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Concise BTS/ACPRC guidelines
Physiotherapy management of the adult, medical,
spontaneously breathing patient

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**CONCISE BTS/ACPRC GUIDELINES
PHYSIOTHERAPY MANAGEMENT OF THE ADULT, MEDICAL, SPONTANEOUSLY BREATHING PATIENT**

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The Physiotherapy Guideline Development Group is a subgroup of the British Thoracic Society Standards of Care Committee and the Association of Chartered Physiotherapists in Respiratory Care, a clinical interest subgroup of the Chartered Society of Physiotherapy.

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CONCISE BTS/ACPRC GUIDELINES

PHYSIOTHERAPY MANAGEMENT OF THE ADULT, MEDICAL, SPONTANEOUSLY BREATHING PATIENT

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WEB APPENDICES

The following appendices are available to download from the British Thoracic Society website (www.brit-thoracic.org.uk/physioguide):

1. Complementary therapies
2. Example of an evidence table
3. Patient information leaflet COPD
4. Patient information leaflet Asthma
5. Patient information leaflet Hyperventilation Syndrome
6. Patient information leaflet Bronchiectasis
7. Patient information leaflet Chest Wall disease
8. Patient information leaflet Spinal Cord Injury
9. Patient information leaflet Neuromuscular Disease
10. Action plan for patients with Neuromuscular Weakness
11. Devices, manufacturers and suppliers of physiotherapy equipment

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SUMMARY OF GUIDELINES

Physiotherapy should be offered to patients with a variety of medical respiratory conditions, with the aim of breathlessness management and symptom control, mobility and function improvement or maintenance, and airway clearance and cough enhancement or support. Strategies and techniques include: rehabilitation, exercise testing and prescription, including for ambulatory oxygen assessment, airway clearance, and positioning and breathing techniques. Physiotherapy may be helpful for postural and/or musculo-skeletal dysfunction and pain, and provide help in improving continence, especially during coughing and forced expiratory manoeuvres. Physiotherapists are usually central to the delivery of pulmonary rehabilitation and may be instrumental in the noninvasive ventilation service. Physiotherapists are frequently involved in the delivery of oxygen and some nebulised substances, as well as providing vital monitoring of e.g., ventilatory function and cough effectiveness. Some complementary therapies may be appropriate in some situations (**Web Appendix 1**).

Further web appendices are available as downloadable documents: an example of an evidence table (**Web Appendix 2**); patient information leaflets (**Web Appendices 3 – 9**), the exceptions being cystic fibrosis (CF), since comprehensive leaflets on physiotherapy treatment are available via the CF Trust <http://www.cftrust.org.uk/aboutcf/publications/fact-sheets> and for restrictive lung disorders, since there was insufficient evidence to warrant it; an action plan for use when managing patients with neuromuscular disease (**Web Appendix 10**) and a list of physiotherapy equipment and suppliers (**Web Appendix 11**).

A short summary with recommendations and good practice points is given for each diagnostic group.

TABLE 1: SCOTTISH INTERCOLLEGIATE GUIDANCE NETWORK (SIGN) LEVELS OF EVIDENCE AND GRADES OF RECOMMENDATIONS

Levels of Evidence	
1++	High quality meta-analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias
1+	Well-conducted meta-analyses, systematic reviews, or RCTs with a low risk of bias
1–	Meta-analyses, systematic reviews, or RCTs with a high risk of bias
2++	High quality systematic reviews of case control or cohort or studies High quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
2+	Well-conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
2–	Case control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not causal
3	Non-analytic studies, e.g. case reports, case series
4	Expert opinion

Grades of Recommendations

- A** At least one meta-analysis, systematic review, or RCT rated as 1++, and directly applicable to the target population; or


A body of evidence consisting principally of studies rated as 1+, directly applicable to the target population, and demonstrating overall consistency of results
- B** A body of evidence including studies rated as 2++, directly applicable to the target population, and demonstrating overall consistency of results; or

Extrapolated evidence from studies rated as 1++ or 1+
- C** A body of evidence including studies rated as 2+, directly applicable to the target population and demonstrating overall consistency of results; or

Extrapolated evidence from studies rated as 2++
- D** Evidence level 3 or 4; or

Extrapolated evidence from studies rated as 2+

Good Practice Points

-  Recommended best practice based on the clinical experience of the guideline development group

Note: The grade of recommendation relates to the strength of the evidence on which the recommendation is based. It does not reflect the clinical importance of the recommendation.

SECTION 1

CHRONIC OBSTRUCTIVE PULMONARY DISEASE (COPD)

Summary

The patient with COPD will benefit from breathlessness management advice, including positioning to fix the shoulder girdle passively and forward lean postures to lengthen the diaphragm and improve its length tension ratio.

A wheeled walking aid (rollator) may help reduce the ventilatory requirements of walking, both in the acute and domiciliary settings.

A variety of breathing techniques can be used to ameliorate dyspnoea and panic at rest and during exertion. Energy conservation strategies can aid activities of daily living.

Pulmonary rehabilitation should be offered to all patients who are symptomatic, with endurance and strength training exercise of both upper and lower limbs, as well as education.

Inspiratory muscle training may help some patients.

Mobilisation and rehabilitation during hospital admission should be encouraged, using a gutter frame if severely dyspnoeic, which may help reduce physical disability following acute exacerbation.

Airway clearance techniques can be used for those that need them, especially during an infective exacerbation.

Noninvasive ventilation (NIV) should be considered in hypercapnic respiratory failure, with staff training and support crucial for the effective delivery of NIV. Both NIV and oxygen therapy should be delivered as per current national guidance.

Intermittent Positive pressure Breathing (IPPB) may be considered in acute exacerbations of COPD where patients present with retained secretions but are too weak or tired to generate an effective cough.

Recommendations and good practice points

1.1 Management of breathlessness: *Positioning*

Recommendations

- ▶ Advise on passively fixing the shoulder girdle for optimising ventilatory muscle efficiency and relief of breathlessness. (Grade D)
- ▶ Assess the effectiveness of forward lean sitting on relief of breathlessness in all patients with COPD, both in the chronic and acute settings. (Grade C)
- ▶ Advise modification of the forward lean position for use in standing and lying, for patients for whom forward lean sitting is effective. (Grade D)

Good practice points

- ▶ Elbows resting on knees or a table when seated or on a suitable surface e.g. a windowsill or wall when standing.
- ▶ Hands/thumbs resting in/on pockets, belt loops, waistband, or an across the shoulder handbag strap when ambulating.
- ▶ Combine shoulder girdle fixation and forward lean positioning

Research recommendation

- ▶ Further research into the effects of position on ventilation, respiratory mechanics and clinical outcomes is required.

1.2 Management of breathlessness: *Walking aids*

Recommendations

- ▶ Assess the effectiveness of a rollator frame for patients with COPD disabled by breathlessness. (Grade B)
- ▶ Assess the effectiveness of a gutter rollator frame in the acute setting, for patients with COPD severely disabled by breathlessness, especially the elderly. (Grade B)

1.3 Management of breathlessness: *Energy conservation*

Recommendation

- ▶ Teach individualised energy conservation techniques to help reduce dyspnoea during activities of daily living. (Grade D)

1.4 Management of breathlessness: *Breathing techniques*

Recommendations

- ▶ Teach patients with COPD breathing control at rest to see if it helps relieve dyspnoea. (Grade D)
- ▶ Diaphragmatic breathing should not be taught routinely to patients with severe COPD. (Grade C)

- ▶ Teach pursed lips breathing during exertion as a potential strategy to reduce respiratory rate and aid recovery in patients with COPD. (Grade C)
- ▶ Teach exhalation on effort (blow as you go!) as a potential strategy to reduce dyspnoea in patients with COPD. (Grade D)
- ▶ Teach relaxed, slower, deeper breathing as a potential strategy to facilitate more effective ventilation during exertion in patients with COPD. (Grade D)
- ▶ Teach paced breathing as a strategy to maintain control of breathing and reduce dyspnoea during exertion in patients with COPD. (Grade D)

Good practice points

- ▶ Breath-holding during exertion should be strongly discouraged.
- ▶ The use of the technique relaxed, slower, deeper breathing should be confined to during activity.
- ▶ Consider combining techniques.

Research recommendation

- ▶ Further research into the use and effectiveness of different breathing strategies is required.

1.5 Ventilation feedback training

Recommendation

- ▶ Ventilation feedback training is not indicated in patients with COPD. (Grade C)

1.6 Managing anxiety and panic

Recommendation

- ▶ Teach patients with COPD positioning, breathing and relaxation strategies to help manage anxiety and panic attacks. (Grade D)

1.7 Pulmonary rehabilitation

Recommendations

- ▶ Pulmonary rehabilitation should include exercise training of the muscles of ambulation. (Grade A)
- ▶ Pulmonary rehabilitation should incorporate strength training of both upper and lower limbs. (Grade A)
- ▶ Information, advice and education should be integral to pulmonary rehabilitation. (Grade A)
- ▶ Pulmonary rehabilitation should be made available to all appropriate patients with COPD. (Grade A)

Good practice points

- ▶ Physiotherapists, trained as they are in exercise, breathing and pacing techniques for patients with a wide range of respiratory disease, should be central to the delivery of effective pulmonary rehabilitation.
- ▶ Physiotherapists involved in the delivery of pulmonary rehabilitation need to be familiar with current published guidance.

