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CLINICAL PRACTICE GUIDELINES

Physiotherapy for cystic fibrosis in Australia and New Zealand: A clinical practice guideline*

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ABSTRACT

Physiotherapy management is a key element of care for people with cystic fibrosis (CF) throughout the lifespan. Although considerable evidence exists to support physiotherapy management of CF, there is documented variation in practice. The aim of this guideline is to optimize the physiotherapy management of people with CF in Australia and New Zealand. A systematic review of the literature in key areas of physiotherapy practice for CF was undertaken. Recommendations were formulated based on National Health and Medical Research Council (Australia) guidelines and considered the quality, quantity and level of the evidence; the consistency of the body of evidence; the likely clinical impact; and applicability to physiotherapy practice in Australia and New Zealand. A total of 30 recommendations were made for airway clearance therapy, inhalation therapy, exercise assessment and training, musculoskeletal management, management of urinary incontinence, managing the newly diagnosed patient with CF, delivery of non-invasive ventilation, and physiotherapy management before and after lung transplantation. These recommendations can be used to underpin the provision of evidence-based physiotherapy care to people with CF in Australia and New Zealand.

Key words: airway clearance, cystic fibrosis, exercise, inhalation therapy, physiotherapy.

Abbreviations: ACBT, active cycle of breathing technique; ACT, airway clearance techniques; AD, Autogenic drainage; CF, cystic fibrosis; CFRD, cystic fibrosis-related diabetes; FEV_1, forced expiratory volume in 1 s; GOR, gastro-oesophageal reflux; PEP, Positive expiratory pressure.

*The extended version of the clinical practice guidelines presented in this paper can be found in the online supplementary information. Any reference to the online supplementary information should cite this Respirology paper as the primary publication.

BACKGROUND

The aim of this Clinical Practice Guideline is to optimize physiotherapy management of people with cystic fibrosis (CF) in Australia and New Zealand. Recommendations for key areas of physiotherapy management are provided, including airway clearance therapy, inhalation therapy, exercise and musculoskeletal management. This Clinical Practice Guideline builds on a previous Consensus Statement for physiotherapy management of CF in Australia[1] and is informed by an evaluation of its uptake and impact.[2]

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METHODS

All physiotherapists who were regularly caring for people with CF across paediatric and adult settings in Australia and New Zealand were invited to participate in guideline development. A systematic literature search was undertaken for each topic area up until June 2014, using MEDLINE, CINAHL, EMBASE and PEDro. Data from each included study were extracted into an evidence table by one reviewer and checked by a second reviewer. Quality was graded according to the NHMRC evidence hierarchy.[3] Recommendations were formulated based on the quality, quantity and level of the evidence; the consistency of the body of evidence; the likely clinical impact; and generalizability and applicability to physiotherapy practice in Australia and New Zealand.[3] Areas of importance to physiotherapy practice, but with insufficient evidence to make recommendations, were highlighted. Updates of each section were prepared by a writing group and circulated to all authors for comment and revision. The draft document was offered to stakeholders for comment, including CF physicians, CF consumers, allied health professionals and physiotherapists who were not part of the writing group.

This guideline provides recommendations for clinical physiotherapy practice and a summary of the evidence that underpins them. More details regarding the evidence underpinning the recommendations and application of the physiotherapy techniques can be found in the Supplementary Appendix S1.

AIRWAY CLEARANCE TECHNIQUES

The aim of airway clearance techniques (ACT) is to clear sputum from the airway, in order to optimize respiratory status and slow disease progression. ACT are frequently described as a 'cornerstone' of CF treatment.[4] A Cochrane review concluded that ACT have short-term beneficial effects on mucus transport in CF.[5] One uncontrolled study evaluated the effects of withdrawing airway clearance for 3 weeks and found a detrimental effect on pulmonary function.[6] Because of ethical concerns regarding withholding such a well-established treatment, it is considered unlikely that more robust controlled trials of the long-term impact of ACT in CF will be conducted.[7]

