TUGAS AKHIR FISIOTERAPI KARDIOPULMONAL

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PROGRAM STUDI FISIOTERAPI

FAKULTAS ILMU KESEHATAN

UNIVERSITAS 'AISYIYAH YOGYAKARTA

Resume Jurnal

1. Physical Therapy Management in Swine-Influenza A (Indian H1N1) Virus – A Case Study

Influenza adalah virus umum pada manusia yang dapat menyebabkan berbagai tingkat infeksi pernapasan mulai dari flu ringan hingga pneumonia berat yang mengancam jiwa, bronkitis, sindrom gangguan pernapasan akut, dan bahkan kematian. H1N1 juga dikenal sebagai "flu babi" dan merupakan virus Influenza A yang berevolusi melalui reassortment genetik. Ini menyebabkan epidemi tahunan dengan tingkat keparahan yang berbeda-beda. Pandemi H1N1 pertama kali muncul pada April 2009, yang dimulai di Meksiko dan segera meluas secara global. Di India, kasus pertama teridentifikasi pada 16 Mei 2009 di Hyderabad. Gejala yang muncul adalah batuk, sakit tenggorokan, flu biasa, demam, sesak napas, sakit kepala, nyeri dada, sputum, muntah, dan hemoptisis pada sebagian besar kasus. Peningkatan laju pernapasan (takipnea), saturasi oksigen rendah (hipoksia), jumlah leukosit dalam darah yang lebih rendah dan juga infliltrasi paru yang diikuti dengan konsolidasi.

Pasien yang dipilih untuk penelitian adalah pasien laki-laki berusia 35 tahun, direkrut dari Casualty ICU dan Day Care Unit Pt.BDS, UHS, Rohtak. Pasien secara klinis didiagnosis dengan virus Swine-Influenza A (Indian H1N1). Pasien diberikan perawatan fisioterapi dengan chest dan limb physiotherapy yang dilakukan dua kali dalam sehari. Manajemen fisioterapi kepada pasien yaitu sebagai berikut :

1. Posisi pasien : postural drainage pada paru kiri lalu lakukan pada paru kanan.

2. Chest Physiotherapy : breathing exercise, bronchial hyiegene therapy dan cough training

3. Limb Exercise : latihan aktif dan pasif

4. Bed Positioning : posisi pasien diubah setiap 2-3 jam untuk mengurangi decubitus dan menjaga mobilitas pasien

Pasien dengan PEEP level 8 cmH2O. diberikan intervensi chest physiotherapy berupa breathing exercise, pembersihan sekresi dengan postural drainage, perkusi dan

segmental vibrasi. Pada hari kesembilan PEEP level menjadi 6 cmH2O lalu pada hari berikutnya pasien di extubasi. Selanjutnya, fisioterapi memberikan cough training dan chest wall mobilization exercises. Hingga 20 hari kemudian pasien sudah dapat bernafas secara normal dengan sendirinya.

Dari studi ini, dapat disimpulkan bahwa physical therapy exercise program efektif diberikan kepada pasien yang mengalami swine-influenza A (Indian H1N1) virus dan juga mempercepat pemulihan.

ORIGINAL RESEARCH PAPER

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PHYSICAL THERAPY MANAGEMENT IN SWINE-INFLUENZA A (INDIAN H1N1) VIRUS –A CASE STUDY

Physiotherapy

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ABSTRACT

Background:- H1N1 is also known as "swine flu" and it is a novel strain of Influenza A virus that evolved by genetic reassortment. It leads to annual epidemics of varying severity. It causes severe illness requiring hospitalization, including severe pneumonia, hypoxemia, lung injury with acute respiratory distress and complications involving renal, liver and cardiac dysfunction. Fever and cough are most common presenting symptoms. Mortality rate is high in H1N1-infected patients with development of ARDS, and patients who require ventilatory support.

Case Report: - We report a case of a young male patient suffering from H1N1 is also known as "swine flu". The patient had a severe infiltration in lungs and was mechanically ventilated on admission. Progressive chest and limb physiotherapy was given to the patient. Physical therapy intervention improved respiratory statuts of the patient as well as aided speedy recovery from this chronic ailment

KEYWORDS

Introduction

Influenza is a common human virus which can lead to varying degree of respiratory infections ranging from mild flu to severe and life-threatening pneumonia, bronchitis, acute respiratory distress syndrome, and even death. H1N1 is also known as "swine flu" and it is a novel strain of Influenza A virus that evolved by genetic reassortment. It leads to annual epidemics of varying severity. Pandemic of H1N1 first emerged in 2009 April, which was started in Mexico and soon expanded globally. In India, the first case was identified on 2009 May 16, in Hyderabad.[1]

The symptoms on presentation are cough, throat pain, common cold, fever, breathlessness, headache, chest pain, sputum production, vomiting, and hemoptysis in majority of cases. Increased respiratory rate (tachypnea), low oxygen saturation (hypoxia), WBC count within normal range, and lower level of procalcitonin in blood are also commonly found. [1]. Commonest radiological presentation on x-ray chest is pulmonary infiltration followed by consolidation.[1]

