

Families facing cystic fibrosis and the need of percutaneous gastrostomy: what role for parental stress?

Abstract

Malnutrition is still a relatively common complication of cystic fibrosis (CF) and has been recognized as a negative prognostic factor of the disease. Therefore malnutrition has to be promptly and adequately corrected and a step wise approach has been recommended including enteral feeding via percutaneously placed gastrostomy (PEG) tube.¹ In our Center less than 6% of patients in regular follow-up, including small children, developed such a severe malnutrition to become candidate to PEG placement. The aim of this study was to investigate in depth the family atmosphere of CF patients undergoing PEG, with particular regard to the stress that is generated in the relationship between parents and children. PEG requires parents at least one night awakening for pancreatic enzyme administration and often involves disturbed sleep in the parent for the child's awakenings or because the infusion pumps is ringing. For these reasons, we assessed whether the positioning and management of PEG in children with CF imposes an increased stress on parents compared to parents who manage a child with CF without the need for PEG. The sample consisted of 13 participants, 4 of them carrying a PEG still active at the moment. The Parenting Stress Index (PSI) was administered to parents. It was found that the Total Stress level is not higher in those families that every night manages nutrition through PEG in their children. On the other hand, it is important to evaluate the amount of stressful events that a family has to face both related and unrelated to enteral feeding. This gives us an important lesson: the level of stress in a family is not only tied to that universe that we share with these patients, that is the disease, but with a pile of existential events, some of which occurred before the discovery/communication and management of the disease.

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Introduction

Cystic Fibrosis (CF) is the most common genetic autosomal recessive, potentially lethal disorder of the Caucasian population; with an incidence of 1: 2000-2500 live births. It is estimated that 1 in 25 people are healthy carriers of the mutated gene, who are completely asymptomatic. The gene encodes for the CF trans membrane conductance regulator (CFTR) protein, a chloride channel located at the apical membrane of epithelial cells. Its absence or dysfunction causes production of dense and viscous secretions. More than 90% of patients are affected by lung disease, the main cause of morbidity and mortality. However, pancreas involvement is also frequent (70-85% of cases) and is most frequently manifested by pancreatic insufficiency, which, if not adequately corrected with pancreatic enzyme replacement therapy (PERT), causes lipid malabsorption and consequent malnutrition.² In recent decades, quality of life and survival of CF patients have progressively increased: according to the CF Foundation Patient Registry, in 2017 the median of life expectancy was 44 years.³ This increase was made possible mainly thanks to early diagnosis through neonatal screening and to the improvement of treatments (availability of new antibiotics, new physiotherapeutic techniques, nutritional interventions aimed at maintaining a proper nutritional status), as well as by patient care delivered by a multidisciplinary team with specialized professionals. To date a wide spectrum of treatment options are available to counteract the progression of the disease at different organ levels, and to maintain a good nutritional status, and more recently a few CFTR modulators have been developed to correct the basic defect in patients carrying specific genotypes.^{4,5} In patients

with pancreatic insufficiency PERT is necessary in order to minimize nutrient malabsorption, by providing an adequate amount of enzymes at the duodenal level.⁶ Patients with CF are on a free diet, so they must learn to manage the dose of enzymes themselves based on the fat content of the meal to be consumed. Therefore dietitians have to provide information regarding the lipid content of the main foods and the number of capsules to be taken at each meal. Malnutrition is defined by the World Health organization (WHO) as a condition of cellular imbalance between nutrient and energy intake and the amount needed by the organism to guarantee growth, development and specific functions. This imbalance can affect specific macro or micronutrients (proteins, vitamins and minerals), or it can affect the nutrients and the energy balance overall, and may occur in excess, that is when the intake is higher than the expenditure, or in default, that is when the food intake is not sufficient to cover the needs. The latter is the case of CF patients. Pediatric malnutrition acts as slowing or stunting growth. Malnutrition by defect in Cystic Fibrosis is mainly due to an imbalance between the following factors: reduced nutrient intake, increased energy loss, increased energy expenditure. The main causes of reduced nutritional intake concern gastrointestinal (gastro esophageal reflux and recurrent abdominal pain due to malabsorption or constipation), respiratory (dyspnea and cough-related vomiting that occur during pulmonary exacerbations) and psychological aspects (the awareness of the chronicity of the disease can worsen negative beliefs about the future). Enteral Nutrition (EN) consists in infusing specific nutritional mixtures into the gastrointestinal tract using nasal-enteric probes or ostomy. The infusion can be intermittent, that is, with bolus administration at fixed time intervals, or continuous during the night

