

Physiotherapy in infants and young children with cystic fibrosis: current practice and future developments

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The manifestation and onset of cystic fibrosis (CF) lung disease is highly variable and the collective wisdom of experienced physiotherapists based in clinical practice is that no standard physiotherapy treatment will suit everyone. Despite this, many physiotherapists keep looking for a standard physiotherapy treatment that will suit all. The number of (apparently) asymptomatic CF neonates diagnosed with newborn screening is increasing worldwide. In the past, physiotherapy in CF was synonymous with airway clearance therapy in order to decrease pulmonary symptoms. The definition of the term 'modern physiotherapy in CF' is much wider. It involves the whole individual in a long-term perspective. The traditional physiotherapy treatment approach in infants has been extrapolated from studies in older patients with chronic sputum production. This approach may be flawed as it fails to consider the uniqueness of the immature infant and small child. During the past two decades a number of different airway clearance techniques have been developed. The choice of an airway clearance technique seems currently to be led by culture, tradition and geographical influences. Development of an optimal individualized physiotherapy regimen requires knowledge about respiratory physiology and pathophysiology, the aims of modern CF care and the rationale for the alternative physiotherapy techniques available. Optimal treatment is not synonymous with maximal treatment. Side-effects of treatment in this vulnerable immature population need to be carefully considered to make sure we do no harm. The aims of modern, individually tailored physiotherapy, as one part of the CF care package, are primarily to retard lung disease and preserve physical function in order to improve quality of life and long-term outcomes. 'Good physiotherapy is the mainstay to clinical well being in CF'.¹

CF LUNG DISEASE

Hypersecretion of viscous, infected secretions and mucus plugging, mucosal inflammation, bronchoconstriction,

airway instability or lung tissue damage can cause pulmonary obstruction alone or in combination, often in older patients with CF. In the lungs of young CF patients the obstruction is likely to be due to hypersecretion of viscous secretions, mucus plugging and inflammation of the peripheral airways. However, considerable disease may be present in the peripheral airways before airflow resistance increases and a decreased flow is measurable.² There is continuing debate about which comes first: inflammation or infection. If inflammation occurs first, the exact cause is unknown, although there are many theories, i.e. ionic balance of the mucus causing inflammation or an increase in inflammatory response to normal respiratory pathogens. One of the markers of airway inflammation is the abundance of neutrophil elastase in the airways. Amitani *et al.* showed that neutrophil elastase damages the respiratory epithelium resulting in impairment of mucociliary transport and thus causing retention of infected secretions.³ The more airway obstruction present, the more ventilation distribution and gas exchange is affected.⁴ The progressive pulmonary obstruction and the lung destruction due to the vicious cycle of chronic inflammation and infection are difficult to eradicate, and result in the development of bronchiectasis, airway wall thickening, cysts and permanent lung damage.^{5–7}

Hypersecretion

Pseudomonas aeruginosa may survive longer in airways abundantly filled with sputum⁸ and airway plug formation is one factor reducing the eradication of pathogens from CF airways by antibiotic therapy.⁹ One factor in the need of high doses of antibiotics is the inaccessibility of *P. aeruginosa* in plugs.¹⁰ Combining intravenous (IV) antibiotic treatment with intensified airway clearance has been shown to improve lung function more than IV antibiotic treatment alone.¹¹ Higher sputum volumes are correlated with a higher degree of inflammation, lung obstruction and destruction. By removing infected secretions the rate of proteolytic tissue damage can be decreased.¹² Inflammatory processes also increase metabolism which in turn increases minute ventilation and the work of breathing.¹³ A care package focusing on minimizing airway obstruction and mucus plugging and performing regular physical exercise has contributed positively to the maintenance of functional status in two different geographical regions. In one CF

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centre, only 51% of all adults¹⁴ and in another centre 20% of all 18-year-olds were colonized with *P. aeruginosa* (McIlwaine, 2003, personal communication).

Breathing mechanics

Pulmonary hyperinflation is a response to the clogging and collapse of airways in an attempt to keep the tidal volume (TV) breathing above closing volume (Figure 1). Most often the degree of hyperinflation follows the degree of obstruction. With hyperinflation, expansion of the chest is accomplished by contracted inspiratory muscles. Expiratory flow is controlled by eccentric contractions of inspiratory muscles and inspiration is accomplished by concentric contractions, but from an unfavourable point on the length/tension curve. The more hyperinflated the chest, the closer the inspiratory muscles get to active insufficiency. Atelectasis occurs if TV is not kept above closing volume, ventilation distribution decreases, and breathing frequency, minute ventilation and the work of breathing increase further. Accessory muscles become involved in the process of hyperinflation. As hyperinflation increases and as ventilation becomes more insufficient these muscles are progressively more involved in the mechanics of breathing. Accessory muscles originate from the chest and insert on the upper limbs, cervical region and skull. Concentric contraction assists in the inspiratory movements required of the severely hyperinflated chest when upper limbs and head are fixed while sitting upright, leaning forward with elbows on a table and the head resting in the hands.¹⁵

As with all soft tissues, the accessory muscles shorten when not stretched to full range regularly. Shortening of the musculo-skeletal tissues in the hyperinflated chest results in elevated and protracted shoulders, a thoracic kyphosis, a cervical lordosis and a decreased mobility of the chest.^{16,17} Many children, adolescents and young adults with CF are reported as having a hyperinflated chest, stiff intervertebral and costovertebral joints, bad posture, thoracic kyphosis and back pain.^{16,18–22} The reported wedging or compression of vertebrae^{19,21} in adult CF populations may be caused by the ergonomic burden of the

thoracic kyphosis in combination with osteoporosis.²³ It has been shown that bad posture, thoracic kyphosis and back pain are partly reversible if properly treated.^{16,17,22,24} But trying to regain what has been lost can be difficult. It is time-consuming, uncomfortable for the patient and an added burden to the CF care package. Thoracic kyphosis due to wedged vertebrae cannot be rehabilitated, but rather increases the load on the adjacent vertebrae. The role of regular physical exercise which maintains normal muscle strength, length and joint mobility as part of the physiotherapy programme from the very beginning will be discussed.

THE UNIQUENESS OF THE INFANT

Infants are developmentally immature and not just little adults. They have a cylindrical and more cartilaginous rib cage and more horizontally aligned ribs causing the diaphragm to be flatter and the sterno-costal angle to be wider. The intercostal muscles are underdeveloped and mechanically less efficient due to the horizontal alignment of the ribs. Therefore infants depend on their diaphragm for respiration. The flatter diaphragm in the infant is mechanically less efficient and has proportionally more type 2 (fast twitch) fibres and fewer type 1 (endurance) fibres and is therefore more vulnerable to respiratory fatigue than in the mature individual.² The wider sterno-costal angle and the flatter diaphragm reduce the effectiveness of the anti-reflux barrier. Infants are obligatory nose-breathers with a larger tongue and a soft palate that extends nearly to the larynx. The suck-swallow-breathing mechanism relies on exquisite controls. Nasal regurgitation is more common in infants.²⁵

Positioning for the purpose of improving ventilation/perfusion matching differs between infants and adults. In adults ventilation and perfusion are preferentially distributed to the dependent lung because of gravitational effects and arterial oxygenation is therefore better in the dependent lung.² In infants the opposite applies: oxygenation is better in the uppermost lung.²⁶ The soft infant chest wall does not support the lungs as well as the adult thorax—thus the infant's resting pleural pressure is closer to atmospheric pressure than that of the adult and therefore airway closure occurs in more dependent regions. In side-lying, ventilation is distributed away from the dependent region to the uppermost lung in the infant.