A number of effective ACT are available. The active cycle of breathing technique (ACBT) consists of breathing control, thoracic expansion exercises and forced expirations.[8] The ACBT is effective for clearance of respiratory secretions[9] with results comparable to other widely used ACT.[10] The ACBT can be performed by all patients who can follow instructions and is useful in all stages of disease. Positive expiratory pressure (PEP) therapy is defined as breathing against a PEP of 10–20 cmH_2O[11] using a mask or mouthpiece. A Cochrane review concluded that PEP was equally effective as other forms of ACT and that patients may prefer PEP.[12] Oscillating PEP combines oscillation of airflow with PEP, in order to loosen secretions.[13] In a 1-year randomised controlled trial (RCT) comparing oscillating PEP with PEP in children with CF, greater deterioration in pulmonary function and more hospitalisations were seen in the oscillating PEP group.[14] However, a more recent 1-year study in adults showed no difference in lung function between groups randomly assigned to PEP or oscillating PEP.[15] Autogenic drainage (AD) uses controlled breathing to achieve the highest possible airflow in different generations of bronchi.[16] Short-term studies have shown that AD is as effective as postural drainage and percussion,[17] oscillating PEP[18] and ACBT.[19] In a long-term comparative study in adolescents with CF, AD was as effective as postural drainage, and participants showed strong preference for AD.[20] Several systematic reviews note that no single ACT is superior,[10,12,21] such that treatment choices should be individualized.

Postural drainage involves use of gravity to drain mucus from the lungs. A number of studies have demonstrated provocation of gastro-oesophageal reflux (GOR) during head-down tilted postural drainage in infants, children and adolescents with CF.[22–24] Two additional studies did not reproduce these results in infants, with no differences in GOR between modified and traditional postural drainage[25,26]; however, the head-down position utilized was not as steep, older infants were studied and they avoided the prone head-down tilted position.[25] Other potential adverse effects of postural drainage with head-down tilt include increased dyspnoea[27] and oxyhaemoglobin desaturation.[28]

Modified postural drainage involves positioning without use of head-down tilt.[22] In a 5-year follow-up of infants randomized to either standard or modified postural drainage, the modified group had fewer radiological changes and significantly better lung function at 6 years of age.[29] In a short-term adult study comparing treatment in head-down versus horizontal positions, there was no difference in the amount of sputum expectorated, but patients reported fewer side effects in horizontal positions.[27]

Physical exercise that increases minute ventilation leads to the mobilization of pulmonary secretions and enhances airway clearance.[30–33] Some people with mild CF lung disease use exercise together with forced expiration and coughing as a stand-alone ACT. Others with more extensive lung disease and larger volumes of sputum use exercise as an adjunct to a formal ACT regimen. Physiological effects of exercise include reduced mechanical impedance of sputum, enhanced expiratory flow rates and inducement of coughing.[34–38] A meta-analysis including three trials found that the addition of exercise to ACT significantly increased forced expiratory volume in 1 s (FEV_1) compared with ACT alone.[39] Whether exercise can be used as an alternative to formal ACT is less clear, with conflicting results across trials.[31,32,38,40]

1 ACT should be performed across the lifespan in CF (C).
2 The ACBT is an effective form of airway clearance and can be used by people with acute and chronic lung disease independently or in conjunction with other ACT (B).

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3 PEP therapy, oscillating PEP and AD are effective forms of airway clearance, which can be performed independently (B).
4 Postural drainage in head-down positions should not be used routinely in infants with CF (B) or in patients of any age with known or suspected GOR (C). Modified postural drainage is recommended in infants and young children where active participation in airway clearance therapy is not possible (B).
5 Physical exercise may be used to reduce mechanical impedance of sputum (B), achieve short-term improvements in pulmonary function (A) and improve ease of expectoration (B).

INHALATION THERAPY

Inhalation therapy is an important treatment for CF respiratory disease. Effective inhalation therapy is integral to the success of ACT and vice versa; as a result, physiotherapists should be adequately skilled in delivery of inhalation therapy in order to maximize the effectiveness of both treatments.

