Drug Schedule for Inhaled Medications in Cystic Fibrosis

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Abstract

In persons with cystic fibrosis the lung deterioration is the primary means of death. The lungs deteriorate through acquisition of microorganisms and the development of pneumonia. The treatment used to prevent or minimize the lung component of cystic fibrosis involves many different inhaled medications. This paper seeks to determine the most effective ordering of inhaled medications through review of the existing literature, recommendations from professional organizations, as well as input from professionals working with individuals with cystic fibrosis and to then develop a policy that can be implemented in hospitals to optimize mucus clearance and lung function. There was found to be a limited amount of research concerning this topic, yet the professional organizations utilize the existing research and knowledge of physiology to recommend that the order of administration be: bronchodilator, Dornase Alfa, Hypertonic Saline, chest physiotherapy, and antibiotic. There is controversy surrounding the benefits and side effects of using of inhaled corticosteroids in the treatment if cystic fibrosis. Collaboration between nurses, respiratory therapists, and physicians is needed to implement and sustain the proposed protocol in the hospital setting.
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Problem to be Studied

Cystic fibrosis is an autosomal recessive genetic disorder that is the most prevalent genetic life threatening disease among Caucasians. The disease is the result of a mutation of the cystic fibrosis transmembrane conductance regulator gene (CFTR), which results in an abnormal expression of the protein CFTR throughout the cells of the body. This protein is responsible for ion transport on the surface of epithelial cells lining the lungs, pancreas, sweat glands, bile duct, and vas deferens (McCance & Huether, 2006). Disruptions in ion transport across the cell membranes of these cells cause the clinical manifestations and organ degeneration in cystic fibrosis.

The most commonly effected organs are the lungs, where the cause of death is usually pneumonia from decreased ability of the lungs to clear mucus and bacteria from the alveoli, bronchioles, and larger air ducts. The survival of individuals with cystic fibrosis is directly linked with their ability to maintain normal or stable lung function and to avoid infections and respiratory exacerbations.

Toward this goal, these individuals are frequently monitored and their respiratory function closely mapped through pulmonary function tests (PFT). These individuals are often on a large regimen of inhaled medications to maintain or increase their lung function. These medications include prophylactic antibiotics (to prevent pulmonary infections and suppress growth of bacterial colonies in the lungs) such as Tobramycin, mucolytics (to thin mucus) such as Dornase Alfa, expectorants (to assist in the removal of mucus) such as Hypertonic Saline,
corticosteroids (to decrease the inflammatory process) such as Fluticasone, and bronchodilators (to allow mucus to drain into larger airways) such as Albuterol. These patients also undergo daily chest physiotherapy, considered a vital part of cystic fibrosis management, which further aids in lung clearance. The purpose of this paper is to determine the best practice in the ordering of administration of inhaled medications, as they relate to each other as well as how they relate to chest physiotherapy, with persons with cystic fibrosis. This paper aims to determine if the ordering of inhaled medication administration alters lung function in persons with cystic fibrosis.

Cystic fibrosis is a complex, and multi-organ disease with treatment that requires many different medications in multiple forms. For this reason, the care and treatment of persons with cystic fibrosis is also a very complicated and involved process. This paper outlines the existing research and professional guidelines for the administration of inhaled medications and recommends the best practice for the order of inhaled medication administration. To this end, a literature review was performed and input on current practice from cystic fibrosis care centers was obtained.

Conceptual Model

This paper follows the conceptual model developed by Rosswurm and Larrabee (1999) for implementing research and evidence into the clinical setting. With this model the major steps include: assessing for a need in practice, link the problem with interventions and actions, synthesize best evidence, design practice change, implement and evaluate change in practice, and finally integrate and maintain change in practice (Rosswurm & Larrabee).

There was found to be no protocol for the ordering of inhaled medications in the treatment of cystic fibrosis, and a need for a consistent and effective ordering system. Improper
ordering of inhaled medications can lead to decreased pulmonary function and increased rates of infection, resulting of accelerated pulmonary decline. This problem can be altered by determining the most effective ordering of inhaled medications and developing a protocol that will guide practitioners in caring for individuals with cystic fibrosis with the aim of increasing lung function. This paper follows the final steps of the Rosswurm and Larrabee conceptual model in order to determine and implement the best evidence-based practice for the ordering of inhaled medications in the treatment of cystic fibrosis.