In the embryo the lungs are an offshoot of the primitive foregut at 6 weeks of gestation. At birth lungs are far from mature. The number of alveoli will increase 15 times and the size of the air-tissue interface more than 25 times (Table 1).²⁷ Lung volumes are proportional to those in the adult but smaller (Table 2).²⁸ The gastric and respiratory systems share a common nerve supply via the vagal nerve

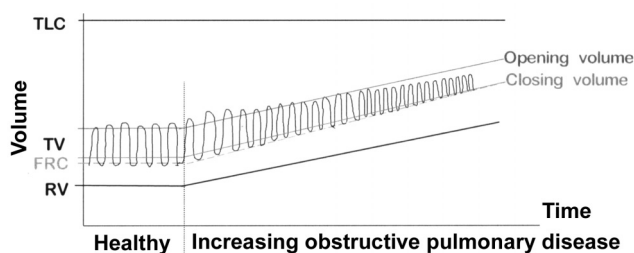


Figure 1 Changes in lung volumes with progressive obstructive pulmonary disease. TLC, total lung capacity; TV, total volume; FRC, functional residual capacity; RV, residual volume

and a common pathway through the oropharynx. Sensory information from the oesophagus is conveyed by the vagus nerve to the nucleus of the solitary tract in the dorsomedial medulla. Interneurons project to the motor neuron pools in the ventrolateral medulla, after which motor efferents located in the vagus nerve project to the tracheobronchial tree.²⁹ The oesophagus is proportionally narrower and shorter in the infant (5–6 mm wide and 7–14 cm long in the infant compared with approximately 25 cm long in the adult). Infants consume frequent liquid feeds of approximately 120 mg/kg per day, which in the average adult would be almost 10 L per day. Their musculature is immature, causing them to spend much time recumbent and thus reducing the effects of gravity and delaying gastric emptying. They tend to ingest a larger meal than their gastric volume, causing them to vomit or spit up. At full term the gastric volume is about 30 mL.³⁰ Poor truncal tone causes infants to adopt a slumped sitting position, increasing intra-abdominal pressure and predisposing them to gastro-oesophageal reflux (GOR).³¹

Infants have an increased metabolic rate for oxygen consumption and therefore hypoxaemia can develop rapidly. The hypoxia response in infants is bradycardia (due to myocardial hypoxia and acidosis), whereas in adults the response is tachycardia and systemic vasodilation. The infant's respiratory rate is high, around 40 breaths per minute. Because of the immature respiratory system, infants are unable to respond by increasing their tidal volume and therefore increase their respiratory rate instead. This can be as high as 50–60 breaths per minute and explains why infants can rapidly develop respiratory fatigue and respiratory failure.²

Cyanosis and nasal flaring during inspiration are further indications of respiratory distress. Expiratory grunting ('auto-PEEP') is an attempt by the infant to maintain positive pressure on expiration to prevent alveolar collapse,

and involves exhaling against a partially closed glottis. Due to hyperinflation and the need for a higher inspiratory flow caused by the increased breathing rate, a higher pressure is generated drawing in the compliant chest wall. With moderate distress slight subcostal and intercostal retractions are seen. As distress increases the retractions become more widespread and obvious. As the infant forcefully contracts the diaphragm to move air, a paradoxical pattern of breathing may be seen.²

NEWBORN SCREENING AND THE RATIONALE FOR COMMENCING PHYSIOTHERAPY AT DIAGNOSIS

Over the past decade, newborn screening has become standard practice in many countries. As a result babies are being diagnosed with CF in the first 2 months of life, often before any recognizable respiratory symptoms appear. Prior to newborn screening the diagnosis of CF was made as a result of symptoms, which (if not due to meconium ileus) appeared at an older age (months to years) at a time of increasing maturity. This has raised the question as to when one should commence physiotherapy in a neonate diagnosed with CF, especially one who is apparently asymptomatic.³²

What do we mean by asymptomatic?

Due to lack of randomized controlled trials justifying the commencement of physiotherapy in neonates with CF, some physiotherapists believe that physiotherapy is unnecessary in the absence of overt respiratory symptoms. Lung symptoms are usually defined by a combination of clinical observation and assessment, and include: breathing rate, breathing pattern, sound of cry or stimulated cough, auscultation, chest radiograph and oximetry. An asymptomatic infant has a clear chest radiography, no abnormal breath sounds on auscultation, a clear-sounding cry or stimulated cough, a normal breathing pattern, a normal respiratory rate, and normal oximetry. A symptomatic infant is usually defined as having one or more of the following: changes on chest radiograph, abnormal sounds on auscultation, a moist-sounding cough or cry, an abnormal breathing pattern, an increased rate of breathing and possibly abnormal oximetry.

Early symptoms

However, when pediatric pulmonologists used these criteria to determine whether a patient was asymptomatic or not, the infants whom they judged to have normal respiratory status had diminished airway function present when full and partial forced expiratory manoeuvres were performed and they responded positively to bronchodilator and physiotherapy.^{33,34} Tepper states, 'In the absence of respiratory symptoms, the clinical exam and radiological evidence may underestimate the degree of lung disease'.³⁵

Table 1 Lung development after birth

Age	No. of alveoli	Air-tissue interface
Birth	20 million	2.8 m ²
8 years old	300 million	32 m ²
Adult	300 million	> 75 m ²

Table 2 Lung volumes in neonates compared with adults

Lung volumes	Neonates	Adults
Total lung capacity	160 mL	6 L
Vital capacity	120 mL	4–5 L
Tidal volume	16 mL (crying)	500 mL

This indicates that the present clinical tools used to assess the degree of respiratory involvement in infants with CF are inadequate in detecting underlying lung pathology. Tepper also reported, 'Assessment of respiratory status is often limited to clinical and radiological methods. Infant pulmonary function tests (PFT) have not yet been standardized and there is no normative data, only comparative between normal infants and infants with CF'.³⁵ Tepper concludes that it is not possible to assess a patient as being asymptomatic on the basis of the clinical examination alone.

High-resolution computed tomographic scans and bronchoalveolar lavage (BAL) offer more sensitive complementary information and give us a more accurate description of the respiratory state in infants with CF.³⁶ However, these techniques are invasive and expensive. Some CF centres now perform BAL routinely on all newly diagnosed CF infants to assess pulmonary status at the time of diagnosis. Khan *et al.* performed BAL in 16 infants with CF, mean age 6 months, and compared them with 11 healthy infants.³⁷ Their findings indicated that airway inflammation was present in infants with CF as young as 4 weeks of age. Infants presenting with airway infection had a more marked inflammatory response. However, an inflammatory response was also present in infants who were free from colonization with common CF pathogens. Despite being clinically asymptomatic, babies with CF frequently have been shown to have bacterial colonization and airway inflammation during the first weeks of life.³⁷⁻⁴¹ Even though the onset and intensity of lung disease is highly variable, most begin to develop a progressive pulmonary obstruction during early childhood.^{33,34,38,42-46} Armstrong *et al.* also reported inflammation and infection present in young infants less than 6 months of age with CF.⁴⁰ They concluded that airway inflammation follows respiratory infection.

Early commencement of treatment

The outcomes of newborn screening and early commencement of the CF care package have been compared with outcomes in a group of patients with a delayed diagnosis and treatment. Connett and Yeatman looked at the result of delayed diagnosis in 73 patients with homozygous DeltaF508 mutation,⁴⁶ comparing outcomes at 1, 3, 6 and 10 years. Treatment included physiotherapy. Patients with delayed diagnosis required increased treatment to maintain their health (intravenous antibiotics, 0 versus 19%; nebulized antibiotics, 57% versus 75%; inhaled steroids, 36% versus 85%). At 10 years the delayed diagnosis group had significantly more radiological changes.⁴⁷

Even though we do know that CF lung disease begins early in life and that mucociliary transport is impaired,

there are physiotherapists who question whether commencing physiotherapy at diagnosis will have any effect on slowing the progression of the disease. Many members of the multidisciplinary care team believe that physiotherapy should be commenced at diagnosis in order to attempt to compensate for the impaired mucociliary clearance, to prevent or at least retard the ensuing processes of inflammation instead of waiting until the process is already established and the treatment goals are targeted towards treating symptoms rather than the cause.^{6,34,48,49} However, in our objective to do no harm, we need to critically evaluate all aspects of airway clearance techniques currently used with infants and small children. It is the opinion of experienced physiotherapists working with infants and young children with CF in a clinical setting that laying the foundation of daily physiotherapy from an early age is essential. Infants accept physiotherapy as a part of their daily routine when this is introduced at a young age. Their parents develop frames of reference and sensitive observational skills to detect early changes and learn to respond and communicate with the team appropriately. Since the quality of the treatment that is carried out by parents is critical to the outcome, manual handling skills need to be developed and maintained. However, what the patient and parents are asked to do must be carefully considered, so that the burden that is placed on the family is not greater than the gain from treatment.

