CLINICAL GUIDELINES FOR THE PHYSIOTHERAPY MANAGEMENT OF CYSTIC FIBROSIS

CYSTIC FIBROSIS TRUST

Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF)

Recommendations of a Working Group

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CLINICAL GUIDELINES FOR THE PHYSIOTHERAPY MANAGEMENT OF CYSTIC FIBROSIS

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1. FOREWORD

1.1 Guidelines development

The development of these clinical guidelines has taken place as the move towards evidence based practice. The core group responsible for development of the guidelines consisted of chartered physiotherapists, both clinical practitioners and academics all of whom were members of the Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF). The group was later joined by other advisors and specialists including people with cystic fibrosis (CF). These guidelines have been developed independently of any funding bodies.

1.2 How to use these guidelines

Clinical Guidelines for the Physiotherapy Management of Cystic Fibrosis aims to be a useful tool and reference document for all physiotherapists and others involved in the delivery of care to people diagnosed with cystic fibrosis from birth and throughout life. They are based on scientific evidence but where this is not available a best practice consensus has been outlined. The endorsement process of the guidelines by the CF Trust has included review by relevant experts as well as peer review. As indicated within the text, the interpretation of scientific evidence and guidelines should be taken in context with a holistic approach to management and patient acceptability.

The recommendations are intended to encourage physiotherapists to develop local guidelines tailored to their specific needs and circumstances.

1.3 Review of the guidelines

The document will be reviewed in 2005 by the ACPCF and updated according to any new evidence available and/or changes in practice.

1.4 Grading scheme for recommendations in the guidelines

The recommendations in this document are graded based on the Scottish Intercollegiate Guidelines Network *Clinical Guidelines: Criteria for Appraisal for National Use** and the Agency for Health Care Policy and Research**.

Levels of evidence

Level	*Type of evidence based on AHCPR 1992*
Ia	Evidence obtained from meta-analysis of randomised controlled trials.
Ib	Evidence obtained from at least one randomised controlled trial.
IIa	Evidence obtained from at least one well designed controlled study without randomisation.
IIb	Evidence obtained from at least one other type of well designed quasi-experimental study.
III	Evidence obtained from well designed non-experimental descriptive studies, such as comparative studies, correlation studies and case control studies.
IV	Evidence obtained from expert committee reports or opinions and/or clinical experience of respected authorities.

Grading of recommendations

Grade	*Type of recommendations based on AHCPR 1992*
A (levels Ia, Ib)	Requires at least one randomised controlled trial as part of the body of literature of overall good quality and consistency addressing the specific recommendation.
B (levels IIa, IIb, III)	Requires availability of well conducted clinical studies but no randomised clinical trials on the topic of recommendation.
C (level IV)	Requires evidence from expert committee reports or opinion and/or clinical experience of respected authorities. Indicates absence of directly applicable studies of good quality.

*Petrie GJ, Barnwell E, Grimshaw J, on and behalf of the Scottish Intercollegiate Guidelines Network. *Clinical guidelines: criteria for appraisal for national use*. Edinburgh, Royal College of Physicians, 1995.

**Agency for Health Care Policy and Research. *Acute pain management, operative or medical procedures and trauma* 92-0032, Maryland, USA, Agency for Health Care Policy and Research Publications, 1992.

2. PROVISION OF CARE

The Cystic Fibrosis Trust, British Paediatric Association (now the Royal College of Paediatrics and Child Health) and British Thoracic Society have published detailed guidelines relating to provision of care in 1996 which have been extensively revised in 2001[1] (available through the CF Trust). All patients should have access to a Specialist CF Centre and be reviewed at least annually. Evidence suggests that involvement of a Specialist CF Centre results in improved survival, clinical status and greater satisfaction with care[1-6]. Any service provision should acknowledge the implications of all health and safety issues, including cross-infection.

2.1 Provision of physiotherapy services

All Specialist CF Centres and CF Clinics should aim to provide the following services:

- Qualified physiotherapy staff to provide treatment as necessary throughout a 24-hour period[3].
- Protected time for outpatient clinics to ensure that all patients have the opportunity to see a physiotherapist.
- Provision of a comprehensive Annual Review.
- Provision of, or access to community physiotherapy for advice and support when required.
- Administrative support to ensure clinical time is fully utilised.
- Physiotherapy managers should provide opportunity for continuing professional development including performance review, education and training.

In addition Specialist CF Centres should aim to provide:

- Physiotherapy staffing levels in a ratio of approximately two whole time equivalent physiotherapists per 50 patients, depending on the role of the physiotherapist within the CF multidisciplinary team, the disease severity of the caseload and the requirement for community support. These physiotherapists should have specific expertise in the care of patients with CF, and have relevant adult and/or paediatric experience depending on their patient caseload[1].
- Physiotherapists with knowledge of and working to the published *Standards for respiratory care* of the Association of Chartered Physiotherapists in Repiratory Care (ACPRC)[7].
- An open access service.
- Acknowledgement of any shared care or local services already available to the patient.
- A shared care service to CF Clinics.
- Exercise and exercise testing facilities.
- Safe and appropriate equipment for patient and therapist use.
- An age appropriate service ensuring smooth transition from paediatric to adult care.
- Resources for education and training e.g. lectures, courses, seminars.
- Opportunity for clinical research and evaluation of new techniques and equipment.

2.2 Guidelines for commissioning and contracting

- Contracts should outline provision of services offered as above.
- Responsibility for provision and maintenance of equipment should be identified.

3. ASSESSMENT

Assessment is a continuous process, which identifies individual problems and needs of the patient, providing information essential for effective clinical reasoning and formulation of treatment plans. It enables the monitoring of clinical management and evaluation of treatment intervention. It is the process that forms the basis of professional autonomy and competence and should include assessment of respiratory status and functional ability.

3.1 Assessment of outcomes

Measurement tools to monitor disease progression and treatment intervention include:

- lung function tests
- oximetry
- blood gas analysis
- sputum or cough swab culture
- exercise capacity
- qualitative assessment of aspects such as breathlessness and pain, using subjective tools
- auscultation
- radiographic imaging
- quality of wellbeing measures
- body weight and Body Mass Index (BMI)
- number of exacerbations.

Recommendations

- *On confirmation of diagnosis the individual should be assessed by an experienced CF physiotherapist in order to formulate an effective management plan (Grade C, Level IV).*
- *Patients should be reviewed at regular intervals and treatment regimens adapted according to their changing needs (Grade C, Level IV).*

4. AIRWAY CLEARANCE TECHNIQUES

Airway clearance techniques (ACT) enhance the clearance of excess bronchial secretions. By removing obstructive secretions they aim, in the short-term, to reduce airway obstruction, airway resistance and improve ventilation. In the long-term chest physiotherapy aims to delay the progression of respiratory disease and maintain optimal respiratory function, as removal of mucopurulent secretions may help to reduce the elastase mediated damage to the airways.