Review of Literature and Synthesis of Best Evidence

Prophylactic Antibiotics

Ramsey et al. (1999) compared the administration of inhaled Tobramycin with a placebo to investigate the efficacy and safety of inhaled Tobramycin for the treatment of Pseudomonas aeruginosa infection in persons with cystic fibrosis. The study consisted of two placebo-controlled, multicenter, randomized, double-blind trials with a total of 520 participants at 69 cystic fibrosis centers in the United States. All participants had been diagnosed with cystic fibrosis, had sputum cultures positive for Pseudomonas aeruginosa, were older than 6 years of age, and had a forced expiratory volume in one second (FEV1) between 25% and 75% of the predicted value. Treatment regimens consisted of 300mg aerosolized Tobramycin administered twice daily via jet nebulizer or the placebo delivered in the same manner over three cycles. Each cycle consisted of 28 days of Tobramycin or placebo administration followed by 28 days when these were not administered. Participants were evaluated for FEV1 and the density of Pseudomonas aeruginosa in sputum. At the end of 20 weeks, the participants treated with Tobramycin had an average of 8% increase in FEV1 while the placebo group had a 1% decrease.
There was also a decrease in the density of Pseudomonas aeruginosa in the lungs of the 
Tobramycin treated group, while the placebo had an increase in Pseudomonas aeruginosa 
density. This study showed the effectiveness of Tobramycin, both in increasing lung function as 
well as decreasing bacterial growth, in persons with cystic fibrosis.

Hudson, Gallagher, and Govan (2002) evaluated the change in lung function (through 
FEV1% predicted) and the change in Pseudomonas aeruginosa density in the sputum of both 
Tobramycin and Colistin in the treatment of cystic fibrosis. 108 participants completed the 
randomized controlled study, which consisted of 4-weeks of twice daily administration of either 
300mg aerosolized Tobramycin or 80mg inhaled Colistin. All participants were 6 years or older, 
had been diagnosed with cystic fibrosis, had a sputum culture positive for Pseudomonas 
aeruginosa, and had a FEV1% predicted of greater than 25%. The Tobramycin treatment group 
showed a 6.70% increase in FEV1% predicted from baseline, while the Colistin treatment group 
showed a 0.37% increase, which was not found to be significant. Both treatment groups showed 
a significant decrease in Pseudomonas aeruginosa sputum density. This study demonstrated that 
while both antibiotic agents decreased the amount of Pseudomonas aeruginosa colonized in the 
lungs, only Tobramycin increased lung function in persons with cystic fibrosis.

These studies demonstrate the effectiveness of chronic use of inhaled Tobramycin in the 
treatment of cystic fibrosis. These findings are in agreement with the Cochrane review (Ryan, 
Mukhopadhyay & Singh, 2008), which found that inhaled antibiotic treatment of persons with 
cystic fibrosis improves lung function and decreases exacerbations of infections. The Cystic 
Fibrosis Foundation (Flume et al., 2007) also recommended the chronic use of aerosolized 
Tobramycin.
While the use of antibiotics, especially Tobramycin, is supported by the literature and recommended by many professional organizations, there is no research, to date, on the optimal timing and order of administration within the existing inhaled medication regimen.

**Mucolytics**

In their review of current literature the Cystic Fibrosis Foundation ((Flume et al., 2007) recommends the chronic use of Dornase Alfa (a mucolytic) to improve lung function and reduce exacerbations.

Fitzgerald, Hilton, Jepson, and Smith (2005) performed a crossover, randomized, controlled trial to determine if the use of Dornase Alfa was equally efficacious if administered before or after chest physiotherapy (CPT) in patients with cystic fibrosis. All participants were between the ages of 5 to 18 years, had clinical evidence of mild to moderate lung disease, had an FEV1 of less than 90% predicted, and a forced vital capacity (FVC) greater than 40% predicted. Patients were randomized to receive Dornase Alfa either 30 minutes before or after chest physiotherapy. 50 participants completed the 6-week study. The study found that 67% (35 participants) showed a greater than 10% increase in FEV1 with Dornase Alfa over one two-week treatment block. The researchers found a statistically non-significant crossover difference of 0.02L when administration of Dornase Alfa before CPT was compared to after CPT and there were no statistically significant differences in the FVC or the mid-expiratory flow (MEF) between the two groups. This study demonstrated that Dornase Alfa is equally as efficacious when applied either 30 minutes before or after CPT in terms of lung function, quality of life, and aerobic fitness in children with cystic fibrosis. The study also stated that patients with Pseudomonas aeruginosa colonization may gain a greater improvement in FEV1 when the
Dornase Alfa was administered after CPT.