MODERN PHYSIOTHERAPY IN CF

In the past the primary aim of physiotherapy in CF was to clear excessive secretions and thus reduce symptoms. The effects of a single treatment session was measured in amounts of expectorated sputum, lung function, oxygen saturation or gas exchange, and pattern of breathing or level of dyspnoea. The term 'physiotherapy' is nowadays used in a much wider sense. Modern physiotherapy is an adequate combination of:

- inhalation therapy
- airway clearance therapy (ACT)
- physical education/exercise
- continuous education about the disease and its treatment.

The aim of modern ACT in CF is to *prevent* clogging of peripheral airways and consequent micro-atelectasis. Immediate effects of physiotherapy may not be measurable.⁴⁷ However, in the longer term, by compensating for the impaired mucociliary clearance the objective is to retard the development of lung disease as much as possible and to preserve lung function and physical capacity.⁴⁹ This is achieved by tailoring a time-efficient treatment programme that places the least possible burden on the patient/family

and makes compliance with the treatment possible. Since the disease cannot yet be cured, preventive daily treatment routines need to be established from the very beginning.⁴⁹ Patients (and families) need to learn to live with the disease as a reality in their everyday life. The treatment programme should be individualized to the needs, personality and lifestyle of the patient, the aim always being to allow them to lead as normal a life as possible. The time a patient/family is prepared to dedicate to treatment must be used as effectively as possible.

Inhalation therapy

Inhalation therapy is increasingly important in CF care, but many questions remain unanswered concerning this treatment, which differs between centres.^{50,51} The effect is dose-dependent in many drugs administered through inhalation, but little is known about the intrapulmonary dosage deposited and the deposition pattern in patients with an obstructive pulmonary disease.⁵¹⁻⁵³ Large individual differences are likely to appear since the intrapulmonary dosage deposited and the deposition pattern depend on several factors that can be influenced only to a certain extent.⁵⁰⁻⁵² The results of inhalation therapy are highly dependent on indication, the administration device, the inhalation technique and the ventilation distribution,^{51,53-56} on possible side-effects, treatment strategy and adherence with the treatment. Delivering aerosolized drugs to infants poses an even bigger challenge.⁵⁷ Often inhalation therapy is prescribed to be administered via a nebulizer system, even though that might not be the most efficient route and not the easiest mode of delivery when considering adherence. The use of inhalation therapy is individually tailored^{58,59} and the effects should be regularly evaluated. Caregivers require knowledge about a number of technical details and need to be up-dated in this developing field to be able to provide patients with an optimal result.⁵⁴ Factors to consider for optimal outcomes of inhalation therapy are:⁵⁵

Administration device (MDI+spacer, powder inhaler or nebulizer system)

For infants and toddlers the choice is between a metered dose inhaler (MDI)+spacer and a nebulizer system. A nebulizer system (i.e. the combination of a nebulizer and driving source) must have the capacity to produce an aerosol with a respirable quality that allows deposition throughout the airways even in infants, as well as being time-efficient (high drug output) to make adherence with the treatment likely. A high-capacity and reliable system is required to achieve a good aerosol quality and a high drug output at the same time. The device(s) chosen must be easy to use, to assemble/disassemble and to clean. Nebulizer systems need maintenance service regularly to be able to produce expected aerosol quality and output. The jet

nebulizer part of the system must be replaced regularly, according to recommendations from the supplier.

Agent(s)

Many different types of drugs can be administered through inhalation in CF care: bronchodilators, mucolytics, steroids, antibiotics, antifungals, antivirals and other agents. A worldwide survey carried out by Button and colleagues in 1999 of 166 centres in 27 countries showed a highly variable inhalation therapy practice in asymptomatic and symptomatic infants (Table 3).⁵⁹ The effects of bronchodilators can be multifactorial,^{60,61} but since it has been shown that bronchodilators can be negative in CF, effects should be followed up regularly.⁶²

Treatment strategy

The aerosolized drug is transported into the lungs by the inspired air. As discussed previously, a decreased ventilation distribution (obstructed airways, hyperinflation and atelectasis) and affected breathing pattern reduce lung deposition and affect deposition pattern. This may seriously alter the potential effects of many inhaled agents. Inhaled bronchodilators (if beneficial) and ACT should therefore precede inhaled agents targeting mucosa, remaining viscous secretions and micro-organisms.¹⁰ The inhalation therapy must be optimized for each drug by designing a treatment strategy (pre-, per- or post-ACT and pre- or post-physical exercise).

Mouthpiece/mask

Use of a mouthpiece or mask should be individually tried. A mouthpiece should be tried first, regardless of age. Some infants and toddlers can manage a mouthpiece; they may find a circular mouthpiece more suitable than an oval or flat one. The mouthpiece must be placed well into the mouth, on top of the tongue, to avoid deposition of aerosol in the oropharynx (Figure 2). If a mask is used it must be held as close to the face as the infant will tolerate.

Table 3 The use of inhalation therapy in 166 centres in 27 countries treating infants with CF (expressed as percentage of centres surveyed)

Inhaled agent	Asymptomatic		Symptomatic	
	Always/ frequently	Rarely/ never	Always/ frequently	Rarely/ never
Bronchodilators	27	50	74	4
Isotonic saline	27	59	38	48
Hypertonic saline	2	95	5	86
Pulmozyme	9	77	30	39
Anti-inflammatory	7	74	36	25
Antibiotics	16	70	49	21
Propylene glycol	0	99	2	95



Figure 2 Correct placement of an inhaler mouthpiece well into the mouth on top of the tongue to avoid oropharyngeal aerosol deposition

Inhalation technique

Patients and parents must be carefully instructed and trained regarding the inhalation technique, which must be regularly re-assessed and optimized regardless of choice of device.^{54,55} An optimal inhalation technique is dependent on each patient’s age, ability, willingness to learn and degree of lung disease.

Airway clearance therapy (ACT)

A Cochrane report 2000 states, ‘There is no evidence to support the efficacy of physiotherapy in the treatment of CF’.⁶³ This was due to the fact that no randomized controlled studies were eligible for inclusion in their report. The authors of the report did suggest, based on studies they did not include in their report, that physiotherapy consisting of ACTs could have a beneficial effect in patients with CF. One of these was a cross-over study in more mature patients which compared physiotherapy consisting of postural drainage and percussion (PD&P) with no physiotherapy (only directed coughing) over a 3-week period. When physiotherapy was discontinued for 3 weeks, forced expiratory volume in 1 second (FEV₁) declined significantly and when physiotherapy recommenced FEV₁ returned to baseline.⁶⁴ In another long-term 3-year prospective study in older children and adolescents, PD&P plus huffing was compared with huffing alone. The FEV₁ declined significantly in the huffing group compared with the PD&P group. It was concluded that PD&P combined with huffing was more effective in maintaining lung function over a 3-year period in patients with CF.⁶⁵

These two studies provide clinical evidence that physiotherapy helps to slow the progression of CF lung disease in children and adolescents.