H1N1 influenza causes severe illness requiring hospitalization, including severe pneumonia, hypoxemia, lung injury with acute respiratory distress and complications involving renal, liver and cardiac dysfunction. Maximum patients are between age group of 20-40 year. Fever and cough are most common presenting symptoms. Common comorbidities are pregnancy, diabetes, hypertension, and obesity. Mortality rate is high in H1N1-infected patients with development of ARDS, associated pregnancy and patients who require ventilatory support.[1-2]

The severe illness and deaths associated with seasonal influenza/ swine influenza infection epidemics are in large part the result of secondary complications, including primary viral pneumonia, secondary bacterial pneumonia (particularly with group A Streptococcus, Staphylococcus aureus, and Strep. pneumoniae) and exacerbations of underlying chronic conditions.[3,4-5]

A diagnosis of confirmed swine flu requires laboratory testing of a respiratory sample (a simple nose and throat swab).[3]

There are very limited number of studies on influenza virus-related diseases and morbidity in India[1].

Present study is an effort to reveal the predictors of mortality for better preparation to handle any such epidemics in future.

There is dearth of literature regarding physical therapy evaluation and management of patients suffering from Swine-Influenza A (Indian H1N1) virus. We present our experience with physical therapy management of a patient suffering with Swine-Influenza A (Indian H1N1) virus with an aim to improve his respiratory status, remove retained secretions and help the patient to recover from illness through physical therapy regime.

Patient Information and Clinical findings:-

The patient chosen for study was a 35 year old male patient, recruited from the Casualty ICU and Day Care Unit of Pt.B.D.S., U.H.S., Rohtak. Patient was clinically diagnosed with Swine-Influenza A (Indian H1N1) virus. Detailed history and physical examination, including the onset and nature of symptoms, general and systemic manifestations was recorded for the patient.

The patient was evaluated thoroughly using an evaluation performa. Primary symptoms of the patient included fever, shortness of breath, cough with expectoration, chest crepitation with atelectasis in both lungs (left lung more than right).

Therapeutic Intervention:-

The patient was given physiotherapeutic treatment including chest and limb physiotherapy. Measurements were taken at two time intervals, one at the time of admission of patient (PRE) and second after ten days of admission (POST). Effects of physiotherapy treatment were studied on dependent variables including static lung compliance (C_{ST}), oxygenation ratio (PaO2:FiO2 ratio), partial pressure of carbon dioxide in arterial blood ($PaCO_2$) , cologarithm of activity of dissolved hydrogen ions in arterial blood (pH) and chest X-rays.

Protocol

The patient was given physical therapy exercise management, which included the following exercise protocols:-

1. Position of Patient- Patient had primarily involvement of left lung

and hereby was positioned accordingly; then postural drainage was being done first for left lung and then for the right lung.

2. Chest Physiotherapy:-Patients was given Breathing exercises, also bronchial hygiene therapy and cough training for clearance of chest secretions.
3. Limbs Exercise:- Limb movements were done including both active and passive range of motion exercises for maintaining joint mobility.
4. Patient bed positioning: - Position of the patients was changed every 2-3 hourly to prevent bed sores and maintain mobility.

Patient's Daily Treatment and Changes in Intervention:-

Initially at the time of admission, the patient was on Pressure control mode with a PEEP level of 8 cmH_2O. He was being given chest physiotherapy in form of breathing exercises, secretion clearance through postural drainage, percussion and segmental vibration techniques along with suction for secretion clearance. After seven days of admission, patient was weaned off to pressure support mode and on 9^{th} day of admission patient was on PEEP level of 6 cmH_2O. On the 10^{th} day of admission patient was extubated and taken on T-piece. Further, non invasive ventilation was given by Hudson mask. The physical therapy regime was progressed to cough training and chest wall mobilization exercises. On further progression to O_2support with nasal prongs and oral diet being allowed to semi solid, the patient was made to walk around the ward with support. The Chest physiotherapy regime was progressed in frequency and repetition including the above mentioned therapeutic interventions and rigorous cough training was given to the patient. Finally, patient was discharged twenty days post admission with normal spontaneous breathing at its own.

Frequency and Repetitions: -

Chest and limb physiotherapy was done twice a day.

Follow-up and Outcomes

Post intervention measures of the variables were recorded at the time of admission and after 15 days of admission. Static lung compliance (CST) readings were recorded from the display on the ventilator. An average of three readings of static pulmonary compliance was taken.Arterial blood gas analysis samples were taken to monitor oxygenation(PaO2:FiO2),partial pressure of carbon dioxide in arterial blood (PaCO2) and cologarithm of activity of dissolved hydrogen ions in arterial blood (pH) (Table 1).

