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Original Article

Nutritional status, nutrient intake and use of enzyme supplements in paediatric patients with Cystic Fibrosis; a European multicentre study with reference to current guidelines

Joaquim Calvo-Lerma [a,*], Jessie M. Hulst [b], Inês Asseiceira [c], Ine Claes [d], Maria Garriga [e], Carla Colombo [f], Victoria Fornés [a], Sandra Woodcock [b], Tiago Martins [c], Mieke Boon [d], Mar Ruperto [e], Sylvia Walet [b], Chiara Speziali [f], Peter Witters [d], Etna Masip [a], Celeste Barreto [c], Kris de Boeck [d], Carmen Ribes-Koninckx [a] on behalf of MyCyFAPP Project

[a] *Instituto de Investigación Sanitaria La Fe, Avenida Fernando Abril Martorell, 106, Torre A, planta 0, 46026 Valencia, Spain*
[b] *Erasmus Medical Center, Sophia Children's Hospital, Rotterdam, the Netherlands*
[c] *Associação para a Investigação e Desenvolvimento da Faculdade de Medicina, Avenida Professor Egas Moniz, 1649-028 Lisboa, Portugal*
[d] *Pediatric Pulmonology and Cystic Fibrosis Unit, Department of Pediatrics, University Hospitals Leuven, Belgium*
[e] *Hospital Universitario Ramón y Cajal, Carretera Colmenar Viejo km 9, 100, 28034 Madrid, Spain*
[f] *CF Center, Università degli Studi di Milano, Fondazione IRCCS Ca' Granda, Ospedale Maggiore Policlinico, Via Commenda 9, 20122 Milan, Italy*

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Abstract

Background: The New European guidelines have established the most updated recommendations on nutrition and pancreatic enzyme replacement therapy (PERT) in CF. In the context of MyCyFAPP project - a European study in children with CF aimed at developing specific tools for improvement of self-management - the objective of the current study was to assess nutritional status, daily energy and macronutrient intake, and PERT dosing with reference to these new guidelines.
Methods: Cross sectional study in paediatric patients with CF from 6 European centres. SD-scores for weight-for-age (WFA), height-for-age (HFA) and body mass index-for-age (BMI) were obtained. Through a specific 4-day food and enzyme-dose record, energy and macronutrients intake and PERT-use (LU/g lipids) were automatically calculated by the MyCyFAPP system. Comparisons were made using linear regression models.
Results: The lowest quartiles for BMI and HFA were between 0 and -1SD in all the centres with no significant differences, and 33.5% of the patients had a SD-score <0 for all three parameters. The minimum energy intake recommendation was not reached by 40% of the children and mean nutrients intake values were 14%, 51% and 34% of the total energy for protein, carbohydrates and lipids respectively. When assessed per centre, reported PERT doses were in the recommended range in only 13.8% to 46.6% of the patients; from 5.6% up to 82.7% of children were above the recommended doses and 3.3% to 75% were below.
Conclusion: Among the 6 centres, a large variability and inconsistency with new guidelines on nutrition and PERT-use was found. Our findings document the lack of a general criterion to adjust PERT and suggest the potential benefit of educational and self-managerial tools to ensure adherence to therapies, both for clinical staff and families. They will be taken into account when developing these new tools during the next stages of MyCyFAPP Project.

Keywords: Cystic fibrosis; Nutritional status; Pancreatic insufficiency; PERT; Nutritional requirements macronutrients; Guidelines; Paediatrics; Self-management; Telemedicine

Abbreviations: BMI, body mass index for age; CI, confidence interval; CF, Cystic Fibrosis; E/S, enzyme to substrate ratio; ECFS, European Cystic Fibrosis Society; ESPEN, European Society for Parenteral and Enteral Nutrition; ESPGHAN, European Society for Paediatric Gastroenterology, Hepatology and Nutrition; FER, food and enzymes record; HFA, height for age; kcal, kilocalorie; LU, lipase units; mHealth, mobile health; PERT, pancreatic enzyme replacement therapy; PI, pancreatic insufficiency; PS, pancreatic sufficiency; SD, standard deviation; WFA, weight for age

* Corresponding author.

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1. Introduction

Antibiotics, physiotherapy and nutrition are considered the three pillars in the treatment of Cystic Fibrosis (CF) according to the most recent standards of care [1]. Whilst therapies for lung disease have been a continuous focus of research in recent years [2] less high-impact studies have addressed the improvement of the nutritional and pancreas-related aspects.

