Validation of a Nutrition Screening Tool for Pediatric Patients with Cystic Fibrosis

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ABSTRACT

Background In cystic fibrosis (CF), nutrition diagnosis is of critical relevance because the early identification of nutrition-related compromise enables early, adequate intervention and, consequently, influences patient prognosis. Up to now, there has not been a validated nutrition screening tool that takes into consideration clinical variables.

Objective To validate a specific nutritional risk screening tool for patients with CF based on clinical variables, anthropometric parameters, and dietary intake.

Design Cross-sectional study. The nutrition screening tool was compared with a risk screening tool proposed by McDonald and the Cystic Fibrosis Foundation criteria.

Participants/setting Patients aged 6 to 18 years, with a diagnosis of CF confirmed by two determinations of elevated chloride level in sweat (sweat test) and/or by identification of two CF-associated genetic mutations who were receiving follow-up care through the outpatient clinic of a Cystic Fibrosis Treatment Center.

Main outcome measures Earlier identification of nutritional risk in CF patients aged 6 to 18 years when a new screening tool was applied.

Statistical analyses performed Agreement among the tested methods was assessed by means of the kappa coefficient for categorical variables. Sensitivity, specificity, and accuracy values were calculated. The significance level was set at 5% ($P<0.05$). Statistical analyses were carried out in PASW Statistics for Windows version 18.0 (2009, SPSS Inc).

Results Eighty-two patients (49% men, aged 6 to 18 years) were enrolled in the study. The agreement between the proposed screening tool and the tool for screening nutritional risk for CF by the McDonald method was good ($\kappa=0.804$; $P<0.001$) and the sensitivity and specificity was 85% and 95%, respectively. Agreement with the Cystic Fibrosis Foundation criteria was lower ($\kappa=0.418$; $P<0.001$), and the sensitivity and specificity were both 72%.

Conclusions The proposed screening tool with defined clinical variables promotes earlier identification of nutritional risk in pediatric patients with CF.

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CYSTIC FIBROSIS (CF) IS A GENETIC DISORDER, potentially lethal, progressive, and a multisystem disease. It is characterized clinically by the presence of chronic pulmonary obstructive disease and pancreatic insufficiency, which lead to a constellation of clinical and nutrition-related manifestations and complications. Airway involvement is progressive and varies in intensity. Decline in pulmonary function is associated with increased morbidity, and is the cause of death in more than 90% of patients with CF.[1] Treatment of pulmonary infections, enzyme replacement therapy, and nutritional support play essential roles in the management of CF.[2]

In patients with CF, poor nutritional status is associated with decreased pulmonary function,[3] and maintenance of adequate nutritional status is a most important goal of the multidisciplinary care of CF. Malnutrition in this patient population occurs due to increased energy requirements secondary to pulmonary inflammation and recurrent infections, as well as to pancreatic insufficiency.[4] Nutrition diagnosis is of critical relevance because the early identification of nutrition-related compromise enables early, adequate intervention and, consequently, influences patient prognosis.[5]

Use of a nutrition screening tool is recommended to identify risk of malnutrition in patients with CF. Such a tool should be able to identify a series of variables related to overall nutritional risk and assess specific aspects associated with the course of CF that interfere with nutritional status. Efficacy of the nutrition screening process is ensured by the use of instruments that employ information available for the entire population of interest and that can be administered quickly. Reliability and validity are also essential components of the clinical and nutrition screening process.[6]

Several tools have been advocated for identification of nutritional risk in hospitalized patients.[7-9] In 2002, the Cystic Fibrosis Foundation (CFF), in its CFF Consensus Report,[10] set forth criteria for nutrition-related diagnosis in patients with CF, taking into account stricter cutoff points

for malnutrition and nutritional risk and recommending early and effective intervention to help recover and slow the decline of pulmonary function in this population. In 2008, McDonald[11] validated the first nutrition screening tool for patients with CF, using body mass index (BMI) percentile data and longitudinal follow-up of weight gain and growth to stratify patients into low, moderate, and high nutritional risk groups. Data from the CFF Patient Registry were analyzed to determine the association between pulmonary function and BMI for age percentile. BMI for age percentile value above the 50th percentile were associated with well-preserved lung function and lower values of BMI were associated with incrementally lower values of forced expiratory volume in 1 second (FEV_1). Thus, CFF recommended that children with CF between the ages of 2 and 20 years maintain a BMI at or greater than the 50th percentile for age and sex.[12]

Along with lung function, nutritional status appears to be among the most important prognostic indicators in CF patients. In addition, other clinical variables may affect nutritional status. The aim of our study was to validate a tool for early nutritional risk identification for patients with CF.

