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Improved lung function using a therapeutic educational multidisciplinary program in a pediatric cystic fibrosis clinic

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Abstract – Background: Pulmonary disease remains the most common cause of morbidity and mortality in cystic fibrosis. The clearing of mucus by chest physiotherapy is a major keystone of treatment. Daily therapy is time consuming, with respiratory therapy, inhalation therapy and a lot of medications to be taken all day long. Treatment burden becomes always a reason for intra-familial conflicts and stress. Adherence to treatment is a main issue, and influences the efficiency of physiotherapy and inhalation **Objective:** The objective was to implement and evaluate TPE in a pediatric cystic fibrosis consultation. **Method:** Open interventional study in patient ≥6 years of age. Patients were followed on their regular 3-month basis over 2 years. Interventional education by cystic fibrosis physician and nurse took place at each visit with assessment once yearly. At visit 1, a contract defining the content of the educational program was discussed. Primary endpoint: Evolution of FEV1 and FEF25-75%. Secondary endpoint: Evaluation of disease knowledge and quality of life (CFQ®). At the end of the study we tested the knowledge about nutritional aspects of the disease in the study group. **Results:** Out of 32 patients from 6 to 16 years, 17 accepted the study. FEV1% predicted increased 5.3% in the intervention group. **Conclusion:** Using TPE, there is a clear trend to improve lung functions (+2.65% annual rate of change of FEV1% predicted). TPE could be an important strategy to stabilize lung function.

Key words: cystic fibrosis / child / therapeutic education / lung function / quality of life

Résumé – Amélioration des fonctions pulmonaires grâce à un programme multidisciplinaire d'éducation thérapeutique dans une clinique pédiatrique de mucoviscidose. Introduction : L'atteinte pulmonaire reste la cause principale de morbidité et de mortalité dans la mucoviscidose. Le drainage du mucus par la physiothérapie respiratoire est un élément clé du traitement. Le traitement quotidien est lourd, avec drainage autogène, inhalations de médicaments ainsi que de nombreux traitements à prendre tout au long de la journée. La charge quotidienne engendre des conflits intrafamiliaux et un stress important. La compliance au traitement est un enjeu majeur qui influence l'efficacité de la physiothérapie et des inhalations. **Objectifs :** L'objectif est de créer une structure d'éducation thérapeutique au sein de notre consultation pédiatrique de mucoviscidose et de l'évaluer. **Méthode :** étude interventionnelle ouverte chez des patients de ≥ 6 ans. Les patients ont été suivis au rythme habituel, chaque 3 mois pendant 2 ans. Une séance d'éducation thérapeutique se faisait à chaque consultation, soit par le médecin en charge de la consultation, soit par l'infirmière référente. À la 1e visite, un contrat éducatif est défini avec le patient. Objectif primaire : évolution du VEMS et du VEF25-75%. Objectifs secondaires : Évaluation de la connaissance de la maladie et de la qualité de vie des patients (CFQ®). À la fin de l'étude, la connaissance des patients a été testée pour les aspects nutritionnels de la maladie dans le groupe étudié. **Résultats :** Sur 32 patients éligibles, 17 ont acceptés l'étude. Le VEMS% prédit a augmenté de 5,3 % dans le groupe intervention.

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Conclusion : L'utilisation d'un programme d'éducation thérapeutique permet un gain net sur les fonctions pulmonaires (gain annuel du VEMS de +2,65 %). L'éducation thérapeutique semble être une stratégie importante pour la stabilisation des fonctions pulmonaires.

Mots clés : mucoviscidose / enfant / éducation thérapeutique / fonctions pulmonaires / qualité de vie

Abbreviations: ATS: American thoracic society, BMI: body mass index, CF: cystic fibrosis, CFCH: Swiss society for cystic fibrosis, CFQ: cystic fibrosis questionnaire, ERS: European respiratory society, FEF: Forced Expiration Flow, FEV1%: Forced Expiration Volume in 1s, expressed in %, GETHEM: Groupe de travail Education THErapeutique et Mucoviscidose, IPCEM: institut de perfectionnement en communication et éducation médicales, RV: residual volume, TPE: therapeutic patient education, QoL: quality of life, TLC: total lung capacity, VEF: volume expiration flow, VEMS: volume flow measurement.

