Description of a standardized nutrition classification plan and its relation to nutritional outcomes in children with cystic fibrosis

Amanda Leonard,1 MPH, RD, CDE, Erin Davis,1 RD, CNSD, Beryl J. Rosenstein,2 MD, Pamela L. Zeitlin,2 MD, PhD, Shruti M. Paranjape,2 MD, Donna Peeler,2 RN, Cynthia Maynard,3 PhD, and Peter J. Mogayzel,2 Jr, MD, PhD
1Division of Pediatric Gastroenterology and Nutrition, 2Eudowood Division of Pediatric Respiratory Sciences, The Johns Hopkins Medical Institutions and 3Department of Behavioral Psychology, Kennedy Krieger Institute

Objective Better nutrition enhances lung function and increases survival for children with cystic fibrosis (CF). Therefore, we developed a standardized strategy to evaluate nutritional status and create individualized treatment plans to ensure that all patients received the same high-quality care in a busy CF Center.

Methods A quality improvement approach was undertaken to develop a novel nutrition classification strategy to identify and treat children with subtle manifestations of nutritional deficits in addition to those with obvious nutritional issues. Results During the 15-month study period, the median body mass index (BMI) percentile increased from 35.2 (0–95.9) to 42.0 (0–97.7), p < .005. Additionally, the number of children with a BMI ≥ 50th percentile increased by 11.8%. Conclusions Adoption of a standardized approach to nutritional assessment and treatment led to significant improvement in nutritional outcomes of CF patients, demonstrating that systematic changes in clinical practice can improve clinical outcomes substantially over a short period of time.

Key words cystic fibrosis; nutrition; quality improvement.

A hallmark of cystic fibrosis (CF) is malnutrition leading to poor weight gain. Many factors contribute to inadequate weight gain and malnutrition in CF (Borowitz et al., 2005). Pancreatic insufficiency, found in ~85–90% of the CF population, can be difficult to overcome, even with pancreatic enzyme replacement therapy. A variety of gastrointestinal disorders, including gastroesophageal reflux, constipation, bacterial overgrowth, and distal intestinal obstruction syndrome (DIOS), can decrease caloric intake and nutrient absorption (Borowitz et al., 2005). Worsening lung disease increases caloric needs while also decreasing appetite in some cases. Undiagnosed, or inadequately treated, CF-related diabetes (CFRD) can cause rapid weight loss (Rolon et al., 2001). Behavioral barriers to optimizing nutrition in CF patients have been well described (Stark & Powers, 2005). Finally, socioeconomic and cultural barriers can also affect nutritional outcomes (Schechter, Shelton, Margolis, & Fitzsimmons, 2001).

Nutrition plays a vital role in the health of individuals with CF. In their classic article, Corey et al. (Corey, McLaughlin, Williams, & Levison, 1988; Emerson et al., 2002; Peterson et al., 2003; Steinkamp & Wiedemann, 2002) demonstrated a marked survival advantage in children with better growth parameters. This relationship has been supported by several studies showing that better nutritional status is associated with better lung function, (Milla, 2007). Additionally, inadequate nutrition early in life has been shown to affect lung function later in life (Konstan et al., 2003). Using data from the US CF Foundation Patient Registry, an expert panel convened by the CF Foundation identified a significant correlation between body mass index (BMI) and lung function (Stallings, Stark, Robinson, Feranchak, & Quinton, 2008). These findings, which were widely disseminated in the CF community prior to their publication, led to an emphasis on maintaining individual BMI at or above the
Achieving good nutritional outcomes is not always easy. Review of clinical parameters, such as BMI, at CF care centers within the US reveals wide variation in results (Quinton & O’Connor, 2007). This disparity in outcomes among CF care centers has led the US CF Foundation to launch a far-reaching quality improvement (QI) initiative (www.cff.org/LivingWithCF/QualityImprovement/) that advocates the use of CF Foundation guidelines, evidence-based medicine, and benchmarking of best care practices to develop standardized treatment algorithms. The goal of this program is to encourage the implementation of best practices to improve clinical outcomes (Quinton & O’Connor, 2007).