Van der Giessen, de Jongste, Gosselink, Hop, and Tiddens (2007) assessed the difference in efficacy between nebulization of Dornase Alfa before CPT and after CPT. Participants were included who were at least 5 years old, had been diagnosed with cystic fibrosis, were on maintenance therapy with Dornase Alfa, and were clinically stable. The study was a randomized, double blind, double dummy, cross-over design. Each participant was placed in one of two groups, these participants were given both nebulized Dornase Alfa and a placebo that resembled Dornase Alfa in appearance and taste. One group was instructed to use the Dornase Alfa before CPT and the placebo after, while the other group was instructed to do the opposite, and these schedules were reversed after 2 weeks. 25 participants were enrolled in the study, and 24 completed the study. After week 3 16 participants had a greater MEF25% (expiratory flow at 25% of the actual FVC) when they nebulized before CPT, whereas 7 patients had a decrease in the before CPT group. There were no significant differences in any other measurements (including FEV1 and FVC). This study stated that improved efficacy may be achieved when Dornase Alfa is inhaled before CPT. This study recommends that Dornase Alfa should be administered 30 minutes prior to CPT.

Van der Giessen, Gosselink., Hop, and Tiddens (2007) compared the efficacy of nebulization of Dornase Alfa before bedtime with nebulization in the morning after waking up. Participants were diagnosed with cystic fibrosis, at least 5 years old, had an FVC greater than 40%, were on maintenance treatment of Dornase Alfa, and were clinically stable. The study was a randomized, double-blind, double-dummy, crossover design where all subjects were nebulized with both Dornase Alfa and a placebo. One group used Dornase Alfa at bedtime and the placebo
In the morning, and the other group did the opposite. The groups reversed these administration schedules after 2 weeks. 25 participants enrolled in the study and 24 participants completed the study. MEF 25% did not significantly differ between bedtime or after waking administration. This study indicates that administration of Dornase Alfa is safe and effective when administered either at bedtime or after waking, and that there is no significant difference in lung function between the two schedules.

There appears to be no consensus on the optimal time for administration of Dornase Alfa. The prevailing thought seems to be that Dornase Alfa has a significant improvement on lung function, and that it is effective when administered either before or after chest physiotherapy. Further research is needed to determine the placement of Dornase Alfa within the medication regimen, apart from chest physiotherapy.

\textit{Expectorants}

Both the Cochrane review (Wark, McDonald, & Jones, 2008) and the study comparing the effectiveness of Hypertonic Saline (an expectorant) with Dornase Alfa (a mucolytic) by Eng, Morton, Douglass, Riedler, Wilson, and Robertson (1996) determined that while Hypertonic Saline is effective in lung function compared to a control, the improvement is less than that of Dornase Alfa. For this reason the Cystic Fibrosis Foundation (Flume et al., 2007) recommends the chronic use of Hypertonic Saline, although this use is not to substitute the use of Dornase Alfa.

Elkins, Robinson, Rose, Harbour, Moriarty, Marks, et al. (2006) investigated the long-term effects of Hypertonic Saline on lung function of persons with cystic fibrosis. This study was a parallel-group, randomized, controlled trial taking place over 48 weeks. All participants were at
least 6 years old and had a diagnosis of cystic fibrosis. Participants were randomly assigned to one of two groups, receiving either 7% saline or 0.9% saline, each with a bronchodilator administered 15 minutes before each dose administration (either the patient’s usual bronchodilator or two 100 µg puffs of Albuterol if the patient did not have a usual bronchodilator). 164 participants were randomized and completed the study. The Hypertonic Saline group showed increases in both FEV1 and FVC during the first 4 weeks, and then plateaued, while the placebo group showed FEV1 and FVC values that were unchanged from baseline. There also appeared to be fewer exacerbations requiring intravenous antibiotic therapy in the Hypertonic Saline group. In this study there was a moderate but sustained improvement in lung function and a marked decrease in the number of exacerbations. The authors concluded that the use of Hypertonic Saline preceded by a bronchodilator is an effective additional therapy for persons with cystic fibrosis.

