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ORIGINAL ARTICLE

Factors impacting the growth and nutritional status of cystic fibrosis patients younger than 10 years of age who did not undergo neonatal screening

Taís Daiene Russo Hortencio, Roberto José Negrão Nogueira, Fernando Augusto de Lima Marson, Gabriel Hessel*, José Dirceu Ribeiro, Antônio Fernando Ribeiro

Universidade Estadual de Campinas (UNICAMP), Campinas, SP, Brazil

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KEYWORDS
Cystic fibrosis/ complications;
Child;
Neonatal screening;
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Abstract
Objective: The aim of this study was to evaluate by clinical and laboratory parameters how cystic fibrosis (CF) affects growth and nutritional status of children who were undergoing CF treatment but did not receive newborn screening.
Methods: A historical cohort study of 52 CF patients younger than 10 years of age were followed in a reference center in Campinas, Southeast Brazil. Anthropometric measurements were abstracted from medical records until March/2010, when neonatal screening program was implemented. Between September/2009 and March/2010, parental height of the 52 CF patients were also measured.
Results: Regarding nutritional status, four patients had Z-scores ≤-2 for height/age (H/A) and body mass index/age (BMI/A). The following variables were associated with improved H/A ratio: fewer hospitalizations, longer time from first appointment to diagnosis, longer time from birth to diagnosis and later onset of respiratory disease. Forced vital capacity [FVC(%)], forced expiratory flow between 25-75% of FVC [FEF_{25-75}(%)], forced expiratory volume in the first second [FEV_1(%)], gestational age, birth weight and early respiratory symptoms were associated with improved BMI/A.
Conclusions: Greater number of hospitalizations, diagnosis delay and early onset of respiratory disease had a negative impact on growth. Lower spirometric values, lower gestational age, lower birth weight, and early onset of respiratory symptoms had negative

*Corresponding author.
E-mail: ghessel@fcm.unicamp.br (G. Hessel).

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impact on nutritional status. Malnutrition was observed in 7.7% of cases, but 23% of children had nutritional risk.

PALAVRAS-CHAVE
Fibrose cística/complicações;
Criança;
Triagem neonatal;
Antropometria;
Estado nutricional

Fatores de impacto sobre o crescimento e o estado nutricional de pacientes com fibrose cística menores de 10 anos que não foram submetidos à triagem neonatal

Resumo
Objetivo: O objetivo deste estudo foi avaliar por meio de parâmetros clínicos e laboratoriais como a fibrose cística (FC) afeta o crescimento e estado nutricional de crianças que foram submetidas ao tratamento para FC, mas não receberam o diagnóstico pela triagem neonatal.
Métodos: Uma coorte histórica com 52 pacientes com FC menores de 10 anos de idade foram acompanhados em um centro de referência em Campinas, Sudeste do Brasil. Peso e altura foram coletados de prontuários médicos até março de 2010, quando a triagem neonatal foi implementada. Entre setembro de 2009 a março de 2010 a altura dos pais foi mensurada.
Resultados: Quatro pacientes tiveram escores Z≤-2 para altura/idade (A/I) e índice de massa corporal/idade (IMC/I). Foram associados com melhor A/I: menor número de hospitalizações, maior tempo entre a primeira consulta e o diagnóstico, maior tempo entre o nascimento e o diagnóstico e início tardio da doença respiratória. Capacidade vital forçada [CVF(%)], fluxo expiratório forçado entre 25-75% da CVF [FEF_{25-75}(%)], volume expiratório forçado no primeiro segundo [VEF_1(%)], idade gestacional, peso ao nascer e início sintomas respiratórios foram associados com melhor IMC/I.
Conclusões: Maior número de hospitalizações, retardo no diagnóstico e início precoce da doença respiratória tiveram impacto negativo no crescimento. Menores valores espirométricos, menor idade gestacional, menor peso ao nascer e o início precoce dos sintomas respiratórios tiveram impacto negativo no estado nutricional. A desnutrição foi observada em 7,7% dos casos, mas 23% das crianças apresentaram risco nutricional.

Introduction

Cystic fibrosis (CF) is an autosomal recessive disease that is highly prevalent among caucasians. CF is characterized by the involvement of multiple organs, especially the pulmonary and gastrointestinal systems; by abnormally high levels of sweat chloride, and by increased incidences of male infertility and diabetes mellitus.[1]

Dietetic care is needed and individualized attention should be given to ensure adequate energy intake among CF patients. To maintain adequate nutritional status, CF children should ingest 110 to 150% of the daily caloric intake recommended for healthy children.[2] Pancreatic insufficiency with chronic malabsorption, recurrent infections, chronic inflammation and energy expenditure, and insufficient nutritional intake are factors exacerbating malnutrition in CF patients. These factors lead to difficulties with weight maintenance and weight gain, and failure to thrive in infancy.

Regardless of the origin and reasons for high energy expenditure, the more clinically pertinent question is the influence of nutrition on the progression of lung disease, because lung function in CF patients is the main predictor of survival.[3,4] Clinical studies indicate that nutritional status plays an important role in the progression of lung disease in CF and it is a survival advantage among patients with good nutritional status.[3,4] These studies consistently support the strong influence of growth and nutritional status on CF-associated lung disease. But from birth, nutritional deficiency is determined primarily by pancreatic insufficiency and malabsorption, so subsequent aggressive nutritional support should facilitate proper growth and preserve lung function.

In Brazil, CF is associated with high morbidity and mortality. However, the survival of affected children in Brazil has increased substantially in the last 50 years due to an interdisciplinary approach to treatment, new medications and progress related to nutritional intervention and control. During the past 20 years, the benefit of early diagnosis on the nutritional status of CF patients has been established.[5,6] However, failure-to-thrive diagnoses remain common despite early identification of CF.[7,8]

In this study, we use clinical and laboratory variables to assess how CF affects the growth and nutritional status of patients younger than 10 years who were undergoing CF treatment but did not receive newborn screening.

Method

A historical cohort study was designed to evaluate CF patients younger than 10 years from the CF Reference