Viscous hypersecretion problems occur in all generations of airways in CF. Due to pulmonary physiology it is not likely that the viscous mucus in the peripheral airways is influenced by coughing and huffing alone.^{64,65} Modern ACT, no matter what technique or combination of techniques is being used, must be built upon a physiological strategy or cycle consisting of different steps. The different steps can be expressed as (Figure 3):

- 1 open up and get air behind secretions
- 2 mobilize and collect secretions from the peripheral airways
- 3 transport secretions towards the central airways
- 4 evacuate secretions.

Physical education/exercise

Osteopenia/osteoporosis, fragility fractures and thoracic vertebral deformity have been reported in adult populations with CF.²¹ Nowadays most CF centres recommend physical activity to their patients. However, children and adolescents with CF, despite having good lung function, have been shown to be engaged in less vigorous spontaneous physical activity than their non-CF peers.⁶⁶ Whether this is due to less spare time caused by time-consuming therapy, to protective parents or healthcare system, or to the disease itself can be discussed. Simply recommending patients to be physically active is obviously not enough. They probably need more active guidance and continuing encouragement to become and remain physically active.

Physical training was gradually introduced as part of the CF care package in the late 1970s/early 1980s, and the collective experience from centres using it shows beneficial effects.^{67–72} Physical activity/exercise can be used as part of the airway clearance regimen, utilizing caused changes in

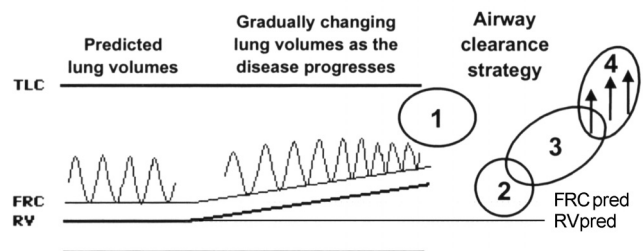


Figure 3 The four steps in the physiological strategic airway clearance cycle: 1, open up airways to get air behind secretions; 2, mobilize and collect secretions from the small airways; 3, transport secretions towards and through the bigger airways; 4, evacuate secretions from the central airways. TLC, total lung capacity; FRC, functional residual capacity; RV, residual volume

breathing pattern and in breathing volumes.^{70,73} The amiloride-sensitive sodium conductance in the respiratory epithelium is partially blocked during moderate-intensity exercise in CF,⁷⁴ which could increase water content of the mucus in the lung during exercise and explain the beneficial effects on airway clearance. Two studies have reported good results of physical exercise as part of the airway clearance regimen replacing the PD&P.^{67,70}

Children need physical activity to develop motor maturity and body awareness. If they are given the opportunity to experience pleasure and satisfaction during physical activity, much may be gained for future outcomes. Good chest mobility allows effective ACT. Good posture probably reduces the risk of back pain and spinal complications. Physical loading in upright positions stimulates bone accretion, reducing the risk of osteopenia/osteoporosis and the risk of spontaneous fractures including wedging and compression of vertebrae. If physical exercise aiming to preserve physical function is included in the treatment, patients can maintain good posture and chest mobility, even if lung disease should progress. They can also maintain a high working capacity in spite of poor respiratory capacity. Good posture contributes to positive body image and self-esteem.

The different types of exercise that should be included in a physical exercise programme from the very beginning are:

- chest mobility activities/exercises using movements around a vertical, sagittal and horizontal axis
- shoulder mobility exercises, especially elevation and external rotation
- muscle-strengthening activities/exercises, especially for postural muscles
- working capacity training/activities/exercises.

Exercises must never be uncomfortable. For infants, toddlers, children and adolescents, the activities/exercises must be stimulating, enjoyable and age-appropriate and should be individualized and provided at appropriate times in different settings. Team sports/activities provide the added benefits of normal social interaction, utilizing the benefits of all aspects of regular treatment.

Continuous education about the disease and its treatment

Education about the disease and its treatment starts at the time of diagnosis and is a never-ending process. Considering cross-infection factors, education can be organized as planned activities in groups or individually, depending on local conditions. These individual education sessions may be fitted into regular clinic appointments or treatment sessions

when a question is raised. Education should incorporate the manifestations of the disease, the progression of lung symptoms, the importance of preventive care and early identification of exacerbations as well as how best to retard the progression of the disease. Patients, parents, siblings, child care/school staff and the local hospital are educated and up-dated as needed. Patients/families may offer relatives and friends information provided by the CF team.

Adherence

No therapy is effective unless it is carried out regularly. Adherence with ACT is variable, but often reported as being low.^{32,75-77} Reasons reported are that patients forget to do it, do not feel any immediate effects, and the airway clearance technique they are asked to do is time-consuming, boring and a burden.^{76,78} Adherence with treatment requires full understanding of the indications and a cooperative approach between patient and physiotherapist. Cleaning teeth to prevent tooth decay is an appropriate analogy for understanding and motivation in children. The more satisfied the patient is with the treatment programme the better the adherence.⁷⁹ Greater optimism, hopefulness and optimistic coping skills are associated with increased adherence.⁸⁰ Different types of physiotherapy may suit different personalities at different times in life. Sometimes a treatment compromise that is adhered to should be considered as the best for the time being. Adherence with the physiotherapy may be improved if:

- alternative technique(s) are available
- a gradually developed treatment package is provided where the physiotherapist meets the patient/family in their current needs, motivation and capability
- patients are given time and assistance to process the information and organize their everyday life.

No-one has examined the effects on adherence of commencing physiotherapy at diagnosis as compared with commencing it when symptoms appear. It is often reported by clinical physiotherapists that when physiotherapy is commenced at diagnosis then incorporated into the family's daily life and made fun, acceptance of treatment and adherence are greater. There are no reported research studies to support this, but physiotherapists find that the most difficult period to try to teach the family to perform physiotherapy is with a 2- to 3-year-old child who does not understand why treatment has been introduced. In contrast when physiotherapy is commenced as a baby, the child accepts it much more readily as part of life, just like eating or cleaning teeth. It was found that parents whose child was diagnosed with CF prior to 3 months of age and treatment commenced at this age, had more adaptive coping styles

than parents whose child was diagnosed later and treatment commenced later.⁸¹

HISTORY OF POSTURAL DRAINAGE AND PERCUSSION

Postural drainage consists of placing the patient in a position that employs gravity to move mucus centrally from the targeted lung unit. There are 12 different postural drainage positions, one for each pulmonary segment. With the patient in a selected position, percussion is applied over the relevant chest for varying periods of time. In mature patients deep-breathing exercises, vibration during expiration and huffing are sometimes incorporated in the technique. In infants the technique is applied without the active participation of the infant. The first reference to the use of postural drainage was in 1901 by Ewart, who referred to it as 'empty bronchus treatment by posture in the bronchiectasis of children'. Ewart advocated continuous drainage for hours at a time with the patient sleeping in these positions.⁸² Traditionally head-down tilted positions of 20°–45° have been used (Figure 4). In 1934 the anatomy of the different lobes and segments of the lungs was described⁸³ and in 1949 the American Thoracic Society appointed an international committee to agree on the uniformity of the nomenclature of the broncho-pulmonary anatomy.⁸⁴

The exact mechanism by which percussion may assist in the removal of secretions is unknown. Mechanical percussion increases intra-thoracic pressure.⁸⁵ But no studies have been performed to examine the effects of manual percussion. It has been hypothesized that the air trapped between the cupped hand and the chest wall creates a vibratory wave that is transmitted through the chest wall and loosens secretions attached to the airway walls.