1.8 Field exercise tests

Recommendation

- ▶ The recommended number of practice walks must be included when assessing exercise tolerance with a field exercise test for either the prescription of exercise or ambulatory oxygen. (Grade C)

1.9 Peri-and postexacerbation pulmonary rehabilitation

Recommendations

- ▶ Consider pulmonary rehabilitation soon after exacerbation for patients with COPD. (Grade B)
- ▶ Consider some form of rehabilitation during exacerbation to maintain mobility and function in patients with COPD. (Grade D)

1.10 Respiratory muscle training

Recommendations

- ▶ Consider adding inspiratory muscle training to a general exercise programme where respiratory muscle weakness is thought to be contributing to the patient's problems. (Grade A)
- ▶ Consider inspiratory muscle training in the management of COPD to improve respiratory muscle strength and/or endurance. (Grade A)
- ▶ Consideration of maintenance of an inspiratory muscle training programme is required. (Grade D)
- ▶ Devices that incorporate control of breathing pattern and flow rate should be considered over devices that do not have this function. (Grade D)

Research recommendation

- ▶ Further research is required to establish the adjunctive and relative efficacy of inspiratory muscle training with pulmonary rehabilitation.
- ▶ Studies are required to establish the optimum frequency and intensity of training modalities and most efficacious maintenance therapy.

Good practice points

- ▶ Inspiratory muscle training should not be used to replace pulmonary rehabilitation.
- ▶ Inspiratory muscle training should be considered for patients who are unwilling or unable to partake in pulmonary rehabilitation in order to improve dyspnoea and exercise tolerance.

1.11 Non-invasive ventilation

Recommendations

- ▶ NIV should be offered to patients with COPD and acute hypercapnic respiratory failure, if they meet recommended BTS criteria. (Grade A)
- ▶ Facilities for NIV should be available 24 hours per day in all hospitals likely to admit such patients. (Grade A)

Good practice points

- ▶ Personnel involved with the delivery and care of patients using non-invasive ventilation should be adequately trained in the principles, assessment and effects of non-invasive ventilation.
- ▶ Physiotherapists involved in the delivery of non-invasive ventilation need to ensure that their practice remains in line with current guidance.

1.12 Intermittent positive pressure breathing

Recommendations

- ▶ Tidal volume must be increased to achieve a therapeutic effect. (Grade C)
- ▶ Care must be taken to ensure settings achieve patient synchrony with the device to reduce work of breathing. (Grade C)
- ▶ Short periods of daytime intermittent positive pressure breathing should not be used to treat chronic respiratory failure in stable COPD. (Grade A)

- ▶ Consider intermittent positive pressure breathing in acute exacerbations of COPD where patients present with retained secretions but are too weak or tired to generate an effective cough. (Grade D)
- ▶ When using intermittent positive pressure breathing in acute respiratory failure, FiO₂ of 0.4 may be used. (Grade B)

Good practice points

- ▶ Intermittent positive pressure breathing may be considered in acute exacerbations of COPD where patients do not have immediate access to NIV and intubation is not an option.
- ▶ Monitor the patient carefully and ensure they are returned to their normal FiO₂ following treatment.

1.13 Oxygen therapy

Recommendations

- ▶ Administer oxygen therapy, both in the acute and domiciliary settings, according to current national guidance. (Grade A)
- ▶ Consider assessing the benefit of a walking aid to transport the ambulatory oxygen, especially for the more disabled patient. (Grade B)

1.14 Airway clearance techniques

Recommendations

- ▶ Consider the active cycle of breathing techniques (which includes the forced expiration technique), autogenic drainage and plain or oscillating positive expiratory pressure for patients with stable COPD who need an airway clearance technique to assist in the removal of secretions. (Grade C)
- ▶ Incorporate postural drainage only if it further aids clearance and has no detrimental effects. (Grade D)

Good practice point

- ▶ Consider patient preference in the selection of airway clearance techniques and devices in patients with COPD.

1.15 Pelvic floor muscle training

Recommendations

- ▶ Patients with COPD should be questioned about their continence status. (Grade D)
- ▶ All patients with chronic cough, irrespective of continence status, should be taught to contract the pelvic floor muscles before forced expirations and coughing ('The Knack'). (Grade D)
- ▶ If problems of leakage are identified, patients should be referred to a physiotherapist specialising in continence. (Grade D)

SECTION 2

ASTHMA AND DISORDERED BREATHING (HYPERVENTILATION SYNDROME) AND VOCAL CORD DYSFUNCTION

Asthma Summary

Some form of breathing retraining (breathing exercises and relaxation) should be considered an appropriate treatment for patients with asthma to reduce symptoms and improve quality of life, along with their prescribed medication. This may be in the form of the Buteyko Breathing Technique.

Physiotherapy Guideline

Exercise is recommended, but there is insufficient evidence to support or refute the use of inspiratory muscle training, or the use of airway clearance techniques in patients with asthma.

There is insufficient evidence to support or refute the use of relaxation therapies in the management of asthma and disordered breathing. However, many asthmatics report the benefit of relaxation therapy.

Modalities that may be considered as complementary therapies can promote physical relaxation and improve posture, as well as encouraging gentle breathing at tidal volume. Such modalities may include yoga, Tai Chi, acupuncture or acupressure, with or without TENS (see **Web Appendix 1**, complementary therapy). Some asthma patients support their use and others may find this approach to the management of their breathlessness appropriate to their needs and personal philosophy.

Recommendations and good practice points

2.1 Breathing exercises

Recommendations

- ▶ Breathing exercises, incorporating reducing respiratory rate and/or tidal volume, and relaxation training, should be offered to patients to help control the symptoms of asthma and improve quality of life. (Grade A)
- ▶ Buteyko Breathing Technique may be considered to help patients to control the symptoms of asthma. (Grade B)
- ▶ The use of suitable tools such as an asthma specific quality of life measure, measures of anxiety and depression and the Nijmegen Questionnaire should be used to establish baseline severity of symptoms and monitor progress with treatment (Grade B).

Good practice points

- ▶ Patients should be advised that breathing strategies are adjunctive to, not replacement therapy for, medication.
- ▶ Consider any cost implications to the patient of the Buteyko Breathing Technique.

2.2 Airway clearance techniques

Research recommendations

- ▶ Further research is required for the evaluation of airway clearance techniques in the management of secretions in asthma.
- ▶ Research into the effects of steam inhalations in asthma is required.

Good practice points

- ▶ Where an airway clearance technique is required, consider trying the simplest technique with the least effect on airway constriction.
- ▶ Monitor the patient's condition carefully throughout treatment.

2.3 Heart rate variability biofeedback in asthma

Research recommendation

- ▶ Further research is required in the evaluation of biofeedback in the form of heart rate variability in asthmatics.

2.4 Exercise training

Recommendations

- ▶ Physical training should be advised for improvements in fitness and cardio-respiratory performance in patients with asthma. (Grade B)

- ▶ Physical training should be advised to help reduce breathlessness and improve health related quality of life in people with asthma. (Grade B)
- ▶ Physical training programs should aim to reach a minimum of activity as per the American College of Sports Medicine guidelines. (Grade A)

2.5 Complementary therapy

Patients and patient representatives on these guidelines report benefit from certain complementary therapies, especially those that use controlled breathing techniques with exercise. For discussion of the use of these techniques in asthma please see **Web Appendix 1**.

Hyperventilation Syndrome summary

Breathing retraining (breathing exercises and relaxation) should be considered a first line treatment for patients with hyperventilation syndrome to reduce symptoms.

Modalities that may be considered as complementary therapies can promote physical relaxation and improve posture, as well as encouraging gentle breathing at tidal volume. Such modalities may include yoga, Tai Chi, acupuncture or acupressure, with or without TENS (see **Web Appendix 1**, complementary therapy).

2.6 Breathing retraining

Recommendation

- ▶ Breathing retraining incorporating reducing respiratory rate and/or tidal volume should be offered as a first-line treatment for hyperventilation syndrome, with or without concurrent asthma. (Grade B)

Good practice point

- ▶ The use of a suitable tool such as the Nijmegen Questionnaire should be used to establish baseline severity of symptoms and monitor progress with treatment.

Vocal cord dysfunction (VCD) summary

No evidence on physiotherapeutic techniques exists in this condition, but expert opinion supports the use of the breathing strategies successful in asthma and hyperventilation syndrome.

2.7 Breathing retraining

Research recommendation

- ▶ Research is required to establish the clinical efficacy of breathing techniques in vocal cord dysfunction.

SECTION 3

CYSTIC FIBROSIS

Summary

Physiotherapy is an integral part of the management of the person with CF. The physiotherapist should provide holistic care including assessment and treatment for cardiovascular fitness (exercise) and airway clearance, with the appropriate use of inhalation therapies, oxygen therapy, and non-invasive ventilation and intermittent positive pressure breathing to enhance or support treatment.

A variety of airway clearance techniques may be offered, including those with and without mechanical assistance. The need for and effectiveness of, appropriate postural drainage should be assessed for benefit on an individual basis, as should the addition of nebulised therapies to enhance clearance.