1 Introduction

Pulmonary disease remains the most common cause of morbidity and mortality in cystic fibrosis (CF). Abnormal airway surface liquid conducting to impaired mucociliary clearance is the basis of lung disease and begins soon after birth [1, 2]. Accumulation of thick mucus in the small bronchi makes the nidus of inflammation and infection, leading to lung destruction. The clearing of mucus by chest physiotherapy is a major keystone of pulmonary treatment.

Daily therapy is time consuming, with respiratory therapy, inhalation therapy and a lot of medications to be taken all day long. Adherence to treatment is, as always, a keystone of efficacy of treatment and a main issue for morbidity and mortality. Treatment burden becomes quite always a reason for intrafamilial conflicts and stress.

Therapeutic patient education (TPE) consists in a way to give more competences to patient and his family to understand and deal with all aspects of his disease, integrated in everyday life of the patient. TPE will change the hierarchy between patient and his medical staff in order to involve patient and his family in the medical decisions. It can be done by acquisition of self-care and adaptation competences.

Educational diagnosis is the first step on the way to education. Its purpose is to asses aspects of patient's life and personality; to identify the needs, attempts and projects; and to evaluate its potentiality. During the 2nd step, an educational contract is discussed with the patient and his family. After this step, periods of training give the opportunity to acquire skills discussed within the contract. Finally, competence testing is used to analyze the progress and to go on with a new contract. It is also the time point for professionals for self-criticism [3].

As CF is a chronic disease, it is predisposed to profit from TPE due to its treatment modalities, which includes amongst others in most patients daily respiratory physiotherapy inclusive inhalation of medications. In addition, the efficacy of physiotherapy and inhalation are particularly dependent of the compliance of the patient, and therefore dependent in parts of the education of the latter.

The aim of our prospective study was the evaluation of the implementation of TPE in our pediatric CF cohort. Our unit was not making any "structured" TPE. The clinical team included a pediatrician pneumologist, a pediatrician, a physiotherapist, a dietetician a nurse and a social worker. Patients were seen at least 4 times a year by the physician and by the other professionals on demand. Two members of the team participated to the IPCEM cursus in Paris and all team had supervisions by the CF team in Nantes.

2 Methods

2.1 Study patients

All patient ≥ 6 yrs of age with confirmed diagnosis of CF (two positive sweat tests [chloride > 60 mmol/l] or a genotype with two identifiable CF-causing mutations), able to perform acceptable-quality spirometry, were eligible to participate in the study. There were no formal exclusion criteria.

2.2 Study design

This was an open interventional prospective study, conducted at the pediatric CF consultation of the university hospital of Lausanne, Switzerland.

The goal of this study was to implement and to evaluate a multidisciplinary program of therapeutic education in pediatric CF patients. The study duration was determined for 2 years time.

We used tools created and validated by the GETHEM (Groupe de travail Education Therapeutique et Mucoviscidose) in France: they allow a large flexibility according to needs and attempt of patients with an organized basis [4–6].

Those tools include flow sheets and some story board used by the investigators, varying according to age groups of children. They cover all CF fields (self-care skills: how to detect a pulmonary exacerbation, adaptation of enzymes replacement according to alimentation, respiratory care, hygiene; adaptative skills: how to react in case of pulmonary exacerbation, in case of midsummer heat, in case of dehydration, how to cope with what other people think; genetics). Each flow sheet is characterized by the age of children, duration of the intervention and extra needs of material. We also used a CD developed by the Swiss Society for Cystic Fibrosis (CFCH): "un voyage à travers l'appareil digestif" dedicated for children, which explain digestion process and needs of enzyme substitution [7].

Patients were followed on their regular 3-month basis in the outpatient clinic, with an annual assessment according to international guidelines [8]. There were no additional visits scheduled. After medical assessment one person of the team was leading an educational session with the patient and his parents. At the first study visit, patient and CF team agreed on an educational diagnosis. This first step allows grasping the different sides of the patient life and personality. It allows to identify is needs, his potentiality and to take in account his attempt.

This led to a contract. This allowed defining the content of next visits, according to patient, family and CF team. Interventional education by the CF physician and the CF nurse took place at each visit. Those interventions were done together with the child and his family, except for adolescent patients where intervention was directed only to the patient. Assessment using specific tools was done every 6 month.