The main determinants of deposition pattern for nebulised medications are the breathing pattern during inhalation, droplet size and age/condition of the lung.[41] A slower breath results in a more desirable peripheral deposition pattern, improved homogeneity of deposition and increased overall drug deposition.[42] Slow steady breaths with occasional deep breaths have traditionally been recommended to promote improved deposition.[43] However, the specific device being utilized may determine the optimal breathing pattern.[44] Given the varying physico-chemical behaviours of the nebulised medications in CF, it is important to use a nebuliser/compressor combination, which is effective for the specific preparation.[41]

Nebulised medication should be taken via a mouthpiece to maximize delivery of the drug to the airways and avoid nasal filtration.[45] Exceptions are in young children who may be unable to use a mouthpiece effectively, or where sinuses are a target of therapy, or those with acute dyspnoea. Bronchodilators should be delivered by metered dose inhaler and spacer[41] except where patients are too dyspnoeic or are unable to follow instructions. Metered dose inhalers with spacer should be used for administration of inhaled corticosteroids. Patients should be encouraged to rinse their mouth with water and gargle afterwards to reduce the risk of thrush.[45]

Combining ACT and inhalation therapy can reduce the time-related burden of care in CF. However, studies have reported conflicting results of this practice on lung deposition, bronchodilation, dyspnoea, cough and mucus production.[46–48] The combination of PEP with inhalation therapy (hypertonic or isotonic saline, salbutamol) is commonly used by physiotherapists. Some also combine inhalation therapy with positioning and breathing techniques such as ACBT or AD. There is insufficient research investigating the combination of inhalation therapy and ACT to make recommendations regarding this practice.

The optimal timing of inhalation of dornase alfa in relation to ACT has been investigated in a Cochrane systematic review.[49] Meta-analysis showed that inhalation of dornase alfa after ACT had similar effects on FEV_1, forced vital capacity and quality of life compared with inhalation prior to ACT. However, forced mid-expiratory flow25% was significantly better with dornase alfa inhalation before ACT, based on the pooled data from two small studies in children with well-preserved pulmonary function.[50,51] A longer time interval between administration of dornase alfa and ACT is more effective than inhalation immediately preceding ACT.[52] There appears to be no detrimental effects on sleep quality or nocturnal cough if this time interval is extended so that dornase alfa is administered before bedtime.[53] In the absence of strong evidence to indicate that one regimen is better than another, the timing of dornase alfa inhalation in relation to ACT or time of day can be based on pragmatic reasons or individual preference.

The timing of hypertonic saline inhalation in relation to ACT (before, during or after) does not appear to have a substantial effect on lung function after a single treatment session.[54] However, participants were more satisfied with the entire treatment session when hypertonic saline was inhaled before or during ACT, and perceived these timing regimens as more effective.

6 Where possible, nebulised medication should be taken via a mouthpiece (C).
7 To optimize dose delivery and treatment time, inhalation technique should be adapted to the device being used, including consideration of body position and concurrent ACT (C).
8 Bronchodilators should be delivered by metered dose inhaler unless there is clinical need for nebulisation (C).
9 Metered dose inhalers with spacer should be used for the administration of inhaled corticosteroids (B).
10 Hypertonic saline may be administered before or during ACT (B).

EXERCISE ASSESSMENT AND TRAINING

Measures of exercise capacity predict survival in children and adults with CF,[55,56] and those with better physical fitness have better quality of life.[57] Structured exercise programmes for people with CF improve fitness and thoracic mobility, maintain bone mineral density and may slow the rate of pulmonary decline.[58–64] A study involving over 200 people with CF conducted over 9 years showed that patients with higher levels of physical activity in daily life (encompassing all activities, not just exercise programmes) had a slower rate of FEV_1 decline than those who were less active.[65]

Exercise testing enables evaluation of exercise capacity, functional capacity, response to treatment and disease progression.[55,66–68] For physiotherapists, exercise tests also provide the basis for exercise prescription. Commonly used field exercise tests in CF are the 6-min walk test, the modified shuttle walk test and the 3-min step test. More details regarding the conduct and choice of exercise test are provided in

Supplementary Appendix S1. The 6-min walk test is considered a particularly important measure during preparation for lung transplantation in CF, providing guidance regarding the timing of referral.[69]

Both aerobic training and anaerobic training are beneficial in CF.[58] Aerobic training results in improved maximum exercise capacity, strength and quality of life.[70,71] Anaerobic training has positive effects on lactate levels, peak power[72] and fat-free mass.[71] Both types of exercise may have positive effects on pulmonary function.[71,73] Exercise programmes have beneficial effects both during admission for acute exacerbations[38,71,74] and for the stable outpatient.[72,73,75] It is unclear whether home-based, unsupervised training programmes are equally as effective as supervised programmes.

