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Evaluating the impact of 2006 Australasian Clinical Practice Guidelines for nutrition in children with cystic fibrosis in Australia

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ABSTRACT

Objectives: To determine the association between the implementation of the 2006 Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis (CF) and the nutritional status of children participating in the Australian Cystic Fibrosis Data Registry (ACFDR).

Methods: This research consisted of a quantitative study using ACFDR data and a survey of clinicians and dietitians treating children with CF. Two independent cohorts of children (2–5 years and 6–11 years) were selected from ACFDR between 1998 and 2014 (N = 2304). Generalised estimating equation model was used to assess weight, height and body mass index (BMI) *z-scores* for each patient before and after the implementation of the nutrition guidelines. A nationwide online survey was sent to 48 clinicians to explore the enablers and barriers to implementation of the guidelines.

Results: Data analysis showed significant increase ($p < 0.05$) in mean weight, height and BMI *z-scores* ranging from 0.06 to 0.18 after implementation of the guidelines in both cohorts of children. Nineteen (39%) clinicians participated in the survey. The majority of the respondents adopted the recommendations into their practice and used the guidelines as part of their professional development. Structural barriers included a lack of adequate staff resources and clinic space for consultations, inappropriate staff classification, high staff turnover and lack of mentoring support.

Conclusion: In children participating in the ACFDR, nutritional status improved after the implementation of the 2006 guidelines. Survey results revealed enablers and barriers to guideline implementation and will inform implementation strategies for the revised Australasian nutrition guidelines for CF, released in 2017.

1. Introduction

Cystic Fibrosis (CF) is a common, genetically acquired, life-shortening chronic illness affecting primarily the lungs and digestive system due to a malfunction in the exocrine system, responsible for producing saliva, sweat, tears and mucus [1]. More than 30,000 people with CF live in the USA and approximately 3300 in Australia [2,3].

Manifestations of the disease often include frequent respiratory infections resulting in progressive scaring of lung tissues and impaired absorption of nutrients resulting in suboptimal weight gain and growth [4]. Both malnutrition and poor lung function are associated with an increased risk of mortality, therefore, treatments and interventions in patients with CF need to aim at maintaining good nutrition and preserving lung function [5–7]. Despite recent advances in management of the disease, poor nutrition remains common due to increased resting energy expenditure, malabsorption and reduced energy intake [4,8,9]. Nutritional status and pulmonary function are closely linked and stunting has been found to be an independent predictor of mortality in patients with CF [10,11]. Good nutrition in early life is particularly important due to rapid physical and cognitive development. Despite improvements, associated with centre-based care, high energy unrestricted fat diets, pancreatic enzyme replacement therapy and newborn screening [12–15], the problems of growth failure and poor nutritional status remain a challenge [10,15,16].

In Australia, following a survey of dietetic practice and management of CF in 2006 [6], the Dietitian Association of Australia published the

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2006 Australasian Clinical Practice Guidelines for Nutrition in CF [17]. The guidelines aimed to reduce variations in practice and improve the nutritional status and quality of life of patients with CF and their families [6,17]. The guidelines made recommendations regarding dietetic staffing levels, nutritional assessment, nutritional requirements, pancreatic enzyme replacement therapy and nutritional support as well as managing complications including pancreatitis and CF related diabetes, and special situations including pregnancy and nutrition for lung transplantation. These guidelines were distributed to dietitians and other health professionals working in specialist CF clinics. They also were published on the websites of professional associations. In addition, workshops for dietitians working in specialist CF clinics were held at the Annual Meetings of the Dietitians Association of Australia CF Special Interest Group during 2007.

Recently, evidence-informed and practice based guidelines on the nutritional care of infants, children and adults with CF have been established including those by the ESPEN-European Society of Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN)-ECFS [9], as well as the Cystic Fibrosis Foundation (CFF) guidelines on enteral tube feeding both published in 2016 [18].

To describe trends and patterns of growth and pulmonary function in people with CF, and to examine longitudinal associations amongst these variables, a rich data source is needed. To collect such information, CF patient registries have been established in the USA, United Kingdom, Europe, Australia and New Zealand [19]. The Australian Cystic Fibrosis Data Registry (ACFDR) is a national registry that was established in 1998, and collects clinical data on patients with CF attending specialist clinics. It captures > 90% CF patients enrolled in the registry [20,21], and at the end of 2015 the ACFDR held records of 3379 Australians diagnosed with CF [2].

The aim of this study was to determine the association between the implementation of the 2006 Australasian Clinical Practice Guidelines for Nutrition in CF and the nutritional status of children participating in the ACFDR between 1998 and 2014.