In healthy individuals ciliary action moves mucus up the airways at a rate of 3–5 mm/min and in the trachea at a rate of 20 mm/min.⁶⁰ In patients with CF, where the mucociliary action is impeded, mucus was found to move up the trachea at the rate of 3–5 mm/min,^{60,86} but approached normal rate when patients were placed in head-down postural drainage positions.⁸⁶ Following an extensive review of the literature it was concluded that the rate of movement of secretions in the small airways in CF during postural drainage is unknown. Theoretically, if the rate of movement in the small airways is similar to that in the trachea in CF (which is doubtful) then to be effective in moving secretions from the basal segments of the lungs to the larynx would require the patient to be placed in a head-down position for 60–100 min. A study of 42 ventilated patients showed an increase in total lung compliance following chest physiotherapy consisting of postural



Figure 4 A toddler during postural drainage and percussion (PD&P) in a 45° head-down position

drainage, percussion and vibration for a mean of 57 min.⁸⁷ It was suggested that chest physiotherapy needs to be of 1 h duration to be effective.

It has been speculated that the redistribution of ventilation, as occurs with a change in body position, might alter the local airway patency and gas/liquid pump.^{88,89} Consequently, it can be hypothesized that the physiological basis on which the concept of postural drainage was originally developed may not be the only mechanism for the improvement seen with changes in position as used in postural drainage positions. This hypothesis is partly supported in a study using clearance of inhaled radioactive aerosol, where more secretions were cleared from the dependent lung rather than from the uppermost lung during postural drainage in children, adolescents and adults with CF.⁹⁰

SIDE-EFFECTS ASSOCIATED WITH POSTURAL DRAINAGE

Side-effects that have been observed and objectively measured by physiotherapists during treatments incorporating postural drainage include desaturation, discomfort/pain and GOR.^{91–98} GOR is an abnormal increased tendency to regurgitate gastric contents into the oesophagus.⁹⁹ The respiratory consequences of GOR may include aspiration of acidic gas, micro-aspiration or macro-aspiration of stomach contents. Oesophageal irritation may lead to vagal nerve stimulation and reflex broncho-spasm/wheeze.¹⁰⁰ Because infants are nose-breathers it can be difficult to manipulate refluxate to a safe place.²⁵ The following behaviours are sometimes associated with GOR in infants: burping, crying, drooling, irritability, frowning, yawning, stretching, mouthing, stridor and in extreme cases Sandifer's sign, a neurobehavioural manifestation of GOR.¹⁰⁰

Gastro-oesophageal reflux in CF

Feigelson and colleagues first described GOR in CF in 1975.¹⁰¹ Vinocur *et al.* in a further study concluded that GOR and its complications can significantly alter the pulmonary courses of some children with CF. GOR should be managed as aggressively in CF as it is in any child with reflux.¹⁰² Scott and colleagues found a higher incidence of GOR in CF children than their siblings and Stringer and co-workers found significantly lower lung function in children with CF and GOR compared with those without GOR.^{103,104} GOR is common in healthy infants under 1 year, with a prevalence of 18–40%.^{100,105} The prevalence of GOR in infants and young children with CF varies between 35% and 81%.^{97,102,106,107} As infants mature and consume a more solid diet and spend more time upright, around 80% of normal infants outgrow GOR at between 18 and 24 months.¹⁰⁰ There is a study in progress showing trends towards a higher reflux index and longer reflux episodes in infants who are *P. aeruginosa*-positive than those who are not colonized.¹⁰⁸ This raises the question of whether GOR¹⁵ is related to early colonization with *P. aeruginosa*.

GOR and postural drainage

Foster and colleagues documented increased GOR during postural drainage in children and adolescents with CF in the early 1980s.⁹¹ Vandenplas *et al.* found that postural drainage provoked increased GOR in a large cohort of infants without CF.⁹⁶ Vandenplas and colleagues noted in their study that ‘There was no evidence for a correlation in time between provoked coughing during physiotherapy and GOR. A reflux episode occurring simultaneously with coughing provoked by physiotherapy was only recorded once’.⁹⁶ Button and colleagues studied the effects of two different regimens on GOR in 20 infants with a mean age of 2 months diagnosed with newborn screening.⁹⁷ They compared standard postural drainage using four common positions (including 30° head-down tilt) with modified postural drainage (avoiding head-down tilt) (Figure 5). There were significantly more episodes of GOR associated

with postural drainage than with modified postural drainage.⁹⁷ Further statistical analysis of data collected during this study showed that during postural drainage there was a significant association between crying and reflux episodes. Oxygen saturation was significantly lower during crying. They concluded that postural drainage was associated with GOR, distressed behaviour and lower oxygen saturation. Therefore postural drainage should not be routinely prescribed in (apparently) asymptomatic infants.¹⁰⁹

Phillips and colleagues had contradictory findings in 21 infants ranging from 1 to 27 months with respiratory disorders including 11 subjects with CF. They avoided the prone head-down position (one of the most reflux-provoking positions in the other studies). Further, they used head-down tilted angles of 15°–20° compared with 30° plus used in the other studies, which together with an older cohort of infants may have accounted for the different results.^{106,110}

AIRWAY CLEARANCE TECHNIQUES CURRENTLY USED IN INFANTS WITH CF

A survey to establish how physiotherapists manage infants with CF was undertaken in 1998–1999 (Table 4).⁵⁹ This is a European Respiratory Society taskforce project endorsed by the International Physiotherapy Group for CF. The airway clearance techniques used have their origins in the techniques developed for use with older patients with chronic sputum and have been adapted for use in infants (Table 4). Many of these techniques have no evidence base for use in infants.

Postural drainage (PD)

In the infant survey referred to above 55% of the 166 centres replied that they frequently or always carried out postural drainage (Table 4). The use of a passive form of therapy which includes postural drainage and manual techniques such as chest percussion is standard practice in infants and small children.⁷⁶ The authors of this publication are currently questioning this approach (personal communication, North American and European CF Conferences,

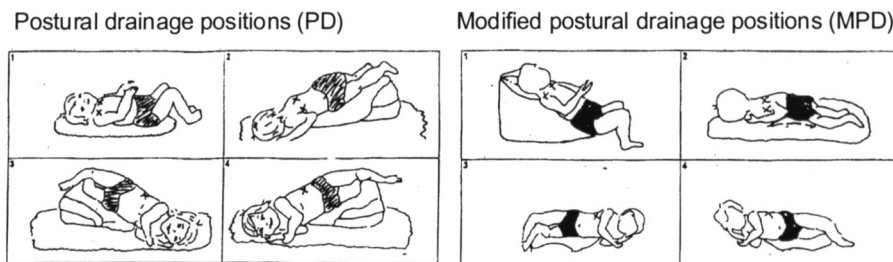


Figure 5 The postural drainage (PD) positions changed to modified postural drainage (MPD) positions to avoid gastro-oesophageal reflux (GOR) in infants (Ref 113)

Table 4 The use of airway clearance therapy in 166 centres in 27 countries treating infants with CF (expressed as percentage of centres surveyed)

Airway clearance technique	Always/frequently	Occasionally	Rarely/never
PD	55	14	31
Modified PD	47	26	27
Clapping/percussion	35	12	53
Manual vibration	52	18	30
Mechanical vibration	9	12	79
Positioning to ↑ ventilation	58	18	24
Passive/assisted AD	19	12	69
French technique	17	9	74
German technique	15	7	78
Swedish technique	19	8	73
PEP	14	5	81
Cough stimulation	31	28	41
Tracheal pressure	13	20	67
Oropharyngeal stimulation	4	15	81
Oropharyngeal suction	7	20	73
Nasopharyngeal suction	9	25	66

PD, postural drainage; AD, autogenic drainage; PEP, positive expiratory pressure

2003). The recommended angles of head-down tilt vary in different texts. Some recommend 15°–20°¹⁷ while others propose 30°–45°.¹¹¹ This kind of airway clearance therapy places a great burden on the patient/family, and adherence is known to be poor.^{32,75,76}

Modified postural drainage (MPD)

In the infant survey referred to above, 47% of the 166 centres replied that they frequently or always used modified postural drainage (Table 4). The randomized controlled study in newly diagnosed infants discussed previously⁹⁷ continued for a further 5 years. The effects of postural drainage incorporating 30° head-down tilt with modified postural drainage (avoiding head-down tilt during infancy) were compared in the longer term (Figure 5). Blowing games, physical activities and ‘bubble PEP (positive expiratory pressure)’, using a bottle with a column of water and wide-bore tubing to provide the PEP,¹¹² were introduced as an adjunct or alternative as soon as infants were mature enough to actively participate. This more active and fun regimen was preferred by many families. At a mean age of 5.5 years the infants randomized to modified postural drainage at diagnosis had significantly better FEV₁ and forced vital capacity (FVC) and fewer radiological changes than those in the postural drainage group.¹¹³ These findings were recently discussed in an editorial, where it was recommended that modified postural drainage be used instead of postural drainage in asymptomatic infants (Figure 5).¹¹⁴

In Canada, modified postural drainage is commenced at diagnosis. Once the child is physically able, blowing games, bubbles, trampoline jumping, swimming and other physical activities together with active coughing and the forced expiration technique are introduced. This approach has been found to be acceptable to parents and young children. Using this approach, adherence to treatment in the 0- to 5-year-old group has been found to be 92% (McIlwaine, 2003, personal communication).