Table 1:- Comparison of before and after intervention

Variables	Time		% improvement
	Pre (before intervention)	Post (after intervention)	
pH	7.38	7.35	-0.41%
PaO_2:FiO_2 Ratio	356.6667	462.6667	29.72%
Cstat (mL/cmH2O)	39.4	47.1	19.54%
$PaCO_2$ (mm Hg)	54.6	33.6	-38.4%

There was 29.72% improvement in the PaO_2:FiO_2 Ratio, 19.54% improvement in the Cstat (mL/cmH2O) values, a reduction by -0.41% in pH values, and a reduction by -38.4% in the $PaCO_2$ (mm Hg) values. All the parameters showed clinical improvement.

Chest X-rays showed marked changes from baseline (Fig.1a and Fig.1b).

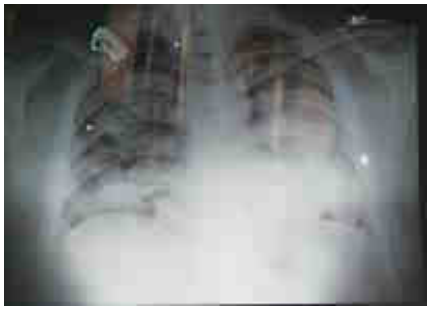

Fig.1 (A):- on the day of admission

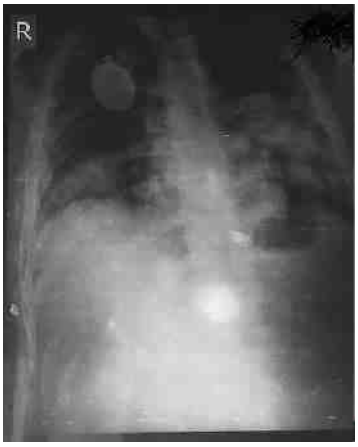

Fig.1 (B):- Two weeks post admission

Discussion

H1N1 influenza causes severe illness requiring hospitalization, including severe pneumonia, hypoxemia, lung injury with acute respiratory distress and complications involving renal, liver and cardiac dysfunction.[1-2]

Respiratory failure being major reason for mortality, careful monitoring, and appropriate management and early recognition of this complication may decrease the mortality rate among these patients. Physical therapy may be indicated for patients in intensive care setting when they have retained secretions and radiological evidence of atelectasis or infiltrate. In addition, mechanically ventilated patients are at risk for retained secretions due to endotracheal intubation disrupting mucociliary escalator, relative immobility of mechanically patient confined to bed can lead to atelectasis, impaired cough, and retained secretions., Physical therapy interventions in patients admitted in ICU's include postural drainage, breathing exercises, percussion, vibration, manual hyperinflation, coughing, huffing, and suction.[4-5]

C_{ST} is considered as important clinical outcome measure, and may be used to predict mortality in patients with respiratory failure. In present study, there was improvement in static lung compliance, after intervention. The application of chest physiotherapy techniques may have facilitated collateral ventilation and effective recruitment of alveoli, thereby improving time-dependent elastic behavior of the lung[6-7].

In addition, PaO2:FiO2 ratio mean values showed percentage improvement and also there was reduction in PaCO2 and pH values post admission. The probable reason is that with increase in the recruitment of functional alveolar units after intervention, there may have been an improvement in ventilation- perfusion ratio; decreased shunting of blood in lungs and improved oxygen transport in blood.[8]

Conclusion

The present study concludes that the physical therapy exercise programme is effective in patients suffering from Swine-Influenza A (Indian H1N1) virus. Physical therapy intervention not only improves respiratory status in patients suffering with Swine-Influenza A (Indian H1N1) virus but also aids in speedy recovery from this chronic ailment.

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2. Effectiveness of physiotherapy on quality of life in children with asthma

Asma adalah penyakit paru-paru kronis yang paling umum di masa kanak-kanak, ditandai dengan hyperactivity dan airflow obstruction. Di seluruh dunia, asma mempengaruhi sekitar 300 juta orang dan prevalensinya masih meningkat, terutama pada anak-anak. Di Amerika Serikat, ada sekitar 6 juta anak yang terkena asma, yang menyebabkan beban berat pada healthcare system. Asma disebabkan oleh faktor lingkungan, paparan rokok, infeksi virus, paparan allergen biologis, selain itu bisa juga karena gen seperti GATA3,KIAA1109 dan MUC5AC yang diidentifikasi bisa menyebabkan asma sedang hingga berat.

Fisioterapi diterapkan untuk anak – anak penderita asma dengan tujuan meningkatkan fungsi paru – paru dan kualitas hidup mereka dengan physical training, breathing exercise dan IMT (inspiratory muscle training yang dilakukan selama kurang lebih 2 minggu.

Untuk evaluasi menggunakan Pediatric Asthma Quality of Life Questionnaire (PAQLQ) yang terdiri dari gejala, keterbatasan aktivitas, dan fungsi emosional kemudian hasil dari evaluasi akan dilaporkan.