Early diagnosis and improved medical management have resulted in a dramatic improvement in longevity. Increasing emphasis has been placed on quality of life and the ability for those with cystic fibrosis to lead independent lifestyles. Several internationally recognised airway clearance techniques[8,9] have been developed in order to facilitate this goal.

Traditionally a twice-daily regimen of postural drainage and percussion has been advocated on confirmation of the diagnosis. The rationale behind routine treatment has been that early intervention may both delay the onset and progression of lung disease and that establishing a routine early on in the child's life may improve long term adherence. The former argument might to some extent be supported by evidence which indicates that infection and inflammation are present in the lungs of some infants from a very early stage in the disease process, long before the manifestation of respiratory signs and symptoms[10,11,12].

The treatment related burden of a daily regimen of airway clearance is significant and adherence to chest physiotherapy has been widely documented as poor[13,14]. The prescription of such an intervention would therefore benefit from supporting scientific evidence, but, to date, the value of routine treatment has not been proven. Few studies evaluate the effect of treatment versus no treatment. A meta-analysis of chest physiotherapy in CF[15] indicated that standard chest physiotherapy comprising postural drainage, percussion and vibration resulted in significantly greater sputum expectoration than no treatment and that standard therapy combined with exercise was associated with an improvement in forced expiratory volume in one second (FEV_1) compared to standard physiotherapy alone. However in a recent systematic review[16] no robust scientific evidence conforming to the strict criteria of the review was found to support the hypothesis that chest physiotherapy, for the purpose of clearing airway secretions, is of value in cystic fibrosis. There was some suggestion from short term cross over trials, which for methodological reasons could not be included in the review, that airway clearance may be beneficial in the short term. Chest physiotherapy is, however, a universally accepted part of the respiratory management of this disorder. To achieve a definitive answer a randomised controlled trial of treatment versus no treatment would need to be undertaken, which would have ethical implications.

Research in airway clearance techniques has therefore primarily focused on comparing the efficacy of two or more treatment modalities. Although many such studies exist, most evaluate short term efficacy and often the results of these comparative studies are contradictory. Confusion in the existing literature can to some extent be explained in terms of study design, protocol and methodology[17]. Overall there does not currently appear to be any evidence which supports one modality as being superior to others. Very few long term studies of airway clearance techniques have been undertaken[18,19,20]. Cystic fibrosis is a life-long disease and data from long term studies should give information on more meaningful outcomes such as frequency of exacerbations/ hospitalisation, quality of life and exercise tolerance. Although long term studies to date do not include a "control" group, the association of their findings has been suggested to indicate that chest physiotherapy does influence the adult consequences of cystic fibrosis[21].

Before treatment is initiated a physiotherapist trained in the techniques should undertake a thorough assessment of the patient's clinical status. In the presence of acute complications, for example an undrained large pneumothorax or severe haemoptysis, airway clearance techniques should be withdrawn until the primary problem has been controlled by medical or surgical management. All techniques will require modification when other complications occur.

4.1 Management of infants and small children

It is generally advocated that a regimen of gravity assisted positioning and percussion should be instituted on a daily basis from the time of diagnosis.

- A general regimen of drainage which includes the apical segments of the upper lobes is usually recommended (alternate side lying and prone with a head down tip, supine flat and supported sitting).
- Treatment is usually recommended between 1-3 times per day (depending on clinical status) and should be timed before feeds.
- In cases where there is proven gastro-oesophageal reflux, positioning may need to be modified to avoid potential exacerbation of reflux.
- There are no long-term trials to date showing that a routine regimen of airway clearance, particularly in asymptomatic patients, is of long-term benefit.
- Although postural drainage and percussion has remained the treatment of choice in the UK, many European CF centres use alternative methods of airway clearance in infants and small children e.g. PEP or a session of physical activity. Although as yet no longitudinal data has been reported in such cohorts, alternative forms of airway clearance may be just as appropriate both in terms of efficacy and adherence.
- By the age of two or three years children can usually begin to play a more active role in their treatment e.g. with breathing techniques. They can then gradually be introduced to an independently performed treatment modality, which is appropriate to their individual circumstances.

4.2 Active cycle of breathing techniques (ACBT)

The active cycle of breathing techniques is the most commonly used treatment modality for CF in the United Kingdom[22]. It was initially described as the forced expiration technique[23], and later documented as ACBT. This technique has been shown to have beneficial effects on both airway clearance and lung function[24,25]. It consists of a cycle of breathing techniques adapted to individual need, but with each component of the cycle clearly defined.

Technique

The active cycle of breathing techniques are principally used in combination with gravity assisted positioning but equally can be used in modified postural drainage (PD) positions or sitting as indicated by the patient's needs. The three individual components of the cycle are:

- Breathing control: gentle relaxed breathing at tidal volume using the lower chest. This is an integral part of the cycle as it minimises any potential increase in air flow obstruction and maintains oxygen saturation[26].

- Thoracic expansion exercises (TEE): three to four deep breaths with emphasis on inspiration, an inspiratory hold, followed by quiet (passive) expiration. Lung volume is increased, reducing resistance to airflow within the distal airways and collateral channels[27], allowing air to assist in the mobilisation of secretions.
- Forced expiration technique (FET): one to two forced expirations with an open glottis from mid to low lung volume to mobilise peripheral secretions. When secretions reach the proximal airways they can then be cleared by a huff or cough at high lung volume. This physiological mechanism is explained using the concept of the equal pressure point (EPP)[28], whereby compression of the airway occurs downstream to the EPP, i.e. where pleural pressure equals intrabronchial pressure at a point dependent on lung volume. The equal pressure point moves distally as lung volume falls, hence mobilising peripheral secretions.

Practical points

- The length of each phase is flexible and should be tailored to individual needs.
- Forced expiratory manoeuvres can lead to an increase in airflow obstruction. Patients with significant airway hyper-reactivity may benefit from longer periods of breathing control to avoid any potential increase in airway obstruction.
- Assisted or self applied chest clapping (percussion) may be performed for short periods during TEE. Many patients find percussion beneficial but it is not an essential component of the cycle[29]. When accompanied by TEE chest percussion has been shown not to have a detrimental effect on oxygen saturation[30].

4.3 Autogenic drainage (AD)

A three phase breathing regimen which uses high expiratory flow rates while avoiding airway closure[31,32]. The utilisation of high airflow produces shearing forces within the airways that may tear mucus from the bronchial walls. There is some evidence to suggest that autogenic drainage is as effective as other airway clearance techniques[33,34].