Given that Hypertonic Saline is a relatively new therapy, there were a very limited number of studies investigating the sequencing of Hypertonic Saline along with other inhaled medications of cystic fibrosis. The preceding study, however, showed a sustained improvement in lung function with the use of Hypertonic Saline when preceded by a bronchodilator.

Corticosteroids

Balfour-Lynn, Lees, Hall, Phillips, Khan, Flather, et al. (2006) performed a multicenter, randomized, double-blind, placebo-controlled, trial to investigate the viability and safety of withdrawing inhaled corticosteroids from persons with cystic fibrosis, as well as determining if certain populations would, in fact, benefit from inhaled corticosteroid use. Participants were 6 years or older, diagnosed with cystic fibrosis, had an FEV1 greater than 40% predicted, and used
inhaled corticosteroids for greater than 3 months with 171 participants completing the study. The study took place over 8 months, including a 2 month run-in period where all participants received Fluticasone, with Salbutamol used as a rescue bronchodilator. During the trial, there was no difference in time to first pulmonary exacerbation, nor was there a significant change in lung function between the two groups. This study demonstrated that during the first 6 months after stopping inhaled corticosteroids there was no apparent impact (adverse or beneficial) on lung function of patients with cystic fibrosis. The placebo was found to be safe with no increase in adverse effects. This study shows that not all persons with cystic fibrosis need an inhaled corticosteroid, and that each patient should be examined for the need, such as lung hyper-reactivity and those persons with cystic fibrosis who have an asthma component to their disease.

In a review of anti-inflammatory therapy in patients with cystic fibrosis, Dinwiddie (2005) found that, while the use of inhaled corticosteroids is widespread, there is little research on the long-term benefits of these medications. Likewise, the Cochrane review (Balfour-Lynn, Walters, & Dezateux, 2008) of inhaled corticosteroids found that there was no evidence supporting the chronic use of inhaled corticosteroids in cystic fibrosis, while there was insufficient data on the effects of withdrawing inhaled corticosteroids from patients already treated with them. The Cystic Fibrosis Foundation (Flume et al., 2007) also recommends against the chronic use of inhaled corticosteroids in cystic fibrosis.

The consensus among the current research is that inhaled corticosteroids do not have an inherent benefit in the treatment of cystic fibrosis, except with specific circumstances. The use of inhaled corticosteroids is wide-spread and normally occurs after chest physiotherapy to decrease hyper-reactivity of the airway and to decrease neutrophil activity within the lungs. Further
research is needed to determine the course of action with persons with cystic fibrosis currently on inhaled corticosteroids, but it is recommended that patients not currently on corticosteroids remain as such.

**Bronchodilators**

While the Cochrane review of inhaled bronchodilators (Halfhide, Evans, & Couriel, 2008) found that there was insufficient evidence to support the use of Albuterol and other bronchodilators in cystic fibrosis, the Cystic Fibrosis Foundation (Flume et al., 2007) recommends for the chronic use of inhaled beta 2-adrenergic receptor agonists to improve lung function in cystic fibrosis. Despite this controversy, the use of bronchodilators in cystic fibrosis is widespread in not only practice, but research as well.

Natale, Pfeofle, and Homnick (1994) investigated the effectiveness of using an intrapulmonary Percussionator ventilator (IPV-1) in facilitating mucus production and mobilization as compared to aerosol and chest physiotherapy. An IPV-1 is a device that delivers rapid minibursts of gas mixture (including a bronchodilator) which is thought to increase bronchodilation and aid in mucus clearance. In this study 9 participants underwent a randomized crossover trial consisting of three treatment regimens, IPV-1 with 2.5mg of Albuterol, high volume aerosol with postural drainage and percussion (PD&P), or standard aerosol and PD&P. In comparing the three treatment groups, there was no significant difference in the change from baseline for PFTs from baseline between the three groups. In this study, it was determined that a single IPV-1 treatment was as effective as standard therapy in improving lung function in patients with excellent to moderate cystic fibrosis severity grades.

