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REABILITAÇÃO DOMICILIAR SUPERVISIONADA OU PARCIALMENTE SUPERVISIONADA PARA CRIANÇAS E ADOLESCENTES COM FIBROSE CÍSTICA: UM PROTOCOLO DE REVISÃO SISTEMÁTICA

SUPERVISING OR PARTIALLY SUPERVISING HOME REHABILITATION IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS: A SYSTEMATIC REVIEW PROTOCOL

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Resumo

Introdução: A fibrose cística (FC) impacta a vida de crianças e adultos com a doença. Assim, várias abordagens de tratamento são utilizadas na prática clínica. O objetivo da revisão proposta é avaliar a eficácia das estratégias de terapia de reabilitação domiciliar para pacientes com FC. Métodos: Será realizada uma busca sistemática em 5 bases de dados para localizar estudos publicados e não publicados. Serão considerados ensaios clínicos randomizados que incluam crianças e adolescentes diagnosticados com FC em reabilitação domiciliar. As etapas de escolha, avaliação e extração de dados dos artigos serão realizadas por dois autores independentes. O desfecho primário será a melhora dos sintomas de dispneia. Os desfechos secundários serão os resultados da intervenção na função pulmonar, sintomas, aptidão física e qualidade de vida relacionada. Study Registry International Prospective Registry for Systematic Reviews (PROSPERO) Número: CRD42021264835.

Palavras-chave: fibrose cística; crianças; adolescentes; reabilitação.

Abstract

Introduction: Cystic fibrosis (CF) impacts the lives of children and adults with the disease. Thus, several treatment approaches are used in clinical practice. The purpose of the proposed review is to assess the effectiveness of home rehabilitation therapy strategies for CF patients. Methods: A systematic search of 5 databases will be performed to locate published and unpublished studies. We will consider randomized clinical trials that include children and adolescents diagnosed with CF who underwent home rehabilitation. The phases of choosing, evaluating and extracting data from the articles will be carried out by two independent authors. The primary outcome will be improvement in dyspnea symptoms. Secondary outcomes will be intervention outcomes on lung function, symptoms, physical fitness and related quality of life. Study Registry International Prospective Registry for Systematic Reviews (PROSPERO) Number: CRD42021264835.

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Keywords: cystic fibrosis; children; adolescents; rehabilitation.

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1. Introduction

Cystic fibrosis CF occurs when a person inherits one (abnormal) copy of CFTR (cystic fibrosis transmembrane conductance regulator) from each parent and is classified as autosomal recessive (SOCKRIDER; FERKOL, 2017). This disease still has no cure, however, life expectancy has steadily improved, reaching an average age of 45 in the USA (SOCKRIDER; FERKOL, 2017; RADLOVIC, 2012). Furthermore, we know that pulmonary exacerbations triggered by the complications of CF continue to significantly impact the lives of children and adults with the disease (GOSS, 2019).

Physiotherapeutic treatment is of paramount importance for patients with CF, acting in the construction of a rehabilitation plan based on airway clearance, elaboration of exercise plans for the upper, lower limbs and trunk, management of possible musculoskeletal sequelae that may arise in the long term and also cardiorespiratory exercises (MCLLWAINE; LEE; RICHMOND, 2014; BUTTON *et al*., 2016).

In addition, it is already present in the literature, through randomized clinical trials, that home-based rehabilitation for children and adolescents triggers positive results for patients with FC, such as, for example, improved exercise capacity, muscle strength and quality of life (DEL CORRAL *et al*., 2018) and increased mucus expectoration (MILLER *et al*., 1995). Thus, the aim of this proposed systematic review is to synthesize the results obtained from randomized clinical trials that used home rehabilitation, either conventional or without rehabilitation therapy, on physical capacity, quality of life, lung function and symptoms.

2. Methods

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2.1 Question systematic review

Question PICO: Supervised or partially supervised home rehabilitation (I) improves lung function (O) in children and / or adolescents with cystic fibrosis (CF) (P) when compared to conventional rehabilitation or no intervention (C)?

2.2 Protocol and registry

The review methodology was registered in the International Prospective Register of Systematic Reviews (PROSPERO: CRD42021264835) and followed the recommendations of the Cochrane Handbook (HIGGINS; GREEN, 2021) and PRISMA statement (MOHER *et al*., 2009).