The Johns Hopkins CF Center pediatric care team embarked on a QI initiative in 2005, which was designed to improve the nutritional outcomes for children with CF. At that time, the median BMI for the Johns Hopkins CF Center was below the national average. Specifically, our goals were to increase the median BMI percentile of our patients and decrease the number of children with suboptimal nutritional status, defined as a BMI <50th percentile. We hypothesized that the systematic application of a standardized evaluation and treatment algorithm would improve nutritional outcomes for children in our clinic. This project was championed by the physicians and dieticians within the CF Center and coincided with our adoption of the CF Foundation’s guideline that the BMI for patients should be ≥50th percentile (Stallings et al., 2008).

Methods

This study was approved by the Institutional Review Board at the Johns Hopkins Medical Institutions. This was a prospective, observational study of a QI initiative within the Johns Hopkins CF Center from September 1, 2005, to November 30, 2006.

Subjects

During the 15 months of the study, all children aged 2–21 years who had not undergone lung transplantation were evaluated. Children <2 years of age were not included in the study because the initial visit for many of these subjects was shortly after establishing the diagnosis of CF. Therefore, improvements in nutritional status would most likely result from initiating CF therapies, such as pancreatic enzyme replacement, rather than the use of our nutrition classification system and treatment algorithm. All subjects were pediatric patients at The Johns Hopkins CF Center.

Planning the Intervention

Although it is well accepted that optimizing nutrition is important in CF care, this goal has proven difficult to achieve at many CF Centers (Quinton & O’Connor, 2007). To date, only a few approaches for optimizing nutrition in CF patients have been studied in a systematic fashion. We used these studies in conjunction with “best practice” observations derived from CF Foundation benchmarking of care centers with excellent nutritional outcomes to develop a standardized approach to nutrition classification and therapy. We began by developing a novel classification system that focused attention on children that had any evidence of nutritional problems. In addition to assigning a nutrition risk category to each patient, a standardized treatment algorithm was created to ensure that all patients received the same high quality care (see Supplementary Material). Treatment was geared toward: (a) maximizing caloric intake; (b) optimizing nutrient absorption; and (c) diagnosing and treating CFRD. The primary goal of this classification and treatment strategy was to identify affected children and establish a therapeutic intervention as early as possible.

A core group from the multi-disciplinary staff attended the Learning and Leadership Collaborative IV sponsored by the CF Foundation to receive training in QI processes (Quinton & O’Connor, 2007) and design an initial project to improve nutritional outcomes. The entire multidisciplinary team attended weekly QI meetings that focused on developing overall goals and specific interventions to improve the nutritional status of children with CF.

The available literature including observations about “best practices” was reviewed. An intervention strategy was developed that entailed systematically categorizing each patient’s nutritional status, developing a treatment algorithm, and implementing this revised practice pattern in a busy CF clinic. The team agreed that in order to achieve the goal of improved nutritional outcomes these algorithms needed to be applied systematically and that frequent follow-up for children with nutritional deficits would be of critical importance.

Initial Assessment

Nutrition risk classification was completed either by the physician with verification by the dietician or by the dietician alone during each clinic visit between September 1, 2005, and November 30, 2006 (see Supplementary Material). The initial assessment and assignment of a nutrition classification category was completed at the first visit.
during the study period. Both sick and routine quarterly clinic visits were included in the study.

**Classification**

During the planning stage of this project, we created five nutrition classification categories (Table I) that were based on a modification of the CF Foundation pediatric nutrition consensus statement (Borowitz et al., 2002). Children were defined as: nutritional “failure” if the BMI was <10th percentile for age, “at risk” if the BMI was 10–24th percentile, “acceptable” if the BMI was 25–49th percentile, and “optimal” if the BMI was ≥50th percentile. Minimal height and weight percentiles were also utilized in the evaluation process (Table I). The unique aspect of our classification schema was the use of a “concerning” category designed to identify and promote early intervention for those individuals with a BMI ≥25th percentile who exhibited any weight loss since the previous clinic visit, no weight gain, or a downward crossing of percentiles over the previous three months. We continued to use the term “acceptable”, rather than a new description such as “sub-optimal”, because a BMI ≥25th percentile had previously been considered adequate in our clinical practice and by the CF Foundation. The use of the term “acceptable” was adopted to encourage patients and parents to aim for a higher BMI percentile without diminishing their previous efforts to maintain optimal nutrition. We did not want patients to feel stigmatized by an unflattering label because the bar had been raised for their nutrition goal. The use of the term “optimal” was also new for our clinic.

**Nutrition Intervention**

A nutrition treatment algorithm was developed to standardize care once the need for intervention was identified, with three primary areas of focus: caloric intake, nutrient absorption, and CFRD (see Supplementary Material).