Marks, Hare, Saunders, and Homnick (2004) investigated the effectiveness of the
PercussiveTech HF devise (PTHF) as compared with standard CPT treatment. Like the IVP-1, the PTHF delivers a high-volume aerosolized bronchodilator along with chest physiotherapy. The study was a single-intervention, randomized, crossover, open-label trial consisting of two treatments, each occurring one week apart (either PTHF or traditional CPT). 10 participants were recruited and 9 finished the study. In this study there was no significant difference between the PTHF device and traditional CPT, showing that PTHF is as effective as CPT.

Both of the preceding studies investigated the integration of administration of inhaled bronchodilators and CPT into one step, thereby simplifying the medication regiment. The only drawback to this technique is that administration of other medications between bronchodilator administration and chest physiotherapy is not possible. As previously discussed, Hypertonic Saline shows a significant improvement in lung function when administered following a bronchodilator. With patients using Hypertonic Saline, it is then advised that the order of administration would be, 1) bronchodilator, 2) Hypertonic Saline, and 3) CPT. The patient not on Hypertonic Saline may see a decrease in medication complexity by using one of the previously discussed devices and combining bronchodilator administration with CPT.

**Professional Recommendations**

The Cystic Fibrosis Foundation examined the ordering of inhaled medications in 2006 (Network News) and found that well-designed research is greatly lacking in finding the best order of inhaled medication administration to optimize mucus clearance and lung function. The Cystic Fibrosis Foundation found that much of the ordering for inhaled medications was grounded on assumptions based on respiratory physiology. At that time, the Cystic Fibrosis Foundation determined that a bronchodilator should be administered first in order to open up the
airways, which will promote more thorough deposition of the other inhaled medications, improve CPT, and prevent bronchospasm. Hypertonic Saline was recommended as the second drug therapy, yet this status is less rigid than the bronchodilator, with the only set stipulations being that the bronchodilator is administered prior to the Hypertonic Saline and that the Hypertonic Saline precede CPT. The Cystic Fibrosis Foundation then advocated for the use of Dornase Alfa third. However, as with Hypertonic Saline, the authors stated that there was no firm evidence for this placement, and the patient’s preference should dictate when this medication is administered. The last medications to be administered were antibiotics and corticosteroids. The reasoning for this was that their effectiveness would be diminished if they were deposited onto mucus that was removed by CPT, thereby removing the medication from the lungs.

In 2007 the Cystic Fibrosis Foundation (Flume et al.) performed a review of literature concerning the inhaled medications, and found again that there was a shortage of research concerning the ordering of inhaled medications. Despite the limited research in the area, this study recommended that the order be: bronchodilator, Hypertonic Saline, Dornase Alfa, CPT, and antibiotics. In this review, the authors did not include the use of corticosteroids, due to the evidence that inhaled corticosteroids are not beneficial for all persons with cystic fibrosis.

Two hospitals responded to a query concerning the ordering of inhaled medication administration in cystic fibrosis sent out to the cystic fibrosis nurses listserv. Both hospitals responding were located in Tennessee and one hospital in Tucson, Arizona was contacted through personal e-mail. The East Tennessee Children’s Hospital (C. Norris, personal communication, December 3, 2008) ordered their medications as: bronchodilators, Hypertonic Saline, Dornase Alfa, CPT, antibiotics, and corticosteroids. The hospital at the University of
Tennessee at Memphis (T. Knight, personal communication, December 3, 2008) ordered their medications as: bronchodilator, Hypertonic Saline, Dornase Alfa, CPT, corticosteroids, and antibiotics. The staff replying on behalf of this hospital stated that their ordering appeared medically and physiologically logical. G. Drake, the CF Respiratory Therapist Care Center Coordinator for the University Medical Center (UMC) in Tucson, AZ (personal communication, December 2008) stated that the UMC orders their medication as: Albuterol/Xopenex (bronchodilator), Dornase Alfa and/or Hypertonic Saline, CPT, antibiotics, and corticosteroids. This individual stated that this ordering was adopted due to the Cystic Fibrosis Foundation recommendations.