The acceptability of airway clearance techniques however is vital and patient preference for techniques must be taken into account. Non-adherence to treatment is one of the major problems in the management of cystic fibrosis. Treatment factors such as amount of time and effort, infringement on daily activities and unpleasantness are factors that may affect adherence, therefore patient preference for techniques must be taken into account.

Physiotherapists should also undertake assessment and treatment for musculoskeletal and postural disorders, and incontinence.

Physiotherapists must be vigilant about hygiene and infection control in this population.

Recommendations and good practice points

3.1 Adherence to treatment

Research recommendations

- ▶ Further research, using validated methods, is required into adherence to physiotherapy interventions.
- ▶ Research into all physiotherapy techniques should include validated outcome measures to assess adherence levels and patient preference for technique.

3.2 Exercise

Recommendations

- ▶ Exercise should be an integral part of the management of patients with cystic fibrosis. (Grade B)
- ▶ Physical training programs should aim to reach a minimum of activity as per the American College of Sports Medicine guidelines. (Grade A)

Research recommendations

- ▶ Further research is required to assess comprehensively the benefits of exercise programmes in patients with cystic fibrosis, particularly long-term effects.
- ▶ Further research is required on the relative benefits of aerobic and strength training for patients with cystic fibrosis.
- ▶ Research is required to assess effects of exercise programs in people with co-morbidities such as osteoporosis and diabetes.

Good practice point

- ▶ Involve the specialist multidisciplinary team in the decision to instigate or progress physical training programs in the adult with cystic fibrosis, especially in the presence of co-morbidities.

3.3 Airway clearance

Recommendations

- ▶ Teach patients with cystic fibrosis an airway clearance technique to increase mucus transport in the short term. (Grade A)
- ▶ Self administered techniques should be the first line airway clearance techniques offered in order to improve adherence to treatment. (Grade B)

- ▶ Patient preference for techniques should be considered in order to improve adherence to treatment. (Grade B)

Good practice points

- ▶ The technique that is simplest and most effective for any individual should be the method of choice.
- ▶ The frequency and duration of the airway clearance technique should be specific to the needs of the individual patient, which may alter with periods of infective exacerbation.
- ▶ When possible, the airway clearance treatment session should be undertaken until most of the excess bronchopulmonary secretions are expectorated.
- ▶ The airway clearance session should not be so long that the patient becomes fatigued.

Research recommendation

- ▶ Further research is required to assess the long-term effects of airway clearance techniques in adults with cystic fibrosis.

3.4 Postural Drainage and manual techniques

Recommendations

- ▶ Individually assess the effect and acceptability of gravity assisted positioning in patients with cystic fibrosis. (Grade B)
- ▶ Individually assess the effect and acceptability of modified gravity assisted positioning in individual patients with cystic fibrosis. (Grade C)
- ▶ If patients using independent techniques are unable to clear secretions effectively, chest wall vibration should be considered. (Grade C)

3.5 Simple airway clearance techniques

Recommendations

- ▶ Consider the active cycle of breathing techniques when recommending an airway clearance technique for adults with cystic fibrosis. (Grade A)
- ▶ Consider autogenic drainage when recommending an airway clearance technique for adults with cystic fibrosis. (Grade A)
- ▶ Consider positive expiratory pressure when recommending an airway clearance technique for adults with cystic fibrosis. (Grade A)
- ▶ Consider oscillating positive expiratory pressure devices when recommending an airway clearance technique for adults with cystic fibrosis. (Grade A)
- ▶ Exercise in isolation should not be used as an airway clearance technique for patients with cystic fibrosis unless adherence to other techniques is problematic. (Grade D)
- ▶ The addition of exercise to an appropriate physiotherapy regimen should be considered to further increase airway clearance. (Grade D)

Research recommendations

- ▶ Further research is required to assess patient preference for PEP as compared to other airway clearance techniques.
- ▶ Further research is required to establish the safety of High Pressure Positive Expiratory Pressure.
- ▶ Further research is required to establish the efficacy of High Pressure Positive Expiratory Pressure relative to other techniques.
- ▶ Further research is required into the effects of manual techniques in patients with cystic fibrosis.
- ▶ Further research is needed to investigate the adjunctive effect

and optimal regimen of exercise for enhancing airway clearance in patients with cystic fibrosis.

Good practice points

- ▶ Caution should be exercised and regular monitoring undertaken with high pressure positive expiratory pressure.
- ▶ If patients using independent techniques with chest wall vibrations are unable to clear secretions effectively, percussion or chest shaking should be considered.

3.6 Mechanical devices for airway clearance

Recommendations

- ▶ Consider high frequency chest wall compression/oscillation when recommending an airway clearance technique for adults with cystic fibrosis. (Grade A)
- ▶ High frequency chest wall oscillation is not recommended during an infective exacerbation. (Grade B)
- ▶ Consider mechanical vibration when recommending an airway clearance technique for adults with cystic fibrosis. (Grade A)
- ▶ Consider intrapulmonary percussive ventilation when recommending an airway clearance technique for adults with mild to moderate cystic fibrosis. (Grade A)

Research recommendation

- ▶ Research is required to assess the effects of intrapulmonary percussive ventilation in patients with severe disease or when experiencing an infective exacerbation.
- ▶ Further research is required in the use of mechanical in-exsufflation as an airway clearance technique in patients with cystic fibrosis.

Good practice Points

- ▶ High frequency chest wall compression/oscillation, intrapulmonary percussive ventilation and mechanical vibration should be considered where adherence with other airway clearance techniques is problematic.
- ▶ Cost implications should be considered when choosing mechanical devices.

3.7 Inspiratory muscle training

Research recommendations

- ▶ Research is needed to support or refute the use of inspiratory muscle training for airway clearance in patients with cystic fibrosis.
- ▶ Research is needed to assess the clinical impact of improving inspiratory muscle strength in this patient group.

3.8 Non-invasive ventilation

Recommendations

- ▶ Try non-invasive ventilation for airway clearance where there is evidence of respiratory muscle weakness or fatigue. (Grade A)
- ▶ Try non-invasive ventilation where desaturation is present during airway clearance. (Grade A)
- ▶ Try non-invasive ventilation when the patient has difficulty clearing bronchial secretions with other techniques. (Grade A)
- ▶ Consider a trial of intermittent positive pressure breathing for airway clearance as an alternative to non-invasive ventilation, where the indications for NIV in this situation exist. (Grade D)

Research recommendations

- ▶ Further research is required into the long-term effects of non-invasive ventilation for airway clearance in this patient group.
- ▶ Further research is required into the short and long-term effects of non-invasive ventilation during exercise in this patient group.
- ▶ Further research is required in the use of intermittent positive pressure breathing to aid airway clearance in this patient group.
- ▶ Further research is required in the use of continuous positive airway pressure to aid airway clearance in patients with cystic fibrosis.
- ▶ Further research is required in the use of continuous positive airway pressure during exercise in patients with cystic fibrosis.

3.9 Suction

Recommendation

- ▶ Suction should not be considered for use as a routine airway clearance technique in non-intubated patients with cystic fibrosis. (Grade D)

Good practice points

- ▶ Suction may be considered during palliative care where all other methods of decreasing secretions have failed and secretions are distressing for the patient.
- ▶ Suction may be considered if the patient is unresponsive but secretions are distressing for the relatives or friends present.

3.10 Oxygen therapy and humidification

Recommendations

- ▶ Administer oxygen therapy, both in the acute and domiciliary settings, according to current national guidance. (Grade A)
- ▶ Assess patients with advanced disease for supplemental ambulatory oxygen therapy. (Grade D)
- ▶ Bubble through humidification should be avoided due to no evidence of clinical benefit and increased infection risk. (Grade A)

Research recommendation

- ▶ Further research into the short and long-term effects of oxygen therapy during airway clearance and exercise is required.
- ▶ Research into the short and longer-term effects of humidification, particularly with supplemental oxygen, is required.
- ▶ Research into the effects of nebulised normal saline on airway clearance in cystic fibrosis is required.

Good practice point

- ▶ Decide on nebulisation-based humidification for the patient with cystic fibrosis on an individual basis.

3.11 Hypertonic saline

Recommendations

- ▶ Consider the addition of hypertonic saline when enhancing the effectiveness of an airway clearance technique is needed. (Grade A)
- ▶ A pre-dose bronchodilator should be used to minimise bronchospasm with inhalation of hypertonic saline. (Grade D)
- ▶ A bronchoconstriction trial should be carried out at the initial dose of hypertonic saline to ensure safety and suitability for the patient. (Grade D)

3.12 RhDNase for physiotherapy

Recommendations

- ▶ RhDNase should be prescribed as per national and local guidelines. (Grade A)
- ▶ Consider the use of inhaled RhDNase for enhancing airway clearance effectiveness. (Grade D)
- ▶ Consider inhalation therapy with RhDNase for increasing exercise capacity. (Grade D)

Research recommendation

- ▶ Specific research into the effect of inhaled RhDNase on airway clearance is required.