2. Material and methods

2.1. Study design

This quantitative study consisted of two components: 1) analysis of registry data of children enrolled in the ACFDR, and 2) survey of clinicians treating children with CF in Australia.

2.2. ACFDR data

The ACFDR contains detailed demographic and clinical information of patients with a confirmed diagnosis of CF, receiving clinical care at twenty-three accredited CF centres in Australia (2, 20). Currently there are ten paediatric specialist CF centres participating in the ACFDR. On a regular basis, for each patient the registry collects data regarding lung function, nutrition, mutation, respiratory microbiology, hospitalisation and treatment. The registry provides detailed annual and centre-specific reports, distributed to the clinicians in the participating sites. A detailed description of the registry is described elsewhere [2,21].

In this study, nutritional data of children from two independent cohorts, 2–5 and 6–11 years of age, were collected longitudinally from 1998 to 2014. We have chosen two different age cohorts due to differences in rates of normal growth, eating behaviour and development of children between these two groups. This choice was based on the Centres for Disease Control and Prevention Growth Charts [22].

2.3. Survey

A simple ten-question anonymous survey was conducted among medical staff (paediatric consultants and fellows in respiratory/thoracic medicine and registrars in an accredited respiratory medicine program) and dietitians across Australian paediatric specialist CF centres aiming to explore enablers and barriers to the implementation of the 2006 nutrition guidelines at their centres. Doctors were identified through their centre's involvement in the ACFDR and dietitians were identified through the Dietitians Association of Australia CF Interest Group network.

The survey was based on the theoretical domains framework [23] to examine current practice and identify enablers and barriers associated with the implementation of the guidelines. The survey consisted of the following four components: 1) demographic characteristics of participants, 2) implementation of the guidelines, 3) guideline recommendations, and 4) evidence for the guidelines. A survey was delivered online via Qualtrics Survey Software [24] between August 2017 and February 2018.

2.4. Outcome measures

Nutritional parameters analysed in this study were weight, height and body mass index (BMI) function measurements *(z-scores)* obtained on the occasion of the best lung function annually. *Z-scores* represent child's or adolescent's weight, height or BMI relative to the distribution of weights/heights/BMI observed in a normal reference population of children and adolescents of the same sex and age, transformed onto a standardised scale representing the signed distances from the population mean divided by the standard deviation. These scores are compiled for children and adolescents aged from 2 to less than 18 years using the tables published by Centres for Disease Control and Prevention [25].

2.5. Confounding factors

Age, sex, pancreatic status, dornase alfa (*Pulmozyme*) therapy, number of ever-colonised positive sample of *Pseudomonas aeruginosa* and presence of a G551D mutation were considered as potential confounders in this study.

Dornase alfa (*Pulmozyme*) was included to the analysis as a possible confounder because of its mucolytic therapy use, which might be an indication of a more severe lung disease and could potentially result in poorer nutritional outcomes in children undertaking this therapy [26]. Dornase alfa (*Pulmozyme*) was listed on the Australian Pharmaceutical Benefits Scheme in 1996 and recommended for children age of six years or older [27]. In 2009 the listing was extended to those under five years of age with more severe lung disease. Compared to the USA, the lower rate of dornase alfa (*Pulmozyme*) use in Australia was likely to be due to strict qualification criteria for government subsidisation of this expensive medication, which in 2003 limited ongoing use to those who had demonstrated an improvement in lung function of at least 10% within a month of initiating treatment [28].

Another major factor associated with increased morbidity and mortality and, therefore, poorer nutritional outcomes in CF patients is chronic *Pseudomonas aeruginosa* infection [29,30]. Starting from the late 1990s, eradication treatment for *Pseudomonas aeruginosa*, to prevent or delay chronic infection, became more widely adopted [31] with the process formally recognised in the 2008 CF Standards of Care in Australia [32]. We accounted for the possible influence of improved treatment and medication in our analysis via adjusting for the number of ever-positive *Pseudomonas aeruginosa* results recorded in the registry.

Recent studies have shown that the use of CF transmembrane conductance regulator (CFTR) modulator *ivacaftor* in responsive gating mutations, improves the nutritional outcomes of CF patients [33–35]. Widespread use of *ivacaftor* in Australia did not occur until 2014 after approval by the Therapeutic Goods Association for use in children of six years of age and more with G551D mutation (https://www.tga.gov.au/auspar/auspar-ivacaftor). A small number of children also had access to the treatment in Phase III clinical trials conducted between 2009 and 2011; however, the exact number of patients is not known, as this was not recorded in the registry [36,37]. In 2015, 6.4% of the total CF