There appears to be a generalized consensus for the overall ordering of inhaled medications in the treatment of cystic fibrosis, governed by the Cystic Fibrosis Foundation recommendations and common medical sense, rather than evidence-based practice. There is still a great deal of research needed to determine if this ordering is the best-practice for optimizing mucus clearance and lung function. For the present, the professionals seem to be at a consensus that bronchodilators are administered first, followed by either Hypertonic Saline, Dornase Alfa, or both, CPT, and then antibiotics. The most controversy seems to revolve around the use of corticosteroids, yet when this medication is included it is administered after CPT. Research suggests that corticosteroids have no beneficial effect for most persons with cystic fibrosis and should only be used in certain circumstances (such as the co-morbid condition of asthma). The research, however, also shows that there appears to be no detrimental effects to continuing inhaled corticosteroid treatment in persons with cystic fibrosis already taking the medication. The best course of action in this case is perhaps evaluating each person individually to determine
the necessity of beginning treatment, continuing treatment, or discontinuing treatment with inhaled corticosteroids.

Proposed Program

Utilizing the above information as well as professional input, the author has developed a written protocol for the ordering of administration of inhaled medications for the treatment of cystic fibrosis for the University Medical Center (UMC) in Tucson, AZ. This protocol follows the latest Cystic Fibrosis Foundation guidelines as well as the available research, while receiving professional direction, given the limited amount of research in many areas of this field, and takes into account the existing ordering.

This protocol will be inserted into the UMC Respiratory Care Services policy governing the care of Cystic Fibrosis Patients (see the Appendix A for full protocol). The ordering protocol will be placed under the heading of Small Volume Nebulizers (5.8) and will include the following:

5.82 Order of Therapy

1. Bronchodilator (Albuterol/Xopenex): for widening the airways and allowing better distribution of other inhaled medications.

2. Dornase Alfa (Pulmozyme): a mucolytic given to break-up the mucus in the lungs to facilitate better mucus clearance.

AND/OR

3. Hypertonic Saline: an expectorant that pulls water into the mucus layer and decreases bonding within the mucus, which leads to decreased mucus viscosity, which ultimately allows for better mucus clearance.
NOTE: It is hypothesized that Hypertonic Saline inactivates Dornase Alfa, and therefore, if both are administered in one session, Dornase Alfa should always be administered prior to Hypertonic Saline.


5. Inhaled Antibiotics (Tobramycin): used after mucus clearance to better deposit on the lungs and decrease/halt bacterial growth within the lungs.

6. Administration of inhaled corticosteroid (i.e. Fluticasone) at the discretion of the healthcare provider.

Provisions will be made for individual medication needs and plans, as well as provider preferences. A decision chart will follow step 6 in order to aid the healthcare provider in determining the necessity for inhaled corticosteroids. For full decision chart see Appendix B.

Program Implementation Plan

In order to implement the new protocol the protocol must first be approved for and added to the previous protocol. To this end, the protocol will be given to the UMC respiratory therapy department for review and approval and will then become a part of the protocol for caring for Cystic Fibrosis Patients. Following this approval, there will be an informative meeting/training session of respiratory therapists, physicians, and nurses who have the responsibility of caring for individuals with cystic fibrosis. This meeting will cover the correct ordering of the inhaled medications as well as the rationales for that ordering, so as to increase understanding and compliance on the part of the providers. This could be accomplished with either a formal educator, who demonstrates and instructs groups, or through a computer training module that
could be developed. The last option would allow for more flexibility in when all members of the healthcare team could access this information. This option could also allow for the healthcare worker to refer back to the training if they have questions or need to review the material.

Following this meeting the ordering protocol will then be added to competency/annual check-off requirements for respiratory and nursing staff. This will maintain correct knowledge of ordering and allow for changes in current recommendations. Due to the need for greater research in this field, this will allow the healthcare providers to keep abreast of changes found in the research and further promote the evidence-based practice model.

After the initial training, it is also important that the patient, nurse, and respiratory therapist all know and adhere to the medication ordering schedule. In order to accomplish this, the nurse and respiratory therapist should confer prior to beginning administration of inhaled medications. To further ensure that all parties are aware of the medication ordering and schedule, the nurse or respiratory therapist will create a poster that shows the ordering of inhaled medications, as well as the times the inhaled medications are to be given. This poster will be placed above the patient’s bed, so that the patient has access to the information as well as the healthcare staff. This also allows the patient to better understand their medication routine and gives them a greater sense of control while in the hospital. The poster provides consistency and allows for joint collaborative teaching (by both the respiratory therapist and the nurse) as to the ordering of inhaled medications, which should increase patient compliance following discharge from the hospital.