Intervensi fisioterapi seperti physical training telah terbukti bermanfaat khusus untuk anak-anak yang sehat dengan meningkatkan muskuloskeletal dan kesehatan mental. Namun, efek pada anak penderita asma masih belum pasti.

Study Protocol Systematic Review

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Effectiveness of physiotherapy on quality of life in children with asthma

Study protocol for a systematic review and meta-analysis

Weijian Zhang, MD[a,b], Lilong Liu, MM[c], Wenhao Yang, MM[a,b], Hanmin Liu, MD[a,b,*]

Abstract

Background: Asthma is the most common chronic lung disease in childhood, leading to a great burden to the healthcare system worldwide. Despite the medication treatment, physiotherapy is now applied for asthmatic children aiming to improve their lung function and life quality. However, the effectiveness of physiotherapy on quality of life (QoL) in children with asthma is not clear. We are aiming to perform this study to provide some evidence to doctors on asthma treatment.

Methods: PubMed, Embase and the Cochrane Library will be searched from their inception to 31 March 2019 for randomized controlled trials (RCTs) published in English, which investigated the effectiveness of physiotherapy on QoL in children with asthma. Besides, additional studies will be searched by scanning the reference lists of studies and the relevant systematic reviews.

Two authors will select the studies, extract the data, and assess the risk of bias independently. Data synthesis and statistical analysis will be performed in Review manager 5.3. Stata 14.0 will be used to assess the reporting bias. Quality of evidence will be evaluated based on the Grading of Recommendations Assessment, Development, and Evaluation (GRADE) system.

Results: The results will provide information on the effectiveness of physiotherapy on QoL in children with asthma and further demonstrate which physiotherapy is more effective and which domain of QoL could be improved significantly.

Conclusion: The findings of this study will provide the latest evidence on the effectiveness of physiotherapy on QoL in children with asthma.

Prospero Registration Number: CRD42019133181.

Abbreviations: CI = confidence interval, GRADE = Grading of Recommendations, Assessment Development and Evaluation, IMT = inspiratory muscle training, MD = mean difference, PAQLQ = Pediatric Asthma Quality of Life Questionnaire, PRISMA = Preferred Reporting Items for Systematic Reviews and Meta-Analyses, QoL = quality of life, RCTs = randomized controlled trials.

Keywords: asthma, children, physiotherapy, quality of life

1. Introduction

Asthma is the most common chronic lung disease in childhood, characterized by airway hyperreactivity and airflow obstruction.[1,2] Worldwide, asthma affects an estimated 300 million people and the prevalence is still increasing, especially in children.[3] In the United State, there are around 6 million children affected by asthma, leading to a great burden on healthcare system.[4] Etiology of asthma is multiple and some environmental factors related to asthma have been reported, including virus infection,[5] smoking exposure,[6] particulate matter, and biological allergens exposure,[7] etc. Besides, genes, such as GATA3, KIAA1109, and MUC5AC, were identified associated with moderate-to-severe asthma.[8]

The asthma care consists of 4 important components according to the current guideline, including assessment and monitoring, education, control of environmental factors and comorbid conditions, and pharmacologic treatment.[9,10] Despite the care mentioned above, physiotherapy is now applied for children with asthma aiming to improve their lung function and life quality. The widely used physiotherapies for asthma are physical training, breathing exercises, and IMT.[11]

Nevertheless, the effectiveness of physiotherapy on QoL in children with asthma is not clear. Moreover, no systematic review and meta-analysis was conducted to compare the 3 widely used physiotherapies for asthmatic children. Therefore, we are planning to perform this study to solve these problems.

2. Methods

2.1. Registration

We have registered this study protocol at the International Prospective Register of Systematic Reviews (registration number: CRD42019133181). The *Cochrane Handbook for Systematic*

WJ Zhang and LL Liu are co-first authors who contributed equally to this study.

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The authors have no conflicts of interest to disclose.

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http://dx.doi.org/10.1097/MD.0000000000016195

Table 1

Preliminary search strategy in PubMed.