The ideal exercise prescription for people with CF has not been established. In the absence of specific guidelines, aerobic exercise prescription should follow the same principles as those used in healthy individuals[76] and patients with other chronic respiratory diseases.[77] Exercise training should occur on at least 3 days (preferably 5 or more days) per week, have a duration of 30 min per session and increase heart rate to 75% of maximum heart rate. A combination of aerobic and resistance training is required to achieve maximum benefits.[58]

Patients with CF may exhibit exercise-induced oxygen desaturation during training, even when pulmonary function is well preserved.[78] Supplemental oxygen during training increases exercise duration[79]; whether this improves clinical outcomes is not clear. Supplemental oxygen is frequently used during training in patients whose oxygen saturation falls below 90% during exercise.[80]

11 Exercise is recommended for people with CF throughout the lifespan (B).
12 An exercise test should be considered to assess response to therapy in the inpatient and outpatient settings and as an assessment tool in the prescription of exercise training programmes (C).
13 A 6-min walk test should be performed as part of the initial assessment for lung transplantation (C).
14 Exercise prescription should be tailored to the individual and comply with recommended exercise guidelines (B).
15 Supplemental oxygen should be considered during training in patients with severe exercise-induced desaturation (C).

MUSCULOSKELETAL COMPLICATIONS OF CF

Musculoskeletal manifestations of CF arise as a result of multifactorial abnormalities in bone mineralization, altered respiratory mechanics and muscular imbalance secondary to pulmonary disease. Between 43% and 94% of individuals with CF experience spinal pain[81–92], which occurs across the spectrum of disease severity. Musculoskeletal pain in CF is associated with decreased quality of life, increased respiratory symptoms, sleep disturbance, anxiety, depression and a reduced ability to perform ACT and exercise.[82,86,88–95]

People with CF have multiple risk factors for inadequate bone mineralisation.[96–100] Longitudinal studies have shown that bone gains during puberty are decreased in CF adolescents compared with healthy controls, resulting in decreased attainment of peak bone mass.[97,98,101–106] A meta-analysis reported that in CF, the prevalence of osteoporosis and osteopenia was 23.5% and 38%, respectively.[107] Individuals with more severe lung disease, decreased physical activity and low aerobic capacity had the lowest bone mineral density, higher prevalence of vertebral fractures and more severe kyphosis.[108]

Weight-bearing exercise is the most effective non-pharmacological method to improve bone mineral density in the healthy population by stimulating bone accretion, preventing bone loss and improving bone structural qualities.[106] As the foundation of bone health begins in childhood and there is some evidence of sustained benefit from early vigorous physical activity,[109,110] children and adolescents should engage in high-impact weight-bearing exercise for 30 min three times a week. The prepubertal and early pubertal years are particularly important to help maximize peak bone mass[101,111,112] as approximately one quarter of peak bone mass is gained in the 2 years around the pubertal growth spurt.[111,113]

Changes in muscle strength, length and neuromuscular recruitment have been demonstrated in CF. Reduced lean muscle mass is associated with malabsorption and deconditioning.[114,115] Peripheral muscle impairment is also noted in response to systemic inflammation and lack of moderate to vigorous physical activity.[116,117] In CF, resistance training programmes of moderate to high intensity and variable duration (19 days to 12 months) have produced significant leg strength gains.[118–120] Strength training for children is still a novel area with protocols not clearly defined; this is an area for future research.[120]

Physiotherapists require the skills to manage a diverse range of musculoskeletal complications including CF-related arthropathy, sports injuries, spinal pain and pain associated with coughing. Early reports regarding the use of manual therapy and exercise in the management of pain and restriction are encouraging.[121,122] Patients with CF-related arthropathy require rheumatological management, which may include physiotherapy interventions targeting pain reduction and muscle strengthening.

16 A musculoskeletal assessment should be included at annual review from approximately age 8 years (prepuberty). Earlier assessment is warranted if pain or functional impairment is reported or bone mineral density risk highlighted (C).
17 Regular physical activity, including weight-bearing exercise, should be encouraged throughout the lifespan in order to optimize bone density (C).
18 Strength training programmes should be prescribed in order to optimize muscle mass (B).

