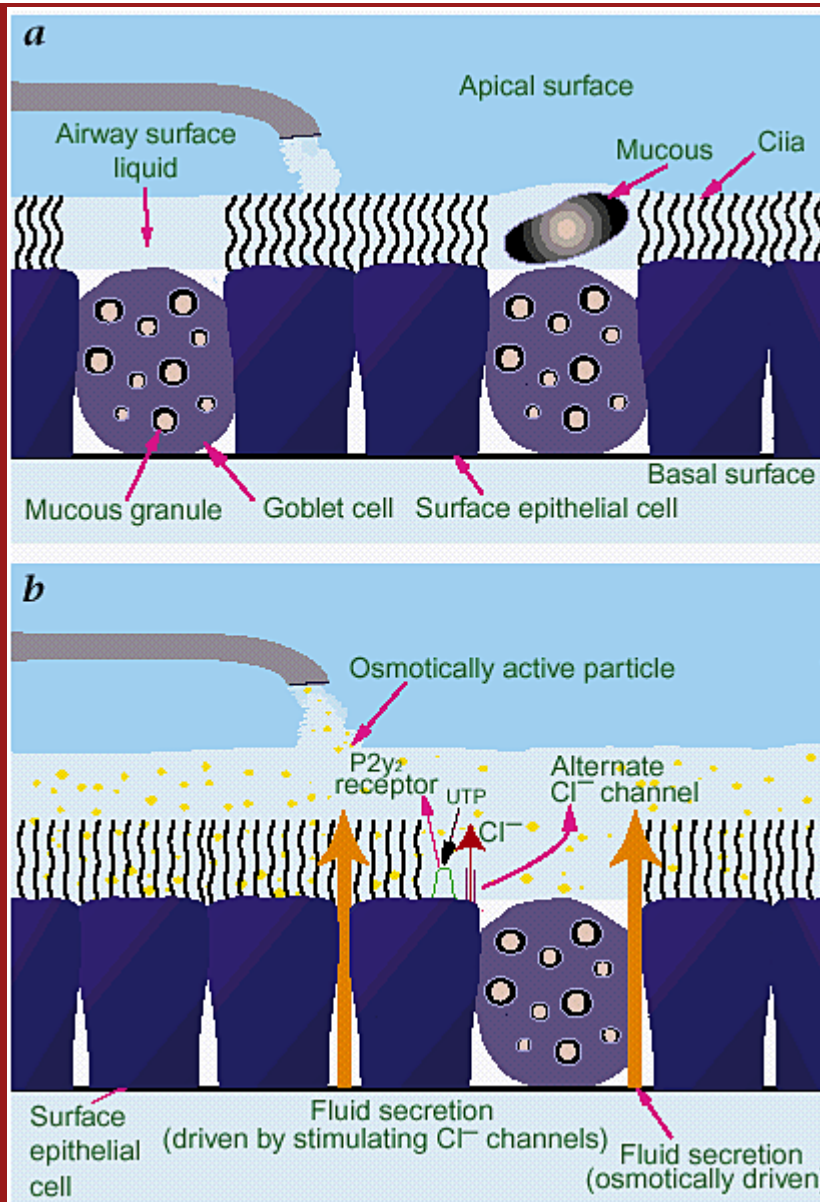


Figure 1. Defective mucociliary clearance in the CF: will hydration therapy correct it?

a, The 'fluid' model of bacterial infection in CF predicts that fluid absorption is enhanced in CF and fluid production driven by the CFTR channel is compromised. This leads to a reduction in the height of the ASL, which impairs mucociliary clearance. If the fluid model is correct, a potential therapy might be to simply add more fluid to the airway. **b**, Tarran *et al.* found that the nasal epithelia of the mutant CF mouse has an increased number of goblet cells and a reduced height of ASL when compared with normal mice. They propose that an effective treatment might be to add a solution containing an osmotically active substance plus a purinergic agonist. This would draw water from the interstitium by osmotic forces and by stimulating alternate chloride channels.

Renee Lucas

An article in the 22 July issue of *Molecular Cell* by Tarran *et al.*⁴ adds support to the fluid model. The authors studied the nasal epithelium of the mouse deficient in the gene implicated in CF as a model for the human disease. They found a substantial increase in the number and size of the goblet cells and a profound decrease in ASL volume in the CF mice compared with wild-type mice. Because in the mouse, goblet cells produce mucous, an increase in the size and number of these cells in the CF mouse might indicate that the nasal epithelium of the mouse responds to a reduction in ASL by producing thicker mucous, similar to humans with CF. However, the authors did not detect any differences in Cl⁻ concentration in the ASL between the mutant and wild-type mice. These data convincingly support the argument that reduced Cl⁻ secretion and enhanced Na⁺ absorption in CF reduce ASL volume without significantly affecting the relative concentrations of these molecules between CF and normal mice. Although the transport abnormalities in CF should lead to a reduction in the total amount of Na⁺ and Cl⁻ in the airway, Tarran *et al.*⁴ show that the nasal epithelium is not able to maintain transepithelial osmotic gradients because of its intrinsically high water permeability. In other words, because water equilibrates rapidly across the nasal epithelium, a reduction in total salt content in the airway leads to a reduction in ASL volume without changing salt concentration. Based on these results, Tarran *et al.* speculate that a reduction of mucous hydration and mucociliary clearance in CF initiates the inflammation and infection that lead to disease.