2.3 Study objectives

The primary endpoint was the evolution of FEV1% predicted and FEF25-75% predicted in the intervention group. The secondary objectives were the evolution of RV/TLC, data on anthropometry, quality of life, needs for emergency antibiotherapy (orally, inhaled or intravenous). At the end of the study a knowledge questionnaire was used [9].

2.4 Lung function measurements

Pulmonary function testing was conducted according to ATS/ERS guidelines [10] at all visits. Results of FEV1% predicted were classified according to the ATS/ERS guidelines [11].

2.5 Anthropometry

Weight, height and body-mass-index (BMI) were measured at all visits.

2.6 Quality of life (QoL) (CFQ®) [12, 13]

Health-related QoL was measured using the Cystic Fibrosis Questionnaire (CFQ®) at first and last visit [13].

2.7 Use of emergency antibiotherapy (orally, inhaled or intravenous)

During the study period, the need for emergency antibiotherapy (oral, inhaled or intravenous) for pulmonary exacerbation ("rescue medication") was noted at all visits.

2.8 Knowledge questionnaire

The knowledge concerning nutrition and pancreatic enzymes was addressed at the final visit using the questionnaire developped by Stapleton *et al.* [9]. This score of knowledge ranges from 0 to 28, the higher the score the better the knowledge. We used only this topic as we couldn't find any other validated questionnaire. Our population was to small to allow a validation of a new questionnaire.

2.9 Statistical analysis

The evolution of FEV1% during the study period was compared by means of two-tailed *t*-tests. The same method was applied to compare the evolution of FEV1% the 2 years preceding the baseline visit of the study (visit 1). Prior to analysis, evolution of FEV1% was z-score transformed. A z-score is the deviation from the sample mean expressed as number of standard deviations (SD).

2.10 Ethics

The study was approved by the local ethics committee at the university of Lausanne. Informed written consent was obtained prior to inclusion.

3 Results

The study was conducted between January 2007 and January 2009.

3.1 Patient demographics

Of 32 patients eligible to participate, 17 patients (11 males) were willingly to participate and were therefore included. Age was from 6 years up to 16 years (median age 11). Patient's demographics at visit 1 are shown in the Table I.

Most of the patients were asking for the nutritional and enzyme therapy aspects of the disease (14 patients, 82.4%), other interest were: "Why do I take pills" (10, 58.9%), hygiene (6 patients, 35.3%), "how to deal myself with treatment" (5 patients, 29.4%) and genetics issues (4, 23.5%). Some topics were considered as fundamental and were proposed to all patients once a year (risk of dehydration, how to detect a pulmonary exacerbation). Adolescent patients were more interested in the genetics aspects.

3.2 Lung function

Two years before beginning of the study, 88% of patients in the intervention group had a normal FEV1% predicted, 6% a moderately severe disease (FEV1%: 70–89% of predicted) and 6% a severe disease (FEV1% < 40%).

Table I. Patients demographics at visit 1. Table shows characteristics of patients in both groups at inclusion. Most patients had normal or mildly decreased lung functions. Only one patient had severe lung disease. – *Caractéristiques des patients à l'inclusion. Le tableau montre les caractéristiques des patients à l'inclusion. La plupart des patients avaient des fonctions pulmonaires normales ou légèrement diminuées. Un seul patient avait des fonctions sévèrement diminuées.*

	Study group
Numbers	17
- Male	11
Age median in years (range)	11.0 (5.6–14.8)
Mean weight in kg (range)	33.5 (20.6–55.7)
BMI percentile	16.6
FEV1 (%)	92
– normal	71%
– mild	18%
– moderate	6%
– moderately severe	0
- severe	6%
– very severe	0

Between the beginning and the end of the study period, FEV1% increased by a mean of 0.36 SD in the study group (two-tailed paired *t*-test: $p = 0.028$). For the FEF25-75%, evolution was positive with a mean fall of 8.3% in the study group. However, this result was not conclusive (paired *t* test: $p = 0.118$). For RV/TLC there was also a benefit, with a mean fall of 25.2% in the intervention group .This result is again not conclusive ($p = 0.100$) (Fig. 1).