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PHYSIOTHERAPY MANAGEMENT OF THE COMPLEX PATIENT

Cystic fibrosis is a complex multisystem disease, and patients often experience complications and comorbidities that have implications for physiotherapy management. These include haemoptysis, pneumothorax, cystic fibrosis-related diabetes (CFRD) and pregnancy.

There are no published data regarding physiotherapy management of patients with haemoptysis or pneumothorax; however, guidelines based on expert opinion are available.[123] When haemoptysis is present, the physiotherapists aim to maintain adequate airway clearance and exercise regimens whilst promoting vessel healing and minimizing the risk of re-bleeding. When a pneumothorax is present, physiotherapists aim to ensure that adequate airway clearance continues whilst minimizing the amount of positive pressure generated inside the patient's lungs. Both of these situations may require alteration to usual ACT.

Cystic fibrosis-related diabetes is a frequent comorbidity in CF, occurring in 5–30% of patients. The American Diabetes Association Clinical Care Guidelines for CFRD[124] state that people with CFRD should perform moderate aerobic exercise for at least 150 min per week, should monitor blood glucose levels before vigorous physical activity and may need to consume extra carbohydrate or alter their insulin dose (level of evidence – expert opinion). During periods of acute illness or courses of corticosteroids, blood sugar levels and insulin requirements may be altered and more careful monitoring required.[124]

Many pregnancy-related physiological changes have implications for optimal physiotherapy care, although little research is available. Women with CF are encouraged to approach pregnancy with a regular ACT routine. Head-down tilted postural drainage should be avoided, along with any ACT that exacerbate nausea. Upright sitting is usually the most comfortable position for airway clearance. Modifications to exercise programmes may be required to accommodate musculoskeletal, respiratory and cardiac changes. Maintenance of adequate hydration during exercise should be emphasized. It is important to consider the need for domestic support during pregnancy and afterwards, to provide sufficient time for regular airway clearance, inhalation therapy and exercise.[125]

PHYSIOTHERAPY MANAGEMENT OF CONTINENCE

The reported prevalence of urinary incontinence in girls and women with CF ranges from 22% to 74%[126–132] in comparison with 13% in healthy women of similar age.[133] There is limited literature in adult men with CF, with a reported prevalence of 8–15%, compared with 7.5% in healthy men.[132,134,135] It is not known whether the cause of urinary incontinence in CF is chronic cough, loading of the pelvic floor during ACT, coughing and physical exercise, or underlying structural differences. People with CF and incontinence report increased anxiety and depression and a negative impact on quality of life.[135,136]

Screening for incontinence should be part of routine physiotherapy care for both male and female patients. Treatment of urinary incontinence in women with CF by a continence physiotherapist with exercise, electrical stimulation, biofeedback and bladder training results in improvements in pelvic floor strength, reduction in leakage and improvement in quality of life.[137] Positive outcomes have also been demonstrated with surgical correction of severe urinary incontinence in women with CF.[138] In order to optimize pelvic floor function, patients should be taught to perform ACT in positions that maintain a neutral lumbar spine,[139] with addition of perineal support in those with urinary incontinence.

19 Women with CF and symptoms of stress urinary incontinence should be taught rehabilitative strength and endurance exercises to provide better control of the pelvic floor (C).
20 Men and women with CF should be screened for symptoms of stress urinary incontinence (C).
21 Airway clearance should take place in postures that maintain a neutral lumbar spine, to optimize pelvic floor function (C).

PHYSIOTHERAPY MANAGEMENT OF THE NEWLY DIAGNOSED PATIENT

Newly diagnosed infants and their families should meet with the CF multidisciplinary team soon after diagnosis.[140,141] The role of physiotherapy in ACT, exercise and active play should be explained, demonstrated and practised.[142,143] Treatment of infants should follow the usual guidelines for physiotherapy in CF, consisting of five modified postural drainage positions performed one to two times daily as appropriate.[22,24,29] In each position, percussion or thoracic compressions should be performed for 3–5 min. Other techniques such as infant PEP or assisted AD can also be introduced.[144] Normal developmental play and prone lying should be encouraged as the first steps towards an active physical lifestyle and routine.[144,145]

Adults and children with a new diagnosis of CF usually have milder disease than those diagnosed in infancy.[142] Some newly diagnosed adults are very well; therefore, physical exercise, huffing and coughing may be appropriate as stand-alone ACT.[34] Those patients with established lung disease should be taught appropriate ACT and exercise regimens and educated about the role of nebulised drugs in their treatment.