The last aspect of implementation would occur just prior to discharge. At this point, the respiratory therapist and the nurse would sit down together and have a joint discharge session
with the patient. In this session medication administration, ordering of medication, and all other respiratory and health issues can be discussed and any questions can be answered. This would given further education and understanding to the patient and promote greater compliance to the treatment plan.

Program Evaluation Plan

Since the drug schedule for the protocol is very similar to what is already in practice, evaluation of the protocol effectiveness will look at individual records, examining improvement, as measured through pulmonary function tests (PFTs), specifically forced expiratory volume in one second (FEV1) levels, rate of improvement, number and duration of hospitalizations, and number of pulmonary exacerbations/exacerbations. This will be accomplished through chart audits, looking at PFTs over time, what medications the patient was previously and currently taking to correlate with PFT levels. The length of time between hospitalizations following implementation of protocol will also be examined. Provisions will be made during this process to account for other factors that could also contribute to degeneration in patient condition. These factors include advancing age (and the advancing illness that accompanies age), non-compliance with treatment, and other compounding conditions. The individual Respiratory Therapist’s understanding of the order of medication administration will be measured via stated understanding as part of the UMC respiratory department competency testing done bi-annually.

If it is evident that, after a period of 12 months, that the protocol has an impact on the improvement of respiratory recovery and a decrease the duration and number of hospitalizations, and an increase in the time between hospitalizations, then there will be an annual or biannual review of existing literature, in order to improve and stay current with research in the field. There
will also be an annual chart audit that seeks to ensure that individuals with cystic fibrosis treated at UMC are still maintaining the improvements that were seen in the first 6 months of the implementation of the inhaled medication ordering protocol.

If, however, there appears to be a consistent decrease in all of the above mentioned indicators of pulmonary improvement, further evaluation of the ordering protocol will be needed. It will be necessary to determine the reason for the decrease, as well as a need for revision of the protocol in order to reflect this new information.

Conclusion

Cystic fibrosis is a complex disease that affects multiple organs and organ systems. The most affected system is the pulmonary system, with death resulting from mucus build-up and bacterial colonization of the lungs. The efforts toward combating this process include multiple inhaled medications that are aimed at decreasing mucus aggregation and stopping bacterial growth and colonization. There has only been a limited amount of research on the ordering of administration of these medications. Through the existing research, professional recommendations, and current healthcare facility practices, the best-evidence ordering of these medications is: bronchodilator, Dornase Alfa and/or Hypertonic Saline, chest physiotherapy, and antibiotic. There remains a lack of consensus surrounding the use of corticosteroids. At UMC, when corticosteroids are used, they are given last in the order. A protocol delineating this ordering has been developed for the UMC, along with an implementation and evaluation plan. There is opportunity for collaboration between nurses and respiratory therapists in the implementation of the protocol.
References


1.0 INTRODUCTION:

1.1 The Cystic Fibrosis patient is considered a high priority patient at UMC. The RCP provides a critical aspect of the ongoing therapy that these patients need. Classically, Cystic fibrosis patients receive bronchodilator, airway clearance, and mucolytic therapies from RCP’s.

2.0 PURPOSE:

2.1 To define the expected level of care for Cystic Fibrosis patients.

3.0 OBJECTIVE:

3.1 To optimize the respiratory function of these patients.

4.0 POLICY:

4.1 The administration of treatment to cystic fibrosis patients may be performed by the following Respiratory Care personnel only:

- Respiratory Care Practitioners Adults and/or Pediatrics, to the level of expertise that they are orientated to
- Respiratory Care Clinical Leaders Adults and Pediatrics
- Respiratory Care Specialists Adults and Pediatrics
- Respiratory Care Director Adults and Pediatrics
- Affiliated Instructors/Students Adults and Pediatrics

4.2 Instruction for administration of treatment to cystic fibrosis patients will be provided by the Respiratory Care Education Coordinator or designee.