Positive expiratory pressure (PEP)

Airway clearance therapy using positive expiratory pressure in combination with the forced expiration technique (FET) in upright sitting in children and young adults was shown to be superior to postural drainage and percussion in terms of sputum clearance.⁹² The PEP technique has been shown to result in improved ventilation and reduced volume of trapped gas.¹¹⁵ While breathing towards an expiratory resistance, clogged and collapsed airways are opened up with the assistance of collateral ventilation, in order to get air behind secretions.⁹² In Denmark, where this technique was developed, it has been adapted to infants and used since the early 1980s.

The technique employs a face mask with two one-way valves to which a variable resistor is attached to the expiratory valve. A manometer can be inserted in the circuit to measure the expiratory pressure achieved. The diameter of the resistor used for treatment is determined for each individual patient to give a steady PEP of 10–20 cmH₂O during the middle part of expiration. This pressure should be maintained during tidal volume breathing with only slightly active expirations. Treatment is carried out in the upright sitting position; babies are carried on the arm during treatment (Figure 6). A treatment session consists of periods of breathing with PEP in order to get air behind and mobilize secretions, followed by the forced expiration technique or cough in order to transport and evacuate the mobilized secretions.⁷³

In a 1-year randomized control trial that compared postural drainage and percussion with PEP in 26 newborns, PEP was found to be equally effective to postural drainage.¹¹⁶ Five infants demonstrated mild GOR in the PEP group, while four had severe GOR in the postural drainage group, from which three were withdrawn from the study because of severe GOR. PEP was concluded to be safe, effective and unanimously preferred by parents and infants. A 1-year randomized study comparing postural drainage and percussion with PEP in children and adolescents showed an improvement in lung function in the PEP group, while the postural drainage and percussion group deteriorated. The differences in lung function were statistically significant.¹¹⁷



Figure 6 PEP in an infant

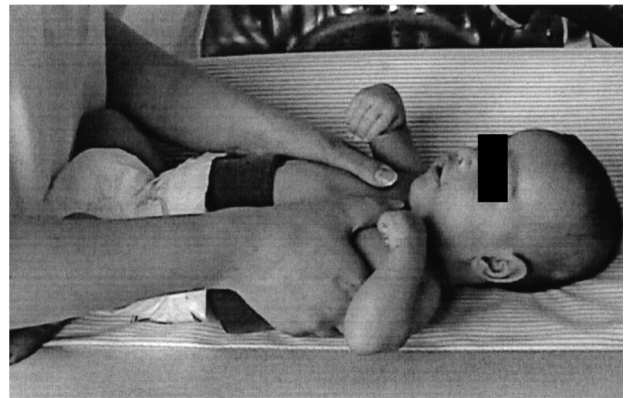
In the infant survey referred to previously, 14% of the 166 centres replied that they frequently or always used PEP for airway clearance while 5% occasionally used this technique with infants (Table 4). At different centres throughout the world, several different ways of achieving a positive expiratory pressure have been developed, as well as other physiological targets of using an expiratory resistance.⁷³

Assisted autogenic drainage (AAD)

Autogenic drainage (AD) was developed by Jean Chevallier in Belgium in the 1970s. Autogenic drainage (self-drainage) is a breathing routine utilizing good body knowledge and expiratory airflow throughout the whole range of breathing from residual volume to total lung capacity. The aim of AD is to achieve an optimal expiratory flow progressively through all generations of bronchi without causing dynamic airway collapse.¹¹⁸ Assisted autogenic drainage (AAD) is the adaptation of autogenic drainage in infants and young children not yet capable of carrying out this technique actively themselves. By placing the hands on the child's chest the therapist manually increases the expiratory flow velocity and prolongs expiration towards residual volume during an individualized number of manoeuvres (Figure 7). AAD is carried out in a gentle way following the child's breathing pattern and stabilizing the infant's abdominal wall. Excessive force is avoided and the number of AAD manoeuvres is limited to the child's response and tolerance. The response of the child to excessive force is protection by resisting the manoeuvres. This includes trying to close the glottis and eliciting a breathhold or at least by activating the inspiratory muscles during expiratory manoeuvres. Passive or assisted autogenic drainage is always or frequently used by 19% and occasionally used by 12% of the 166 surveyed centres in the care of infants (Table 4).

Van Ginderdeuren and colleagues have evaluated AAD to establish whether this form of therapy provokes GOR in infants. No provocation of GOR has been associated with

(a)



(b)

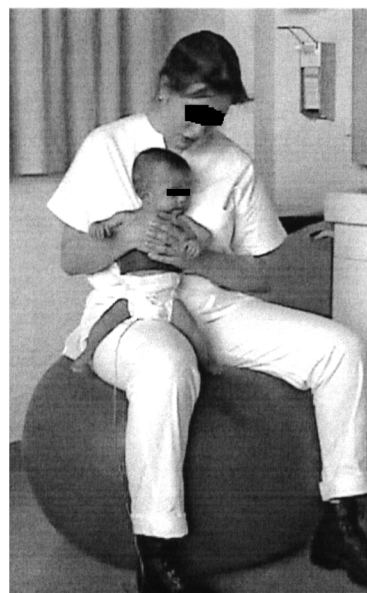


Figure 7 (a) Assisted autogenic drainage (AAD) in an infant (Refs 119, 120) (Photo: van Ginderdeuren) (b) An infant in well-supported upright sitting during assisted autogenic drainage (AAD) (Refs 118, 120) (Photo: van Ginderdeuren)

this technique. The infant is well supported in upright sitting, avoiding a slumped sitting position which may in turn predispose to GOR during treatment (Figure 7b).^{119,120} Sometimes AAD is combined with bouncing (a rhythmic up-and-down movement on a physio ball) to relax the baby (Figure 7b).¹²⁰

Other techniques

A number of alternative techniques are used worldwide (Table 4). Only a few of these have been evaluated for safety or efficacy. A French study analysed the tendency to GOR in different manipulations used during pulmonary physical therapy consisting of nasal suction, oropharyngeal suction or acceleration of expiratory flow in the supine horizontal and supine 35° head-up positions. They found that oropharyngeal suction and treatment in the supine

horizontal position provoked more GOR than nasal suction or treatment in the 35° head-up position. This has resulted in avoidance of oropharyngeal suction and treatment in the supine horizontal position.¹²¹

Physiotherapy for infants and young children with CF as practised in many regions requires them to assume recumbent positions while manual techniques are applied. Recognizing that these activities can be tedious for the children and their parents a study was undertaken to evaluate the benefits of music therapy used during physiotherapy in infants and toddlers with CF. The results of the study indicate that the young children's, and their parent's, enjoyment of physiotherapy significantly increased after the introduction of specifically composed and recorded music as part of their daily physiotherapy routine. It was therefore recommended that parents use the prescribed music in order to assist the establishment of a positive routine.¹²²

The treatment philosophy practised at the Lund CF centre

At the Lund CF centre in Sweden physiotherapy is introduced from the day of diagnosis. The aim is to develop an effective daily routine for each patient that maintains good physical function and prevents lung disease and destruction of lung tissue for as long as possible. Based upon individual needs and results achieved, adequate content and dosage of modern physiotherapy is tailored in cooperation with each patient/family. The cooperation between physiotherapist and patient/family is lifelong and must be founded upon trust, where patients are working partners who actively participate in the discussion and treatment decisions made. The different options are presented to the patient together with the expected outcomes. After adequate discussion the patient is involved in the choice of the treatment. The patient agrees to inform the caregiver if the treatment does not seem to be effective, so that an alternative solution can be found. The atmosphere around the disease and its treatment is kept positive.