Considering the time span between 2 years before intervention until the end of the intervention study FEV1% predicted had a mean fall of 0.14 SD in the intervention group (Fig. 1). FEF25-75% had a mean decrease of 2.8% in the intervention group (Fig. 1). RV/TLC decreased of 57.07% ($p < 0.001$)(Fig. 1).

3.3 Anthropometry

Mean weight at beginning of study was of 33.4 kg. Gain of weight on the study time was of 6.4 kg in the study group.

Mean size at beginning of study was of 139.9 cm. Gain of height on the study time was of 9.0 cm in the study group.

3.4 Number of emergency hospitalization / use of oral rescue antibiotics

During the study period, patients were hospitalized a mean time of 3.5 days pro 6 month period in the study group. Only two patients had a decrease of hospitalization time in the study group.

They took 23.2 days of oral rescue antibiotic therapy. Six patients decreased their antibiotic needs over study time.

3.5 Knowledge questionnaire

Scores obtained were between 22 and 28 (between 78.6% and 100% of correct answer). Median was 26 and mean 25.9 (92.5% of correct answers). For comparison, in the Stapleton study, mean correct score was 54% between 5 and 8 years and 72% between 9 and 12 years old children [9]. It was not possible to make statistical correlations between those results and the gain in lung functions because of the low number of patients. Nevertheless, all patients had a better knowledge score than shown in the original article by Stapleton.

TPE is a long term process, and adherence to treatment is not only (unfortunately) understanding of treatment. Some patients were able to explain very clearly how and why to use pancreatic extract but were not yet able to change their way of doing.

3.6 Quality of life (QoL) (CFQ®)

It was not possible to sort out a trend in either items of the CFQ® questionnaire.

4 Discussion

Using TPE, there was a clear trend to improve lung functions (+2.65% annual rate of change of FEV1% predicted) as well as disease-knowledge. Lack of statistical significance in some clinically relevant associations is likely to be due to the small sample size. In motivated patients and families, TPE could be an important strategy to stabilize lung function.

TPE can help to solve adherence difficulties by a better disease understanding as well as a more responsible position of the patient ("understanding better to act better" [3]). This will probably induce a change over time in most treatment issues, with, at the end changes in some physical parameters, as lung functions. FEV1% was about 10% better in the intervention group 2 years before start of the study, moreover, all pulmonary testing parameters were getting worst in the intervention group before inclusion. The finding of a more important decrease in the intervention group who had better FEV1% (≥90%) in the two-years period prior to the study is coherent with the recent findings by de Boeck et al who described a significant higher annual rate of decline of 1.9% predicted in a group of individuals with an FEV1% ≥90% predicted compared with 0.4% predicted in a group with FEV1% < 90% predicted [14]. In this regard of a faster decline of FEV1% predicted in the intervention group prior to the intervention, with an improvement during the study period, most likely due to the intervention (TPE), becomes even more important.

Skills acquisitions on nutritional issues and pancreatic enzyme substitution were made by two different supports: one tool from the GETHEM ("*Expliquer le rôle du pancréas dans la digestion des graisses. Identifier le mode d'action des enzymes pancréatiques*" with a true/false questionnaire) [14] and a CD ("*Un voyage à travers l'appareil digestif*" [7]).