Search	Query
1	"Asthma"[Mesh] OR asthma*[Title/Abstract]
2	"Child"[Mesh] OR children[Title/Abstract] OR school age[Title/Abstract] OR "Adolescent"[Mesh] OR adolescen*[Title/Abstract] OR teen*[Title/Abstract] OR youth*[Title/Abstract]
3	#1 AND #2
4	physiotherapy[Title/Abstract] OR physical therapy[Title/Abstract] OR physical intervention[Title/Abstract] OR physical rehabilitation[Title/Abstract] OR pulmonary therapy[Title/Abstract] OR pulmonary intervention[Title/Abstract] OR pulmonary rehabilitation[Title/Abstract] OR respiratory therapy[Title/Abstract] OR respiratory intervention[Title/Abstract] OR respiratory rehabilitation[Title/Abstract]
5	(breath*[Title/Abstract]) AND (exercise*[Title/Abstract] OR train*[Title/Abstract] OR retrain*[Title/Abstract] OR educat*[Title/Abstract] OR re-educat*[Title/Abstract] OR physiotherap*[Title/Abstract] OR physical therap*[Title/Abstract] OR respiratory therap*[Title/Abstract] OR buteyko[Title/Abstract])
6	IMT[Title/Abstract] OR inspiratory muscle train*[Title/Abstract] OR inspiratory muscle strength[Title/Abstract] OR inspiratory muscle endurance[Title/Abstract] OR respiratory muscle train*[Title/Abstract] OR respiratory muscle strength[Title/Abstract] OR respiratory muscle endurance[Title/Abstract]
7	physical training[Title/Abstract] OR physical activity[Title/Abstract] OR rehabilitat*[Title/Abstract] OR exercis*[Title/Abstract] OR fitness*[Title/Abstract] OR train*[Title/Abstract] OR aerobic[Title/Abstract] OR swim*[Title/Abstract] OR bik*[Title/Abstract] OR joy*[Title/Abstract] OR walk*[Title/Abstract] OR run*[Title/Abstract] OR sport*[Title/Abstract] OR danc*[Title/Abstract] OR motor[Title/Abstract]
8	#4 OR #5 OR #6 OR #7
9	quality of life[Title/Abstract] OR QoL[Title/Abstract] OR pediatric asthma quality of life questionnaire[Title/Abstract] OR PAQLQ[Title/Abstract]
10	(((((((randomized controlled trial [pt]) OR controlled clinical trial [pt]) OR randomized [tiab]) OR placebo [tiab]) OR groups [tiab]) OR randomly [tiab]) OR trial [tiab])
11	#3 AND #8 AND #9 AND #10

Reviews of Interventions[12] will be used as a guideline to solve any problem when performing the systematic review and meta-analysis. After finishing the study, We will report it according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement.[13] No ethical statement will be required due to no direct involvement of human in this study.

2.2. Eligibility criteria

2.2.1. Types of studies. Only RCTs will be included. Those studies should be published in English before or on 31 March 2019. We will exclude the study not written in English due to language bias.

2.2.2. Types of participants. The population included should be less than 18 years old without gender or ethnicity limitations. We will exclude the studies with participants aged ≥18 years old. The diagnosis of asthma should be clearly defined on all participants in the included studies.

2.2.3. Types of interventions. Physiotherapy intervention for asthma includes physical training, breathing exercises, and inspiratory muscle training (IMT). We will not consider the studies with interventions concerning pharmacology, psychology, or behaviors. Physiotherapy should be conducted for ≥2 weeks.

2.2.4. Types of outcome measures. QoL will be compared between the experimental groups and the control groups. In order to evaluate the life quality, Pediatric Asthma Quality of Life Questionnaire (PAQLQ) should be used in studies and the score should be reported. PAQLQ consists of 3 domains, including symptoms, activity limitations, and emotional function.

2.3. Search methods

PubMed, Embase, and the Cochrane Library will be searched from their inception to 31 March 2018. The search strategy will involve the following terms: child, asthma, physiotherapy, QoL, and RCTs. The preliminary search strategy in PubMed is shown in Table 1. We will use the same search strategy for search in Embase and the Cochrane Library in accordance with different specific requirements. Additional studies will also be searched by scanning the reference lists in included studies and the relevant systematic reviews.

2.4. Study selection and data extraction

2.4.1. Study selection. Two authors will conduct study selection independently. The records identified through database searching and other sources will be sent to Endnote X7. After duplicates removed, records will be screened by reading the title and abstract first. If records not excluded, full-text articles assessed for eligibility will be necessary. We will include a study based on the eligibility criteria. Reasons for full-text articles excluded should be noted. Any dispute between the 2 authors will be solved by discussion or a third author. The study selection process will be summarized and reported in a PRISMA flow diagram (Fig. 1).

2.4.2. Data extraction. Data will be extracted from each included study by 2 authors independently, including study characteristics (author, publication year, country, etc.), participant characteristics (number, age, severity of asthma, etc.), type and duration of intervention, score of QoL (symptoms, activity limitations and emotional function domain irrespectively). After data extracted, 2 authors will check the result with each other and any different data will be re-extracted from the study and re-checked. If different opinions between the 2 authors cannot reach an agreement, a third author will be sought for help. If data is not complete, we will contact the author. If we cannot obtain the data still, we will exclude the study depending on whether the existing data could be transformed.