2.3 Participants

This review will consider studies carried out among children and/or adolescents who have a clinical diagnosis of CF. Studies that include a mixed population, in which people with CF have other respiratory comorbidities will be excluded if the data cannot be obtained separately.

2.4 Intervention (s)

The intervention will include any type of physical activity and / or rehabilitation approach within the home pulmonary rehabilitation programs that is supervised or partially supervised. Physical activity, resistance exercises, aerobic training, alternative therapies (Tai Chi Chuan, Yoga, Acupuncture, among others) and breathing exercises will be considered.

2.5 Comparator (s)

Comparators will include any type of physical activity and / or rehabilitation approach within the conventional supervised pulmonary rehabilitation program performed in clinics and / or outpatient clinics to treat CF patients or no rehabilitation therapy.

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2.6 Results

This review will consider studies that include the results:

Symptoms: subjective assessments of stress symptoms that is, dyspnea and fatigue (ie, Borg scale); Lung function: respiratory rate, forced expiratory volume in 1 second, forced vital capacity; Physical capacity: will be performed based on an analysis of the results obtained in tests chosen and carried out during the studies (i.e., Ergospirometry, 6-min walk test); Health status: will be evaluated through the results obtained in questionnaires that assess health-related quality of life (i.e., Cystic Fibrosis Questionnaire).

2.7 Types of studies

This review will consider only randomized controlled trials (RCTs) comparing a supervised or partially supervised home rehabilitation program for CF patients with conventional pulmonary rehabilitation or without rehabilitation therapy. There will be no language restriction or year of publication. Studies published in the databases of the Virtual Health Library (VHL) (www.bvsalud.org), Medline (via PubMed) (www.ncbi.nlm.nih.gov/pubmed), Cochrane Library (www.cochranelibrary), EMBASE (https://www.embase.com/login) and EBSCOhost (https://search.ebscohost.com/) (CINAHL database). Duplicate studies, studies with a design not compatible with the objective of the study and those that do not make clear the method used in the study will be excluded. In addition, conference abstracts, letter to the editor and case reports will be excluded.

2.8 Search strategy

An initial search on MEDLINE (via PubMed), Cochrane Database of Systematic Reviews and PROSPERO was carried out in order to verify whether the subject of this review had already been produced. The words of the text contained in the titles and abstracts of the relevant articles, and the index terms used to describe the articles, were used to develop a complete search strategy detailed in

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Appendix I. The final step of the search strategy will be to review the reference lists of articles included in the review to identify additional articles. In order to guarantee the saturation of the literature, comprehensive surveys were constructed of both index terms (MeSH terms), “free text” terms and synonyms.

2.9 Selection of studies

Electronic searches will be carried out by two independent authors (DMX and RALA). The articles found in the databases will be exported to Rayyan, a digital and mobile application for systematic reviews (http://rayyan.qcri.org) (OUZZANI *et al*. 2016), for management, removal of duplicates and selection of manuscripts (SAWYER *et al*., 2018). First, the manuscripts will be selected according to their titles and abstracts based on the eligibility criteria. Then, the selected studies will be read in full to confirm your eligibility. Disagreements about the inclusion process will be resolved by a third reviewer (LFS). The reasons for excluding full-text studies that do not meet the inclusion criteria will be recorded and reported in the systematic review. The results of the research will be described in full in the final systematic review and presented in a flow chart of Preferred Reporting Items for Systematic Reviews and Meta-analyzes (PRISMA) (MOHER *et al*., 2009).

2.10 Bias risk assessment (quality)

Two independent researchers (DMX and RALA) will assess the methodological quality of the tests using a PEDro scale (0-10), with higher scores meaning higher methodological quality. The discrepancies will be resolved by a third investigator (LFS). When available, scores are already available in the PEDro database (https://pedro.org.au/).

2.11 Data extraction

Data will be extracted from studies included in the review by two independent reviewers (DMX and RALA) using a standardized data extraction tool. The extracted

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data will include specific details about the sources, study methods, processing and results obtained for the purpose of the review. Any disagreements that arise between the reviewers will be resolved through discussion or with a third reviewer (LFS).