Technique

Autogenic drainage can be performed in any position effective and comfortable for each individual. The technique involves:

- A slow inspiration using diaphragm and/or lower chest, with the upper airways open. This should reduce the degree of uneven distribution of ventilation.
- An inspiratory hold of three to four seconds maintaining open glottis, this improves even filling of all lung parts, resulting in air behind mucus obstructions.
- Expiratory flow should reach the highest possible velocity without causing airway compression. Expiration should be as fast as possible with an open glottis.
- Tidal volume breathing is carried out at differing lung volume levels, thus maximising the expiratory flow at that particular generation of the bronchial tree.
- Low lung volume level - peripheral airways ‘unstick’ phase.
- Mid lung volume level - middle airways ‘collect’ phase.
- High lung volume level - proximal airways ‘evacuate’ phase.

- The tidal volume breath is raised from low up to high lung volume level breathing according to feedback (audible and palpable crackles). High frequency crackles indicate secretions in peripheral airways. Low frequency crackles indicate secretions in proximal airways. The secretions once collected in proximal airways can be expectorated with one to two effective huffs or coughs.

Practical points

- The avoidance of airway closure as described in AD may be beneficial particularly in patients with significant hyper-reactivity[34,35].
- Autogenic drainage is often performed in conjunction with inhalation therapies and also can be used in conjunction with oscillatory positive expiratory pressure (flutter)[36] or positive expiratory pressure (PEP).

Precautions

- Some patients may find breathing at low lung volume level during the initial phase difficult; particularly those with advanced lung disease.

4.4 Modified autogenic drainage (M AD)

This is a modified form of AD which places less emphasis on the three separate phases of the breathing.

Technique

- Performed in sitting or supine.
- Slow inspiratory phase with an inspiratory hold.
- Fast passive expiration up to expiratory reserve volume, followed by continued active expiration into expiratory reserve volume.
- The length of expiration is determined by the amount of mucus in the airways i.e. the less mucus in the proximal airways the longer the expiration, the more mucus in the proximal airways the shorter the expiration.

Practical points

- With increasing disease severity, self applied PEP in the form of pursed-lip breathing can be utilised to avoid bronchial collapse[37].
- Modified AD is often used in combination with inhalation therapies.

4.5 Oscillating positive expiratory pressure - Flutter®

The Flutter consists of a plastic pipe like device that contains a stainless steel ball bearing. During expiration through the device, the steel ball initially impedes airflow. As expiratory pressure increases and exceeds the effect of gravity, the ball is displaced, the pressure is released and the ball then drops back into the cone. This produces a cyclic oscillatory PEP and a vibratory effect during expiration. Though some evidence exists to suggest that this device has a beneficial effect on lung function and sputum clearance in patients with CF[38,39] other studies suggest no benefit over other techniques such as ACBT[40] and it may be less effective in terms of maintaining pulmonary function[20].

Technique

- Treatment is usually performed in a relaxed sitting position, though gravity assisted positions can also be used.
- Ten to fifteen breaths are carried out inhaling through the nose and exhaling through the flutter following an inspiratory hold of two to three seconds.
- The angle of the flutter is altered until maximal effect of vibration is felt in or on the chest.
- The cheeks are kept flat and firm in order to maximise the vibrations within the chest.
- Coughing is suppressed until the last expiration, which is carried out at approximately twice the speed. The cycle is then repeated as necessary.

Practical points

- Flutter is sometimes applied during a regimen of AD as described previously, though there is little documented on the efficacy of these combined techniques. During the initial phase the angle of the Flutter is tipped upward from horizontal to increase PEP. As secretions move proximally, tidal volume breathing is raised from low to high volume level breathing and the device is lowered to the horizontal position. Coughing is suppressed until secretions have collected in the proximal airways.
- The mechanical oscillatory effect may be advantageous where patients encounter difficulties in mobilising and clearing tenacious secretions[39].
- Flutter can only be introduced at an age where patients are able to co-operate in order to achieve the oscillatory effect during expiration.
- It is important, as with all mechanical devices, that the Flutter is thoroughly cleaned and dried after use and should be used on a single patient basis only.

4.6 Oscillating positive expiratory pressure - R-C Cornet® (Cornet)

The Cornet[41] is a curved tube that contains within its plastic casing a flexible inner tube. During expiration through the device, there is a slight positive expiratory pressure and an oscillation of the air within the airways. The pressure and flow can be varied, by rotating the mouthpiece, until an optimal effect is felt to facilitate airway clearance.

Technique

- Treatment is usually performed in a relaxed sitting position, though gravity assisted positions can be used.
- Start by breathing in through the nose or mouth and out through the Cornet at a normal rate and depth.
- Intersperse normal breathing with a few deeper breaths.
- Breathing control is used in between the more active breathing manoeuvres.
- Huffing and coughing are used to clear secretions mobilised to the upper airways.
- Use the Cornet for 10 to 15 minutes.

Practical points

- The Cornet can be introduced from the age of about 2 years.
- It can be used in any position as it is independent of gravitational forces.
- It is important, as with all mechanical devices, that the Cornet is thoroughly cleaned and dried after use and should be used on a single patient basis only.

4.7 High frequency chest wall oscillation (HFCWO)

A mechanical device (e.g. ThAIRapy® bronchial drainage system, Hayek Oscillator). The ThAIRapy vest which consists of an inflatable jacket and an air pulse generator, to create external chest wall oscillation. The vest fits snugly over the entire thorax and two ports are located on the front panel, which connect to the air-pulse generator. Improvements in lung function measurements have been reported when this technique is compared to conventional chest physiotherapy (PD and percussion) and significant increases in sputum clearance have also been documented[42].

Technique

- Increasing oscillatory frequencies are used during the course of one treatment (range 5-20 Hz). Transient increases in airflow are produced at each compression and are said to result in an alteration in the physical properties and the mobilisation of secretions.
- The device is activated at full inspiration and a five minute period of oscillation at a low frequency applied.
- The patient then performs a forced expiratory manoeuvre before the device is deactivated for expectoration of secretions.
- This cycle is repeated 5-6 times at gradually increasing frequencies.

Practical points

- Treatment is often accompanied by the inhalation of nebulised saline.
- The cost of providing this equipment for each individual is significant and may need careful consideration.
- It is not extensively used as yet in the United Kingdom, but widely used throughout the United States of America.

Additional precautions

- Uncontrolled thoracic chest pain.
- Osteoporosis/osteopaenia.

4.8 Intra pulmonary percussive ventilation (IPV)

Intrapulmonary percussive ventilation combines aerosol inhalation and internal thoracic percussion applied via a mouthpiece. Intrapulmonary percussion is achieved by delivering rapid bursts of air along with a side stream of room air and nebulised solution via a jet venturi. There are few studies to date that evaluate this technique. Intra pulmonary percussive ventilation is reported to be as effective as 'conventional therapy', however no significant differences in lung function, use of intravenous antibiotic therapy or hospitalisation are documented[43]. Further studies need to be undertaken to establish the place of IPV in airway clearance in patients with cystic fibrosis.