Airway clearance therapy based upon physical activity

A long-term study carried out between 1981 and 1983 showed that the effects of physical activity/exercise combined with FET was equally as good as postural drainage with percussion and FET combined with physical activity/exercise.⁷⁰ Since 1983 no patients have used postural drainage or percussion in any form, regardless of age. Infants, toddlers, children, adolescents and adults with mild CF lung disease use an airway clearance regimen based upon physical activity/exercise/training.¹²³ The respiratory aim with the physical activity in the infants is to influence

the breathing pattern and ventilation distribution, and as soon as the child can participate actively, to increase minute ventilation. Theoretically, the altered breathing pattern and ventilation distribution can enhance mobilization of secretions and counteract microatelectasis. For infants and toddlers, the choice of exercise activities is dependent on individual motor maturity. Parents are carefully taught about the types of exercise to use, the rationale for their use and how to combine them. A choice of treatment programmes is provided by the physiotherapist. Choosing which programme to use for each session gives the child (or sibling) a feeling of empowerment and control. Rather than discussing whether to do the treatment or not, the child makes a choice between alternatives provided. For the infants, toddlers and young children the physical activity is interspersed with assisted autogenic drainage manoeuvres (AADs).¹²³ The aims with the AADs are to prolong the expirations towards residual volume and increase the expiratory flow velocity, in order to enhance mucus transport towards the larger airways. AADs are carried out with the child in upright supported sitting and bilateral horizontal side-lying on the lap, gently squeezing the chest following the child's breathing pattern. If possible, three to five AADs are carried out in a sequence. Parents are carefully taught how to perform AADs; their technique is frequently assessed. The AADs must never be uncomfortable or frightening for the child, since that will force them to defend themselves. The defence, contracting the inspiratory muscles, counteracts the aims of the AADs. Physiotherapy is always carried out in a way that makes future cooperation possible.

The physical activity part can be carried out to rhythmic music, which is interrupted for AADs. This choice of physiotherapy regimen is active and fun. Laughter influences the breathing pattern in a similar way to that described above and is incorporated in the treatment. This technique

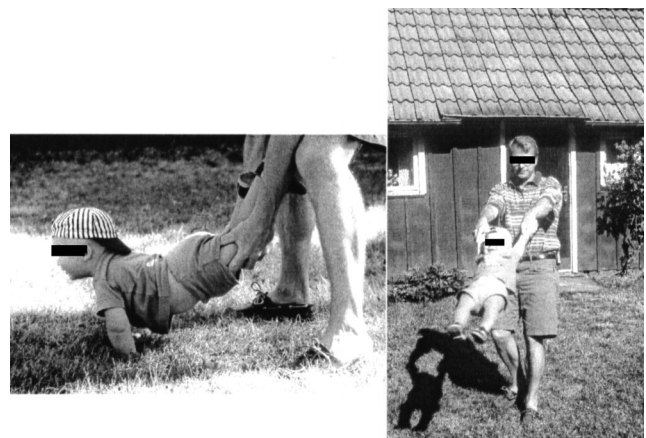


Figure 8 With maturation toddlers actively participate in fun physical activity/games that promote altered breathing patterns, mobility and muscle strength (Ref 124)

not only focuses on airway clearance, but it involves musculo-skeletal care from the beginning, which aims at maintaining chest mobility and the development of good posture (Figure 8).¹²³ Siblings often participate in the treatment, which is a positive psycho-social side-benefit and an opportunity to build positive relations between child, siblings and parents in an enjoyable and positive treatment atmosphere. Patients develop an optimal physical capacity which often makes them excel at gymnastics at school, giving them an unshakeable self-esteem that is positive for the future.

Other airway clearance techniques used

The physical activity/exercise/training is interspersed with the FET, as soon as the child masters the technique properly.¹²³ As patients get older or if adherence with the therapy decreases and/or pulmonary symptoms increase, the most commonly used airway clearance techniques are PEP, autogenic drainage, Hi-PEP, oscillating PEP and controlled cough.¹²³ An individually tailored regimen which often comprises a combination of techniques is developed for each patient, all in accordance with the physiological airway clearance strategy (Figure 3). The ACT can be carried out in sitting only or in a combination of sitting and bilateral horizontal side-lying. Head-down tilted positions have not been used since 1983. Physical exercise is always included, either as part of the airway clearance regimen or as an adjunct.^{73,123}

Results

Lung function and peak working capacity data collected retrospectively between 1990 and 2002 for all 38 7-year-old CF patients in the south of Sweden are summarized in Table 5.¹²⁴ Sweden does not have a newborn screening programme for CF. All 38 patients have attended Lund CF centre from diagnosis; the median age at diagnosis was 3 months (range 1–60 months). Of all 38, 36 (95%) have a DeltaF508 mutation of whom 22 were homozygous; 90% were pancreatic-insufficient. In this 12-year retrospective review, data were collected from each 7-year-old’s extended annual review. Nine individuals (23%) were

chronically colonized with *P. aeruginosa* and one with *Burkholderia cepacia*. All parts of CF care at the Lund CF centre are in accordance with international guidelines, but the physiotherapy part (as described above) is emphasized. None of the patients have ever used postural drainage, percussion and vibration. Their summarized lung function and peak working capacity demonstrate that this treatment philosophy is successful in terms of long-term outcomes (Table 5).

ROLE OF THE PHYSIOTHERAPIST

The factors considered when choosing an airway clearance technique or a combination of techniques are age, reactivity of airways, stability of airways, whether atelectasis is present, degree of lung tissue damage, cysts, bronchiectasis, ability to learn, personality, surroundings, achieved treatment quality, adherence with the airway clearance programme chosen and long-term results (lung function and number of exacerbations). While teaching and trying different techniques or combinations, the immediate results such as breathing sounds, wheezing, sounds of mucus moving, type of cough, cough pattern, cough sounds and muscular fatigue are the most important tools used. Attention to the quality of the treatment is emphasized and is continuously being optimized. Adherence is frequently assessed and discussed with the patient and the regimen is modified as required.

Patients have regular and frequent contact with the physiotherapist, who sees the patients for a treatment session at all 6-weeks to 3-monthly outpatient clinics visits and in between if needed, during admission to hospital and intermittently during home visits if needed. Each patient sees the same physiotherapist, building up an effective working relationship. During treatment sessions in hospital the same devices (for inhalation and ACT) are used as the ones patients use at home. The most efficient inhalation administration devices are chosen and the most efficient nebulizer system, if nebulization therapy is required. Spirometry is frequently carried out by the physiotherapist. During spirometry the measurements of volumes and flows together with observations of breathing sounds, sounds of mucus moving, amount and type of coughing, degree of breathlessness generated, time needed for rest in between manoeuvres and ability to handle dyspnoea are all essential tools for assessing results of the therapy, for motivating patients to adhere to current treatment strategies and for developing future treatment rationales.