2.12 Data synthesis

Depending on the availability of the data, if possible, we will do the meta-analyzes. All analyzes will be performed in software R version 3.6.2 using meta and conducted packages. If the heterogeneity test is performed, the results will be grouped using a meta-analysis of random effects if $I^2 \geq 30\%$ and fixed effects if $I^2 > 30\%$. An appropriate subgroup meta-analysis will be used to determine the impact between types of rehabilitation programs (supervised or partially supervised, conventional and non-rehabilitated home rehabilitation) in relation to CF. In addition, funnel charts will be generated when more than ten articles are included in the meta-analysis to identify possible risks of bias.

We anticipate that there will be a limited scope for meta-analysis due to the variety of different measured results and the small number of existing studies, especially in home pulmonary rehabilitation for CF. In this case, only a narrative summary of the findings of the included studies, structured around the treatment approach options, may be possible.

The quality of the evidence generated will be assessed using the GRADE system (Classification of Recommendations, Evaluation, Development and Assessments) (ATKINS *et al.*, 2004) to provide recommendations for clinical practice.

3. DISCUSSION

Evidence-based health empowers health professionals to make important clinical decisions. The results of the systematic review will help us to better

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understand the effects of home pulmonary rehabilitation for children and adolescents with CF. This knowledge is useful for the clinician to understand the rehabilitation techniques used for patients with CF and, above all, to help reduce the morbidity and mortality of the disease. In addition, updated information can help prescribe activities for patients in clinical care, who need to perform activities in the home environment for better results.

To maintain high methodological quality, our systematic review will follow the PRISMA (MOHER *et al*., 2009) statement and the recommendations of the Cochrane Handbook (HIGGINS; GREEN, 2021). The use of a broad search strategy and inclusion criteria will result in a complete synthesis of current evidence regarding home rehabilitation on physical capacity, lung function, quality of life and symptoms.

4. CONCLUSION

It is expected that the results of this systematic review will contribute to the existing literature and help in clinical practice regarding home rehabilitation for children and adolescents with CF.

5. APPENDIX

Search conducted on May 26th, 2021.

Search	Query	Records retrieved
MEDLINE (Pubmed)	((Fibrosis, Cystic OR Mucoviscidosis OR Pulmonary Cystic Fibrosis OR Cystic Fibrosis, Pulmonary OR Pancreatic Cystic Fibrosis OR Cystic Fibrosis, Pancreatic OR Fibrocystic Disease of Pancreas OR Pancreas Fibrocystic Disease OR Pancreas Fibrocystic Diseases OR Cystic Fibrosis of Pancreas) AND (Home Care Services OR Home Care Service OR Service, Home Care OR Care Services, Home OR Domiciliary Care OR Care, Domiciliary OR Services, Home Care OR Home Care OR Care, Home)) AND (Rehabilitation OR Habilitation OR Remedial Exercise OR Exercise, Remedial OR Exercises, Remedial OR Remedial Exercises OR Therapy, Exercise OR Exercise Therapies	151

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	OR Therapies, Exercise OR Rehabilitation Exercise OR Exercise, Rehabilitation OR Exercises, Rehabilitation OR Rehabilitation Exercises)	
BVS	(fibrose cística) AND (Reabilitação OR Habilitação OR Exercício corretivo OR Exercício, Corretivo OR Exercícios, Corretivo OR Exercícios corretivos OR Terapia, Exercício OR Terapias de exercício OR Terapias, Exercício OR Exercício de reabilitação OR Exercício, Reabilitação OR Exercícios, Reabilitação OR Exercícios de reabilitação)	1994
Cochrane Library	cystic fibrosis in Title Abstract Keyword AND Rehabilitation OR Habilitation OR Remedial Exercise OR Exercise, Remedial OR Exercises, Remedial OR Remedial Exercises OR Therapy, Exercise OR Exercise Therapies OR Therapies, Exercise OR Rehabilitation Exercise OR Exercise, Rehabilitation OR Exercises, Rehabilitation OR Rehabilitation Exercises in Title Abstract Keyword - (Word variations have been searched)	252

