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Review

Physiotherapy in cystic fibrosis: optimising techniques to improve outcomes

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EDUCATIONAL AIMS

- To highlight that CF physiotherapy management has needed to adapt to a more dynamic and individualised approach over the last 2 decades.
- To outline some alternative treatment regimen options for patients with CF and their families, which can be an effective method of optimising adherence.
- To outline how the introduction of newborn screening has provided new challenges for physiotherapists working in CF.
- To discuss how physical activity and exercise now plays an important role in the management of individuals with CF.

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SUMMARY

Optimisation of physiotherapy techniques to improve outcomes is an area of cystic fibrosis (CF) care, which has developed considerably over the last two decades. With the introduction of newborn screening and an increase in median life expectancy, the management of individuals with CF has needed to adapt to a more dynamic and individualised approach. It is essential that CF physiotherapy management reflects the needs of a changing cohort of paediatric CF patients and it is no longer justifiable to adopt a 'blanket' prescriptive approach to care. The areas of physiotherapy management which are reviewed and discussed in this paper include inhalation therapy, airway clearance techniques, the management of newborn screened infants, physical activity and exercise.

INTRODUCTION

As the range of treatments for CF increases, the clinician is faced with the dilemma of developing a treatment schedule that provides optimal benefit, takes into account the impact of an increasingly complex therapeutic regimen on patient adherence and which works within the financial restraints imposed on the system.[1] Median life expectancy in CF has increased significantly over the last two decades.[2] The challenge faced by clinicians is to balance CF treatment needed to maintain optimal health status (thus allowing people with CF to achieve personal and professional milestones) with the burden that treatment imposes.[3] It is therefore essential that physiotherapy treatments are optimised to ensure clinical efficacy and optimal outcomes. In essence, the aim of paediatric CF management is to transition 'healthy' children to adult care with normal lung function and nutritional status. The purpose of this review is to discuss some of the methods available to optimise physiotherapy techniques in order to potentially improve outcomes.

INHALATION THERAPY

Inhalation therapy is an important part of CF physiotherapy care and is aimed at altering the properties of the airway surface liquid and/or reducing the viscosity of mucus to facilitate mucociliary clearance.[1] The most common mucoactive agents pertinent to CF physiotherapy care are dornase alfa (rhDNase) and hypertonic saline (HTS). When incorporating inhalation therapy into CF therapeutic routines careful consideration should be paid to the timing of treatment, the delivery method of these nebulised medications and adherence.

Timing of inhalation therapy

Hypertonic saline

Hypertonic saline (HTS) is a sterile salt-water solution, delivered via inhalation therapy, usually at a concentration of between 3% and 10% in volumes of 3 ml to 10 ml via a nebuliser.[4] HTS has been shown to accelerate mucociliary clearance in the CF

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airway[5,6] and three mechanisms are believed to contribute to this improvement. Firstly, restoration of the depleted airway surface liquid volume occurs which peaks almost immediately after a dose, but which may be sustained for several hours.[7] Secondly, there is an improvement in mucus rheology[8] and thirdly stimulation of cough.[5,6] Significant improvements in lung function, quality of life and ease of expectoration with short and long-term use of HTS have been reported.[4,9]

The clinical trials which have demonstrated the clinical efficacy of HTS, have all delivered the medication before airway clearance therapy. As a result the currently accepted clinical practice for nebulised HTS is that each dose is nebulised immediately prior to airway clearance treatment. Alternative timing regimens have been suggested and are empirically used in clinical practice. These include the proposed use of hypertonic saline after airway clearance therapy when the excess bronchial secretions have in theory been mobilised. A recent systematic Cochrane review reported no current evidence to indicate that alternative timing regimens are any more efficacious than the current practice.[10] A recent study of CF adults evaluated the use of HTS before, during and after airway clearance therapy. There was a non-significant trend towards an improvement in lung function when HTS was inhaled before airway clearance compared to during or after treatment. Satisfaction and perceived effectiveness were highest when HTS was inhaled before or during airway clearance. Tolerability was not affected by the timing regimen and satisfaction was lowest when HTS was inhaled after airway clearance therapy.[11] The differences in perceived effectiveness and satisfaction shown in this study may have important implications for long-term adherence.[4]

The advice therefore remains that patients' nebulise HTS before airway clearance treatments as this is the only regimen which has demonstrated clinical efficacy to date.[10] The best time of day for inhalation of HTS has also not been established and clinical practice should therefore be based on convenience and tolerability rather than on a "one size fits all" approach.[10]