The importance of a good nutritional status has clearly emerged over the last few decades with its direct relationship to a better lung function and consequently a better overall prognosis and survival [1,3]. Obtaining and keeping a good nutritional status is, however, a challenge in most patients with CF. This has several reasons, i.e. the increased energy requirements secondary to chronic inflammation and pancreatic insufficiency (PI) (present in almost 80% of patients) on one side and the decreased energy intake and loss of nutrients due to maldigestion and malabsorption on the other side [4].

Therefore, the main nutritional goal in children with CF is to avoid inadequate nutrient intake and to follow a normal growth pattern according to age. Nutritional follow-up and intervention is essential and should aim at achieving an optimal adjustment of pancreatic enzyme replacement therapy (PERT) to correct pancreatic insufficiency as well as at prescribing a balanced diet according to nutritional needs [5,6]. The recently published guidelines on nutrition care for infants, children and adults with CF compile the most recent scientific evidence and establish recommendations regarding nutritional goals and PERT dosage [7].

In comparison with antibiotics or physiotherapy treatments, nutrition and PERT are more likely to be mainly managed by the patients or parents themselves, outside the care of the CF team. Eating and drinking is a daily need and patients are supposed to comply with a balanced diet and adjust the dose of enzymes accordingly. However, if an incorrect nutritional behaviour or an inadequate enzyme dosage occurs, it will most likely not be detected and corrected until the next contact or visit to the CF centre. Consequently, one of the ultimate goals of the medical care in CF – achieving and maintaining an adequate nutritional status – will be jeopardised rather than potentiated.

Therefore, nutritional education and treatment in CF can be considered as one of the ideal targets of mobile health (mHealth) and patients' self-management. These concepts have been pointed out as main priorities in the current European Union's research and innovation programme, Horizon 2020. They strongly suggest that the current and future lines of medical investigation should be focused on this area [8].

In this scenario, MyCyFAPP Project (www.mycyfapp.eu) came to fruition as an innovative approach towards the self-management of nutrition and PERT in paediatric patients with CF by means of a mobile APP. The whole project includes various studies in order to generate educational resources and supporting tools addressed to patients, to ultimately achieve the overall goal.

The present study was carried out to obtain baseline information about nutrient and PERT intake and identify possible discrepancies with the current recommendations. This information will help to develop CF-specific, tailored evidence-based tools to allow for the self-management of nutrition and PERT in order to facilitate adherence to current recommendations. We therefore assessed nutritional status, daily energy and macronutrient intake, and enzyme dosing of a European population of children with CF and related these findings to the new ESPGHAN/ECFS/ESPEN nutritional guidelines [7].

2. Methods

2.1. Subjects and study design

Patients with CF considered for enrolment in this cross-sectional study were regularly followed in the 6 MyCyFAPP participating European CF Centres: Lisbon (Portugal), Madrid (Spain), Valencia (Spain), Milan (Italy), Leuven (Belgium) and Rotterdam (The Netherlands). Inclusion criteria required a confirmed diagnosis of CF for at least 6 months and age between 1.0 to 17.9 years. Patients who had undergone organ transplantation were excluded. Presence of pancreatic sufficiency (PS) was not considered an exclusion criterion.

The anthropometric measurements were taken and clinical data registered. Prospectively, participants completed a newly developed 4-day food and pancreatic enzymes record. The collected information was securely stored in the online MyCyFAPP project system, allowing for data storage and automatic calculations. Participants followed their usual diet and took their regular enzyme dose as prescribed by the specialists in their centres.

The study protocol was approved by the ethical committee of each CF centre and conducted according to the Declaration of Helsinki guidelines. All parents and patients were informed about the purpose and ultimate aim of the study and were asked to sign the informed consent.

2.2. Clinical and anthropometric data

The anthropometric data were obtained by trained personnel of the CF centres. After calibration of the equipment, weight was measured using a digital scale to the nearest 0.1 kg and height was obtained with either a measuring board (supine length) or stadiometer (standing height). Body mass index was calculated as weight (kg)/height (m^2). Anthropometric measurements were converted to standard deviations scores e.g. "height for age" (HFA), "weight for age" (WFA), and "body mass index for age" (BMI) using the CDC references [9].

2.3. Nutritional data collection and calculation

The specific 4 days food and enzymes record form (FER) was developed, based on current recommendation that a 3 to 5 day diet record is necessary for a quantitative evaluation of nutrient intake [7]. The FER was structured in six sections: 3 main meals (breakfast, lunch and dinner) and 3 snacks (morning, afternoon and night). Patients were asked to indicate the exact time of each meal, the name of the dish, ingredients/food products it contained and the amount taken, as well as the PERT dose. The FER also included a household measure table with equivalences in grams