**Caloric Intake**

The caloric intake arm assessed frequency of meals and snacks, use of high-calorie foods/additives, nutritional supplement use, and identification of any barriers to intake. The number of meals and snacks were evaluated as well as the types of foods consumed. If the patient was not already using a high-calorie oral supplement, one was recommended. Barriers to intake were identified (e.g., food availability, time for breakfast before school) and potential solutions were discussed with the patient and family. A referral to a pediatric psychologist was suggested for behavioral intervention when deemed appropriate.

**Nutrient Absorption**

The absorption arm assessed the proper administration of pancreatic enzyme supplements (e.g., appropriately mixed if unable to swallow pills, no generic formulations, dosage, and timing) and acid suppression to maximize pancreatic enzyme efficacy.

A referral to a pediatric gastroenterologist was made if caloric intake and absorption had been optimized and weight gain continued to be inadequate. Referral was also considered if the patient exhibited symptoms of severe gastroesophageal reflux, severe constipation, bacterial overgrowth, or if placement of a gastrostomy feeding tube was indicated.

**CF-related Diabetes**

The CFRD arm assessed premorbid symptoms of diabetes (e.g., polydipsia, polyuria, unexplained weight loss or unexplained decline in lung function). Additionally, a diagnostic evaluation for CFRD was conducted if the patient had a history of casual blood glucose values that were abnormal or if caloric intake and nutrient absorption...
had been maximized and weight gain remained suboptimal. Individuals with possible CFRD underwent an oral glucose tolerance test and a glucometer was prescribed for home use. Blood glucose values (fasting and 2 hr post-prandial) were collected daily for 2 weeks, after which a decision was made regarding referral to a pediatric endocrinologist.

**Behavioral Psychology Intervention**

If behavioral concerns were identified by the care team, referral was made to a pediatric psychologist who evaluated the family and identified specific barriers (e.g., family and psychosocial stressors, family functioning, parent–child interactions, structure of meals, child behaviors, etc.) that could negatively impact the child’s adherence to behavioral and nutritional interventions. Recommendations were provided to parents that focused on behavioral modifications, with an emphasis on differential attention, contingent reinforcement, time-out procedures, stickers, token economies, and direct commands. Parents were also encouraged to refrain from participating in behaviors which included feeding, coaxing, bartering, and physical prompting. Parents were educated on ways to promote structure during meal times and were encouraged to set strict rules and expectations at meals, to limit meals to no longer than 20 min, and to implement contingencies for eating and non-eating.

**Nutrition Education**

Patients and families received education on the importance of nutrition for the health of CF Patients. Education was individualized to address the needs of the each patient and family. Examples of topics covered include tips for adding calories, suggestions for meal planning, assistance with enzyme dosing and administration, and trouble shooting for barriers to nutrition intervention identified during the visit. At each visit, patients received a trend of their weight, height and BMI percentiles over the preceding 1–2 years as well as written goals.

**Implementation**

The team realized that the introduction of this QI project would require several changes in clinic. For example, the dietician needed to evaluate the trend of weight gain for every patient that was seen in clinic, not just those that lost weight or had obvious poor weight gain. Additionally, more frequent follow-up would be required for many children, thereby increasing the number of patients being seen in each clinic session.

To pilot this new nutritional intervention program, we first introduced it in one of the 3 weekly CF clinic sessions utilizing a single dietician. This allowed several, short plan, study, do, act (PDSA) cycles to be performed to refine the program before it was introduced into all the clinic sessions. These analyses caused us to change the template of appointment times and make adjustments to clinic flow to better utilize the dietician’s and other providers’ time. Clinic paperwork was simplified and a white board was installed to better track the location of patients and providers. Additionally, shorter “nutrition” follow-up visits were scheduled at the beginning and end of each clinic session.

Another change to our previous clinic practice included the use of an algorithm to provide structure to the treatment interventions (see Supplementary Material). Finally, the use of the “concerning” category focused our attention on children with an otherwise “acceptable” or “optimal” BMI who had subtle growth issues. These changes in practice were agreed upon by all the physicians and dieticians seeing children with CF.

**Measuring the Response to Nutrition Interventions**

Response to nutrition intervention was evaluated by assessing change in BMI percentile for individual patients as well as for the entire clinic population using run charts. Additionally, the number of children in each nutrition risk category was assessed. Run charts were utilized to track the changes in the median BMI of the clinic and the number of patients in each nutrition risk category over time. Median BMI percentile for those patients with at least two clinic visits during the study period was compared before and after the intervention using the Wilcoxon Signed-Rank test.