3.13 Musculoskeletal problems and back pain

Recommendations

- ▶ Question patients with cystic fibrosis about musculoskeletal problems and back pain. (Grade D)
- ▶ Assess the problem if present and institute appropriate posture correction, chest wall mobility and stretching exercises or manual therapy treatments where indicated. (Grade D)

Good practice point

- ▶ Effective coughing with appropriate positioning advice should be advocated.

Research recommendation

- ▶ Further research is needed to establish the effectiveness of postural correction and exercise in reducing thoracic pain and deformity.

3.14 Pelvic floor muscle training

Recommendations

- ▶ Question patients with cystic fibrosis about their continence status. (Grade D)
- ▶ All patients with cystic fibrosis, irrespective of continence status, should be taught to contract the pelvic floor muscles before forced expirations and coughing ('The Knack'). (Grade D)
- ▶ If problems of leakage are identified, patients should be referred to a physiotherapist specialising in continence. (Grade D)
- ▶ Therapeutic interventions should include an element of endurance training of the pelvic floor muscles to meet the demands of prolonged coughing. (Grade D)

3.15 Infection control

Recommendation

- ▶ Physiotherapists caring for patients with cystic fibrosis should be aware of consensus documents regarding infection control. (Grade C)

Good practice points

- ▶ Physiotherapists need to be aware of local infection control policies in addition to consensus documents, particularly for the provision of physiotherapy equipment.
- ▶ Physiotherapists should help provide guidance on the importance of infection control and equipment maintenance to patients with cystic fibrosis and their carers.

SECTION 4

NON-CYSTIC FIBROSIS RELATED BRONCHIECTASIS

Summary

Physiotherapy has a key role in the management of the person with non-CF related bronchiectasis, providing holistic care.

This should include assessment and treatment for cardiovascular fitness (exercise) and referral to pulmonary rehabilitation if dyspnoea is impacting on exercise tolerance or functional activities.

A variety of airway clearance techniques may be offered and taught for use as necessary, with accurate and appropriate postural drainage assessed for benefit, as well as the addition of nebulised therapies to enhance clearance.

The acceptability of airway clearance techniques however is vital and patient preference for techniques must be taken into account.

Early review after initiation of therapy is warranted to ensure adjustment of treatment can be made if necessary, taking account of factors such as amount of time and effort, infringement on daily activities and unpleasantness which may all affect adherence.

Use of non-invasive ventilation and intermittent positive pressure breathing to enhance or support treatment should be considered if needed.

Physiotherapists should also undertake assessment for incontinence.

Recommendations and good practice points

4.1 Pulmonary rehabilitation

Recommendations

- ▶ Offer pulmonary rehabilitation to individuals with non-cystic fibrosis related bronchiectasis with breathlessness affecting activities of daily living. (Grade A)
- ▶ Consider the use of inspiratory muscle training in conjunction with conventional pulmonary rehabilitation to enhance the maintenance of the training effect. (Grade B)

4.2 Airway clearance

Recommendations

- ▶ Teach all patients with bronchiectasis and a chronic, productive cough, and/or evidence of mucus plugging on CT, an airway clearance technique for use as necessary. (Grade D)
- ▶ Review the effectiveness and acceptability of the chosen airway clearance technique within approximately 3 months of the initial visit. (Grade D)
- ▶ Patients should be made aware of other available airway clearance technique options. (Grade D)
- ▶ Patient preference and adherence to treatment must be taken into account. (Grade B)

Good practice points

- ▶ If available, use the CT scan to identify affected bronchopulmonary segments to facilitate effective treatment.
- ▶ Use an airway clearance technique that allows independent treatment where possible.

- ▶ Teach individuals with a non-productive cough an appropriate airway clearance technique to use during exacerbations of pulmonary infection.
- ▶ The frequency and duration of the airway clearance technique should be specific to the needs of the individual patient, which may alter with periods of infective exacerbation.
- ▶ Where possible, the treatment session should continue until most of the excess secretions are expectorated but should not be so long that the patient becomes fatigued.

4.3 Postural drainage

Recommendations

- ▶ Where it is found to enhance airway clearance and has no unwanted side-effects, postural drainage should be taught and encouraged. (Grade B)
- ▶ Patient preference and adherence to treatment must be taken into account. (Grade B)
- ▶ Take co-morbidities, and contra-indications and precautions to head-down tilt positions into account. (Grade D)
- ▶ Consider offsetting the increased load of breathing by the use of non-invasive ventilation or intermittent positive pressure breathing where postural drainage is essential for clearing secretions in a breathless patient. (Grade D)

Research recommendations

- ▶ Research is required into the long-term effects of techniques incorporating postural drainage compared with those that do not.
- ▶ Further research is required on the efficacy of modified postural drainage in patients with non-cystic fibrosis related bronchiectasis.

Good practice points

- ▶ Use the CT scan to aid selection of postural drainage positions.
- ▶ Offer modified postural drainage positions (no head-down tilt) as an alternative, only if as effective as the correct postural drainage position.
- ▶ Assess the effectiveness of the position on airway clearance.
- ▶ Offer modified postural drainage positions (no head-down tilt) as an alternative if using the correct postural drainage position is problematic for the patient in any way.

4.4 Manual techniques

Good practice points

- ▶ Consider manual techniques when patients using independent techniques are unable to clear secretions effectively.
- ▶ Offer manual techniques as part of an airway clearance regimen during an acute exacerbation, or when the patient is more fatigued than usual.

Research recommendation

- ▶ Further research is required into whether manual techniques enhance the efficacy of independent airway clearance in patients with non-cystic fibrosis related bronchiectasis.

4.5 Simple airway clearance techniques

Recommendations

- ▶ Consider the active cycle of breathing techniques when recommending an airway clearance technique for adults with non-cystic fibrosis related bronchiectasis. (Grade A)

- ▶ Consider oscillating positive expiratory pressure when recommending an airway clearance technique for adults with non-cystic fibrosis related bronchiectasis. (Grade A)
- ▶ The test of incremental respiratory endurance should not be considered as a first-line airway clearance technique. (Grade B)
- ▶ The inclusion of postural drainage should be considered for all airway clearance techniques. (Grade B)
- ▶ The inclusion of the forced expiration technique should be considered for all airway clearance techniques. (Grade B)

Good practice points

- ▶ Autogenic drainage may be offered as an alternative airway clearance technique if other techniques are less effective or acceptable to the patient with non-cystic fibrosis related bronchiectasis.
- ▶ Positive expiratory pressure may be offered as an alternative airway clearance technique if other techniques are less effective or acceptable to the patient with non-cystic fibrosis related bronchiectasis.

Research recommendations

- ▶ Further research is required to assess the effectiveness of autogenic drainage in adults with non-cystic fibrosis related bronchiectasis.
- ▶ Further research is required to assess the effectiveness of positive expiratory pressure in adults with non-cystic fibrosis related bronchiectasis.
- ▶ Further research is needed to investigate the relative efficacy of different airway clearance techniques in non-cystic fibrosis related bronchiectasis.
- ▶ Further research is required to establish whether the interrupter technique is a valid outcome measure for use in adults with bronchiectasis.

4.6 Humidification

Recommendations

- ▶ Consider nebulised sterile water inhalation before treatment to enhance sputum clearance. (Grade B)
- ▶ Consider nebulised β_2 -agonists before treatment to enhance sputum clearance. (Grade B)
- ▶ Consider nebulised normal saline before airway clearance to increase sputum yield, reduce sputum viscosity and improve ease of expectoration when hypertonic saline is not suitable or available. (Grade B)

4.7 Hypertonic saline

Recommendations

- ▶ Consider nebulised hypertonic saline before airway clearance to increase sputum yield, reduce sputum viscosity and improve ease of expectoration. (Grade B)
- ▶ When first administered, FEV1 or PEFr should be measured pre and post nebulised hypertonic saline to assess for possible bronchoconstriction. (Grade D)
- ▶ Pre-treat with a bronchodilator, particularly for those with bronchial hyper reactivity. (Grade D)

Research recommendations

- ▶ Research is required to determine the long-term effects of hypertonic saline.
- ▶ Research is required to determine effectiveness in patients who produce >10g sputum per day.

4.8 Non-invasive ventilation and intermittent positive pressure breathing

Recommendation

- ▶ Consider NIV or intermittent positive pressure breathing to augment tidal volume and reduce the work of breathing in patients who are becoming fatigued and finding airway clearance difficult. (Grade D)

4.9 Pelvic floor muscle training

Recommendations

- ▶ Patients should be questioned about their continence status. (Grade D)
- ▶ All patients with chronic cough, irrespective of continence status, should be taught to contract the pelvic floor muscles before forced expirations and coughing ('The Knack'). (Grade D)
- ▶ If problems of leakage are identified, patients should be referred to a physiotherapist specialising in continence. (Grade D)

SECTION 5

RESTRICTIVE LUNG CONDITIONS

Summary

There is a paucity of evidence on physiotherapy for these conditions.

Recommendations and good practice points

5.1 Lung fibrosis: Pulmonary Rehabilitation

Recommendation

- ▶ All patients with chronic restrictive conditions, such as pulmonary fibrosis should be considered for pulmonary rehabilitation. (Grade B)

Good practice points

- ▶ Patients with restrictive lung disease should be referred for pulmonary rehabilitation as early as possible in the disease process.
- ▶ The content of education sessions should be adjusted accordingly.