2.5. Risk of bias assessment

The methodological quality of all included studies will be evaluated independently by 2 authors in accordance with the Cochrane Collaboration's tool.[14] The following aspects will be assessed: selection bias (random sequence generation and

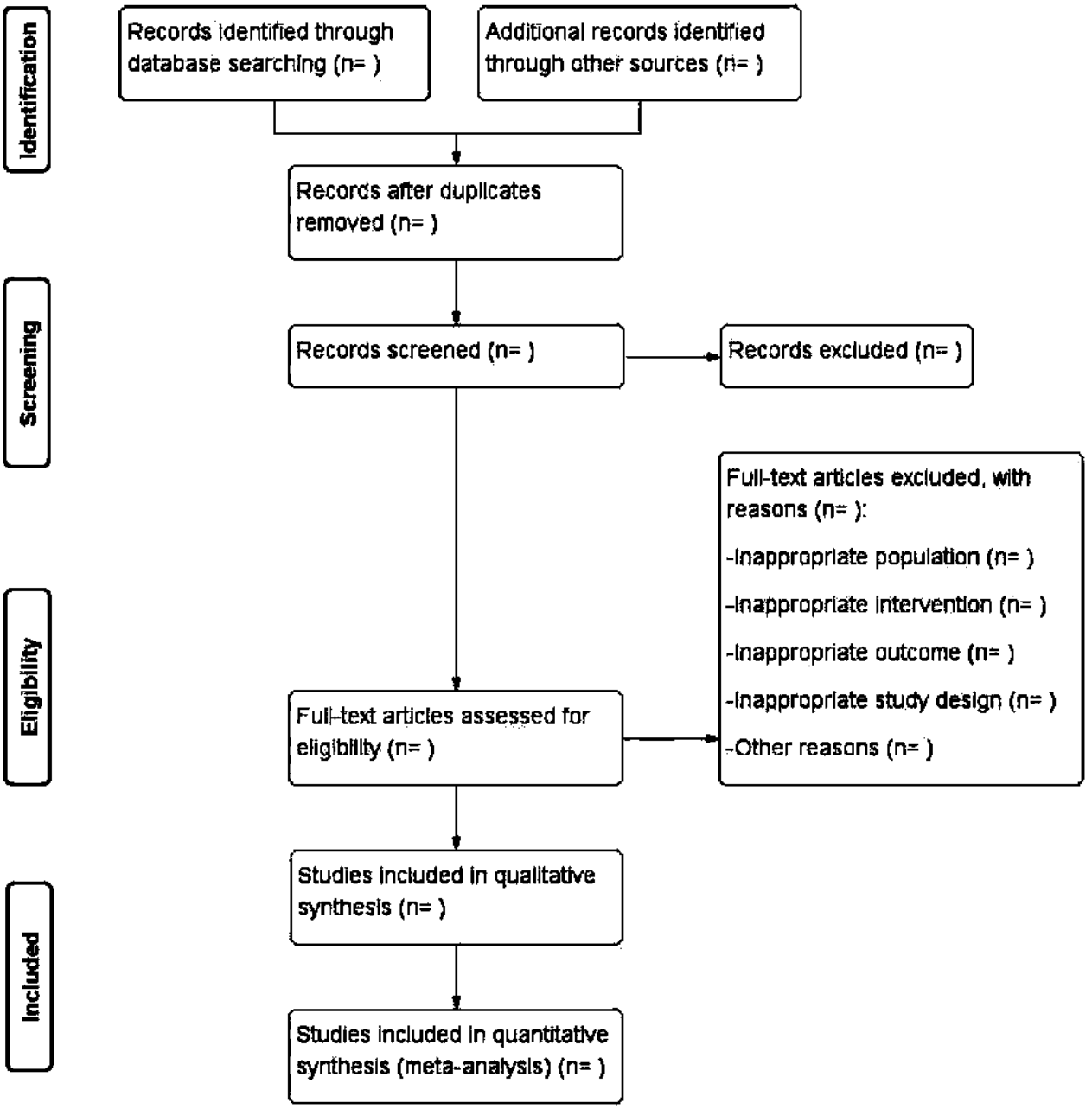

Figure 1. Study selection summarized in a PRISMA flow diagram.

allocation concealment), performance bias, detection bias, attrition bias, reporting bias, and other biases. The risk will be categorized into 3 levels: high level, low level, or unclear level. Any disagreement will be solved by discussion or with a third author.

2.6. Data synthesis and statistical analysis

2.6.1. Data synthesis. Review-Manager 5.3 will be used to conduct the meta-analysis. To evaluate the effectiveness of physiotherapy on QoL, the pre- and the post- intervention score should be reported. The post-intervention score will be extracted and used for the mean difference (MD) and the 95% confidence interval (CI) calculating. The results will be weighted by using a random effect model if studies have a large diversity. We will consider it statistically significant if $P < .05$.

2.6.2. Assessment of heterogeneity. We will assess the heterogeneity by the χ^2 test and the I^2 test. The χ^2 test with $P < .10$ indicates statistical significance, and the I^2 test with $I^2 > 50\%$ indicates moderate-to-high heterogeneity.[15] Reasons for the high heterogeneity will be analyzed.

2.6.3. Subgroup analysis. Subgroup analyses will be conducted to investigate the effectiveness of physiotherapy on life quality based on the type of physiotherapy (physical training, breathing exercise, and IMT), and the 3 domains of QoL (symptoms, activity limitations, and emotional function).