**Results**

**Subjects**

There were 247 children evaluated, of whom 213 had had at least two clinic visits during the study period. The median age (range) for all subjects was 12.4 (2.1–21.5) years, with equal distribution by gender of 123 females (49.8%) and 124 males (50.2%). Twenty-three (9.3%) of the patients were pancreatic sufficient.

**Initial Assessment**

The initial median BMI percentile of all 247 patients in this study was 37.8 (0–95.3). A nutrition risk category was assigned to each patient during their initial clinic visit (Fig. 1). Seventy-nine (31.9%) of the patients were classified as “optimal”, 53 (21.5%) of the patients as “acceptable”, 46 (18.6%) of the patients as nutritionally “at risk”, and 52 (21.1%) of the patients in nutritional “failure.” There were 17 (6.9%) patients classified as
“concerning” at the initial visit during the study period. The most common cause of weight loss or poor weight gain in this group was poor appetite, which occurred in 7 (41.2%) of the patients. Other reasons for the “concerning” classification included: weight loss associated with acute illness occurred in 6 (35.2%), skipping meals in 2 (11.8%), malabsorption in 1 (5.9%), and increased activity in 1 (5.9%).

### Age Distribution

There was an age-related trend in the distribution of nutrition classifications (Fig. 1). The 2- to 5-year-old group had the highest percentage of “optimal” and “acceptable” patients (66.7%). The percentage of subjects within these categories decreased in the older cohorts so that only 18% of the ≥16-year-old group was considered “optimal”. These results confirm previous findings that nutritional status varies with age (Lai, Cheng, & Farrell, 2005).

### Nutritional Intervention

Patients classified as “acceptable”, “concerning”, “at risk”, or in nutritional “failure” received an intervention according to a treatment algorithm that included three arms as detailed above. Interventions varied based on patient needs and individual treatment plans were developed for each patient, which included frequent follow up in clinic and by phone.

The response to our nutritional intervention over time was monitored using run charts. Figure 2 demonstrates the change in median BMI percentile over time for all patients seen in the clinic and for the cohort of 213 patients who were seen more than once during the study period. The median BMI for the patients who had at least two clinic visits during the 15-month study period increased 19.3% from 35.2 (0–95.9) to 42.0 (0–97.7), $p < .005$.

A run chart demonstrating the nutrition classification at each clinic visit is shown in Figure 3. During the first 2 months of the study, 10–11% of the patients were not evaluated. With refinement of the clinic flow we were able to assess every patient by the 5th month of the study. During the course of the study there were 120 visits when a child was classified as “concerning.” At the subsequent visit, 37 (30.8%) of the children were classified as “optimal”, 40 (33.3%) were “acceptable”, 27 (22.5%) remained “concerning”, 8 (6.7%) were “at risk”, and 8 (6.7%) were in “failure.”

Figure 4 shows nutrition classification at the beginning and the end of the study period for the cohort 213 patients with at least two clinic visits during the study. The number of patients in the “optimal” and “acceptable” ranges increased while the number of patients in both the “at risk” and “failure” categories decreased. The greatest improvement was observed in...
children moving from “at risk” to “optimal” or “acceptable” groups. The number of children in the “nutritional failure” group remained relatively stable, which was not an unexpected finding because their heights, which were often <5th percentile, were unlikely to change during the study period. The percentage of children considered either “optimal” or “acceptable” increased from 61.5% to 70.4% during the study period. The number of children with a BMI ≥50th percentile increased from 76 to 85, an increase of 11.8%. Patients were not classified as “concerning” for this analysis.

**Discussion**

Maintaining optimal nutrition is a challenge for both CF patients and caregivers. We have shown that a systematic approach to classifying and treating nutrition in a busy CF clinic can improve outcomes in a short period of time. Our goal of creating a standardized approach to both nutritional assessment and treatment of CF patients improved the median BMI percentile in our clinic by 19% in just 15 months.