5.2. Community acquired pneumonia: Physiotherapy Management

Recommendations

- ▶ Medical condition permitting, patients admitted to hospital with uncomplicated community acquired pneumonia should sit out of bed for at least 20 minutes within the first 24 hours and increase mobility each subsequent day of hospitalisation. (Grade B)
- ▶ Patients admitted with primary uncomplicated pneumonia should not be treated with traditional airway clearance techniques routinely. (Grade B)
- ▶ In uncomplicated community acquired pneumonia patients admitted to hospital, the regular use of positive expiratory pressure should be considered. (Grade B)

Research recommendations

- ▶ Research is required into the effects of physiotherapeutic strategies for the management of dyspnoea and cough in this patient group.
- ▶ Further research is required on the effects of physiotherapy in patients with pneumonia.

Good practice points

- ▶ Patients should be offered advice regarding expectoration if sputum is present.
- ▶ Consider airway clearance techniques if the patient has sputum and difficulty with expectoration or in the event of a pre-existing lung condition.

5.3 Community acquired pneumonia: NIV / IPPB / CPAP

Recommendations

- ▶ Continuous Positive Airway Pressure (CPAP) should be considered for patients with pneumonia and type I respiratory failure who remain hypoxaemic despite optimum medical therapy and oxygen. (Grade C)
- ▶ Non-invasive ventilation (NIV) can be considered for selected patients with community acquired pneumonia and type II respiratory failure, especially those with underlying COPD. (Grade C)
- ▶ Patients admitted with primary uncomplicated pneumonia should not be treated with traditional airway clearance techniques and intermittent positive pressure breathing (IPPB) in combination. (Grade B)

Good practice points

- ▶ Patients on CPAP or NIV should be carefully monitored for signs of deterioration and appropriate action taken.
- ▶ Physiotherapists involved in the delivery of NIV need to ensure that their practice remains in line with current guidance.
- ▶ Personnel involved with the delivery and care of patients using NIV should be adequately trained in the principles, assessment and effects of NIV.

SECTION 6

NEUROMUSCULAR DISEASES AND MUSCULO-SKELETAL DISORDERS OF THE CHEST WALL

6.1 Chest wall disorders

Summary

Patients with chest wall deformity from any cause with reduced exercise capacity and/or breathlessness on exertion should be offered Pulmonary Rehabilitation. Need for ambulatory oxygen should be assessed.

Respiratory muscle training may be offered, but there is insufficient evidence to support or refute the use of breathing exercises and thoracic mobility exercises in this client group.

6.1.1 Pulmonary rehabilitation and ambulatory oxygen

Recommendations

- ▶ Offer patients with chest wall restriction post tuberculosis pulmonary rehabilitation. (Grade B)
- ▶ Offer patients with chest wall deformity from other causes, who have reduced exercise capacity and/or breathlessness on exertion, pulmonary rehabilitation. (Grade C)
- ▶ Assess patients with moderate to severe kyphoscoliosis who desaturate on exercise for ambulatory oxygen. (Grade D)

Good practice point

- ▶ Pulmonary rehabilitation sessions for patients with chest wall restriction should include relevant education sessions.

6.1.2 Respiratory muscle training and breathing exercises

Recommendation

- ▶ Consider respiratory muscle training in patients with kyphoscoliosis. (Grade D)

Research recommendations

- ▶ Further research into the use of breathing exercises and their effects should be undertaken in this client group.
- ▶ Further research into the use of resisted inspiratory and expiratory breathing exercises (including formal respiratory muscle training) should be undertaken in this client group.
- ▶ Further research into the use of both resisted and unresisted, inspiratory and expiratory, breathing exercises should be undertaken in this client group when undergoing surgical correction of kyphoscoliosis with a Harrington rod.
- ▶ Further research into the use of thoracic mobility exercises and their effects on vital capacity, total lung capacity and clinical outcome should be undertaken in this patient group.

6.2 Spinal cord injuries

Summary

The patient with spinal cord injury will have a good deal of physiotherapy input post trauma, which should not cease when the acute phase is over.

Monitoring of ventilatory function by simple vital capacity measurement must be made regularly. Cough effectiveness must be regularly assessed and monitored. Lack of vigilance to spot indications of impending problems with either ventilation or cough effectiveness can lead to major respiratory problems, the commonest cause of mortality in this client group.

Appropriate and timely care is required. Ventilatory function can be improved with positioning and the use of abdominal binders.

Patients may have difficulty clearing secretions primarily due to expiratory muscle weakness and the use of strategies, such as manually assisted coughing (MAC) and mechanical insufflation-exsufflation (MI-E) must be considered and introduced where indicated.

Patient and carer preference should be considered when performing or teaching manually assisted coughing.

Functional electrical stimulation of expiratory muscles can be used to enhance vital capacity and may lead to improved cough effectiveness. 13.7 Exercise should be encouraged and respiratory muscle training can enhance muscle strength or vital capacity although the clinical significance of this is not yet established.

Recommendations and good practice points

6.2.1 Monitoring

Recommendations

- ▶ Monitor the spinal cord injury patient for the signs and symptoms of respiratory problems and take appropriate action if abnormal or changing. (Grade A)

- ▶ Measure VC routinely in the upper spinal cord injury patient and take appropriate action if falling. (Grade D)
- ▶ Alert medical staff if VC falls to 1 litre or less. (Grade D)

Good practice point

- ▶ Consider the use of an incentive spirometer to monitor vital capacity at home.

6.2.2 Positioning

Recommendations

- ▶ Consider the supine position to maximise vital capacity. (Grade B)
- ▶ Assess the head up 30° position for improving pulmonary function. (Grade C)
- ▶ The head down position should only be used where there is a demonstrable need and only with extreme caution. (Grade D)
- ▶ Any patient, especially those with early spinal cord injury should be carefully monitored for signs of hypoxaemia in head down positions. (Grade D)
- ▶ Take co-morbidities and contra-indications and precautions to head-down tilt positions into account. (Grade D)

Good practice points

- ▶ Spinal cord injury patients with resting hypoxaemia should be given supplemental oxygen if placed in the head down position.
- ▶ Patient comfort and preference should be taken into account with any position.
- ▶ The effect of an abdominal binder, if used, should be taken into consideration.

6.2.3 Abdominal binders

Recommendations

- ▶ Assess the effect of an abdominal binder for upright sitting where improvement in either vital capacity or respiratory muscle function is required. (Grade D)
- ▶ Patients using non elastic binders should be monitored closely. (Grade D)
- ▶ When using an abdominal binder, the optimal position for the individual patient should be determined. (Grade D)

Research recommendation

- ▶ Further research into the use of abdominal binders should be undertaken.

6.2.4 Spontaneous cough

Good practice points

- ▶ Try the forward lean position to enhance the effectiveness of spontaneous coughing.
- ▶ Try 'hooking' one arm over the back of the wheelchair for added stability and leverage during spontaneous coughing.

6.2.5 Assisted coughing

Recommendations

- ▶ Try manually assisted coughing for patients with an ineffective cough. (Grade D)
- ▶ The upright seated position should be considered initially. (Grade D)
- ▶ The abdominal thrust (Heimlich-style manoeuvre) should be considered initially. (Grade D)

Good practice points

- ▶ Alternative body positions and thrusts should be tried if these fail to produce an effective result.

6.2.6 Mechanical insufflation-exsufflation

Recommendations

- ▶ Mechanical insufflation-exsufflation should be considered for individuals with upper spinal cord injury, if simpler techniques fail to produce an adequate effect. (Grade D)
- ▶ Where cough effectiveness remains inadequate with mechanical in-exsufflation alone, combine it with manually assisted coughing. (Grade D)

Good practice points

- ▶ Caution should be observed in acute upper spinal cord injured patients who may be susceptible to bradycardia or cardiovascular instability.
- ▶ Mechanical insufflation-exsufflation pressures should be set to obtain the optimal airway clearance effect in an individual, but avoid using high pressures where possible.
- ▶ End the treatment session with an insufflation to minimise airway closure.

6.2.7 Functional electrical stimulation

Recommendation

- ▶ Consider electrical stimulation of the abdominal muscles as a possible means of enhancing lung volumes and cough effectiveness. (Grade C)

Research recommendation

- ▶ Further research is required into the clinical effects of functional electrical stimulation and the optimum electrode placements and electrical frequency.

6.2.8 Exercise

Recommendation

- ▶ Active exercise should be encouraged in patients confined to a wheelchair as a result of spinal cord injury. (Grade D)

6.2.9 Breathing exercises

Recommendation

- ▶ Deep breathing exercises should be encouraged in patients with spinal cord injury. (Grade D)

Research recommendations

- ▶ Further research is required into the both the nature and comparative effects of deep breathing and resisted breathing exercises in patients with spinal cord injury.