2.6.4. Sensitivity analysis. We will carry out a sensitivity analysis by excluding individual study sequentially and compare the pooled results by using a random effect model and a fixed effect model. If necessary, low-quality studies will be excluded and the meta-analysis will be repeated to test the stability of pooled results.

2.7. Assessment of reporting bias

If 10 or more studies are included, we will construct a funnel plot and use the Egger test in Stata 14.0 to examine the reporting bias. The results will be calcified according to the *Cochrane Handbook for Systematic Reviews of Interventions*.[12] If $P < .1$, we will consider it a significant reporting bias.

2.8. Confidence in cumulative evidence

We will assess the quality of evidence by using the GRADE system. Quality of evidence will be adjusted to a high level, moderate level, low level, or very low level.

3. Discussion

Physiotherapy such as physical training has been proved to be beneficial for healthy children by improving their musculoskeletal health and mental health.[16,17] However, the effects of physiotherapy in asthmatic children remain uncertain. Previous publications have reported the effects of physiotherapy in patients with asthma, but these studies included both children and adults, or adults only.[11,18,19,20] Although some systematic reviews did not include adults, they only investigated the effects of one type of physiotherapy.[21,22,23,24] Thus, we are going to include different types of physiotherapy and asthmatic children only to conduct a systematic review and meta-analysis. This systematic review will calcify the effectiveness of physiotherapy on QoL in asthmatic children and demonstrate which physiotherapy is more helpful. It will provide evidence directly to the doctors for making a choice on asthma treatment.

Some limitations will exist because blinding of participants involving physiotherapy is not possible, leading to a decrease in the quality of evidence in this study probably. Besides, language bias may exist since studies published in a language other than English will be excluded.

Author contributions

WJ Zhang put forward the concept and drafted this protocol. WJ Zhang, LL Liu, and WH Yang will conduct the study search, study selection, and data extraction. WJ Zhang and LL Liu will analyze the data and assess the risk of bias. HM Liu will help to solve any dispute in this study. WJ Zhang and LL Liu are co-first authors who contributed equally to this study. All authors reviewed and approved this final manuscript.

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

Tujuan dari clinical practice guideline ini adalah untuk mengoptimalkan manajemen fisioterapi pada cystic fibrofis di Australia dan New Zealand dengan teknik airway clearance theraphy, inhalation therapy, exercise dan musculoskeletal management.

Airway Clearance

Tujuan dari airway clearance technique (ACT) adalah untuk membersihkan sputum dari jalan napas, untuk mengoptimalkan status pernafasan dan memperlambat perkembangan penyakit.

Active Cycle of Breathing Technique (ACBT) yang terdiri dari breathing control, thoracic expansion exercises dan forced expiration merupakan teknik yang efektif untuk pembersihan sekresi pernafasan dengan hasil yang sebanding dengan ACT banyak digunakan lainnya. ACBT dapat dilakukan oleh semua pasien yang dapat mengikuti petunjuk dan berguna pada semua tahap penyakit. Terapi positive expiratory pressure (PEP) didefiniskan sebagai bernapas terhadap PEP 10 - 20cmH2O menggunakan masker atau mouthpiece. PEP merupakan teknik lainnya yang efektif untuk pembersihan jalan napas.

Postural Drainage memanfaatkan gravitasi untuk mengurangi mucus pada paru – paru dengan positioning head-down tilt kepada pasien CF balita, anak – anak hingga remaja. Penilitian terbaru mengatakan bahwa positioning dengan posisi horizontal dapat mengurangi sputum dengan efek samping yang lebih sedikit daripada posisi head – down.

Beberapa penderita penyakit paru CF ringan menggunakan olahraga bersamamaan dengan forced expiration dan coughing.Dan untuk penderita penyakit paru-paru yang lebih luas dan volume dahak yang lebih besar menggunakan olahraga sebagai tambahan untuk rejimen ACT formal. Efek fisiologis dari latihan meliputi penurunan mechanical impedance of sputum, peningkatan expiratory flow rates dan inducement of coughing.

Inhalation Theray

Terapi inhalasi merupakan pengobatan penting untuk penyakit pernapasan CF. Terapi Inhalasi yang efektif sangat penting untuk keberhasilan ACT dan sebaliknya; Akibatnya, fisioterapis harus cukup terampil dalam memberikan terapi inhalasi untuk memaksimalkan keefektifan kedua perawatan. Penentu utama pola deposisi untuk obat nebulised adalah

pola pernapasan saat menghirup, ukuran droplet dan usia / kondisi paru. Hasil napas yang lebih lambat dalam pola deposisi perifer yang lebih diinginkan, peningkatan homogenitas deposisi dan peningkatan deposisi obat secara keseluruhan. Nafas pelan yang stabil dengan sesekali menarik napas dalam-dalam secara tradisional direkomendasikan untuk meningkatkan pengendapan.

