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Nutritional Status and GI Symptoms in Pediatric Patients with Cystic Fibrosis

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NUTRITIONAL STATUS AND GI SYMPTOMS IN PEDIATRIC PATIENTS WITH CYSTIC FIBROSIS

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Background

- Cystic Fibrosis (CF) is a genetic disorder that affects many organ systems including the lungs and the gastrointestinal tract. It is caused by a mutation in the CFTR gene which creates a defective protein that poorly regulates the flow of chloride ions across the cell membrane. This defect causes viscous mucus to accumulate, decreasing lung function and providing an environment for bacteria to colonize. In the GI tract, this defect can lead to serious complications including poor digestion, poor nutrient absorption, increased inflammation, and abnormalities in the gut microbiota[1]. Without treatment, these problems can lead to malnourishment and adverse GI symptoms.
- Approximately 60 to 90% of people with CF suffer from exocrine pancreatic insufficiency (EPI) due to mucous blockage which, in turn, inhibits the release of enzymes needed for digestion[1]. Pancreatic Enzyme Replacement Therapy (PERT) provides patients with the necessary enzymes (amylase, lipase, and protease) that help improve digestion and absorption. Despite PERT, CF patients often have difficulty absorbing enough calories, fat, protein, and fat-soluble vitamins to maintain healthy weight. To account for deficiencies in these nutrients, higher dietary intakes (DRIs) are recommended for people with CF. Maintaining healthy nutritional status can improve the overall health of the lungs and digestive system. Adequate nutritional status leads to less severe symptoms and increased ability to fight off infection[2].

Materials and Methods

- Participants with CF and their siblings without CF were recruited for this study. Each participant completed a food frequency questionnaire and a GI symptom questionnaire, prompting them to answer questions about the frequency and severity of their symptoms.
- The data from the food frequency questionnaire was analyzed to determine the approximate nutrient composition of each food or drink that was reported. These nutrients were then compiled and summarized for each participant. A food intake summary was constructed to contain average daily amounts for 30 different nutrients. These included: calories, carbohydrates, proteins, fats, fibers, vitamins, and minerals.
- The food intake summary was categorized by gender and age group (Ages 3-8; 4-9; 9-13; 14-18). These averages were then directly compared to a chart of daily reference intakes (DRIs) unique to guidelines for children with CF[3] and healthy children without CF[4]. Based on DRIs for each gender and age group, average nutrient intakes were deemed sufficient, deficient, or excessive.
- Nutrient intake averages for CF participants and non-CF siblings were compared using t-tests with unequal variances.
- Frequency of GI symptoms was collected for CF participants and non-CF siblings. These symptoms included presence of: diarrhea, constipation, bloated, oily stool (steatorrhoea), excessive flatus, abdominal pain, and decreased appetite. Relative frequency of GI symptoms was compared for CF participants and non-CF siblings and for CF participants alone based on genotype (homozygous ΔF508 and heterozygous ΔF508).

Objective

To evaluate the nutritional intake and frequency of GI symptoms in CF participants compared to their non-CF siblings.

DEMOGRAPHICS	CF (n=23)	Non-CF Siblings (n=10)
Age (years)	8.7 [3.2]	10.3 [3.6]
Age @ Enrollment		
3-8 years	17 (74%)	5 (50%)
9-13 years	4 (17%)	4 (40%)
14-18 years	2 (9%)	1 (10%)
Gender (male)	15 (65%)	9 (90%)
BMI %tile	56.9 [23.6]	47.6 [29.4]
Caucasian	23 (100%)	10 (100%)
Hispanic	1 (4%)	0 (0%)
Not Hispanic	22 (96%)	10 (100%)
Sweat chloride (mmol/L)	80.0 [19.1]	21.5 [11.4]
Fecal elastase (mcg/g)	104.8 [139.4]	
CF genotype		
Homozygous F508del	13 (57%)	
Heterozygous F508del	9 (39%)	
Other	1 (4%)	

Data presented as mean [SD] or frequency (%)

Table 1: Demographics

Results

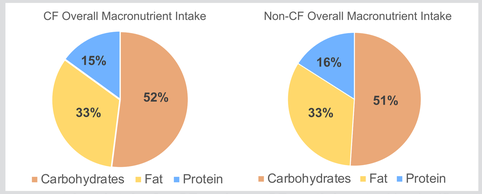

Figure 1: Overall macronutrient intake for CF and non-CF participants. No significant difference was observed between macronutrient intake for CF and non-CF groups. No significant difference in mean total calorie intake was observed between CF and non-CF groups (mean 2749 kcal vs 2733 kcal, respectively).

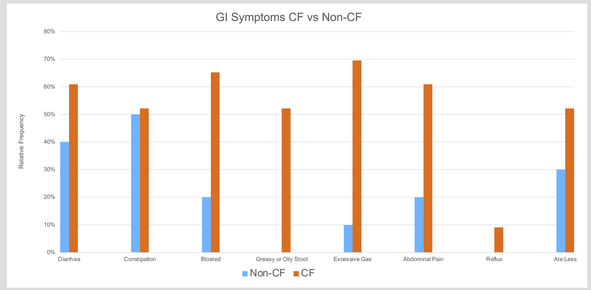

Figure 2: Summary of GI symptoms for CF and non-CF participants. CF participants experienced GI symptoms more frequently than non-CF participants. Bloated: 65% of CF participants reported feeling bloated while only 20% of non-CF participants did; Oily Stool: 52% of CF participants reported having oily stool while zero non-CF participants did. Excessive Gas: 69% of CF participants reported excessive gas while only 10% of non-CF participants did ; Abdominal Pain: 61% of CF participants reported abdominal pain while only 20% of non-CF participants did.