6.2.10 Respiratory muscle training

Recommendations

- ▶ Inspiratory muscle training may be considered for patients with upper spinal cord injury to improve respiratory muscle strength. (Grade C)
- ▶ Inspiratory muscle training may be considered for patients with upper spinal cord injury to improve vital capacity and residual volume. (Grade C)

- ▶ Training of the accessory muscles of respiration with progressive loading should be considered. (Grade D)

Research recommendations

- ▶ Further research is required to establish the clinical benefit of inspiratory muscle training for patients with upper spinal cord injury.
- ▶ Further research is required in the use of respiratory muscle training in patients with spinal cord injury to establish the optimum type, frequency and duration.
- ▶ Further research is required in the use of specific training for the clavicular portion of the pectoralis major muscle in patients with spinal cord injury.

6.3 Neuromuscular disease

Summary

The patient with neuromuscular disease has great need of physiotherapy to provide vital monitoring of ventilatory function and cough effectiveness, as well as actual assistance with airway clearance. Lack of vigilance and appropriate and timely care can lead to major respiratory problems, the commonest cause of mortality in this client group, particularly when the disease and muscle weakness progress.

Oxygen therapy should be administered with great care in patients with neuromuscular disease with evidence of diaphragm dysfunction. NIV should be considered in those at risk of developing hypercapnia in response to oxygen therapy. The use of respiratory aids when oxygen saturation falls below 95% may prevent or delay the need for tracheostomy and used in conjunction with non invasive ventilation may increase survival in this client group.

Patients may have difficulty clearing secretions due to inspiratory, expiratory and or bulbar muscle weakness. Peak cough flow and vital capacity must be regularly measured and monitored to indicate impending problems with either ventilation or cough effectiveness.

Conventional physiotherapy techniques are unlikely to be effective and suction is not usually tolerated or successful in this client group. The use of strategies, such as glossopharyngeal breathing (frog breathing), or respiratory aids, to breath stack and/or increase maximal insufflation capacity (MIC) must be considered and introduced where indicated.

The use of strategies, such as manually assisted coughing (MAC) or mechanical insufflation-exsufflation (MI-E) can be used to increase Peak Cough Flow to effective levels and enhance cough effectiveness and must be considered and introduced where indicated.

Careful consideration must be given to the ability of the patient to perform the techniques, such as the presence of bulbar dysfunction. Patient and carer preference should be considered when performing or teaching manually assisted coughing.

Maximal insufflation capacity should be used regularly as a means of maintaining range of movement to the lungs and chest wall.

Some patients with early disease may benefit from respiratory muscle training but caution is advised in Duchenne muscular dystrophy.

Recommendations and good practice points

6.3.1 Monitoring

Good practice points

- ▶ Monitor vital capacity in patients with neuromuscular disease to guide therapeutic interventions.
- ▶ When vital capacity falls to <50% take appropriate action to minimise the risk of respiratory failure and cough impairment.
- ▶ Monitor oxygen saturation in patients with neuromuscular disease to guide therapeutic interventions.
- ▶ Routinely assess for signs of hypoventilation in this patient group (see **Appendix B**).

6.3.2 Oxygen therapy and non-invasive ventilation

Recommendations

- ▶ Low flow (high FiO₂) oxygen therapy should be avoided or used with extreme caution due to the risk of carbon dioxide retention in patients with neuromuscular disease. (Grade A)
- ▶ Consider non-invasive ventilation as an initial intervention in patients with, or at risk of developing, hypercapnia. (Grade D)

Good practice points

- ▶ Repeat blood gases analysis, or end-tidal CO₂ monitoring if available, should be performed 30 minutes post administration of newly administered low flow oxygen therapy in the at risk patient group.
- ▶ The delivery of non-invasive ventilation must remain in line with current guidance.
- ▶ Personnel involved with the delivery and care of patients using non-invasive ventilation should be adequately trained in the principles, assessment and effects of non-invasive ventilation.

6.3.3 Peak cough flow monitoring

Recommendations

- ▶ Peak cough flow should be measured regularly in patients with neuromuscular disease. (Grade D)
- ▶ Measure peak cough flow additionally at the time of an acute respiratory tract infection. (Grade D)
- ▶ When peak cough flow is equal to or less than 270L/min in a medically stable patient, introduce strategies for assisted airway clearance to raise it above 270L/min. (Grade D)
- ▶ When peak cough flow is equal to or less than 160 L/min, additional strategies to assist secretion clearance must be used. (Grade D)
- ▶ If peak cough flow remains equal to or less than 160 L/min despite additional strategies, contact medical colleagues to discuss ventilation and/or airway management needs. (Grade D)

Good practice point

- ▶ The peak cough flow values above are a useful guide only and physiotherapists should ensure that the patient's cough is sufficient to clear secretions.

6.3.4 Airway Clearance

Recommendation

- ▶ When oxygen saturation falls below 95% the use of non-invasive ventilation and/or strategies to aid airway clearance should be considered. (Grade D)

Good practice point

- ▶ Careful assessment is needed to determine if assistance with inspiration, expiration, or both, is required.

6.3.5 Maximal insufflation capacity

Recommendations

- ▶ Use some form of maximal insufflation strategy to improve effective cough generation when vital capacity falls below 1500mls or 50% predicted. (Grade D)
- ▶ Use single maximal insufflation techniques for patients with bulbar dysfunction who are unable to breath stack. (Grade D)
- ▶ Teach patients without bulbar muscle involvement unaided breath stacking to improve cough effectiveness independently where possible. (Grade D)
- ▶ Regular breath stacking (10-15 times three times per day) to maximal insufflation capacity should be performed by patients with vital capacity of less than 2000ml or 50% predicted. (Grade D)

Research recommendation

- ▶ Further research is required to establish the relative efficacy of different maximal insufflation strategies in patients both with and without bulbar muscle involvement to improve cough effectiveness.

6.3.6 Glossopharyngeal breathing

Recommendations

- ▶ Consider teaching glossopharyngeal breathing to patients with reduced vital capacity to maintain range of chest wall movement and pulmonary compliance. (Grade D)
- ▶ Consider teaching glossopharyngeal breathing as one of the means of achieving maximal insufflation capacity in patients who have difficulty in clearing secretions. (Grade D)
- ▶ Consider teaching glossopharyngeal breathing to ventilator dependent patients to allow some ventilator free breathing time. (Grade D)
- ▶ Consider teaching glossopharyngeal breathing to patients with decreased voice strength. (Grade D)

Good practice points

- ▶ Physiotherapists involved in the long-term care of patients with neuromuscular disease should consider learning the technique of glossopharyngeal breathing.
- ▶ Physiotherapists should include glossopharyngeal breathing more widely in their rehabilitation plan to ensure a more holistic and active programme for the neuromuscular patient.

Research recommendation

- ▶ Further study of the effects of glossopharyngeal breathing is required.

6.3.7 Manually assisted coughing

Recommendations

- ▶ Manually assisted coughing should be used to increase peak cough flow in patients with neuromuscular disease. (Grade C)
- ▶ Combine manually assisted coughing with a maximal insufflation capacity strategy. (Grade D)
- ▶ Abdominal thrusts should be performed standing in front of the patient where possible to assist communication. (Grade D)

6.3.8 Mechanical in-exsufflation

Recommendations

- ▶ Consider mechanical in-exsufflation as a treatment option in patients with bulbar muscle involvement who are unable to breath stack. (Grade D)
- ▶ Consider mechanical in-exsufflation for any patient who remains unable to increase peak cough flow to effective levels with other strategies. (Grade D)
- ▶ Where cough effectiveness remains inadequate with mechanical in-exsufflation alone, combine it with manually assisted coughing. (Grade D)

Good practice points

- ▶ Mechanical in-exsufflation pressures should be titrated to suit the individual to optimise the insufflation and exsufflation required to achieve an effective cough,
- ▶ If secretions require loosening to facilitate removal, other strategies must be employed prior to using mechanical in-exsufflation.

Research recommendations

- ▶ Further research is required to establish the effect of mechanical in-exsufflation in patients with neuromuscular disease and acute respiratory infection.
- ▶ Further research is required to establish the relative effectiveness of the mechanical in-exsufflator compared with other combinations of techniques.

6.3.9 Intrapulmonary percussive ventilation

Recommendations

- ▶ Intrapulmonary percussive ventilation may be considered for patients with neuromuscular disease to aid loosening of secretion prior to removal where there is evidence of sputum retention and other techniques have failed. (Grade D)
- ▶ In patients with ineffective cough assisted cough strategies must be used additionally to increase cough effectiveness. (Grade D)
- ▶ Patients using intrapulmonary percussive ventilation must be monitored closely during and after treatment for any adverse response. (Grade D)

Research recommendation

- ▶ Further research is required to evaluate the safety and efficacy of intrapulmonary percussive ventilation in the care of patients with neuromuscular disease.

6.3.10 Respiratory muscle training

Research recommendations

- ▶ Further research is required to determine whether respiratory muscle training is safe and beneficial in patients with Duchenne muscular dystrophy and spinal muscle atrophy.
- ▶ Further research is required to determine whether respiratory muscle training is beneficial in patients with other types of neuromuscular disease.