FUTURE PHYSIOTHERAPY IN CF

Inhalation therapy

Antimicrobial agents for inhalation will continue to develop and new chemicals will be introduced.⁵¹ Inhaled gene

Table 5 Lung function and peak working capacity in the 7-year-olds who have been exposed to the Lund CF centre treatment philosophy since the day of diagnosis

	Median %p (Mean)	Range
Vital capacity (VC)	100 (98)	70–129
Forced expiratory volume in 1 min (FEV ₁)	100 (98)	64–135
Total lung capacity (TLC)	101 (99)	71–134
Residual volume (RV)	103 (112)	50–200
Functional residual capacity (FRC)	100 (99)	64–144
Peak working capacity	110 (118)	77–228

therapy will eventually be available, even though it will take longer to develop than was predicted. If the target is the respiratory epithelium, the therapy needs to be repeated regularly since the effect will be time-limited to the treated cells which are replaced by a new cell generation. Depending on the pharmacological dynamics of the agent and of the function of the new untreated cell generation, pulmonary symptoms may appear between administration occasions and may need treatment. Poor distribution of the inhaled drug occurs in patients with severely affected pulmonary disease (Figure 9), which probably makes inhalation therapy more beneficial to those with lung function as normal as possible.¹²⁵ The results of the therapy are highly dependent on the administration device, the inhalation technique and the ventilation distribution.^{51,54,55} An administration device is required that can deliver a precise dosage without loss into the environment, an optimal aerosol quality and a high drug output simultaneously.⁵¹

Airway clearance therapy

Compensating for impaired mucociliary clearance will still be a requirement in the future. With increasing knowledge about the pathophysiology in CF and the physiology of the different techniques utilized, continuing improvements can be anticipated by tailoring treatment regimens to individual patients. New airway clearance techniques will be developed while currently available techniques will continue to improve as our understanding of the use of these techniques develops through further research studies.

Physical education/exercise

Physical exercise is part of the care package for all patients, in order to maintain physical function and to develop good body awareness. Patients/parents are also taught how to recognize and respond to changes in cardiopulmonary status which tend to be highlighted during physical effort.

Lung transplantation

Lung transplantation is the last treatment option for severely ill patients with progressive lung disease. Without increased organ donation and an improvement in long-term survival,¹²⁵ a careful selection of patients is necessary. To what extent the decreased lung function is due to clogged airways, partly reversible with an optimized physiotherapy regimen, or to irreversible lung tissue damage must be carefully analysed before accepting patients onto the transplant list.

Scientific studies

Infants and small children must be protected from unnecessary or harmful interventions and offered the best

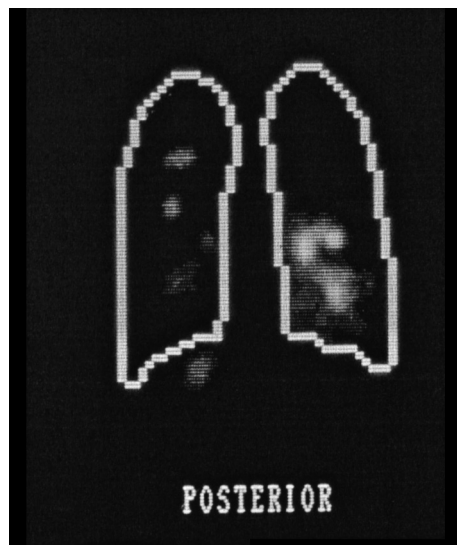


Figure 9 Severely affected ventilation distribution results in a poor distribution of inhaled drug

opportunity for optimizing lung function and long-term quality of life and survival. Therefore studies evaluating the safety and efficacy of airway clearance techniques used with infants and small children need to be undertaken. The study protocol, measurement parameters used⁴⁹ and grading of studies should consider the aims of treatment and the population studied. Short-term studies (weeks to months) are useful in evaluating the effects of therapy on acute exacerbations or if patients have been sub-optimally treated. In such studies an increase in lung function and a decrease in symptoms and expectorated sputum (in older patients) are expected outcomes.⁴⁹ In long-term studies (12 months or more) where treatment has been deemed to be optimal, the expected outcomes are stable lung function and physical status or a decrease in the rate of decline of lung function expected in a chronic life-limiting disease,⁴⁹ together with good quality of life. Modern physiotherapy in CF consists of continuing optimization of multidisciplinary treatment incorporating new therapies and individual needs. There are ethical issues related to studies where a group of patients may be randomized to a potentially harmful long-term fixed protocol which may result in irreversible lung damage.

It has been suggested that physiotherapists carry out double-masked randomized control trials.¹²⁶ Masking of airway clearance techniques in CF in studies is by their nature impossible, as are placebo-controlled trials.^{32,49,77} The value of case series or $n=1$ studies should not be underestimated as these types of study often result in new observations which lead to new therapies or improved understanding of current therapies.

The manifestation of CF lung disease is highly variable and there are many other factors to consider when treating patients with CF. Some of these factors include physical development, age, maturity, family situations and dynamics, personality, interests, culture, aspirations and surroundings. Instead of aiming to find one technique that will suit all patients of all age groups (which is unlikely), we should rather aim at identifying physiological and psychological factors that make one technique or regimen of combined techniques more effective than another. Therefore subgroup analysis and trends in different individuals in randomized controlled trials and cross-sectional studies should be analysed and given a higher recognition than is currently the case.

CONCLUSION

The onset and intensity of the progressive lung tissue destruction in CF is highly variable. But inflammation and infection is present before 4 weeks of age in apparently asymptomatic infants. The chronic infection and the inflammatory response is the reason for lung tissue damage, and since the destruction of lung tissue is not reversible, the most logical objective must be pro-active treatment which attempts to prevent or at least slow the progression of lung disease. We do not yet have the knowledge to identify individual risk factors in infants and young children who need more intense treatment from the start. Because of this, a daily treatment should be established from the very beginning at the time of diagnosis. Patients and families need to learn to live with the disease and incorporate it in their daily routine, since CF is a reality in the family. But, when we ask parents of a newly diagnosed CF patient to initiate any form of treatment, we must recognize that we are placing the responsibility on the family. Therefore, our aim as physiotherapists managing young children with CF should be: (1) to define and teach a time-efficient treatment programme that aims to retard lung disease, (2) to preserve physical function. The treatment regimen prescribed should be one that puts the least physical and psycho-social burden on patients/families and that promotes adherence. Optimal treatment is not synonymous with maximal treatment.

Many new airway clearance techniques have been developed. Several studies comparing the different techniques have been carried out, showing only slightly different effects. Many authors conclude that no single airway clearance technique is the best for all patients and that treatment regimens therefore should be individualized. It is the responsibility of physiotherapists to understand the physiology and learn the craft of the different techniques so that patients can be correctly instructed and the techniques used optimally. The physiological aims of parts of the techniques can be achieved in the youngest patients if they

are adapted to the young child's immature pulmonary anatomy and physiology, motor maturity and inability to follow instructions. Furthermore, treatment should as far as possible be stimulating and incorporate siblings in activities that are therapeutic at the same time as being fun. Physiotherapy can be performed enjoyably and safely and should fit into the family's lifestyle. By introducing tailored music to the physiotherapy regimen enjoyment can be increased and adherence promoted even further.

Modern physiotherapy is a much wider concept than just airway clearance. All the following should be considered when developing a physiotherapy programme: inhalation therapy, airway clearance therapy, physical fitness and function, body awareness and posture. In developing the individual regimen, with the patient/family as an active participant, the following factors will influence the elements that form the programme: age, motor maturity, degree and components of lung disease, ability to learn, achieved treatment quality, personality, likes/dislikes, adherence to treatment, long-term results, understanding of the disease and its treatment, personal aspirations of the patient/family, socio-economic considerations, culture and more. Infants and young children should be treated as unique immature individuals and not as little adults. Frequent follow-up, review of agreements and continuing optimization of the programme are essential for an optimally effective and time-efficient daily treatment regimen aimed at preventing or at least slowing the progression of lung disease.

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