Additional precautions

- Uncontrolled thoracic chest pain.
- Osteoporosis/osteopaenia.

4.9 Positive expiratory pressure (PEP)

Positive expiratory pressure can be applied via a facemask or mouthpiece. The PEP mask system comprises a facemask, one way valve with inspiratory and expiratory ports and a selection of resistors. A resistor is placed in the expiratory port to achieve PEP, the level of which can be measured by inserting a manometer between the port and resistor.

Positive expiratory pressure therapy increases intrabronchial pressure in central and peripheral airways splinting the airways open and preventing compression induced by airway collapse. This promotes inflow of air behind mucus obstructions either via a bronchial route or collateral airway channels. Smaller bronchial airways are prevented from collapse thus permitting the continuing upward movement of secretions[44,45]. Several studies to date report PEP to be an acceptable and effective treatment regimen[45,46,47].

Technique

- Before treatment is initiated the correct resistor must be selected. The patient is asked to breathe through the mask using each resistor in turn. The correct size will be that which achieves a steady mid-expiratory PEP of between 10-20 cm H_2O where this is comfortably maintained during a cycle of breaths for 2 minutes.
- Treatment is performed in sitting position. The patient breathes through the mask at tidal volume for approximately 10-12 breaths. Expiration should not be prolonged or forced. Breathing control, huffing and expectoration follows. The cycle is then repeated as necessary.

Practical points

- Expiration will tend to be active against the resistance in the device, but should not be forced. Full expiration should be avoided.
- May be useful in patients with unstable airways, where airway collapse leads to significant unproductive coughing.
- May be useful in patients with significant, proven gastro-oesophageal reflux exacerbated by a head down tipped position.
- It may be less effective in patients producing large volumes of sputum[45].
- It is essential that the mask/mouthpiece system is thoroughly cleaned and dried regularly.
- PEP mask therapy is sometimes combined with ACBT or AD, however there is as yet little scientific evidence available which evaluates such combinations.

4.10 High pressure positive expiratory pressure

High pressure PEP is a modification of the original PEP technique[48]. As disease severity increases, hyperinflation (due to obstructive secretions) and airway instability (due to airway damage) become increasing problems. Trapping of secretions distal to areas of airway collapse during forced expiration may have a negative effect on clearance. By performing forced expiration against a fixed resistance this effect may be negated.

Forced expiration against a resistance creates a constant expiratory flow during early and mid expiration as muscular effort is balanced with the limitation to flow offered by the resistance.

Long and short-term studies have shown beneficial effects on both lung function and sputum clearance[48,49].

Technique

- The size of resistor required is assessed by the patient performing forced vital capacity (FVC) manoeuvres through each level of resistance while the mask is connected to a rolling spirometer or pneumotachograph. The correct resistor will be that which gives a flow volume curve with a maximal FVC, demonstrates a long spell of constant expiratory flow and no curvilinearity (indicating that no airway obstruction has occurred).
- Treatment is performed in supported forward lean sitting. Six to10 breaths of tidal volume breathing through the mask are followed by a full forced expiration through the mask.
- Coughing is usually stimulated at low lung volume, secretions expectorated and the cycle repeated as necessary.

Practical points

- Meticulous regular assessment and monitoring with the use of pulmonary function testing equipment is essential to ensure that the correct size of resistor is being used so as to provide optimal therapeutic value.
- During forced expiration, high expiratory pressures may be generated (40-100 cm H_2O). The safety of the technique has been well documented[50].
- If high pressure PEP induces bronchospasm, pre-medication with bronchodilator therapy should be given or an alternative technique considered.

4.11 Postural drainage (PD) and percussion

This is the most traditional form of chest physiotherapy associated with CF and was introduced soon after the disease was first described. It is still used and often referred to as "conventional physiotherapy". There is no standard definition and this term describes a passive form of treatment which usually requires an assistant and is most commonly used in infants and children. It comprises a regimen of manual or mechanical techniques (chest percussion or vibration) performed in gravity assisted positions and interspersed with huffing and coughing.

Technique

- Postural drainage: gravity assisted positioning utilises the force of gravity to drain specific lobes/segments of the lungs, mobilising mucus from the peripheral to more central airways[51]. Eleven postural drainage positions can be used to drain the various lobes and lung segments[26]. In infants and small children a general regimen of drainage (head down tip in prone and alternate side lying and supine flat) is usually most suitable. Particular attention should be paid to the upper lobes and treatment performed in a sitting position. In older patients a PD regimen which includes one or two areas of the lungs as a specific treatment is more appropriate (e.g. basal zones during one treatment, middle and upper zones at a later treatment). In the presence of specific areas of pathology the relevant PD position should be taught and progress monitored carefully.
- Manual techniques: the inclusion of manual techniques i.e. percussion and vibrations can facilitate mucociliary clearance, enhancing flow rate and sputum production[52]. This may be further improved by incorporating thoracic expansion exercises (TEE) with forced expiratory manoeuvres[53].

- Percussion: assisted or self-applied chest percussion is widely used to enhance sputum clearance in cystic fibrosis[26]. Percussion has been associated with a fall in oxygen saturation[44,54], however when used in combination with ACBT there is no significant detrimental change[30]. Single or double handed rhythmical chest wall percussion is applied comfortably with a cupped hand. The rate, depth and force of the technique should be adapted to each individual. Slow percussion may be better tolerated in patients with hyper-reactive airways[26].
- Percussion can also be applied using a mechanical device. Mechanical percussion has been shown to increase intrathoracic pressure and assist in sputum clearance[55]. However no overall benefit has been shown in comparison to PD with manual percussion and FET on lung function or sputum clearance[56].
- Vibrations: A fine oscillatory pressure thought to produce a mechanical effect assisting in peripheral sputum clearance when combined with PD. There is however little consistent evidence to support this[57]. Vibrations are applied during the expiratory phase of TEE. The force and depth of the technique are adapted to meet individual needs.

Practical points

- In infants and small children when active participation is not possible, passive therapy such as PD and percussion is usually the mainstay of chest physiotherapy care.
- Utilising numerous positions of postural drainage for short periods in one treatment session is ineffective[44].

Additional precautions

When used in conjunction with PD care should be taken in patients with the following:

- significant dyspnoea
- pulmonary oedema
- clinically proven gastro-oesophageal reflux
- late stages of pregnancy when the foetus compromises diaphragmatic excursion
- enlarged spleen and liver.

