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Title
Cystic fibrosis in children: Clinical practice and research.

Abstract

Cystic fibrosis (CF) is the most common genetically inherited disease among the Causasian population (Spoonhower and Davis 2016). The disease is caused by mutations in the long arm of Chromosome 7, which codes for the cystic fibrosis transmembrane conductance regulator (CFTR) protein, a c-AMP mediated chloride ion channel, which regulates the function of other chloride and sodium channels in the intestinal epithelium (Riordan et al. 1989). Consequently, thickened mucous secretions are formed, mostly in the respiratory and intestinal systems and the losses of sodium and chloride in the sweat result in nutritional deficits, poor appetite and developmental disorder in children (Culhane et al. 2013). Thus, the maintenance of good nutritional status is of vital importance for CF patients.

According to the current guidelines, the energy requirements of children with CF correspond to 120-200% of the estimated average requirements of healthy children with corresponding age, sex and body type, with 35-40% of the required energy coming from fat, 20% from proteins and the remaining 40-45% from carbohydrates (Turck et al. 2016, Castellani et al, 2018). Most CF Centers focus on a high energy density diet combined with pancreatic enzyme replacement therapy, in case of pancreatic insufficiency (Oliver 2016).

Although guidelines have focused on the need of achieving the nutritional needs of CF patients, there is no data on the dietary habits and dietary patterns of children with CF, and how these standards are correlated with clinical and laboratory characteristics of CF patients.

The present presentation is focused in presenting results from the combination of the experience of dietetic clinical practice and research in the field of paediatric cystic fibrosis.

Key references

1. Castellani C, Duff AJA, Bell SC et al. (2018) ECFS best practice guidelines: the 2018 revision. J Cyst Fibros 17, 153-178.
2. Turck D, Braegger CP, Colombo C et al. (2016) ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis. Clin Nutr 35, 557-577.

Key messages

1. The maintenance of good nutritional status is of vital importance for CF patients.

2. Assessment of exocrine pancreatic insufficiency and pancreatic enzyme replacement therapy should be directed by CF-experienced dietitians.

3. Supplementation of sodium when necessary and also fat soluble vitamins based on laboratory results.