The rationale for developing our classification system was to identify not only those children with obvious nutritional issues, but also to recognize those children who had subtle manifestations of nutritional deficits regardless of their nutritional category. Any child with a BMI <50th percentile was thought to need more aggressive intervention. Additionally, those children classified as “concerning” may have had an otherwise “optimal” BMI, but had additional problems such as weight loss or slower than anticipated weight gain. We believe that the increased attention given to these children allowed us to alter weight trends before these children developed significant nutritional deficiencies, which would be more difficult to treat and reverse. However, the improvements in nutrition were not limited to those children with subtle problems since the number of children in the “at risk” and “failure” groups also decreased during the study.

Suboptimal nutrition in CF patients is often multifactorial. Therefore, we chose to develop treatment plans that encompassed three aspects known to affect nutritional outcomes in CF: caloric intake, nutrient absorption, and CFRD. The interventions were coupled with education about nutrition and its importance in improving CF patient outcomes.

Increasing caloric intake can be a challenge in CF patients. Since CF patients have a higher energy expenditure compared to unaffected individuals, they require increased caloric intake to maintain adequate growth.
Additionally, these requirements are increased in children with more advanced lung disease (Moudiou, Galli-Tsionopoulou, Vamvakoudis, & Nousia-Arvanitakis, 2007). Clearly, modifying behavior is critical to the success of nutritional interventions (Stark, 2003). Several studies have demonstrated that behavioral interventions can improve caloric intake in children with CF (Stark et al., 1996, 2000). However, barriers to increasing caloric intake may be difficult to overcome. For example, many families find it difficult to alter their typical routines and set aside time for additional snacks during the day. The availability of Behavioral Psychology services was of great benefit in identifying and overcoming behavioral barriers.

Nutritional supplements, either oral or delivered by gastrostomy tube, are frequently prescribed to enhance caloric intake. However, these supplements have not been shown to consistently improve nutritional outcomes, especially when compared to behavioral interventions (Koretz, Avenell, Lipman, BraunSchweig, & Milne, 2007; Smyth & Walters, 2007). Additionally, there are no randomized control trials to support the use of enteral feeding tubes in CF patients (Conway, Morton, & Wolfe, 2008). Appetite stimulants are also widely prescribed, but there are no large trials confirming their usefulness. Gastrointestinal problems, including gastroesophageal reflux, constipation, bacterial overgrowth, and DIOS can affect appetite and the ability to gain weight (Borowitz et al., 2005). Caloric needs of CF patients may be further increased due to acute illness, making weight gain even more difficult. Enhancing absorption of calories by appropriate use of pancreatic enzyme supplements is vitally important. Drugs that block gastric acid production are frequently used to enhance the function of these enzyme preparations, but there are no large studies to support this practice. CFRD has been associated with worse lung function and survival, especially for females with CF (Marshall et al., 2005; Milla, Billings, & Moran, 2005). Undiagnosed CFRD may contribute to poor weight gain, and once treated, may still have a negative impact on overall nutritional status (Rolon et al., 2001). The targeted interventions outlined in this study allowed us to identify the reasons for weight loss, or slower than expected gains, and determine an action plan with the family to improve nutritional status.

Since preventing malnutrition can be difficult in CF patients, care teams must strive to develop the best possible approach to this problem. This study demonstrates that QI techniques can be used to bring about improved nutrition in a CF center. Involvement of the entire care team in identifying an important clinical problem ensured the success of this project. A core group of care providers received training in QI techniques and championed this project. However, the entire care team was involved in its planning and implantation. One of the biggest challenges to successful implementation of this project was to make more efficient use of the dietician’s time. At the beginning of the study, 10–11% of the children were not assessed, however, we were able to assess every patient by the 5th month of the study. This was accomplished by streamlining the clinic paperwork and tracking the location of patients and care providers. Additionally, we strove to decrease the length of time that patients spent in clinic by decreasing nonvalue added time when patients were in rooms without being seen by a member of the team. Achieving this goal allowed us to see more patients in each clinic session without sacrificing patient care or the amount of time that any individual care provider spent with a patient.

Clearly, there are limitations to this study since it is not a randomized trial but rather a reflection of current clinical practice. Although this study was carried out at a single center, we believe that its results can be duplicated at other centers, not necessarily by adopting our exact strategy, but by identifying and systematically addressing an important clinical problem. We have shown that QI techniques can be used to develop a standardized approach to classification and treatment of CF patients with suboptimal nutrition, thereby leading to substantial improvement in a short period of time.

Supplementary Data

Supplementary data can be found at: http://www.jpepsy.oxfordjournals.org/.

Conflicts of interest: None declared.

Received July 22, 2008; revisions received and accepted March 20, 2009

References