Chest percussion and vibrations should be avoided or applied with care in the following:

- osteopaenia or osteoporosis (low bone mineral density)
- uncontrolled thoracic pain
- rib fractures/flail segment.

Manual techniques should not be used:

- directly over incisions
- directly over implantable venous access devices
- in those at risk of or known to have low bone mineral density.

Conclusion

Most of the studies undertaken do not conform to the 'gold standard' of randomised controlled trials. Trials which compare different treatment modalities have addressed short term issues in what is a life long illness. Studies have been undertaken mainly in an adult population with results extrapolated to children and subject numbers within studies are generally small with variable methodology and descriptions of techniques. Existing evidence to date does not show any of these techniques to be superior to all others. The grading of the recommendation has therefore been based on the consensus opinion of experts working within this field.

Recommendation

- *Choice of treatment should be dictated by individual requirements (Grade C, Level IV).*

5. EXERCISE

The importance of exercise in maintaining a healthy lifestyle is well recognised both in health and disease[58,59]. Short-term studies of exercise training programmes in cystic fibrosis have been shown to have considerable therapeutic benefit and the majority of patients wish to include it in their routine self care. Everyone can exercise: patients with mild to moderate disease ($FEV_1 \geq 55\%$ predicted) can exercise to the same level as their peers. Those with more severe disease ($FEV_1 < 55\%$ predicted) will require careful assessment with exercising testing and supervised exercise programmes[60]. Exercise programmes must therefore be tailored to the individual, based on disease severity, level of fitness and patient preference. The patient's baseline exercise capacity must be assessed to define their fitness and exercise limitations and provide guidance for safe and effective prescription. It is useful to establish their patterns of habitual activity.

5.1 Benefits of exercise

- increased cardiorespiratory fitness[61]
- increased ventilatory muscle endurance[62]
- decreased breathlessness[63]
- enhanced sputum clearance[64]
- improved body image through increased muscle mass and muscle strength[65]
- improved morale[63]
- enhanced quality of life[66].

5.2 Types of exercise

- An exercise programme should combine endurance and strength training exercises for upper and lower body[67].
- Endurance exercise aims to improve the capacity to endure more exercise without discomfort. Examples include swimming, running, cycling, skipping, aerobic classes, step aerobics, trampolines[61,68,69,70].
- Strength training aims to increase muscle mass and strength. Examples include weights and sprint training[65].
- Interval training may be useful for those patients unable to sustain long periods of exercise. Short bursts of exercise at higher rates will enhance a training response. It may be of benefit to those patients with prolonged periods of desaturation.
- Weight bearing exercises may be useful to prevent or delay loss of bone mineral density.
- Ventilatory muscle training. The evidence for inspiratory muscle training is conflicting[71,72] and to date has shown no advantage over general upper body muscle training[62].
- 'Lifestyle change'. There are no studies to date evaluating the benefits of a lifestyle change. It is important to establish from the time of diagnosis the importance of the contribution of exercise to a healthy lifestyle and encourage the participation of the whole family.

5.3 Exercise programmes

An exercise programme should include exercise of sufficient intensity, frequency and duration to improve cardiorespiratory fitness[67,73,74]. There is no specific information for CF and the general recommendations for the normal population are used. The intensity will be derived from the results of exercise testing. Useful guidelines are:

- 50% of peak work capacity[75]
- 50-60% maximum oxygen uptake (VO_2 max)[76]
- 70-85% of peak heart rate[61]
- 'breathlessness without distress'[77].

Frequency and duration

- Start at a tolerable time and progress to 20-30 minutes 3-4 days a week.

5.4 Precautions

There are no absolute contraindications to exercise but some sports may carry a medical risk (*see below) and careful consideration should be given during certain circumstances:

- acute exacerbation
- arthropathy
- breathless hypoxic patient
- caesarean section
- exercise induced asthma (rare in CF)
- intestinal obstruction
- pneumothorax
- severe haemoptysis
- surgery
- pyrexia.

***Some sports may carry a medical risk:**

- Contact sports and for example bungee jumping for those patients with portal hypertension and significant enlargement of the spleen and liver[78].
- Patients with diagnosed low bone mineral density are at increased risk of facture.
- Scuba diving for patients with air trapping and sinus disease[78].
- Fierce aerobic and anaerobic exercise at altitude for hypoxic patients[79].

All patients undertaking endurance sports should be well hydrated[80] and may require additional salt tablets.

5.5 Exercise and children

Children are not mini-adults. They have different capabilities for, and adaptations to, exercise. They are different in relation to growth, muscles and fat[81]. Children have higher respiratory and heart rates, inferior cooling mechanisms, increased energy expenditure and increased reliance on fat metabolism. All of these factors have important implications for children with cystic fibrosis.

Strength training is not necessarily contraindicated in children; but poorly performed, unplanned and over-strenuous resistance training is as dangerous for children as adults. Growing bones are sensitive to stress, especially repetitive loading and the epiphysial plate is susceptible to injury before full growth is complete. It is important to provide a varied programme and ensure joints are not subjected to repetitive stress.

5.6 Exercising the patient with advancing disease

There is no evidence that exercise is harmful in these patients and they should not be excluded from a training programme[82,83]. For patients with severe disease and those awaiting transplantation maintaining mobility is crucial.

- A maximal exercise test will define the limits.
- Strength and endurance exercises should be continued.
- Oxygen may be required to ease the symptom of breathlessness and increase exercise performance[82].
- For those patients who are dependent on nasal intermittent positive pressure ventilation, exercise should also be performed using the equipment. This can take the form of weight training in sitting, and step-ups in standing or walking on a treadmill.
- It is essential to maintain mobility and careful assessment is required to provide a comfortable level of exercise without undue breathlessness, with or without oxygen[83].

5.7 Exercise testing

Clinical exercise testing[84] can be maximal or submaximal. Types of progressive maximal tests include; treadmill, cycle ergometry, and the modified shuttle test[85]. Submaximal tests include treadmill, cycle ergometry and the 'step test'[86]. Strength testing utilises an isokinetic dynamometer, isometric dynamometer or the maximal weight that can be lifted comfortably.

The value of exercise testing is to:

- assess functional capacity and limitations
- determine level of fitness
- allow safe and effective exercise recommendations
- provide a baseline for further testing
- monitor progress
- evaluate an intervention.

Assessment should consider:

- type of test
- measurements.

Choice of protocol depends on:

- information required
- facilities available
- patient's clinical condition.

5.8 Exercise outcomes

Exercise tolerance

- exercise testing, field tests, strength, flexibility, posture
- diaries
- Habitual activity, Activities of Daily Living (questionnaires)
- activity monitors
- respiratory exacerbations
- hospitalisations
- outpatient visits
- time off work/school.

