

Review

# Nutritional intervention in patients with Cystic Fibrosis: A systematic review

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Received 25 July 2012; received in revised form 12 November 2012; accepted 12 November 2012  
Available online 9 December 2012

## Abstract

**Background:** To systematically assess the literature published after 1997 describing the effectiveness of nutritional interventions in Cystic Fibrosis patients.

**Methods:** An online search in PUBMED, EMBASE and COCHRANE databases was conducted. Original studies with 4 patients or more, describing a nutritional intervention and giving at least weight as an outcome parameter were included.

**Results:** The inclusion criteria were met by 17 articles, focusing on respectively behavioural interventions (n=6), oral supplementation (n=4) or enteral tube feeding (n=7). This latter intervention was universally successful to induce weight gain. One behavioural study and 2 oral supplementation studies also reported significant weight gain.

**Conclusion:** Enteral tube feeding is effective to improve nutritional status, while the described effects of behavioural intervention and oral supplementation are not consistent at present.

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**Keywords:** Diet; Body size; Gastrostomy; Cystic Fibrosis; Nutrition assessment

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**Abbreviations:** CF, Cystic Fibrosis; FEV<sub>1</sub>% pred., Forced expiratory volume in 1 s expressed as % of predicted; RDA, Recommended daily allowances; BMI, Body-mass-index.

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## 1. Introduction

Cystic Fibrosis (CF) is the most common lethal genetic disorder in Caucasians, affecting 1 in 4750 live births [1]. It is characterized by a gradual decline in pulmonary function, intestinal malabsorption and often an impaired nutritional status. Lung disease and nutritional status are tightly intertwined [2] and both are strong predictors of morbidity and mortality in patients with CF [1,3,4]. Malnutrition, due to a negative energy balance, is a common problem caused by a combination of faecal fat losses and increased energy requirements due to chronic infections [5]. Therefore, dietary guidelines prescribe that patients with CF should attain up to 200% of the recommended daily caloric intake [6,7]. However, this can be difficult to achieve because patients may have reduced appetite, infection-related anorexia, gastro-oesophageal reflux or abdominal pain. In this respect nutritional interventions can be helpful to increase caloric intake. In 1997, Jelalian et al. described in a meta-analysis that all nutritional interventions aimed at gaining weight were successful, including behavioural modifications, oral supplementation, enteral tube feeding as well as parenteral nutrition [8]. As CF treatment, and thus the nutritional status of patients has changed during the last 15 years [9,10], effectiveness of nutritional interventions might have changed too. Therefore, we have conducted a systematic review of the literature published after 1997, describing the current effectiveness of interventions aimed at enhancing nutritional status in patients with CF.

## 2. Methods

An online search in PUBMED, EMBASE and COCHRANE Central Register of Controlled Trials was carried out for all available articles published from the 1st of January 1997 up to April 30th, 2012. The search query was: ‘Cystic Fibrosis’ [MESH] AND ‘Nutritional Status’ [MESH], ‘Cystic Fibrosis’ [MESH] AND ‘Diet’ [MESH], ‘Cystic Fibrosis’ [MESH] AND (‘Body Size’ [MESH] OR ‘Body Weight’ [MESH]), ‘Cystic Fibrosis’ [MESH] AND ‘Gastrostomy’ [MESH] OR ‘Enteral Nutrition’ [MESH]. With this latter search term also studies using (nasogastric) tube feeding were identified. The reference lists of eligible articles and review articles were examined for additional studies. Excluded were articles concerning animals,

non-English or non-Dutch articles, editorials, reviews, meta-analyses, articles with no abstract available and articles with a minimal sample size of three subjects or less. The search yielded 361 articles which were screened on title and abstract, and considered suitable if a nutritional intervention, with the aim to improve weight in CF patients, was described. Studies conducted in subgroups only, such as patients with CF related diabetes, were excluded. This resulted in 119 publications that were potentially eligible, which were subsequently screened on full text. To pass this final screening it was necessary that the clinical outcome included a weight variable, either absolute weight, z-score weight, weight-percentile, weight percentage, weight-for-height or body-mass-index (BMI), as a result of the treatment applied. Finally 17 articles were appropriate and included in this review. These studies described interventions involving behavioural modification aimed at increasing caloric intake, prescription of oral supplements or enteral tube feeding through a gastrostomy.

The following data were extracted: the name of the first author, country and year of publication, study design, the intervention offered for nutritional rehabilitation, duration of the intervention, size and, if available, gender and age distribution of the study population, initial weight, caloric intake, the duration of follow-up and, if described, pulmonary function assessed as forced expiratory volume in 1 s, expressed as percentage of predicted (FEV<sub>1</sub>% pred.). The primary outcome measurement was the change in weight, either expressed as absolute weight in kilogramme, weight-for-age z-score, weight percentile, percentage weight-for-age, percentage of ideal-body-weight, percentage of weight-for-height, absolute body-mass-index (BMI) in kg/m<sup>2</sup>, percentage BMI or BMI z-score. The secondary outcome measurement was the change in caloric intake per day and/or forced expiratory volume in 1 s expressed as % of predicted (FEV<sub>1</sub>% pred.), if described.

## 3. Results

Nutritional interventions were subdivided into behavioural intervention (n=6) [11–16], oral supplementation (n=4) [17–20] and enteral tube feeding (n=7) [21–27]. The treatment length of the behavioural interventions ranged from 7 weeks [13–15] to one year [11] and the follow-up period from 1 year [11,12,16] to 2 years [13–15]. In two oral supplementation

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