Obat nebulisasi harus diminum melalui mulut untuk memaksimalkan pengiriman obat ke saluran udara dan menghindari nasal fi ltrasi. Pengecualian terjadi pada anak kecil yang mungkin tidak dapat menggunakan mouthpiece secara efektif, atau di mana sinus menjadi target terapi, atau mereka dengan dispnea akut. Bronkodilator harus diberikan dengan meter dosed inhaler dan spacer kecuali pasien yang terlalu dyspnoeic atau tidak dapat mengikuti instruksi. Meter dosed inhaler dan spacer harus digunakan untuk pemberian inhaled kortikosteroid. Pasien harus didorong untuk membilas mulutnya dengan air dan kemudian berkumur untuk mengurangi risiko sariawan.

Exercise Assasment and Training

- Olah raga dianjurkan bagi penderita Cystic Fibrosis semua umur .
- Tes latihan harus dipertimbangkan untuk menilai respon terhadap terapi pada rawat inap dan rawat jalan dan sebagai alat penilaian dalam dosis program pelatihan latihan.
- Tes jalan 6 menit harus dilakukan sebagai bagian dari penilaian awal untuk transplantasi paru.
- Dosis latihan harus disesuaikan dengan individual dan mematuhi pedoman latihan yang direkomendasikan.
- Oksigen tambahan harus dipertimbangkan selama pelatihan pada pasien dengan desaturasi berat akibat olahraga.

Musculoscletal Complications of Cystis Fibrosis

- Penilaian muskuloskeletal harus disertakan di review tahunan dari sekitar usia 8 tahun (prapubertas). Penilaian lebih awal diperlukan jika nyeri atau gangguan fungsional dilaporkan atau risiko kepadatan mineral tulang merosot.
- Aktivitas fisik teratur, termasuk weight - bearing exercise, harus didorong sepanjang umur untuk mengoptimalkan kepadatan tulang.
- Program pelatihan kekuatan harus ditentukan untuk mengoptimalkan massa otot

Physiotherapy Management of Continence

Wanita dengan Cystic Fibrosis dan gejala stress urinary incontinence harus diajarkan latihan kekuatan dan ketahanan rehabilitasi untuk memberikan kendali yang lebih baik pada otot panggul. Pria dan wanita dengan Cystic Fibrosis harus diskrining gejala stress urinary incontinence. Pembersihan jalan napas harus dilakukan dalam postur yang dapat mempertahankan tulang belakang lumbar, dan untuk mengoptimalkan fungsi otot panggul.

Physiotherapy Management of Newly Diagnosed Patient

Perawatan untuk bayi yang baru didiagnosis mungkin termasuk perkusi untuk 3 - 5 menit di setiap posisi postural drainase an permainan fisik harian sesuai usia. Perawatan fisioterapi untuk yang baru didiagnosis anak dan orang dewasa harus memasukkan latihan fisik secara teratur, bentuk lain dari pembersihan jalan nafas harus ditambahkan sesuai kebutuhan.

Non-invasive Ventilation for Cystic Fibrosis

- NIV harus dipertimbangkan pada semua pasien dengan akut gagal pernapasan yang terdaftar untuk transplantasi.
- Pada pasien dengan gejala kegagalan ventilasi nokturnal, percobaan NIV nokturnal mungkin dilakukan.
- NIV adalah tambahan yang berguna untuk pembersihan jalan napas di pasien dengan penyakit parah dispnea dan kelelahan sehingga membatasi pembersihan jalan napas yang efektif.
- NIV mungkin merupakan tambahan yang berguna untuk latihan pada pasien dengan penyakit parah dispnea dan kelelahan sehingga berkontribusi pada penurunan kondisi dan membatasi pelatihan yang efektif.

Physiotherapy and Lung Transplantation

- Pasien dengan cystic fibrosis harus melakukan program latihan yang dirancang untuk mengoptimalkan fungsi fisiknya saat berada dalam daftar tunggu transplantasi.
- Pasien dengan cystic fibrosis yang telah menjalani transplantasi paru harus berpartisipasi dalam program rehabilitasi formal yang diawasi pasca operasi

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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.

*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.

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.

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*.
Any reference to the online supplement should cite this *Respirology* paper as the primary publication: Button BM, Wilson C, Dentice R, Cox N, Middleton A, Tannenbaum E, Bishop J, Cobb R, Burton K, Wood M, Moran F, Black R, Bowen S, Day R, Depiazzi J, Doiron K, Doumit M, Dwyer T, Elliot A, Fuller L, Hall K, Hutchins M, Kerr M, Lee A, Mans C, O'Connor L, Steward R, Potter A, Rasekaba T, Scoones R, Tarrant B, Ward N, West S, White D, Wilson L, Wood J, Holland AE. Physiotherapy for Cystic Fibrosis in Australia and New Zealand: A Clinical Practice Guideline. *Respirology* 2016; **21**: 656-667

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