Symptoms

- dyspnoea (Visual Analogue Scales and Borg scale), cough, sputum, weight gain/loss
- quality of life
- psychological tests.

Recommendations

- *Exercise should be included as part of the therapeutic regimen (Grade A, Level Ib).*
- *Subjective measures of perceived exertion, breathlessness or fatigue may be useful complementary measures when performing exercise tests or in order to gauge levels of activity during exercise programmes (Grade B, Level IIa).*
- *Exercise testing is useful to assess functional capacity, determine the impact of disease process on everyday function, to monitor progress and allow safe and effective exercise recommendation (Grade B, Level III).*
- *Exercise testing on an annual basis is recommended in the CF Trust's Standards for the Clinical Care of Children and Adults with Cystic Fibrosis[1] (Grade C, Level IV).*
- *Exercise programmes should be tailored to the individual taking into consideration disease severity, level of fitness and the patient's preferences to exercise activities (Grade B, Level III).*
- *Supplementary oxygen during exercise may be necessary to prevent exercise induced desaturation and relieve symptoms of breathlessness (Grade B, Level IIb).*

6. MANAGEMENT OF SPECIFIC PROBLEMS

6.1 Advanced cystic fibrosis

Increasing loss of independence with deteriorating health may be difficult for the adult with CF to accept. Heart/lung or double lung transplantation may be considered. In the terminal stages the patient may need help in coming to terms with death. Bereavement counselling should be available for the family and carers, and should continue after the death of the patient.

A feature of the advanced stage is cardiorespiratory failure in spite of optimal medical and physiotherapy management. Cardiorespiratory failure may simply be hypoxic respiratory failure or hypoxic respiratory failure complicated by hypercapnia. Respiratory failure leads to pulmonary artery hypertension and cor pulmonale. Where indicated management should include:

- Regular assessment and monitoring of the requirement for supplemental oxygen both at rest and during exercise. An appropriate delivery system must be selected and consideration given to humidification.
- Treatment of hypoxia and hypercapnia which are the most common cause of death in those with advanced lung disease.
- Treatment of airflow obstruction - bronchodilators, antibiotics, steroids, rhDNase, modified airway clearance techniques.
- Treatment of haemoptysis - refer Section 6.7 'Haemoptysis'.
- Treatment of pneumothorax - refer Section 6.12 'Pneumothorax'.
- Treatment of chronic headache[87].
- Use of adjuncts to physiotherapy e.g. bi-level positive airway pressure (BiPAP) or intermittent positive pressure breathing (IPPB).
- Use of non-invasive ventilation may be included in the management of respiratory failure.

Recommendations

- *Physiotherapy becomes an integral part of the life of a patient with cystic fibrosis and in the terminal stages treatment will need to be modified but contact with a physiotherapist should be continued (Grade C, Level IV).*
- *Those who wish to remain at home and in the community for as long as possible should be provided with domiciliary nursing and physiotherapy support (Grade C, Level IV).*

6.2 Allergic bronchopulmonary aspergillosis (ABPA)

An association between ABPA and CF has been documented[88]. The reported incidence has varied from 0.6% to 11%[89]. There is a marked inflammatory reaction within the lung parenchyma and often fleeting shadows on the chest radiograph. Tenacious mucus plugs, often brown in colour, cause occlusion of the bronchi. These plugs may cause lobar or segmental collapse with subsequent bronchiectasis[90]. Tightness in the chest is often described and associated with an increase in airflow obstruction and deterioration in lung function[91].

Recommendations

- *Airway clearance techniques should be adapted if there is marked airflow obstruction (Grade C, Level IV).*
- *Exercise tolerance may be reduced[91] and programmes may need to be modified (Grade C, Level IV).*

6.3 Arthropathy and joint pain

Cystic fibrosis related arthropathy may be:

- Hypertrophic pulmonary osteoarthropathy (HPOA).
- Periodic arthritis[92].

Clubbing is considered a form of HPOA and the degree of clubbing is linked to the degree of pulmonary disease. Hypertrophic pulmonary osteoarthropathy typically involves the distal parts and adjoining joints of the tibia, fibula, femur and sometimes the long bones of the arms. There is usually pain, swelling and warmth of the involved areas and occasionally small joint effusions. There may be radiological evidence of alterations of the long bones[93]. Most cases of CF arthritis will resolve spontaneously[93]. The appropriate therapy for pulmonary exacerbation may lead to an improvement in joint symptoms[94].

Recommendations

- *The joints affected should be rested, but gentle mobilisation should be continued as possible (Grade C, Level IV).*

6.4 Cystic fibrosis related diabetes

Changes in glucose metabolism are well documented in CF and the incidence of CF related diabetes increases with age[92]. A symptom of hyperglycaemia is polyuria. This may cause dehydration which increases mucus viscosity making expectoration difficult, potentially leading to a decline in respiratory function. Exercise may reduce insulin requirements and can sometimes improve overall blood glucose control. With well controlled diabetes, the main side effect of exercise is hypoglycaemia. Many patients exercise less in hospital resulting in hyperglycaemia.

Recommendations

- *To optimise airway clearance medical management should include control of blood sugar and rehydration (Grade C, Level IV).*
- *To avoid hypoglycaemia during exercise the nutritional intake and insulin requirements should be tailored to each individual following discussion with the diabetes team (Grade C, Level IV).*
- *Activity should be maintained during an infective exacerbation following close liaison with the diabetes team (Grade C, Level IV).*

6.5 Distal intestinal obstruction syndrome

Distal intestinal obstruction syndrome (DIOS) or meconium ileus equivalent (MIE) is used to describe intestinal obstruction occurring after the neonatal period[95]. The patient may present with abdominal pain, distension, vomiting, palpable faecal masses and partial or complete intestinal obstruction.

Medical management may include fluid replacement possibly intravenously, gastrografin by mouth, a balanced electrolyte solution if gastrografin is unsuccessful and review of pancreatic enzymes.

Recommendations

- *Airway clearance techniques may have to be modified until the immediate problem is resolved (Grade C, Level IV).*
- *Continuing mobilisation may help bowel movement (Grade C, Level IV).*

6.6 Gastro-oesophageal reflux (GOR)

Gastro-oesophageal reflux is well documented in patients with cystic fibrosis. Medical management includes the use of drugs which stimulate gastric emptying or reduce gastric acid output, thickening of feeds, and appropriate positioning. Physiotherapy techniques in the first few years of life often include postural drainage and percussion and current evidence is divided on whether the head down position increases gastro-oesophageal reflux[96,97].