or during the day. When an artificial supplementation is necessary, EN should always be the first choice whenever the gastrointestinal tract is functioning, since it represents the most physiological route of administration; compared to Parenteral Nutrition it is more easily manageable, has a low probability of side effects, it maintains the anatomical-functional integrity of the intestinal mucosa, and is less expensive. Gastrostomy is a gastro-cutaneous fistula between the anterior wall of the stomach and abdominal skin which represents the access route indicated for long-term nutritional treatments (>30 days).⁷ It can be positioned surgically or percutaneously, using a radiological or endoscopic approach; the endoscopic ally positioned gastrostomy is called Percutaneous Endoscopic Gastrostomy (PEG). As reported by the ESPEN-ESPGHAN-ECFS Guidelines, EN is considered appropriate: in children younger than 2 years with a persistent growth defect (weight and length <10th percentile), in children and adolescents between 2 and 18 years with a BMI (Body Mass Index) constantly <10th percentile or with a weight loss of at least two points in percentile compared to the last visit and stunting, in adults (>18 years) with a BMI constantly <18.5 kg/m² or with a continuous weight loss of more than 5%.⁸ In CF enteral supplementation is recommended to improve anthropometric indices in individuals who are not otherwise able to take an adequate caloric and protein intake to achieve growth and weight maintenance goals.^{9,10} The greatest short-term benefits were observed on weight gain^{11–13} and BMI^{14–18} in both children and adults, while a significant increase in height occurred only in prolonged treatments.^{13,14} As far as the improvement of lung function is concerned, the results are discordant: although some report an increase in FEV₁,^{12–16} most show a stabilization of values.^{15–18}

Need of the study

Patients needing PEG constitute a small minority in CF population. Nonetheless it is crucial for dieticians and psychologists to outline how much the home treatment of this group of patients is even burdened by PEG management requirements. This narrow aim should have to be considered just as a part of larger studies aimed to understand why, for this group of patients, usual interventions fail and give relevant insights about appetite. Ultimately, as health professionals daily in contact with the adherence difficulties and sufferings of patient, we are interested in deepening our knowledge on the relationship between CF children and food in order to prevent as much as possible the need of PEG and develop targeted early interventions and counseling programs.

Objectives

The present study aims to assess if the incidence of Total Stress level in families with a child with CF and carrying a PEG is higher compared to families with a child with CF and without PEG.

Hypotheses:

1. Families with a child with CF and carrying a PEG will express a higher level of Total Stress compared with families with a child with CF and without PEG;
2. To be informed about a chronic, incurable even treatable, disease in a child is considered a stressful/potentially traumatic experience for parents.

Very often health professionals forget that the story of a couple of parents could be studied with other highly stressful/potentially traumatic experiences (both as individuals and as a couple) so that the communication of the diagnosis in the child could come as the umpteenth experience of suffering. Following this premise, it is crucial to know, by means of clinical interviews, if the Total Stress level

expressed by parents is tied to the stress caused by the management of the disease or to stressful/traumatic experiences not linked with the disease itself.