22 Treatment for newly diagnosed infants may include percussion for 3–5 min in each of five modified postural drainage positions (B) and daily age-appropriate physical play (C).
23 Physiotherapy treatment for the newly diagnosed child and adult should include regular physical exercise (B); other forms of airway clearance therapy should be added as required (C).

NON-INVASIVE VENTILATION FOR CF

A number of descriptive studies report the successful use of non-invasive ventilation (NIV) to stabilize patients with CF and acute respiratory failure,[146–152] with reduced hypercapnia, respiratory rate and dyspnoea. Although NIV does not reverse the respiratory deterioration inherent in end-stage disease, it may allow the patient to be stabilized for long enough for donor lungs to become available for transplantation. The use of NIV for CF patients who are not awaiting lung transplantation has also been reported[151] where it may be useful for palliation of dyspnoea in end-stage disease.

Hypoxia and hypercapnia occur commonly during sleep in moderate to severe CF[153,154] and may result in daytime respiratory failure.[155] Positive short-term effects of NIV during sleep in CF have been reported.[156,157] Longer-term outcomes of NIV for chronic respiratory failure may include improvements in daytime PaCO2, reduction in the number of days spent in hospital and improvement in symptoms.[158,159] In a randomized controlled trial, domiciliary NIV over a 6-week period resulted in significant improvements in quality of life, respiratory symptom scores, dyspnoea, nocturnal ventilation and increased exercise performance.[160]

Airway clearance techniques are onerous for patients who are unwell, because of increased ventilatory demand,[161] alterations in gas exchange[28,162] and dyspnoea.[27] Two randomized crossover trials report that a single session of NIV can unload the respiratory muscles during ACT in both adults and children with CF, resulting in decreased dyspnoea and less desaturation.[162,163] Similar effects have been observed with the application of NIV during exercise, including improved ventilation, reduced desaturation and increased functional walking performance.[164] This may be useful in patients bridging to transplantation, in whom maintenance of exercise capacity is an important goal.

Non-invasive ventilation delivers air at high flow rates and low relative humidity, which may overwhelm the capacity of the upper airway mucosa to warm and humidify inspired air.[165] Humidity levels during NIV are low enough to cause airway drying.[166] This is of greatest concern in patients with excessive secretions, who are at high risk of sputum retention. Hence, consideration should be given to heated humidification when NIV is used in CF.

24 NIV should be considered in all patients with acute respiratory failure who are listed for transplantation (C).
25 In patients with symptomatic nocturnal ventilatory failure, a trial of nocturnal NIV may be undertaken (B).
26 NIV is a useful adjunct to airway clearance in patients with severe disease in whom dyspnoea and fatigue limit effective airway clearance (B).
27 NIV may be a useful adjunct to exercise in patients with severe disease where dyspnoea and fatigue contribute to deconditioning and limit effective training (B).
28 Heated humidification should be incorporated into the circuit for all applications of NIV in CF (C).

PHYSIOTHERAPY AND LUNG TRANSPLANTATION

There are many systemic features of CF that have the potential to impact on lung transplant suitability and outcomes, including skeletal muscle weakness[167] and poor bone health.[63] Most adult transplant centres offer dedicated preoperative exercise training classes for transplant candidates, in order to optimize physical fitness and strength. A recent large, retrospective study that included 70 people with CF[168] showed that 6-min walk distance was well maintained from listing to transplantation in those who undertook thrice-weekly supervised exercise training. Furthermore, those with a greater 6-min walk distance prior to transplantation had a shorter hospital stay post-transplantation.

Exercise rehabilitation is an established therapy for lung transplant recipients.[169] Although studies in CF are uncontrolled, 3 months of post-transplant rehabilitation has been associated with improvements in functional exercise capacity, strength and quality of life in adults[170] and children.[171] A recent randomized controlled trial of 3 months of rehabilitation in lung transplantation recipients with other respiratory disorders, performed immediately following hospital discharge, showed significant improvements in daily physical activity, quadriceps force and exercise performance at 1 year following transplantation.[172] The content of post-transplantation rehabilitation programmes generally includes aerobic and resistance exercise, performed at least three times per week.[169] Although most rehabilitation takes place in a group setting, patients with resistant organisms may require isolation from other immune-suppressed patients.