MATERIALS AND METHODS

This cross-sectional study included patients aged 6 to 18 years with a diagnosis of CF confirmed by two determinations of elevated chloride level in sweat (sweat test) and/or by identification of two CF-associated genetic mutations who were receiving follow-up care through the outpatient clinic of the Cystic Fibrosis Treatment Center at Hospital de Clínicas de Porto Alegre (HCPA), Rio Grande do Sul, Brazil. Patients with acute respiratory infection, acute organ/system decompensation, or end-stage disease were excluded from the sample. The study was approved by the HCPA Research Ethics Committee (protocol no. 09.637) and informed consent was obtained before participation.

For sample size calculation, we estimated 86% sensitivity for screening of patients at nutritional risk, considering a 7.5-percentage point difference between the nutrition assessment tools being tested and a 95% confidence level. Sample size was determined to be 82 patients.

All participants underwent weight and height measurement as recommended by Lohman and colleagues[13] for calculation of BMI. Nutrition-related data were analyzed as percentiles according to the World Health Organization equations. Dietary intake was assessed by a registered dietitian by 24-hour food record intake, and nutrition facts—as total calories, protein, lipids, and carbohydrate—were calculated using NutWin software (2002, Unifesp). It was compared with the Recommended Dietary Allowances (RDA).[14] Variables such as age, age at diagnosis, time since diagnosis, pancreatic insufficiency (denoted by use of enzyme replacement therapy), route of nutrition, genetic mutation analysis, presence of cystic fibrosis-related diabetes (CFRD) (based on the oral glucose tolerance test when fasting plasma glucose is >125 mg/mL [6.94 mmol/L] and/or 2-hour postprandial glucose challenge is >199 mg/dL [11.04 mmol/L][15]), bacterial colonization, albumin levels, and percent predicted FEV_1 were obtained from patient charts. Specifically, the latter two parameters were obtained from the results of each patient's latest physical examination.

Presence or absence of bacterial colonization with *Staphylococcus aureus*, methicillin-resistant *S aureus* (MRSA), *Pseudomonas aeruginosa*, mucoid *P aeruginosa*, and *Burkholderia cepacia* complex (BCC) was evaluated by patient record, based on colonization of sputum during the past 12 months.

Nutritional risk diagnosis was assessed simultaneously by the same investigator, using three different tools: the proposed study tool, the nutrition risk screening tool proposed by McDonald,[11] and the CFF Consensus Report criteria.[10]

Nutrition Assessment Instruments

Proposed Nutritional Risk Identification Tool. This tool was based on the nutritional risk screening tool proposed by McDonald[11] and on the protocol that we use at HCPA for pediatric nutrition assessment[16] to determine nutritional risk taking into account clinical and nutrition-related indicators that interfere directly with dietary intake and/or nutritional status. Ten indicators believed to constitute risk factors for malnutrition in patients with CF were included in the instrument (Figure 1).

BMI <50th and <10th percentile. BMI percentile is an important indicator of nutritional status. In CF, it has a stronger association with percentage of predicted FEV_1,[17] and is thus considered a risk factor. According to the CFF Subcommittee on Growth and Nutrition recommendations[12] and McDonald,[11] patients with a BMI below the 50th percentile were scored 1 point, and those with a BMI below the 10th percentile—which defines nutritional compromise in patients with CF—were scored 2 points.[10]

Pancreatic insufficiency. Patients with pancreatic sufficiency have a significantly longer median of life expectancy than patients with pancreatic insufficiency (PI).[18] Therefore, the presence of PI was considered a risk factor. Patients with PI (defined as those receiving pancreatic enzyme replacement therapy) were scored 1 point, and those with signs of poorly managed PI such as flatulence, bloating, distension, rectal prolapse, abdominal pain, and self-reported noncompliance with pancreatic enzyme replacement therapy, were scored 2 points.

P aeruginosa*, BCC, or MRSA colonization.* Progressive decline of lung function because of chronic bacterial infection of the respiratory tract by microorganisms such as *P aeruginosa,* BCC, or MRSA is closely related to nutritional status, so it was considered a risk factor for malnutrition (1 point).[19-21]

Dietary intake <100% of RDA. Dietary intake below recommended allowances for sex and age was considered a risk factor for nutrition compromise (1 point), taking into account that recommended daily intake for patients with CF corresponds to 110% to 200% of RDAs.[12]

Weight gain below minimum, zero weight gain, or weight loss. Assessment of weight gain was included as a risk factor on the basis of the nutritional risk screening tool proposed by McDonald.[11] Patients with weight gain below the recommended minimum or zero weight gain were scored 1 point, whereas patients who lost weight were scored 2 points. The evaluation was performed by the weight