4.3 Direct supervision of Respiratory Care staff members during initial administration and usage is not necessary subsequent to orientation. Afterwards, supervision of student’s and staff will be provided on an "as needed" basis. 4.31 Indirect supervision is available through the shift Lead Respiratory Care Practitioner.
5.0 PROCEDURE:

5.1 Upon admission to hospital, the physician will usually order the following studies. These will be
referred to the proper department by the unit assistant.
5.11 Routine lab work
5.12 Pulmonary function testing
5.13 Chest x-ray
5.14 Pulse oximetry spot check

5.2 These tests may be repeated during the hospital stay and upon discharge of the patient from the hospital.

5.3 Treatments will be administered according to physician orders, usually a QID basis on the following
schedule: 0700, 1100, 1500, 1900.

5.4 The delivery of respiratory procedures for Cystic Fibrosis patients are a priority activity.

5.5 The care of Cystic Fibrosis patients is superseded only by:
5.51 Code or trauma calls
5.52 Critical ventilator care (excludes routine ventilator checks)
5.53 Emergency room care
5.54 Stat blood gases

5.6 In the event any respiratory care may be delayed the charge person must be notified to reduce or avert the
delay.

5.7 Practitioners assigned to other areas of the hospital may be assigned a Cystic Fibrosis patient to assure the
treatments will be given on a timely basis. The treatments should be completed before breakfast, lunch and
provide adequate time lapse after dinner (i.e.: 2 hours)

5.8 Small Volume Nebulizers:
5.81 All small volume nebulizer (SVN) treatments will be administered with 100% Oxygen unless
otherwise ordered.
5.811 SVN equipment for CF patients should be changed out Q24 hours

5.9 Chest Physiotherapy (CRT):
5.91 All CF patients should be placed in a "Hill Rohm 4 rail adult mechanical bed".
5.911 Communication with the unit UA will be necessary if a bed needs to be ordered.
5.92 Postural drainage & percussion shall be administered in the appropriate positions for a period of 30-
45 minutes. (See CPT Policy)
5.93 Patients with CPT ordered QID and TID will receive q-other treatments with an alternative method
of postural drainage other than manual CPT with hands. This policy will be followed regardless of
the number of CF patients admitted to the hospital.
5.931 Alternative treatments can be suggested, such as:
5.9311 High Frequency vest
5.9312 Vibramatic
5.9313 Flutter valve
5.9314 Quake
5.932 The schedule of alternative CPT treatments will be delivered at 0700 and 1500.
5.933 Respiratory staff should follow this treatment schedule whenever possible in order to provide
therapy that is consistent to all patients.

5.94 For patients that we are providing their own equipment:
5.941 BME should be notified at 4-7500 to request an "in coming electric check" on their
compressor.
5.942 Both the compressor and vest belonging to the patient is to remain in their room at all times.

5.95 Hospital issues equipment (compressors) may be used between patients provided appropriate
infection control policies are followed for the cleaning of the tubing and compressor between
patients.
5.951 Patients with b-cepacia must have the compressor left in their room

5.10 Oxygen Therapy:
5.101 Oxygen shall be humidified and administered according to physician orders
5.102 Oxygen may be administered during postural drainage & percussion to relieve shortness of breath.
5.103 Oxygen shall be administered at night if ordered.

5.11 Treatments will be discontinued only with a physician's order or if the patient is discharged from the hospital.

5.12 In the event the patient refuses any part of the therapy the physician shall be notified immediately.

5.13 Antibiotics may be aerosolized per physician order, utilizing a separate nebulizer after bronchodilator and CRT.

6.0 REFERENCES:

6.1 Chest Physiotherapy in the Intensive Care Unit, 2nd edition, Mackenzie
6.2 Clinical Application of Respiratory Care, 2nd edition Shapiro
6.3 Respiratory Care, 2nd edition, Burto
Appendix B

Decision Tree for the Use of Inhaled Corticosteroids

Current use of corticosteroids?

NO

Evidence of concurrent asthma, or asthma-like symptoms (i.e. airway reactivity, reversible airway obstruction, recurrent wheezing)/

YES

Evaluate patient condition, and, when stable, discuss with patient and slowly decrease the corticosteroid, carefully monitoring patient for previously described symptoms.

NO

Continue use of corticosteroids as currently manages the patient’s symptoms.
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