Cystic Fibrosis Research News

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

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What was your research question?
Are patients from European CF centres meeting the current guidelines of nutrition? We addressed this question within the frame of MyCyFAPP Project (www.mycyfapp.eu)

Why is this important?
Because fulfilling the most updated recommendations is related to better nutritional and health status. Ensuring patients adhere to the recommendations is the main goal of the guidelines.

What did you do?
We asked 207 patients from 6 European centres (Lisbon, Madrid, Valencia, Milan, Leuven and Rotterdam) to complete a 4-day food record, in which all the meals consumed and the
doses of enzymes were registered by the participants. Then the calculation system of MyCyFAPP Project provided the average consumption of energy, nutrients and enzymes in each centre. Weight and height of each participant was measured. Finally statistical models were then applied to analyse all of the data.

**What did you find?**
The nutritional status was similar for all participants. We found that at least 25% of the patients were below the target weight, height and body mass index for age. 40% of the patients reached the minimum energy intake target. The average nutrient intake values were 14%, 51% and 34% of the total energy for protein, carbohydrates and fats respectively. The mean doses of enzymes varied greatly among the centres, only 14% to 47% of the patients dosages fell into the recommended range. We also observed that the relation of enzymes/grams of lipids varied a lot in the patients from one meal to another.

**What does this mean and reasons for caution?**
Among the 6 centres, extensive variability and inconsistency was identified in regard to the new guidelines on nutrition and enzymes-use. Our findings document the lack of a general and evidence-based criterion to adjust enzymes; this suggests the potential benefit of educational and self-managerial tools to ensure adherence to therapies, both for clinical staff and families.

**What’s next?**
The next stage of MyCyFAPP Project is to address in vitro digestion. This will create a guideline in which to adjust enzyme dosing. Simultaneously, we are developing educational tools to specifically address the nutritional intake imbalances found in the community. This will all conform the self-management APP, which will be tested in a clinical trial to understand the community impact.

**Original manuscript citation in PubMed**
https://www.ncbi.nlm.nih.gov/pubmed/?term=Nutritional%2C+nutrient+intake+and+use+of+enzyme+supplements+in+paediatric+patients+with+Cystic+Fibrosis%3B+a+European+multicentre+study+with+reference+to+current+guidelines