Recommendation

- *Careful consideration should be given to positioning in patients with recognised reflux (Grade C, Level IV).*

6.7 Haemoptysis

Blood streaking of sputum is a frequent occurrence in CF and there is no indication to alter the physiotherapy regimen. In cases of frank haemoptysis physiotherapy should be discontinued temporarily until the bleeding settles. When assessing the patient, dependent positioning of the affected lobe should be avoided during chest clearance procedures as this may exacerbate the bleeding and result in a deterioration of oxygenation.

The principles of physiotherapy management in the presence of haemoptysis are outlined in the table below[98,99].

Physiotherapy

Mild: Streaking	● Normal airway clearance regimen.
Moderate: < 250mls blood	● Careful positioning. ● Thoracic expansion exercises. ● Gentle huffing. ● Minimise coughing. ● Airway clearance techniques should minimise increases in intrathoracic pressure. ● Exercise advice (avoid sudden increases in heart rate).
Severe: > 250mls blood	● Oxygen/humidification ● When the bleeding has subsided resume treatment as for moderate.
Embolisation	● Chest clearance can resume after the procedure[26], in consultation with the radiologist.

6.8 Heart/lung and double lung transplantation

Deteriorating chronic respiratory failure and severely impaired quality of life despite optimal management may be indicators for transplantation[100]. Patients should be referred to a transplant centre for assessment.

Recommendations

Pre-transplantation

- *An individualised exercise programme to optimise muscle strength and cardiovascular function should be formulated (Grade C, Level IV).*
- *Airway clearance techniques should be continued (Grade C, Level IV).*

Post-transplantation

- *Patients need to be taught to assess the presence or absence of secretions in the airways because the airways are denervated below the site of the anastamosis (Grade C, Level IV).*
- *Airway clearance techniques should be used as necessary (Grade C, Level IV).*
- *An active life style should be encouraged to obtain and maintain optimum fitness (Grade C, Level IV).*
- *Daily recording of forced expiratory volume in one second (FEV_1) and forced vital capacity (FVC) by the patient are important for early detection of infection and rejection (Grade C, Level IV).*
- *Close liaison between the referring hospital and transplant centre is important (Grade C, Level IV).*
- *Attention to the other aspects of cystic fibrosis care must be continued (Grade C, Level IV).*

6.9 Infection control

The spectrum of CF pathogens includes bacteria, viruses and fungi, but those most important to infection control are *Burkholderia cepacia, Pseudomonas aeruginosa*, and Methicillin-Resistant *Staphylococcus aureus*[101]. The CF Trust has published guidelines for prevention and control of both *Burkholderia cepacia* and *Pseudomonas aeruginosa* infection in cystic fibrosis[102,103].

Recommendations

General

- *All staff and patients should wash and dry hands frequently especially after coughing and before/after having contact with others. The importance of rigorous hand washing must be emphasised (Grade C, Level IV).*
- *Each patient should have their own equipment as indicated e.g. air compressor and nebuliser system, oxygen therapy equipment, airway clearance devices[104, 105] (Grade C, Level IV).*
- *All equipment should be cleaned, dried[106] and maintained according to local health and safety policies[107,108] (Grade C, Level IV).*
- *Stethoscopes should be wiped before and after use with alcohol based solution (Grade C, Level IV).*

- *It is recommended that lung function equipment should be separate for Burkholderia cepacia and non-cepacia patients (Grade C, Level IV).*
- *The "bag in a bottle" system offers protection from dangers of cross-infection[109] as do bacterial filters (Grade C, Level IV).*
- *Respiratory secretions should be handled with care, tissues should be disposed of immediately and sputum pots at least daily (Grade C, Level IV).*
- *Patients should not share eating and drinking utensils or other personal items [102] (Grade C, Level IV).*
- *Close and intimate contact among CF patients should be discouraged[102] (Grade C, Level IV).*

Inpatients

- *Staff and patients should follow local health and safety policies, particularly strict hand washing procedures (Grade C, Level IV).*
- *It is recommended that each patient has a single room (Grade C, Level IV).*
- *Patients infected with B. cepacia should be segregated according to specific strains and segregated from others with cystic fibrosis. B. cepacia has been found in the air following physiotherapy sessions[109,110] (Grade C, Level IV).*
- *MRSA patients should be nursed separately in a single room, with own toileting facilities and local infection control policies must be adhered to. Hospital admissions for MRSA patients should be limited but social contact outside hospital does not appear to be a major risk factor for contamination[111] (Grade C, Level IV).*

Outpatients

- *Clinics should be segregated into B. cepacia and non-cepacia (Grade C, Level IV).*
- *Sub grouping of MRSA, Pseudomonas aeruginosa, non-P. aeruginosa, multiple resistant P. aeruginosa and transplant recipients should be considered (Grade C, Level IV).*

6.10 Liver disease

Liver disease in cystic fibrosis is well recognised and its incidence increases with age. It characteristically presents as biliary cirrhosis, although fatty infiltration of the liver is also a recognised feature. Portal hypertension may result in oesophageal varices. Further acute pressure increases in the portal system may cause variceal rupture and bleeding, which can be life threatening[112]. When required variceal haemorrhage will be treated by sclerotherapy via endoscopy. Rarely gross splenomegaly may require surgical removal.

Recommendations

- *Physiotherapy should be discontinued during acute haemorrhage (Grade C, Level IV).*
- *Physiotherapy techniques will need to modified and attention given to increased breathlessness as respiratory function may be compromised by hepatosplenomegaly and or ascites (Grade C, Level IV).*
- *Modifications to ACT should include careful consideration of positioning (Grade C, Level IV).*

6.11 Persistent significant atelectasis

Lobar or segmental lung collapse may occur, which may be caused by mucus plugging. Medical management may include bronchodilators, antibiotics, high dose oral corticosteroids and bronchoscopy. Rarely, a lobectomy is performed if the patient has good respiratory function and only one lobe is affected.

Recommendations

- *Particular attention should be given to the frequency and effectiveness of airway clearance techniques (Grade C, Level IV).*
- *Additional inhalation therapy (e.g. mucolytics) may further enhance airway clearance (Grade C, Level IV).*
- *Consider administering physiotherapy at time of bronchoscopy (Grade C, Level IV).*
- *Consider physiotherapy adjuncts e.g. intermittent positive pressure breathing (IPPB), non- invasive ventilation (NIV) (Grade C, Level IV).*

6.12 Pneumothorax

A pneumothorax may occur due to rupture of a sub-pleural bleb through the visceral pleura or, rarely, as a result of misplacement of a central line. The incidence increases in adolescent and adult life and is more common in adult males[113]. A small, asymptomatic pneumothorax may be treated conservatively. Larger pneumothoraces will necessitate intercostal drainage. A persistent pneumothorax may require surgical intervention with video assisted thoracoscopic surgery (VATS procedure). Chemical or talc pleurodesis should, where possible, be avoided in potential transplant recipients, but its use is not an absolute contra-indication to subsequent transplantation.