SECTION 7

PHYSIOTHERAPY WORKFORCE CONSIDERATIONS

Workforce planning is an essential part of service delivery, although physiotherapy services in the UK are usually independent of medical directorates and often adhere to an historical and rather complex system. The system creates difficulties in the identification of numbers of whole time equivalent (WTE) physiotherapists required to provide a service to any given speciality, e.g. respiratory medicine, vital in today's commissioning climate. Being in such small numbers, specialist respiratory physiotherapists cannot provide 7 day or 24 hour cover. For this reason, physiotherapists from other specialities, in particular junior staff, are used to cover 'out of hours' care. Every effort is made to train these individuals to an acceptable degree, but it remains a continuing challenge to provide a competent on call workforce.

Given the complexity of the task it is beyond the scope of this document to provide comprehensive recommendations for WTE physiotherapists for a population of other respiratory patients at this stage. However, a consensus was reached among the steering group, chairs of the guidelines working party and some external experts in managing physiotherapy services, on the clinical component on each physiotherapy intervention and the time needed to undertake the treatment listed in both uncomplicated and complex situations (*Table 2*).

CONCLUSION

This is the first extensive systematic literature review undertaken of the existing evidence surrounding comprehensive physiotherapy management of the spontaneously breathing medical respiratory adult patient and providing graded recommendations for practise.

The full BTS/ACPRC Guideline is published in Thorax Vol 64 Suppl1 Available online at: <http://thorax.bmj.com/content/vol64/issueSuppl1> and at: <http://www.brit-thoracic.org.uk/physioguide>

TABLE 2: TIME (HOURS) REQUIRED FOR PHYSIOTHERAPY INTERVENTIONS IN UNCOMPLICATED AND COMPLEX SITUATIONS

Intervention	Uncomplicated patient	Acutely unwell or complex patient
Initial assessment of all patients includes:		
• History		
• Physical examination		
• Investigations and results	0.5 - 0.75	1
• Interpretation of imaging		
• Tests of mobility, function or exercise capacity		
Airway clearance techniques Initial exploration and teaching of appropriate technique	0.5 - 0.75	1
Airway clearance techniques Follow up/review	0.25 - 0.5	0.5 - 1
Breathlessness management	0.50	0.75
Self management / education	0.25	0.50
Individual help with mobility, physical activity / exercise	0.50	1
Formal exercise test +/- mobility aids	0.25 - 0.50	0.75 - 1
Ambulatory oxygen assessment	1	1.25 - 1.45
Non-invasive ventilation CPAP, NIV, IPPB : Set Up	1	1
Non-invasive ventilation CPAP, NIV, IPPB Ongoing	0.50	1
Special interventions e.g.:		
• Bronchoconstriction trials	1	1
• Induced sputum		
• Hypertonic NaCl trial		
Pulmonary rehabilitation assessment	1 per patient or 10 per group	
Pulmonary rehabilitation 8 week programme	10 / patient / programme, or 100 / group or programme	

APPENDIX A - GLOSSARY OF PHYSIOTHERAPY TERMS

Adapted with permission from the International Physiotherapy Group for Cystic Fibrosis (IPG/CF) 2007
www.cfww.org/IPG-CF/index.asp

Terms used in physiotherapy for respiratory conditions. Common physiological terms and expressions, defined in respiratory physiology books, are not included.

Term	Condition
Active Cycle of Breathing Techniques (ACBT)	An airway clearance technique. A cycle of the techniques of breathing control (BC), thoracic expansion exercises (TEE) and the forced expiration technique (FET).
Air flow	Expressed volume / time, (l/min).
Air flow velocity	Expressed distance / time, (m/min), speed.
Airway Clearance Technique (ACT)	An airway clearance strategy (with or without a device) used to support mucus clearance by loosening, mobilizing, transporting and evacuating airway mucus.
Assisted Autogenic Drainage (AAD)	Autogenic Drainage (AD) carried out with assistance in infants, toddlers or individuals unable to follow instructions or to participate actively.
Autogenic Drainage (AD)	An airway clearance technique utilizing optimal expiratory flow rate at different lung volume levels.
Bi-level Positive Airway Pressure (BiPAP)	Assisted ventilation with independent settings for positive inspiratory and expiratory pressures.
The 'Bird'	See Intermittent Positive Pressure Breathing.
Blow-as-you-go!	A term to help remind the patient to exhale on effort, stretching and bending to improve respiratory mechanics during activity.
Bottle-Blowing / Bubble PEP	"Positive Expiratory Pressure" generated by blowing through a narrow tube into water.
Breathing Control (BC)	Normal tidal breathing encouraging relaxation of the upper chest and shoulders.
Breathing Exercises	Exercises designed to alter breathing for a particular purpose; e.g., increasing lung volumes, decreasing lung volumes, airway clearance.
Buteyko Breathing Technique (BBT)	A compilation of "reduced breathing" exercises and other strategies for control of asthma symptoms; is a more intensive and broader therapy than the conventional physiotherapy technique but with similar results.

Appendix A

Term	Condition
Chest clapping/percussion	Rhythmical percussion (tapping) of the chest wall using either the hand/s with a flexion / extension action of the wrist/s or a mechanical device, with the aim of loosening secretions.
Chest compression	Firm manual or mechanical compression of the chest during expiration in the direction of the normal expiratory movement of the ribs i.e., down and in to enhance “air flow” or “cough peak flow”. Commonly combined with “chest shaking/vibrations” to enhance airway clearance. Can be used as a “manually assisted cough” technique.
Chest mobility exercise(s)	Physical flexibility exercises to maintain or increase the mobility of the chest wall.
Chest physiotherapy	Historical ambiguous term used to define airway clearance therapy. Commonly in the USA refers to “postural drainage” with “percussion”, with or without “chest compression”.
Chest shaking/vibrations	Shaking or vibrating the chest wall during expiration, in the direction of rib movement; usually combined with “chest compression”
Continuous Positive Airway Pressure (CPAP)	Assisted ventilation with the same positive pressure setting during the whole breathing cycle.
Cough technique	Using cough in a controlled way, at specific lung volumes, to check for and / or assist the removal of bronchial secretions.
Cough Control	Being able to control the cough, to prevent unproductive paroxysms of coughing or coughing attacks.
Delta Rollator frame	A triangular “rollator frame”; highly manoeuvrable and may have a carrying bag attached for ambulatory oxygen.
Diaphragmatic Breathing	Breathing using abdominal movement; reducing the degree of chest wall movement as much as possible. Not advocated in patients with hyperinflation. Commonly used in complementary therapies.
Directed coughing	Coughing under instruction; direction given on technique, timing, frequency and duration.
Energy Conservation (EC)	A method of performing tasks and activities to utilise breathing, pacing and positional strategies to reduce the work of a task/activity.
Expiratory Muscle Training (EMT)	Breathing out against a resistance as a means enhancing strength or endurance of the expiratory muscles. Can be against a fixed load or via a threshold load resistor.
Expiratory Resistance Breathing (ERB)	Breathing out against a resistance. Type and size of resistance chosen dependent on physiological strategy, aims and individual needs. May be for e.g., airway clearance or “respiratory muscle training”.
Forced Expiration Technique (FET)	Huffs/forced expirations interspersed with periods of “breathing control”.