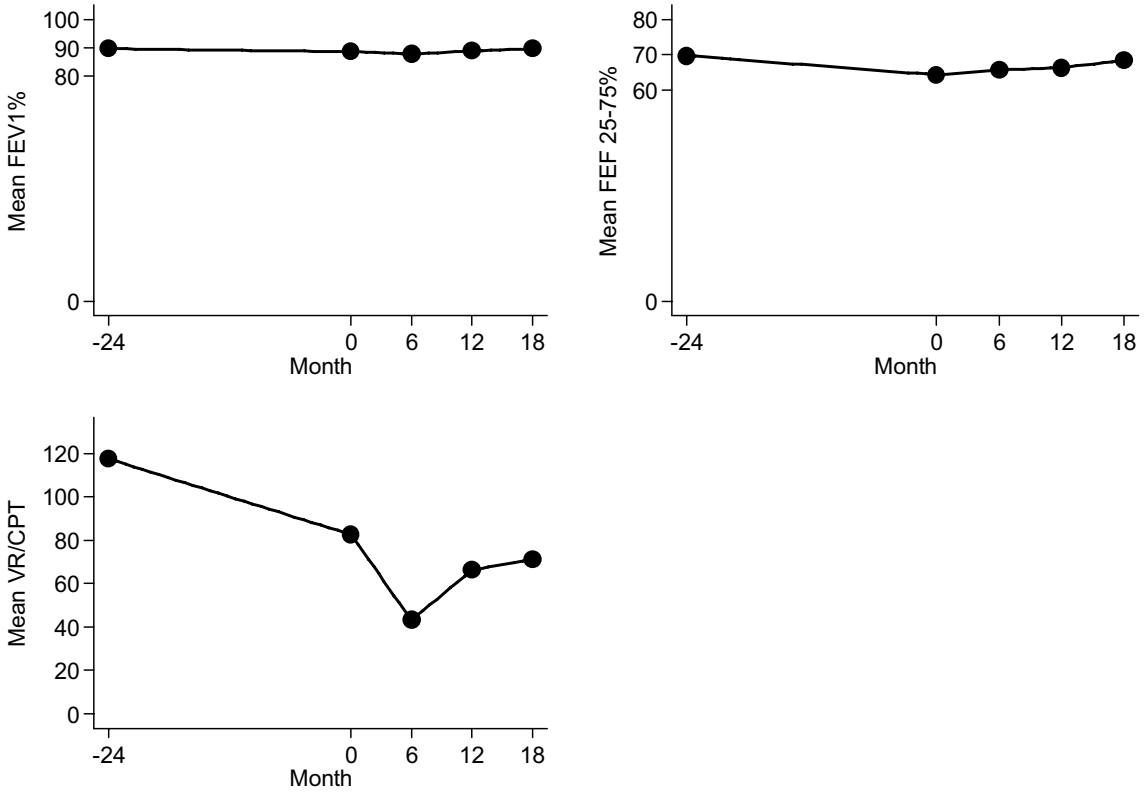

Figure 1. Evolution of pulmonary function during intervention and on a 2 years period before the beginning of the study. Considering the time span between 2 years before intervention until the end of the intervention study, FEV1% predicted had a mean increase of 0.11 SD (paired *t*-test: $p = 0.685$), FEF 25–75% had a mean decrease of 2.8% ($p = 0.636$) and VR/CPT a mean decrease of 57.07% ($p < 0.001$). – *Évolution des fonctions pulmonaires durant l'intervention et sur une période de 2 ans avant le début de l'étude. Sur la période totale, le VEMS prédit a augmenté de 0,11 DS en moyenne (*t*-test apparié:* p = 0,685*), le DEM 25–75 % a diminué de 2,8 % (*p = 0,636*) et le VR/CPT a diminué de 57,07 % en moyenne (*p < 0,001*).*

In regards of quality of life, no trend was seen in the CFQ®. It seems that a longer time of study would be necessary but even with such an intervention; the burden of the disease is always present.

There was as well no significant difference in emergency hospitalizations respectively use of rescue antibiotics (neither orally nor inhaled), probably due to the short term of study and by the fact that all patients were followed regularly at least three-monthly and as needed.

At beginning of the study, there were no dietitian fix accorded to the consultation, reason why it was planned initially to have a dietitian involved for those in the intervention group. However, due to circumstances not related to the study, a fix CF-dietitian was us accorded for all patients, reason why it would have been unethical to deprive the patients not in the intervention group from the regularly follow-up by the dietitian.

The weak point in our study is certainly the lack of a randomized control group, and the number of children recruited was too small to allow adjusting for confounding variables in multiple regressions. However, the same multi-disciplinary approach was continued in all other patients than those participating in the intervention group, without depriving them from the "as usual" therapeutic education according to international guidelines [8]. We clinicians probably overestimate to often our impact when there are no objective measurements ("bench marketing") available [15]. The link between acquired skills and change in way of doing (or being) is a key issue, but difficult to show. It was quite astonishing to see some patients not able to go from the cognitive level to the practical level ("how to change my habits). Nevertheless, we are sure that things are changing with the time. Compliance was not assessed in this study. In conclusion, a system of TPE might stabilize lung function parameters in interested patients and family. Although there is a lack of a randomized control group in our study, it seems important to at least standardize TPE in CF consultations.

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