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REVIEW ARTICLE

Association of growth and nutritional parameters with pulmonary function in cystic fibrosis: a literature review

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KEYWORDS
Cystic fibrosis;
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Abstract
Objective: To review the literature addressing the relationship of growth and nutritional parameters with pulmonary function in pediatric patients with cystic fibrosis.
Data source: A collection of articles published in the last 15 years in English, Portuguese and Spanish was made by research in electronic databases – PubMed, Cochrane, Medline, Lilacs and Scielo – using the keywords cystic fibrosis, growth, nutrition, pulmonary function in varied combinations. Articles that addressed the long term association of growth and nutritional parameters, with an emphasis on growth, with pulmonary disease in cystic fibrosis, were included, and we excluded those that addressing only the relationship between nutritional parameters and cystic fibrosis and those in which the aim was to describe the disease.
Data synthesis: Seven studies were included, with a total of 12,455 patients. Six studies reported relationship between growth parameters and lung function, including one study addressing the association of growth parameters, solely, with lung function, and all the seven studies reported relationship between nutritional parameters and lung function.
Conclusions: The review suggests that the severity of the lung disease, determined by spirometry, is associated with body growth and nutritional status in cystic fibrosis. Thus, the intervention in these parameters can lead to the better prognosis and life expectancy for cystic fibrosis patients.

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PALAVRAS-CHAVE
Fibrose cística;
Pulmão/fisiopatologia;
Crescimento;
Nutrição

Associação dos parâmetros de crescimento e nutricionais com função pulmonar na fibrose cística: revisão da literatura

Resumo

Objetivo: Revisar a literatura que aborda a relação entre os parâmetros de crescimento e nutricionais com a função pulmonar em pacientes pediátricos com fibrose cística.

Fontes de dados: Dados foram coletados de artigos publicados nos últimos 15 anos em Inglês, Português e Espanhol através de pesquisa nas bases de dados eletrônicas – PubMed, Cochrane, Medline, Lilacs e Scielo – usando as palavras-chave: fibrose cística, crescimento, nutrição, função pulmonar utilizando combinações variadas. Os artigos que analisaram a associação de longo prazo entre parâmetros de crescimento e nutricionais, com ênfase em crescimento, com doença pulmonar em fibrose cística, foram incluídos, sendo excluídos aqueles que analisaram apenas a relação entre os parâmetros nutricionais e fibrose cística e aqueles em que o objetivo era descrever a doença.

Síntese dos dados: Sete estudos foram incluídos, com um total de 12.455 pacientes. Seis estudos relataram relação entre parâmetros de crescimento e função pulmonar, incluindo um estudo que analisou apenas a associação de parâmetros de crescimento com a função pulmonar, e todos os sete estudos relataram associação entre parâmetros nutricionais e função pulmonar.

Conclusões: A revisão sugere que a gravidade da doença pulmonar, determinada por espirometria, está associada com crescimento corporal e o estado nutricional em fibrose cística. Assim, a intervenção nesses parâmetros pode contribuir para um melhor prognóstico e expectativa de vida em pacientes com fibrose cística.

Introduction

Cystic Fibrosis (CF) is the most common lethal genetic disease in Caucasian populations. It is caused by a mutation in a gene that encodes the Cystic Fibrosis Transmembrane conductance Regulator (CFTR) protein, which is expressed in many epithelial and blood cells, functioning mainly as a chloride channel.[1] Pulmonary disease is the most important manifestation in CF and the main factor acting in morbidity and mortality of the disease. The response in pulmonary disease is mediated by abnormal CFTR,[2] modifier genes,[3–8] airway infections and inflammation,[9] probably affecting weight and height due to appetite suppression and enhanced energy expenditure.

Malnutrition and growth restriction are also frequent and are related to the impairment of the pulmonary function in a vicious circle: malnourished patients tend to present worst pulmonary function and patients with severe pulmonary disease tend to grow up less. Although these relationships have been already reported,[10,11] there are a few long term analyses in regard to the achievement of growth and nutrition goals for the course of pulmonary function from infancy to adulthood.

In this context, the aim of this study was to analyze long term studies comparing growth and nutrition parameters (with emphasis in growth) with pulmonary function in CF patients, evaluating the relationship among these factors.

Method

A literature review of the last 15 years (2000–2015) about the relationship between growth and nutritional parameters and lung function was made. The search for references in English, Spanish and Portuguese was performed through electronic databases – PubMed, Medline, Cochrane, Lilacs and Scielo – using the descriptors: CF, growth, body growth, pulmonary function and lung function in varied combinations and in their correspondent translations to Portuguese and Spanish. Reviews addressing the theme were also consulted, as well reference lists of all articles, to search for new studies.

After this stage, we started the screening of papers, by analyzing titles and abstracts. The first inclusion criterion was the identification of potentially relevant studies, considering those in which the report compared growth parameters with lung function. In this case, we excluded studies in which the aims were to compare weight and/or height gain, without relationship with pulmonary function, and those in which the aims were to describe CF only.

In the first search, a total of 104 articles were found. By evaluating titles and abstracts, the following recuperation criteria for complete articles were: studies of cohort, longitudinal, cross-sectional, descriptive and prospective, which results evaluated the relationship among pulmonary function and growth parameters in CF patients, excluding those which, despite of appearing in the search results, did not address the subject under this point of view. In this stage, 27 papers were screened. The review was concluded with the reading of the complete articles, and, in the final manuscript, seven articles were included,[12–18] all of them in English (Fig. 1; Tables 1 and 2).

Data synthesis

A total of 12,455 patients were assessed. Considering the start up period of each study, the age range of the patients

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