29 Patients with CF should undertake an exercise programme designed to optimize their physical function whilst on the transplant waiting list (C).
30 Patients with CF who have undergone lung transplantation should participate in a formal, supervised rehabilitation programme post-operatively (B).

END OF LIFE CARE

There is no published literature specifically addressing physiotherapy treatment in the terminal stages of CF. However, many people with CF die of respiratory failure,[173] and physiotherapists are often involved with provision of end of life care. The aims of physiotherapy treatment will be influenced by whether the patient is actively waiting for transplantation. Care should focus on comfort and dignity and be tailored to each patient's goals and values.[174] Minimizing the work of breathing during ACT is an important consideration in those approaching the end of life. Some patients may require therapist-assisted ACT such as percussion or thoracic compressions. Airway clearance may be continued even in the palliative stage if the patient finds it beneficial to relieve symptoms. Comfort

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measures such as soft tissue massage and positioning can also be considered.

INFECTION CONTROL

Respiratory pathogens have a significant impact on morbidity and mortality in CF,[175] and good infection control practices are critical to preventing their transmission. Segregation and cohorting of inpatients and outpatients according to respiratory organisms are now routinely practised.[176,177] Physiotherapists are encouraged to be familiar with their local infection control policies. Because of considerable variation in local policies, formal recommendations are not provided here.

Both *Pseudomonas aeruginosa* and *Burkholderia cepacia* may be spread in droplet form by coughing and can survive on dry surfaces for a number of days.[178–180] There is also potential for airborne transmission.[181] *B. cepacia* has been isolated from the hospital rooms and hands of patients following airway clearance[182–184] and the outside surfaces of sputum cups.[182] These findings reinforce the need to segregate patients whilst performing ACT and inhalation therapy, as well as the importance of hand washing. Stethoscopes should be cleaned with alcohol wipes between patients.[185]

Bacterial contamination of home nebulisers has been documented, and sharing equipment has been associated with transmission of *B. cepacia*.[186–188] Under no circumstances should any respiratory equipment be shared between patients with CF. There is no consensus regarding the use of gloves, gowns and masks during physiotherapy treatment in CF. Physiotherapists should consult their local infection control policy with regard to when these measures are required.

Coughing is common during exercise, and droplet spread of organisms is possible. These droplets may be transmitted within 1 m of an infected patient.[178] It has been reported that contamination can still occur between 1 and 2 m, albeit with lower probability (1.7%).[189] As a result, patients with different organisms, or in different cohorts, do not exercise together. When people considered suitable for cohorting are sharing the gym, universal precautions should be practised. Patients should be educated to maintain a 2-m distance from other patients at all times, hands should be washed on entering and leaving the gym and patients should be taught to wipe down all exercise equipment with an alcohol-based solution before and after use.

PHYSIOTHERAPY SERVICES FOR CF

There is no published research regarding the optimum structure of physiotherapy services for people with CF. For inpatients, expert clinical opinion suggests that physiotherapy assessment and treatment starts on the day of hospital admission.[190] The physiotherapy treatment plan should address inhalation therapy, ACT and exercise. Patients admitted with an acute exacerbation with increased and/or retained secretions will need to carry out more frequent ACT sessions than their baseline daily regimen. The number of treatments will range from two to three or more treatments in 24 h. A graduated physical exercise programme incorporating cardiorespiratory exercise should be commenced as soon as possible.

Patients attending the outpatient department of a Cystic Fibrosis Service should have access to a physiotherapist with expertise in CF management at each clinic visit. It is suggested that each patient be assessed three to six monthly so that their physiotherapy programme can be reviewed and optimized. Complex patients may require more frequent and detailed review. A formal annual review by the CF team, including physiotherapy review, has been advocated for people with CF.[190,191]

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Supplementary Information

Additional Supplementary Information can be accessed via the *html* version of this article at the publisher's website:

Appendix S1 *Physiotherapy for Cystic Fibrosis in Australia and New Zealand: A Clinical Practice Guideline - comprehensive version*.
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