Sample and method

First step: a retrospective cohort study on PEG effects in a larger group

A retrospective cohort study on the whole group of CF patients who underwent PEG between 2000 and 2016 was performed. Sixteen patients regularly followed at the Regional Reference Center for CF (Milan), out of 579 patients in regular follow-up at the end of 2015, were eligible and included in the study. The weight and length/height values were collected and the Z-score of the W/L (based on the growth curves of the WHO) and the BMI by age (using growth reference values for the Italian pediatric population¹⁹) were calculated. The anthropometric data were recorded 6 months prior to placement of PEG, at the time of placement and 6 months after placement were collected. Weight was obtained through the use of electronic scales; length and height were detected respectively through the use of infantometers and stadiometers. Patients were classified with respect to the nutritional goal recommended by the Cystic Fibrosis Foundation (CFF), i.e. the achievement of the 50th percentile of W/L or BMI in children and adolescents (age <20 years) and the maintenance of a BMI $\geq 22\text{kg/m}^2$ for older females and $\geq 23\text{kg/m}^2$ for older males. To identify the state of malnutrition, the cut-offs used were the 10th percentile of BMI for patients up to 19 years and for patients over 20 years 18.5kg/m^2 of BMI. We also found the values of the concentration of albumin and hemoglobin at the time of PEG placement and 6 months later. FEV₁ (% of predicted) was measured by spirometry and data recorded 6 months before the placement of the PEG, at placement and 6 months after placement were used. To evaluate the dietary intake during the period of enteral supplementation, patients were asked to fill in a food diary for 3 days.

Sample characteristics of the retrospective study

Twelve female and 4 male patients, who constituted the intact group of eligible patients were enrolled in the study; median age of 13 years with pancreatic insufficiency in 94% (15 patients). The F508del mutation was present in homozygosity in 7 cases and in heterozygosity in 3. Two patients were affected by CF-related diabetes.

Psychological sub-study: Sample

A sub-sample for a psychological study was selected on the basis of availability and the fact that the family lived near the Center (in Milan or in the nearest hinterland area) The level of parental stress was investigated only in those families with a child within the age of 12 and more adherent to follow-up using a self-administered questionnaire: PSI-SF (Parenting Stress Index-Short Form). Results were expressed in percentiles. Percentiles from 1 to 15 constitute the lowest range while the percentiles from 85 to 100 constitute the range of clinical interest; the percentiles from 20 to 80 constitute the area around the average which is fixed at 50. [20] This evaluation was carried out during periodic meetings of psychological monitoring of patient and family adjustment. 15 parents of 13 CF patients participated in the psychological investigation. A group of control subjects was created evaluating the total stress level also in families with a CF child but not needing EN.

Data analysis

With regard of medical variables, continuous variables were expressed as medians and interquartile ranges, while categorical

variables were absolute frequencies and percentages. The Wilcoxon ranks test was used to verify the significance of the 6-month variations of the Z-score values of W/L and BMI. It is used to compare repeated measures on a single sample to assess whether their population means ranks differ. The McNemar test was used to verify the significance of the 6-month variation in the proportion of patients suffering from malnutrition. In fact, this test is used on paired nominal data. It is applied to 2x2 contingency tables with matched pairs of subjects. The significance threshold of the 2-tail hypothesis tests is 0.05. As far as the psychological results are concerned, because of the small number of subjects, only descriptive analysis was performed.

Results

PEG effects

Malnutrition is present in almost all patients who underwent PEG, with the exception of 3 cases in which the positioning of this device

was necessary due to total aversion to oral feeding and in one case for cleft palate. These 3 patients were younger than 2 years of age and were previously fed via Nose Gastric Tube (NGT). In the period prior to the placement of PEG, nasogastric supplementation was performed in 4 patients, all under the age of 2 years, by means of caloric supplements. The analysis of the collected data shows a significant improvement in BMI: after 6 months of EN: the median values of the Z-score of W/L or BMI increases by about 1 standard deviation. In the 6 months prior to the placement of PEG and at the time of its positioning, 13 patients (81.3%) were classified as malnourished; 6 months after EN by PEG, the proportion of malnourished subjects decreased to 31.3% (N=5). For 8 patients (1 male and 7 female) it was possible to analyze the food diary, compiled during the period of enteral supplementation. During the time of observation, median values of lung function tests did not change significantly. The complications which occurred in the studied group are reported in Table 1 & 2.