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EMBASE	('fibrosis, cystic' OR (fibrosis, AND cystic) OR 'mucoviscidosis'/exp OR mucoviscidosis OR 'pulmonary cystic fibrosis' OR (pulmonary AND cystic AND ('fibrosis'/exp OR fibrosis)) OR 'cystic fibrosis, pulmonary' OR (cystic AND fibrosis, AND pulmonary) OR 'pancreatic cystic fibrosis'/exp OR 'pancreatic cystic fibrosis' OR (pancreatic AND cystic AND ('fibrosis'/exp OR fibrosis)) OR 'cystic fibrosis, pancreatic' OR (cystic AND fibrosis, AND pancreatic) OR 'fibrocystic disease of pancreas' OR (fibrocystic AND ('disease'/exp OR disease) AND of AND ('pancreas'/exp OR pancreas)) OR 'pancreas fibrocystic disease'/exp OR 'pancreas fibrocystic disease' OR (('pancreas'/exp OR pancreas) AND fibrocystic AND ('disease'/exp OR disease)) OR 'pancreas fibrocystic diseases' OR (('pancreas'/exp OR pancreas) AND fibrocystic AND ('diseases'/exp OR diseases)) OR 'cystic fibrosis of pancreas'/exp OR 'cystic fibrosis of pancreas' OR (cystic AND ('fibrosis'/exp OR fibrosis) AND of AND ('pancreas'/exp OR pancreas))) AND ('rehabilitation'/exp OR rehabilitation OR habilitation OR 'remedial exercise' OR (remedial AND ('exercise'/exp OR exercise)) OR 'exercise, remedial' OR (exercise, AND remedial) OR 'exercises, remedial' OR (exercises, AND remedial) OR 'remedial exercises' OR (remedial AND exercises) OR 'therapy, exercise'/exp OR 'therapy, exercise' OR (therapy, AND ('exercise'/exp OR exercise)) OR 'exercise therapies' OR (('exercise'/exp OR exercise) AND therapies) OR 'therapies, exercise' OR (therapies, AND ('exercise'/exp OR exercise)) OR 'rehabilitation exercise' OR (('rehabilitation'/exp OR rehabilitation) AND ('exercise'/exp OR exercise)) OR 'exercise, rehabilitation' OR (exercise, AND ('rehabilitation'/exp OR rehabilitation)) OR 'exercises, rehabilitation' OR (exercises, AND ('rehabilitation'/exp OR rehabilitation)) OR 'rehabilitation exercises' OR (('rehabilitation'/exp OR rehabilitation) AND exercises)) AND ('home care services'/exp OR 'home care services' OR (('home'/exp OR home) AND ('care'/exp OR care) AND services) OR 'home care service'/exp OR 'home care service' OR (('home'/exp OR home) AND ('care'/exp OR care) AND service) OR 'service, home care' OR (service, AND ('home'/exp OR home) AND ('care'/exp OR care)) OR 'care services, home' OR (('care'/exp OR care) AND services, AND ('home'/exp OR home)) OR 'domiciliary care'/exp OR 'domiciliary care' OR (domiciliary AND ('care'/exp OR care)) OR 'care, domiciliary' OR (care, AND domiciliary) OR 'services, home care' OR (services, AND ('home'/exp OR home) AND ('care'/exp OR care)) OR 'home care'/exp OR 'home care' OR (('home'/exp OR home) AND ('care'/exp OR care)) OR 'care, home' OR (care, AND ('home'/exp OR home)))	177
EBSCOhost/ CINAHL	((Fibrosis, Cystic OR Mucoviscidosis OR Pulmonary Cystic Fibrosis OR Cystic Fibrosis, Pulmonary OR Pancreatic Cystic Fibrosis OR Cystic Fibrosis, Pancreatic OR Fibrocystic Disease of Pancreas OR Pancreas Fibrocystic Disease OR Pancreas Fibrocystic Diseases OR Cystic Fibrosis of Pancreas) AND (Home Care Services OR Home Care Service OR Service, Home Care OR Care Services, Home OR Domiciliary Care OR Care, Domiciliary OR Services, Home Care OR Home Care OR Care, Home)) AND (Rehabilitation OR Habilitation OR Remedial Exercise OR Exercise, Remedial OR Exercises, Remedial OR Remedial Exercises OR Therapy, Exercise OR Exercise Therapies OR Therapies, Exercise OR Rehabilitation Exercise OR Exercise, Rehabilitation OR Exercises, Rehabilitation OR Rehabilitation Exercises)	26

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Total	2600

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