Recommendations

Small pneumothorax

- *Physiotherapy should be continued, but positive pressure devices should be used with caution (Grade C, Level IV).*
- *Adequate pain relief should be ensured if necessary (Grade C, Level IV).*
- *Prolonged, forced expirations and paroxysmal coughing should be avoided (Grade C, Level IV).*
- *Mobility should be encouraged, but intensive exercise avoided (Grade C, Level IV).*

Large pneumothorax

- *Physiotherapy is contra-indicated until a patent intercostal tube is in situ (Grade C, Level IV).*
- *Adequate analgesia is essential (Grade C, Level IV).*
- *Airway clearance techniques should be re-introduced as indicated on assessment (Grade C, Level IV).*
- *Mobilisation and gentle exercise should be encouraged (Grade C, Level IV).*

- *Following removal of the intercostal tube, normal activities may be resumed but heavy exercise and lifting are not recommended immediately (Grade C, Level IV).*
- *Patients should be cautioned about travelling by air (Grade C, Level IV).*

Following surgery

- *Adequate analgesia is essential (Grade C, Level IV).*
- *Physiotherapy techniques should be re-started as soon as the patient's condition allows (Grade C, Level IV).*

6.13 Pregnancy

Pregnancy is well tolerated by patients with an FEV_1 > 60% predicted, but associated with increased maternal and foetal complications in those with an FEV_1 < 60% predicted[114]. As pregnancy can stress the pulmonary, nutritional and cardiovascular reserves of the patient with cystic fibrosis, knowledge of the normal changes[115,116] and their implications in CF[117] is useful.

Recommendations

Physiotherapy

- *Airway clearance techniques should continue throughout pregnancy and be modified as pregnancy progresses with consideration to the degree of breathlessness and discomfort (Grade C, Level IV).*
- *Attention should be paid to the clearance of the lung bases due to a reduction in functional residual capacity (FRC) and early closing volumes, potentially causing trapping of secretions (Grade C, Level IV).*
- *Gentle exercise should be encouraged. Non-weight bearing exercises e.g. swimming and cycling have less energy cost than those which are weight bearing[117]. Established aerobic classes normally continue. Weight training should not be introduced during pregnancy. however an established programme should be modified and discussed with the obstetric team (Grade C, Level IV).*
- *Patients should be encouraged to attend antenatal classes and the importance of pelvic floor exercises stressed. This is more important due to the added strain of coughing on the pelvic floor muscles (Grade C, Level IV).*
- *Patients with low back pain or sacroiliac strain should be given postural advice and gentle exercise and, where appropriate, an abdominal support (Grade C, Level IV).*
- *Gastro-oesophageal reflux should be identified and treated if present (Grade C, Level IV).*

Post natal care

- *Airway clearance should be continued but modified as necessary (Grade C, Level IV).*
- *Pelvic floor exercises should be continued and patients should be encouraged to resume physical activity as soon as possible (Grade C, Level IV).*
- *Any activity limited by abdominal or perineal discomfort should be managed with adequate analgesia (Grade C, Level IV).*

6.14 Reduced bone mineral density

Osteopaenia (low bone mass for age) and osteoporosis (low bone mass and architectural deterioration of bone tissue, which may result in traumatic fractures) are potential complications of CF in children and adults[118,119]. Low bone mineral density can occur as a consequence of decreased exercise, glucocorticoid therapy, malabsorption, low body weight and chronic infection. Physical inactivity is known to contribute to osteopaenia, but this has not yet been proven in cystic fibrosis[120,121].

Recommendations

- *Weight bearing activities should be encouraged (Grade C, Level IV).*
- *Care should be taken with some of the manual techniques associated with airway clearance e.g. chest shaking, chest percussion as there is an increased risk of rib fractures[118] (Grade C, Level IV).*

6.15 Surgery

With increasing longevity the incidence of complications requiring surgery are more prevalent[122]. Post-operative pulmonary complications in patients with pre-existing respiratory problems have been documented.

Recommendations

- *Pre-operative assessment and treatment should be given to those undergoing elective surgery (Grade C, Level IV).*
- *Adequate analgesia should be given to facilitate treatment (Grade C, Level IV).*
- *The patient should be adequately hydrated to minimise retention of viscous secretions (Grade C, Level IV).*
- *Airway clearance techniques should be resumed in the early post-operative period[123], although may require modification (Grade C, Level IV).*
- *Early mobilisation should be encouraged (Grade C, Level IV).*

6.16 Tenacious secretions

Causes of tenacious secretions include dehydration, severe infection, and diabetes.

Recommendations

- *The patient should be adequately hydrated and inspired gases should be humidified (Grade C, Level IV).*
- *Inhaled trial of a mucolytic agent eg; rhDNase or expectorant eg; hypertonic saline should be given, as this may facilitate airway clearance and expectoration (Grade C, Level IV).*
- *Adjuncts, for example intermittent positive pressure breathing, should be considered (Grade C, Level IV).*

6.17 Urinary incontinence

Urinary incontinence is an established problem in women and children and to a lesser extent men with cystic fibrosis[124-126]. It impacts on daily life both socially and during treatment. Although pelvic floor exercises may help with the problem[127] further studies are required to determine the mechanisms of leakage.

Recommendations

- *It is important to identify the problem in both children and adults (Grade C, Level IV).*
- *Patients should be referred to a physiotherapist specialising in incontinence (Grade C, Level IV).*
- *The CF multidisciplinary team should provide encouragement for assessment and sensitivity and support with treatment (Grade C, Level IV).*

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8. GLOSSARY OF ABBREVIATIONS

ACPCF	Association of Chartered Physiotherapists in Cystic Fibrosis
AHCPR	Agency for Health Care Policy and Research
ACPRC	Association of Chartered Physiotherapists in Respiratory Care
ACT	Airway clearance techniques
ACBT	Active cycle of breathing techniques
AD	Autogenic drainage
ABPA	Allergic bronchopulmonary aspergillosis
BiPAP	Bilevel positive airway pressure
DIOS	Distal intestinal obstruction syndrome
EPP	Equal pressure point
FET	Forced expiration technique
FEV1	Forced expiratory volume in one second
FRC	Functional residual capacity
FVC	Forced vital capacity
GOR	Gastro-oesophageal reflux
HFCWO	High frequency chest wall oscillation
HPOA	Hypertrophic pulmonary osteoarthropathy
IPPB	Intermittent positive pressure breathing
IPV	Intrapulmonary percussive ventilation
MIE	Meconium ileus equivalent
M AD	Modified autogenic drainage
PD	Postural drainage
PEP	Positive expiratory pressure
TEE	Thoracic expansion exercises

Notes

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INFORMATION

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