Table 1 Effects on

Time Line	Number of malnourished patients	percentage
6 months prior PEG	13	81.30%
6 months after EN by PEG	5	31.30%
Larns Recommended Caloric Intake	Number of patients who filled in the 3 day food diary	Percentage
>100%	7	87.50%
<77%	1	12.50%
percentage of total daily caloric intake provided by en	number of patients	Percentage
<40%	3	37.50%
41% to 55%	5	62.50%

Table 2 Complications which occurred in the studied group

Complication	Number
Mechanical	
Button removed	5
Gastroenterological	
Vomiting	1
Diarrhea	2
Metabolic	
Hyperglycemia	1
Other	
Granuloma	1
Peristomal swelling	1

Psychological results

The studied group includes: 4 mothers of patients currently carrying PEG, 5 fathers of patients not carrying PEG, 6 mothers of patients not carrying PEG. In two cases, both parents of the same patient were interviewed to verify if there were differences in the perception of stress between mother and father. The average age of parents is 39, with a minimum of 31 and a maximum of 54 years. 60% (N=9) received a high school diploma, 27% (N=4) has an eighth grade license and the remaining 13% (N=2) received a degree. All the interviewed subjects, with the exception of one, have a paid job: 8 of them work as workers, 3 as employees, 2 as technicians and 1 as a nurse. The mean age is higher in the group of parents not carrying

PEG (40.5 years against 36). 14 subjects out of 15 received regular psychological support at our Center. The average of patients is 7.3 years; it is lower in non-PEG carriers (6.4 years) than in carriers (9.5 years). At the time of evaluation, the youngest patient is 2 years old, the oldest 12. Out of 13 subjects surveyed, 5 did not attend compulsory school, 5 attended the first year of primary school, 3 secondary school. Pancreatic insufficiency is present in 67% of patients without PEG and in all patients with PEG. The Total Stress levels reported by all participant parents are shown in Table 3. The mean value of Total Stress level was 58.75 with a SD of 9.36 for the group of parents of children with PEG and 31.25 with a SD of 22.88 for the group of parents of children without PEG. The min percentile value recorded in the PEG group of parents is 40 and the max 90, while in the non-PEG group the min percentile value recorded is 5 and the max is 70. Even if the Total Stress levels reported are higher in the PEG group, in both groups the mean percentile value falls within the norm. In the two cases where both parents were interviewed only the parent with worse scores were included in order to avoid a too favorable attitude considering also that in the non-PEG group we have a higher number of participants.

Only one mother reported a percentile value (90) which is in the range of clinical interest. The main source of stress for this mother was the fact that she was unable to find a paid job coupled with the fact that hers was the child with the most important impact of the disease. The value, above average, is justifiable in light of the patient's clinical history: the coexistence of several conditions, in addition to CF, including premature birth (more than a month spent in neonatal pathology at the beginning of his life) and a genetic syndrome. The difficulties that burden the parent are superior to that of other

subjects. It is therefore obvious that perceived stress is greater. Quite paradoxically as far as the domains “Dysfunctional parent-child interaction” and “Difficult child” are concerned, parents of children without PEG reported higher values. In “Dysfunctional parent-child interaction” domain all parents in the PEG group, except the cited mother who scored 95, scored within the norm (between 20 to 70). As far as the “Difficult child” domain is concerned almost all parents in the PEG group scored in the clinical interest area except one mother who scored 80 (min 80, max 100). The mean value for “Dysfunctional parent child interaction” was 61.25 with a SD of 27.01 and in “Difficult child” domain the mean value was 92.5 with a SD of 7.5. In the No-PEG (or control) group the mean value of “Dysfunctional parent-child interaction” was 42.62 with a SD of 34.58, while the mean value in the domain “Difficult child” was 70.62 with a SD of 22.14. This is another evidence that suggest that the perception of the difficulties is not only closely related with medical/treatment needs of the child even if most of the difficulties are placed on the child. In addition, interviewing parents it became clearer that the perceived burden seemed more linked with marriage, family and job dissatisfaction in addition with traumatic past experiences occurred in the life of parents. There were no differences between the stress perceived by mothers and by fathers. Anecdotally, we have to add that in the control group is included a child who obtained an increase in BMI from the 4th percentile (Z-score: -1.77) to the 11th (Z-score: -1.23) within the first 6 months of psychotherapeutic treatment, getting to the 24th percentile (Z-score: -0.71) in the following 6 months of psychological intervention. The crucial element in this case was the highest adherence of the parents to the program indicated and their willingness to get involved for the child and to change behaviors and attitudes that negatively affected the child’s eating behavior.

