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Hypertonic Saline Therapy in Cystic Fibrosis

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HYPERTONIC SALINE THERAPY IN CYSTIC FIBROSIS

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Abstract

Cystic fibrosis is an autosomal recessive genetic disorder that severely affects the lungs as well as other organs in the body. The pathophysiology of cystic fibrosis lung disease involves the production of thick and sticky mucus that accumulates leading to obstruction and inflammation of the airways. This disease is characterized by abnormal transport of sodium and chloride across an epithelium by defective genes, resulting in thick mucus secretions.

Nebulized hypertonic saline (3% NaCl) improves mucociliary clearance through rehydration of the airway surface liquid of the lungs. The purpose of this study is to evaluate the effects of nebulized hypertonic saline in improving lung function and decreasing exacerbation rates in older children (10-18 years of age) and adults with stable cystic fibrosis compared to normal saline (0.9%).

The review of literature will focus on comparing the use of nebulized hypertonic saline in enhancing mucociliary clearance in CF patients compared to other methods. The expected outcome is that nebulized hypertonic saline treatment will improve lung function and decrease exacerbation of CF in these groups.

Introduction

- Cystic fibrosis characterized by abnormal transport of sodium and chloride across an epithelium by defective genes, resulting in thick mucus secretions.
- Enhancement of therapy practices to advance outcomes is an area of cystic fibrosis care, which has progressed significantly over the last few decades. As the choices of treatment for CF increases, the clinicians are confronted with the predicament of developing a treatment plan that provides optimal benefit, addresses the complex therapeutic routine on patient compliance and is cost effective.
- Today, CF patients are living longer and clinicians need to recognize and implement early treatment to maintain optimal outcome. An early efficient management of children with CF is a road map to increasing the quality of life and health status of these patients as they transition to adulthood.

Statement of the Problem

- With the practices of newborn screening and an increase in life expectancy, the management of individuals with CF must adapt to a more vigorous and individualized methodology and CF physiotherapy management needs to reflect the shifting landscape, with increasing life expectancy and decreasing disease burden in CF patients.

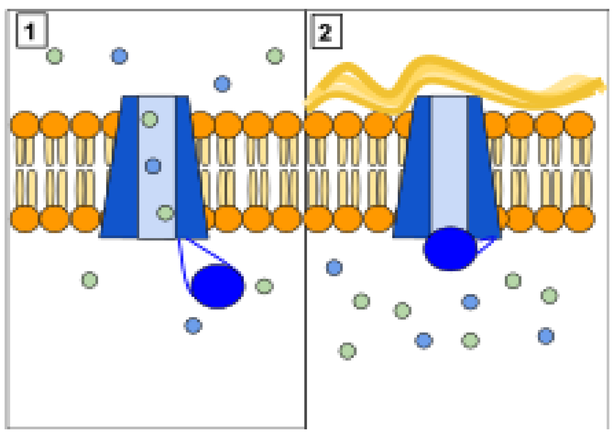
Research Question

- In older children and adults with stable cystic fibrosis who have failed to show improvement with recombinant human deoxyribonuclease (rhDNase) does treatment with nebulized hypertonic saline improve lung function, forced expiatory volume in one second (FEV1) and pulmonary exacerbation compared to NS?

Literature Review

- Gupta et al. (2012) conducted a randomized control trial in which 31 children diagnosed with cystic fibrosis were administered either 3% saline or 7% saline for 28 days.
  - Peak expiratory flow rate improved from 172.7 l/min at a baseline to 195.3 l/min on day 28 of the trial in the group that received the 3% hypertonic saline and from 194l/min to 212 l/min in the group that received the 7% saline.
- Eng et al. (1996) conducted a 48 week double-blind study in which patients were randomly assigned to inhale 4ml of either 7 percent hypertonic saline or 0.9 percent saline twice daily for 48 weeks.
  - The group that received the hypertonic saline treatment had significantly higher FVC (at 82ml; 95% confidence interval, 12, to 153) and FEV1 (at 68ml; 95% confidence interval, 3 to 132) values, but parallel FEF25-75 values.
  - Hypertonic saline had significantly greater numbers of patients without pulmonary exacerbation (76% compared with 62% in the control group; P=0.03).

Figure 1

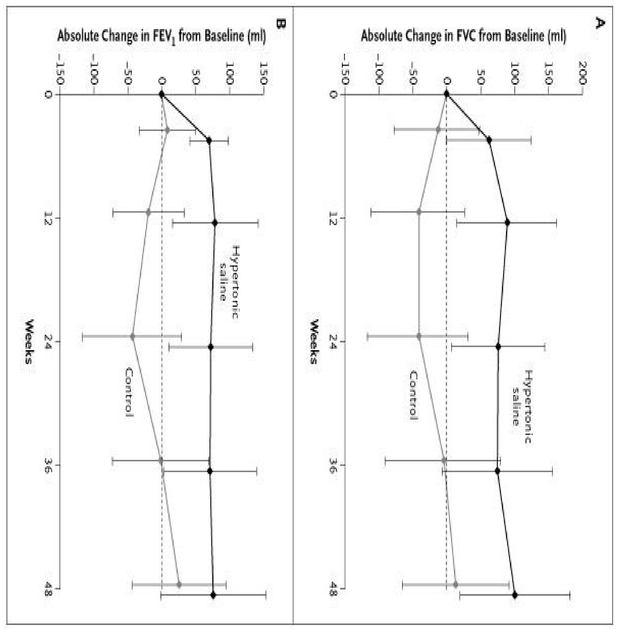


The CFTR protein is a channel protein that controls the flow of H2O and Cl- ions. When the CFTR protein is working correctly (Panel 1) ions freely flow in and out of the cells. However, when the CFTR protein is malfunctioning (Panel 2) Ions cannot flow out of the cell due to a blocked channel (Karl Kunzelmanet, 2000).

Discussion

- In a controlled long-term trial, treatment with hypertonic saline for one year had an adequate yet continued improvement in the level of lung function.
- More interesting, however, were the apparent reduction rates of exacerbations, antibiotic use, and overall improvement of quality of life that were associated with use of hypertonic saline.
- Eng et al. (1996) observed improvement of 15% in FEV1 from baseline with 6% hypertonic saline compared to isotonic saline..
- Hypertonic saline treatment is inexpensive and safe as an additional therapy in patients with cystic fibrosis and can play an essential role in the clinical setting.

Figure 2



Absolute Change from Baseline in Forced Vital Capacity (FVC) (Panel A) and the Forced Expiratory Volume in One Second (FEV1) (Panel B) (Elkins, 2006).

Applicability to Clinical Practice

- Nebulized hypertonic saline illustrates tangible improvement in mucus clearance without major adverse effects by reducing the number of exacerbation, use of antibiotics, and improving lung function.
- Hypertonic saline achieves this function by its direct effect on mucus, breaking the ionic bonds leading to lower viscosity and elasticity.
- Hypertonic saline has been shown to improve mucociliary clearance in patients with cystic fibrosis and recent studies provide indication for clinical efficacy for hypertonic saline in cystic fibrosis.

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