Dornase alfa

Dornase alfa (rhDNase) is a genetically engineered version of a naturally occurring enzyme, which was developed to reduce the visco-elasticity of sputum. Daily inhalation of dornase alfa has been shown to reduce the number of pulmonary exacerbations, improve pulmonary function and is well-tolerated and safe in patients with mild, moderate and severe CF.[12] Therapy with dornase alfa over a one, six or 12 month period is associated with an improvement in lung function.[13] Alternate day treatment with dornase alfa has also been shown to be equally effective as daily treatment in some patients and in those patients using this simplified alternate day treatment regimen may help to reduce treatment burden.[14]

As with HTS there are some recognised practices with regards to timing of dornase alfa. Traditionally, clinicians advise nebulisation 30 minutes prior to airway clearance therapies (ACTs) and that at least one hour should be allowed between the nebulisation of dornase alfa and a nebulised antibiotic to avoid denaturation.[3] This recommendation is based on evidence that dornase alfa makes sputum more pourable within 30 minutes.[15] However alternative timings have been suggested. Inhalation after airway clearance treatments may facilitate drug deposition in the more peripheral airways.[16] Inhalation in the morning may capitalise on faster daytime mucociliary clearance and on the clearance effects of daytime activities such as exercise. Inhalation in the evening, may increase the dwell time in the airways since mucociliary clearance and cough are suppressed overnight.[16]

A recent Cochrane review concluded that the current evidence from a small number of subjects does not indicate that inhalation of dornase alfa after airway clearance is more or less effective than the traditional recommendation of inhalation 30 minutes prior to airway clearance techniques (ACTs).[16] It has also been hypothesised that for children with well-preserved lung function inhalation before airway clearance may be more beneficial for small airway function as two small studies reported FEF_{25} to be significantly worse when dornase alfa was inhaled following airway clearance.[17,18] Clinicians are often anxious that evening inhalation may induce cough and impair sleep quality and in practice this is therefore not routinely recommended.[16] In general it can be concluded that the timing of dornase alfa can be largely based on pragmatic reasons and individual preference with respect to the time of airway clearance and time of day.[16] Providing alternative regimens to patients and families can be a very effective method of optimising adherence to treatments in CF.

Delivery methods

In order to balance treatment demands with lifestyle, therapies have been developed which allow patients to administer treatments independently and often within the home environment.[3] Optimising inhalation techniques is essential to ensure optimal deposition and drug delivery with each dose. The success of inhalation therapy is highly dependent on indication, the administration device and the inhalation technique. New technologies have been developed to produce more homogenous particle sizes and to increase the speed of nebulisation.[3] These include vibrating mesh technology (VMT) systems and adaptive aerosol devices (AAD). These devices are silent, battery powered, portable and have faster delivery times. In addition AADs also reduce wastage of medication by using breath actuated technology and allow clinicians to download technique/usage data which can facilitate optimisation of technique and adherence.[19]

In paediatrics the use of a facemask for inhalation therapy is the first line approach for the infant and younger child, however some toddlers may be able to manage a mouthpiece.[20] As children become older their tolerance of facemasks may reduce and their ability to nebulise effectively through a mouthpiece improves. The use of a nose-clip and mouthpiece in the interim periods can be useful to ensure optimal deposition of the inhaled medication in the lungs. Deposition of medication in the nasal space/sinuses may have some efficacy in patients with nasal obstruction and/or sinus disease and therefore a facemask may be appropriate for these individuals regardless of age.

Patients and parents/carers must always be carefully instructed and trained regarding the optimal inhalation technique and this should be regularly re-assessed and optimised regardless of the choice of device.[20] In essence an optimal inhalation technique is dependent on the individuals age, ability, willingness to learn and the degree of lung disease[20] and these factors need to be taken into consideration at all times.

Appropriate cleaning and maintenance of nebuliser equipment is essential to avoid bacterial contamination of equipment, to decrease the potential risk of acquiring pathogens and to ensure efficiency of the device.[21] Written cleaning and sterilisation guidelines should be provided to all patients and families and cleaning practices should be reviewed on a regular basis.

Adherence and tolerance

The median number of daily treatments prescribed for patients with CF is seven (inter-quartile range (IQR) 5–9).[22] Poor adherence has been identified as the greatest cause of treatment failure. Although technological improvements have reduced the inhalation time considerably, adherence to inhalation therapy remains poor.