The study was taken one step further to investigate whether hydrating the surface fluids modifies the course of disease. The authors aerosolized 3–10% hypertonic saline or mannitol into the nasal passages of the CF mice. The idea was to rehydrate the airway both by directly adding fluid and by drawing water from the blood side of the airway to the epithelial surface (Fig. 1b). Unfortunately, osmolytes administered *in vivo* did not increase the ASL volume. Realizing that there might be several reasons why osmolytes failed to alter ASL height in mice—most likely among them the relative inefficiency of depositing an aerosol into the mouse nose—the authors performed similar experiments on human airway cells in culture. In this setting, osmolytes alone or in combination with pharmacological agents such as amiloride or adenosine triphosphate were effective in producing isotonic volume responses in human airway epithelia. However, the restorative effects were typically short-lived and not as effective on CF cell cultures because of the tendency of CF cells to hyper-absorb fluid and accumulate mucus. Critical experiments did show that a combination of raffinose and uridine triphosphate was most effective in enhancing mucous clearance in CF cells. In this case, enhanced mucous clearance was evident up to 72 hours after treatment. The data showed that it is possible to normalize ASL volume in CF; however, efficacy will likely depend on development of longer-acting pharmacologic agents along with increased efficiency of osmolyte delivery.

Although these experiments might provide an approach to normalize ASL volume in CF, the bigger question remains whether defective ASL volume in CF is the cause of compromised bacterial killing in the CF lung. Several theories in addition to the fluid and the salt models have been discussed to explain why CF patients are susceptible to chronic lung infection in general and *Pseudomonas aeruginosa* in particular. Prince and colleagues⁵ have proposed that CF bronchial epithelial cells bind *P. aeruginosa* to a greater extent than do normal cells. Pier *et al.*⁶ have suggested a defect in bacterial internalization. It is unclear how many of these proposed mechanisms are important *in vivo* and whether they act in combination. Although the manuscript by Tarran *et al.*⁴ provides further evidence supporting the fluid model, the question of whether simply hydrating the ASL will prevent bacterial colonization *in vivo* or whether some other factor particular to the CF lung will have to be corrected to prevent lung infection is still unresolved.

However, what is undisputed is that once bacteria such as *P. aeruginosa* colonize the lung, they are difficult to eradicate. This is especially true when they become mucoid and establish what is referred to as a biofilm⁷. Clearly, for a therapy to be effective, it should be applied early in life before bacterial colonization occurs. Although on the one hand, a resolution of why CF patients are susceptible to bacterial colonization may seem far off, on the other hand, the completion of the *P. aeruginosa* genome⁸ is already providing new tools that might soon

provide the answer. When that answer does come, the CF community will certainly celebrate.