Table 3 Percentile scores reported by parents.

Mother/Father	Child sex	Total stress %ile	PEG Yes/No
Mother	F	45	Y
Mother	F	60	Y
Mother	M	90	Y
Mother	F	40	Y
Father	M	45	N
Mother	F	15	N
Mother*	F	10	N]
Father*	F	15	N
Mother	F	60	N
Mother**	M	70	N**
Father**	M	25	N**
Mother	M	5	N
Father	M	30	N
Father	M	10	N

*and ** parents of the same child.

Discussion

The present study was carried out to investigate the efficacy and possible complications of home EN through PEG in patients with cystic fibrosis, regularly followed at our Center. Although we have examined a limited sample of patients, nutritional therapy has been effective in improving nutritional status as indicated by a significant increase in BMI (+1 Standard Deviation) in just 6 months

of supplementation. However, we did not observe a significant change in respiratory function (FEV₁). The complications reported in half of the patients, in almost all cases were not severe and easily resolvable. The psychological investigation carried out involves a small number of patients, without having any claim to inference to the entire population. However, it is interesting to note that, even considering its limitations, this study reinforces the concept that adequate support to children’s development and growth makes parents feel supported and less stressed, despite the difficulties that the management of a garrison like the PEG imposes them (awake at night for the administration of pancreatic enzymes, management of possible mechanical or gastrointestinal complications). In fact, the stress values were, as a whole, within the normal range both in the group of parents of PEG carriers and in the group of parents of non-carriers. It should be added that the fact that the selected sample represents a homogeneous population, regularly followed up with their health status and supported from the psychological point of view, contributes to the good obtained values.

We can conclude that Hypothesis a (families with a child with CF and carrying a PEG express a higher level of Total Stress compared with families with a child with CF and without PEG) is not proven as true since the Total Stress values reported are within the norm in the two groups except one. On the other side it becomes clearer that the Total Stress level expressed by parents is more tied to stressful experiences like job, and marital dissatisfaction rather than exclusively related to the disease itself. Very interesting is the fact that this frame changes if we consider how difficult the children are perceived by their parents. This feature suggests that some more research is needed to understand if parents feel stressed by the disease or by the burden due to the load tied to rising one or more children and, at the same time, try to maintain a job position, keeping on the house and take care of their couple relationship.

Conclusion

Cystic Fibrosis is a complex disease that requires the involvement of a multidisciplinary team including specialized doctors, nurses, dieticians, physiotherapists, psychologists and social workers. From the experience of our Center and the analysis of the available literature it emerges that the home EN via PEG is safe and free from serious complications to implement the daily caloric intake, as well as a valuable aid for achieving the ideal weight in patients underweight and/or unable to eat sufficiently by mouth. Patients with PEG must be evaluated from a nutritional point of view at every clinical check and supported in the management of this type of nutritional support, considering the important therapeutic load to which they are subjected daily. The primary purpose of the home treatment program in cystic fibrosis is to achieve normal growth and development preventing complications and exacerbations. To get this aim it is crucial for CF patients to have and maintain an adequate nutritional status. Sometimes psychological difficulties negatively interfere with the achievement of this goal. As soon as the Team becomes aware of this situation it is indispensable to set up a psychotherapeutic treatment in order to prevent the necessity of any kind of artificial supplementation, through PEG or NGT. This way many goals can be obtained:

1. By means of saving patients from surgically positioning of PEG also the quality of life of parents is saved;
2. We save on health costs and
3. Patients and families experience more control over their lives and illness and

4. Patients improve their knowledge of their body and the impact of the disease on it as well.

Following Layard and Clark's conclusions of their crucial trial on increased access to psychotherapy (IAPT),²¹ we can say that psychological interventions can not only be of help but also do not cost-effective given the money savings they allow. Periodic evaluations of the results obtained allow the Team members to sharpen the intervention strategies even in the most complex cases.

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Conflict of interest

The author declares that there is no conflict of interest.

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