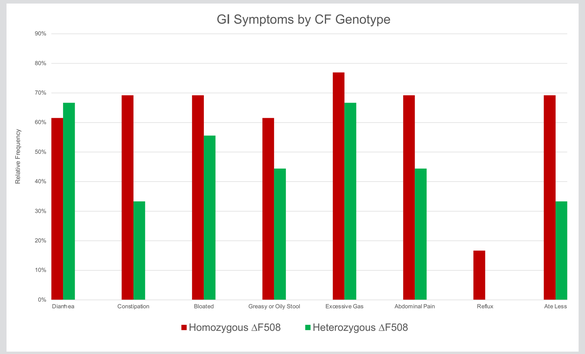

Figure 3: Summary of GI symptoms between homozygous ΔF508 and heterozygous ΔF508 CF participants. On average, homozygous ΔF508 participants experienced GI symptoms 20% more frequently than heterozygous ΔF508 participants. **Homozygous ΔF508 reported:** Constipation & Ate less: 35% more; Abdominal pain: 25% more; Bloated, Oily Stool, Reflux: 15% more; Excessive gas: 10% more.

Nutritional Status Evaluation
Scan QR code to view!
- Contents: Food Intake Summary and DRI Charts.

Results

- Our CF participants were deficient in non-fermentable fiber (52%), vitamin A (48%), vitamin E (96%), and vitamin K (100%). The non-CF siblings were deficient in nonfermentable fiber (60%), vitamin E (40%), and vitamin K (30%).
- Both CF participants and non-CF participants consumed excessive amounts of several nutrients. CF participants consumed excessive calcium (87%), zinc (87%), sodium (87%), iron (91%), and magnesium (83%). Non-CF participants consumed excessive calcium (90%), zinc (70%), sodium (100%), magnesium (70%), and vitamin A (50%). Significant differences were observed between CF and non-CF nutrient intakes of zinc (p=0.05), iron (p=0.04), vitamin A (p=0.03), vitamin E (p=0.003), vitamin C (p=0.05), and vitamin B-12 (p=0.03).

Discussion

- This study measured nutritional intake and symptomatology of CF participants and their siblings. No difference was observed in macronutrient intake between CF participants and their siblings. This could likely be attributed to similar meals being eaten among all members of the household. However, differences in micronutrient intake were present. According to the CFF, it is important that CF patients consume more calories, fat, fiber, zinc, sodium, and fat-soluble vitamins than an individual without the disease[3]. This was achieved in the participants studied.
- Deficient intake of fat-soluble vitamins was most common in CF participants, likely because their RDAs were four to nine times higher than the RDAs of healthy individuals. Both CF and non-CF participants consumed excess amounts of the same nutrients: calcium, zinc, sodium, and magnesium. Excessive values for calcium and zinc came largely from consumption of milk, cheese, and pizza. Excessive values for magnesium came from milk, peanut butter and jelly sandwiches, and energy bars. Lastly, excessive iron levels can be traced back to frequent consumption of cereal and pancakes.
- An obvious difference in GI symptom frequency was observed between CF and non-CF participants. The presence of cystic fibrosis explains the prevalence of these GI symptoms. Studies have shown that patients with the homozygous f508del mutation can have higher degrees of exocrine pancreatic insufficiency (EPI), potentially contributing to more severe GI symptoms[5]. Fecal elastase levels below 184 mcg/g are diagnostic of pancreatic insufficiency. Our homozygotes were more depleted in fecal elastase compared to the heterozygotes (mean 83.1 mcg/g vs 133.4 mcg/g, respectively). These differences in fecal elastase were not statistically significant (p = 0.25). Sweat chloride levels varied significantly by genotype (p = 0.02). Sweat chloride levels of 60 mmol/L or higher are diagnostic of cystic fibrosis. In our sample, homozygotes had higher sweat chloride levels compared to heterozygotes (mean 88.7 mmol/L vs 68.3 mmol/L, respectively). Previous studies have shown that higher sweat chloride levels are associated with failure to thrive (FTT) and more frequent steatorrhoea[6]. Our observations of GI symptoms by genotype support this.
- The main limitation of our study came from the ambiguity of RDA values found for healthy individuals and individuals with cystic fibrosis. Inconsistencies in published RDA values from various sources caused us to choose wider ranges to assess "adequate" intake.

Conclusion and Future Directions

- All participants were eating well and meeting their macronutrient goals.
- Some dietary deficiencies existed in CF participants, including non-fermentable fiber, and vitamins A, E, and K. Exploring their clinical effects will be a future area of focus.
- GI symptoms were common in participants with CF, highlighting the need for interventions in this area.
- Not unexpectedly, GI symptoms in homozygous f508del patients were more frequent than heterozygous f508del patients.
- These findings will be used in larger analyses to assess links between dietary intake and composition of the gut microbiome. Future directions may include using dietary modification to benefit the gut microbiome and GI symptoms in kids with CF.

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