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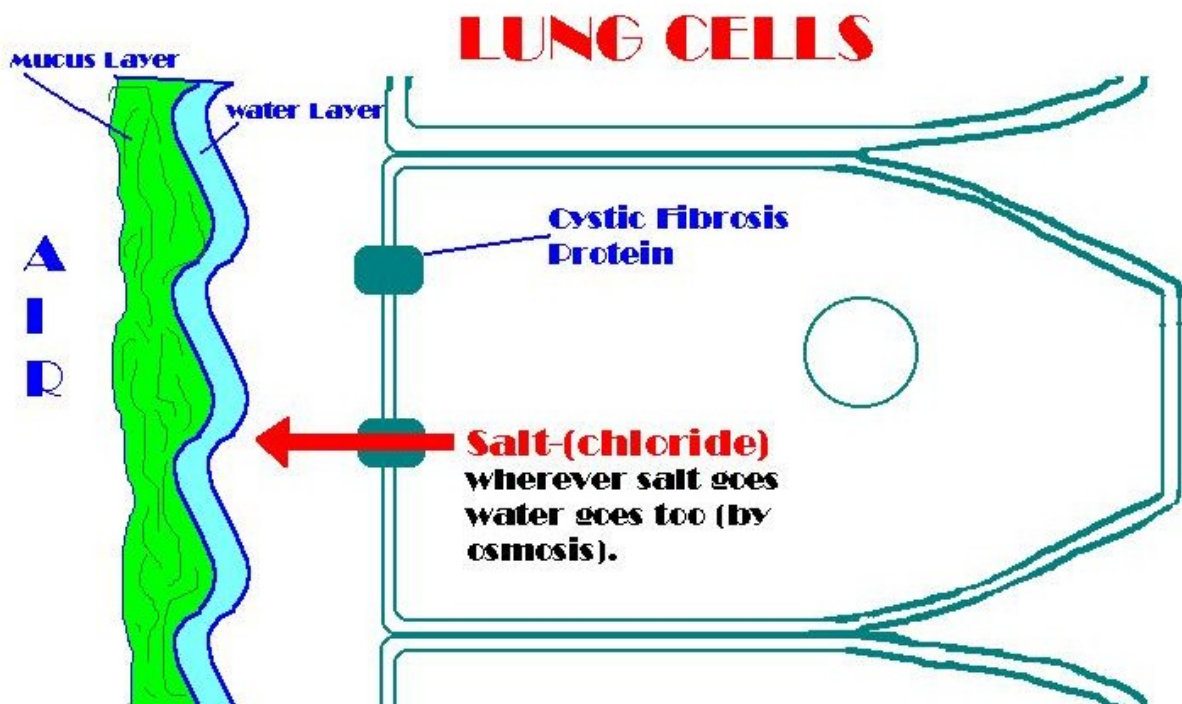
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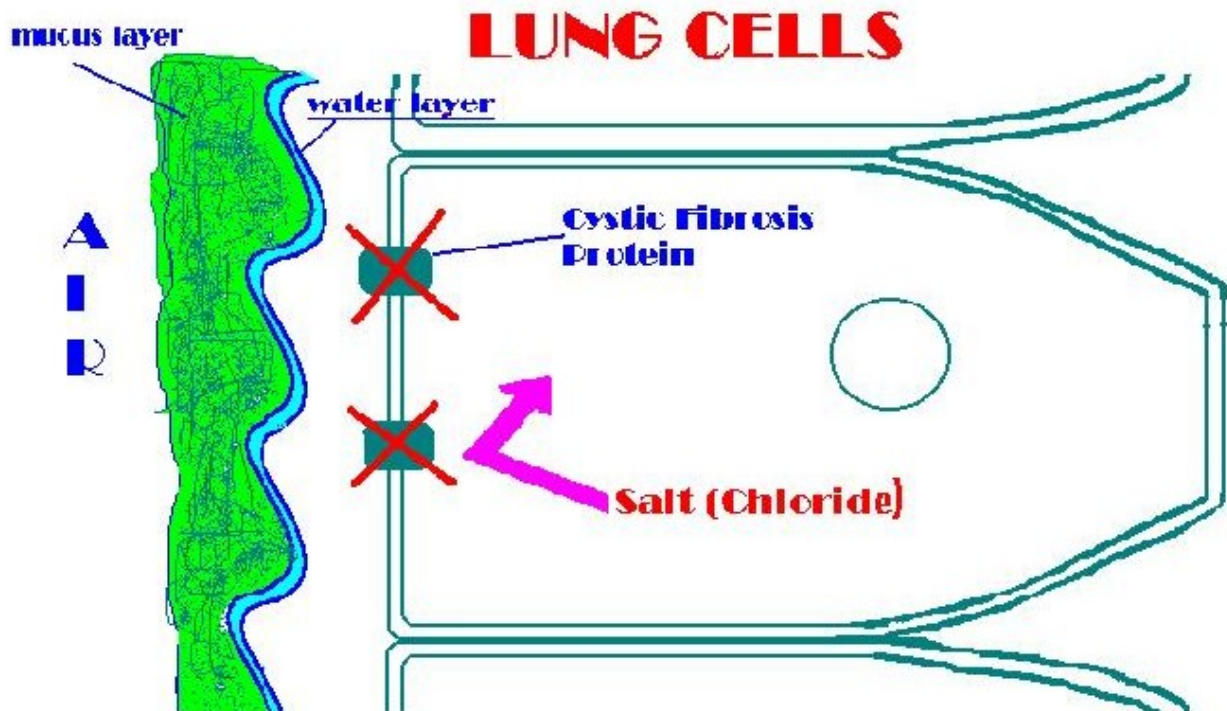
By: Dr. Barb Goodman

Cystic fibrosis is an inherited disease that is relatively common in the U.S. Cystic fibrosis affects multiple parts of the body including the pancreas, the sweat glands, and the lungs. When someone has cystic fibrosis, they often have lots of lung problems. The cause of their lung problems is directly related to basic problems with diffusion and osmosis in the large airways of the lungs.

People without cystic fibrosis have a small layer of salt water in the large airways of their lungs. This layer of salt water is under the mucus layer which lines the airways. The mucus layer in the airways helps to clear dust and other inhaled particles from the lungs.



In people without cystic fibrosis, working cystic fibrosis proteins allow salt (chloride) to enter the air space and water follows by osmosis. The mucus layer is dilute and not very sticky.



In people with cystic fibrosis, non-working cystic fibrosis proteins mean no salt (chloride) enters the air space and water doesn't either. The mucus layer is concentrated and very sticky.

People with cystic fibrosis have lung problems because:

~Proteins for diffusion of salt into the airways don't

- work. (less diffusion)**
- ~Less salt in the airways means less water in the airways. (less osmosis)**
 - ~ Less water in the airways means mucus layer is very sticky (viscous).**
 - ~Sticky mucus cannot be easily moved to clear particles from the lungs.**
 - ~Sticky mucus traps bacteria and causes more lung infections.**
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Therefore, because of less diffusion of salt and less osmosis of water, people with cystic fibrosis have too much sticky mucus in the airways of their lungs and get lots of lung infections. Thus, they are sick a lot.

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