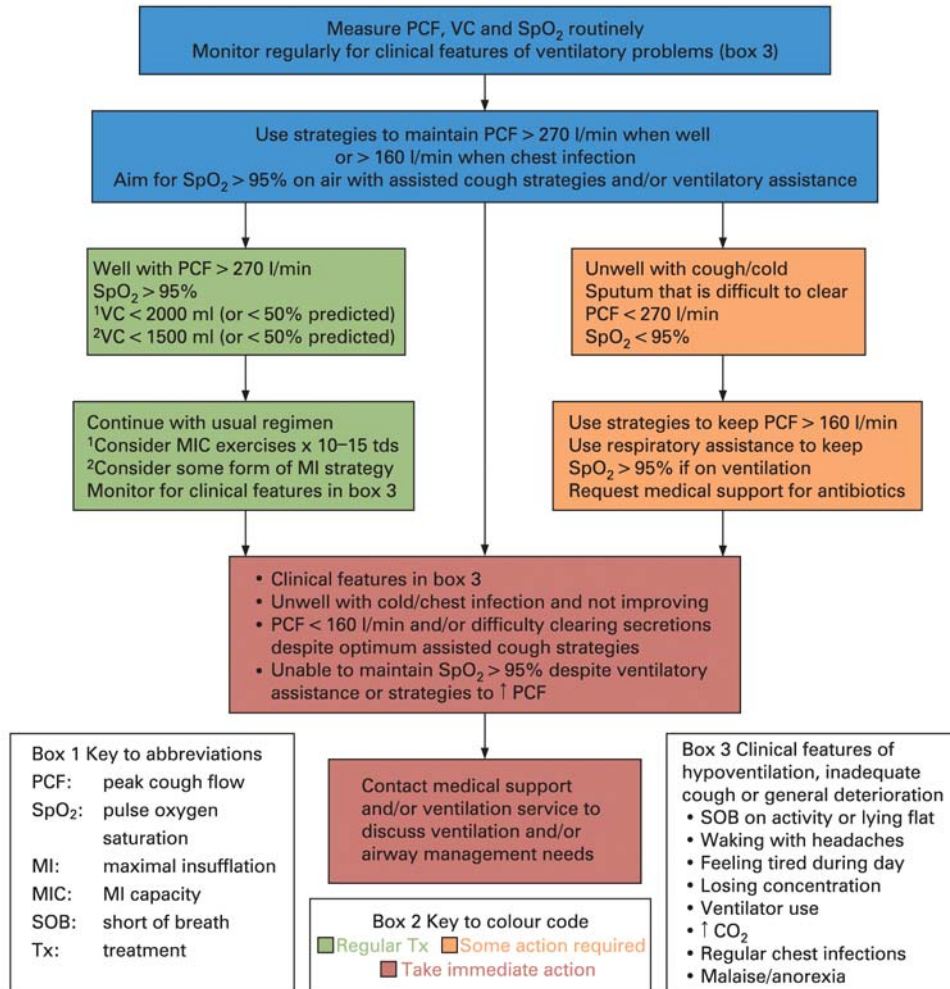
Term	Condition
Forward Lean Sitting (FLS)	Seated, leaning forwards, relaxed; supporting the elbows either on a table or own knees. Enhances respiratory muscle function by loading the diaphragm and by passive fixation of the shoulder girdle. Commonly used where there is hyperinflation of the lungs and increased FRC, as in COPD.
Glossopharyngeal Breathing (GPB) (Frog breathing)	A method of breathing using the tongue and soft palate (as a frog does) to push air into the lungs for enhancing inspiration in an individual with weak inspiratory muscles.
Gutter Rollator frame	A "rollator frame" with a shoulder height support for the forearms; used for severely breathless patients to allow mobility otherwise very difficult.
High Frequency Chest Wall Oscillator/Oscillation (HFCWO)	A device/technique to externally oscillate the chest wall by means of a pneumatic jacket to aid loosening of secretions.
High Positive Expiratory Pressure (Hi-PEP)	An airway clearance technique combining "Positive Expiratory Pressure" with forced expirations against the resistor.
Huff, huffing	A huff is a forced expiration with an open glottis; performed from a large lung volume moves central secretions; from a mid lung volume peripheral secretions.
Humidification	Adding moisture to inhaled air or oxygen to prevent drying of mucosa and/or secretions and to improve gas exchange; may be sterile water or normal saline via nebuliser chambers, both large and small volume and via heated water bath.
Hypertonic Saline (HTS)	A solution of (commonly 7% in the UK but may be 9%) saline to increase fluid flux from the airways into the mucus to improve secretion clearance; usually advocated pre airway clearance.
Inhalation device	A device through which aerosolized or powdered drugs can be inhaled.
Inhalation therapy	Delivery of aerosolized or powdered drugs to the airways through inhalation.
Inspiratory Muscle Training (IMT)	Breathing in against a resistance as a means enhancing strength or endurance of the inspiratory muscles. Can be against a fixed load or via a threshold load resistor.
Intermittent Positive Pressure Breathing (IPPB)	The original form of "NIV"; pressure cycled, powered by compressed gas with integral nebuliser; flow rate, sensitivity and pressure are adjustable. Mouthpiece most commonly used interface but may be delivered via a port-free mask (without a fixed leak), as closed circuit system.
Intra Pulmonary Percussive Ventilation (IPV)	A device to internally oscillate/percuss the chest to aid loosening of secretions, by means of high-frequency bursts of gas. Powered by compressed gas and can be used to deliver nebulised drugs during treatment.
Manually Assisted Cough (MAC)	Firm compression of the chest wall or abdomen during expiration to enhance a weak cough in an individual with weak/paralysed expiratory muscles. Often combined with a "maximum insufflation capacity" technique.

Appendix A

Term	Condition
Manual techniques	The collective term for chest compression, chest shaking and chest wall vibrations; see individual techniques.
Manual therapy	The treatment of joints and muscles by specific mobilization, manipulation and stretching.
Maximum Insufflation Capacity (MIC)	Enhancing inspiration prior to a cough in an individual with weak inspiratory muscles; via “chest compression”, “NIV”, “IPPB”, a bag, or “Glossopharyngeal breathing”. Commonly used with a “manually assisted cough”.
Mechanical In-Exsufflation (MI-E)	A device to provide both positive and negative pressure in alternating cycles to enhance “MIC” and/or “cough peak flow” to enhance airway clearance. Can be combined with other techniques, most commonly a “manually assisted cough” technique.
Modified postural drainage	The adaptation of “postural drainage” i.e. to eliminate head-down positions.
Mucociliary clearance	The physiological movement of airway mucus by the mucociliary transport system, in a cephalad direction (towards the head).
Nebuliser	A device that aerosolises a liquid.
Nebuliser system	Equipment comprising of an energy source and a nebuliser. These function as a unit.
Nijmegen Questionnaire	A self-complete tool for measuring symptoms of hyperventilation.
Non-invasive ventilation (NIV)	Assisted ventilation applied non-invasively via a mask or mouthpiece for spontaneously breathing patients.
Oscillating Positive Expiratory Pressure (OscPEP)	An airway clearance technique which utilises the effects of oscillating “positive expiratory pressure” and oscillating flow, combined with cough or “FET”.
Paced Breathing	Breathing to a rhythm, e.g. in time with walking or stairs, to help maintain control of breathing and thereby reduce dyspnoea.
Peak Cough Flow (PCF)	The peak flow an individual can generate with a cough through a peak flow meter. Used to gauge cough effectiveness in individuals with respiratory muscle weakness.
Percussion	See “Chest clapping/percussion”.
Physical activity	Used to influence breathing pattern, ventilation and ventilation distribution and to preserve physical function and flexibility; sports, activities of daily living (ADL), etc
Physical exercise	Targeted exercise(s) to preserve/improve a specific physical function.
Physical training	A prescribed programme of physical exercise to improve/maintain exercise capacity and endurance, mobility, muscle strength and posture.

Term	Condition
Positioning	The use of different body positions to maintain joint and/or soft tissue length and improve the mechanics of breathing; utilising the effects of gravity to facilitate drainage of bronchial secretions, stimulate both skeletal and smooth muscle postural tone and increase regional ventilation and/or perfusion.
Positive Expiratory Pressure (PEP)	An airway clearance technique which utilises the effects of tidal volume breathing towards an expiratory resistance, combined with FET or cough.
Postural Drainage (PD)	The use of gravity for drainage of secretions guided by bronchial anatomy.
Pursed Lips Breathing (PLB)	The generation of a positive pressure within the airways by expiration against partially closed lips, as in whistling.
Reduced Breathing	A technique using smaller than usual tidal volume and/or lower respiratory rate and increasing relaxation; used for patients with hyperventilation syndrome or stable asthma for control of symptoms; also a key part of the "Buteyko Breathing Technique".
Respiratory Muscle Training (RMT)	Breathing in or out against a resistance as a means enhancing strength or endurance of the inspiratory or expiratory muscles respectively. Can be against a fixed load or via a threshold load resistor.
Reverse Trendelenburg position	Supine position without flexing or extending, with the head higher than the feet.
Rib-springing	A term used to describe a form of "chest compression" with overpressure at the end of expiration to enhance inspiration via stretch reflexes in individuals with weak inspiratory muscles or unable to co-operate.
Rollator frame	A walking frame with wheels back and front for ease of use by breathless patients as it allows fixation of the shoulder girdle.
Self-percussion	Performing "Chest clapping/percussion" independently.
Shaking	See "Chest shaking / vibrations".
Slow, deep breathing	A technique used during exertion/exercise to help maintain control of breathing and reduce dyspnoea in tachypnoeic patients.
Trendelenburg position	Supine position with the feet higher than the head.
Thoracic Expansion Exercise (TEE) (Deep breathing)	Deep inspiration towards inspiratory capacity; the independent means of achieving "MIC".
Vibrations	See "Chest shaking/vibrations".
Visual Analogue Sacle	An arbitrary linear score of 10cm to represent the range of possible symptom perception.

ALGORITHM FOR THE MANAGEMENT OF PATIENTS WITH NEUROMUSCULAR WEAKNESS



INSTRUCTIONS FOR PERFORMING PEAK COUGH FLOW (PCF) MEASUREMENT

Equipment

- Peak flow meter
- Mouth piece
- Full face mask with a good seal for a patient with weak facial muscles

Method

- Select a position of comfort for your patient

Performing Unassisted Peak Cough Flow

- Ask the patient to take a maximal deep breath in
- Ask them to seal their lips tightly round the tube or apply the mask firmly to the face
- Ask them to COUGH as hard as possible into the peak flow mouthpiece or mask

Performing Assisted Peak Cough Flow

- Patients who are able to breath stack (without bulbar weakness)
 - Aim for maximal insufflation capacity (MIC) by breath stacking via either:
 - A volume preset ventilator
 - Glossopharyngeal breathing
 - A manual resuscitator bag
- Patients who are unable to breath stack due to bulbar insufficiency
 - Aim to achieve inspiration to MIC in a single breath by insufflating via either:
 - A volume or pressure preset ventilator
 - A manual resuscitator bag
 - A mechanical insufflator/exsufflator

When the patient has achieved Maximal Insufflation Capacity

- Ask them to seal their lips tightly round the tube or apply the mask firmly to the face
- Ask them to COUGH as hard as possible into the peak flow mouthpiece or mask, with manual or mechanical assistance as required



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