Time invested in the global respiratory care of cystic fibrosis paediatrics patients

Gaudenz M. Hafen[1]*, Yann Kernen[1]* and Quentin M. De Halleux[2]

1 Department of Paediatrics, Respiratory Unit, Lausanne University Hospital, Switzerland
2 Department of Paediatrics, Physiotherapy Unit, Lausanne University Hospital, Switzerland

Abstract

Introduction: Respiratory therapy is a keystone of the treatment for cystic fibrosis (CF) lung disease, but it is time consuming.

Objectives: We aimed to assess the total time spent on respiratory therapy, including chest physiotherapy (CPT) and physical activity (PA), as well as inhalation therapy (IT) and maintenance of materials (MM) to rationalise and optimise treatment.

Methods: A cross-sectional prospective study in a paediatric CF cohort. A questionnaire was developed to look at the time spent on respiratory care over 3 months. Enrolled in this study are all CF patients aged from 6 to 16 years (the exclusion criterion was lung transplantation).

Results: Of the 40 enrolled patients, 22 participated (13 boys and 9 girls), with a mean age of 11 years. The patients spent approximately 19.46 h per week (standard deviation ± 7.53, 8.00–35.25 h) on therapy: CPT (30.58%), IT (15.11%), PA (50%) and MM (4.32%), without statistical significance between sexes.

Conclusion: In our cohort, CF patients spent an average of nearly 20 h a week in respiratory therapy, within a wide range of between 8 h to almost 36 h a week. PA consumes almost half of the time. Physicians have to take into consideration the burden of the treatment, to optimise the therapy.

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Correspondence
Gaudenz M. Hafen, MD, Department of Paediatrics, Respiratory Unit, Lausanne University Hospital, Rue Bugnon 46, 1011 Lausanne, Switzerland.
Tel: +41 79 556 18 25
Fax: +41 21 314 36 45
email: gaudenz.hafen@chuv.ch

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Authorship and contributorship
GMH designed study and wrote paper.
YK designed study, analysed data and wrote paper.
QMDH designed study and analysed data.

Ethics
The study was approved by the local ethics committee at the University of Lausanne, Switzerland. Informed written consent was obtained by all participants prior to inclusion.

Conflict of interest
GMH has no conflict of interest.
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*Equal contribution.

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 (CPT) is a major keystone of pulmonary treatment.

Respiratory therapy includes CPT in its own right, as well as inhaled medications and physical activity (PA) (3). PA is defined as any kind of body movement resulting in an increase of energy expenditure above a basal level (4). PA is considered as having a positive effect on health (5).

As it is known that daily respiratory therapy in CF is time consuming, in this study, we aimed to assess the total time spent on CPT and PA, as well as inhalation therapy (IT) and the maintenance of material (MM), to rationalise and optimise treatment.

Materials and Methods

Study design

It is a cross-sectional prospective study in a cohort of paediatric CF patients.

Study population

All the CF patients followed in the outpatient paediatric CF centre (Lausanne, Switzerland), aged between 6 and 16. The exclusion criterion was lung transplantation.

Questionnaire

A specific questionnaire was developed to look at the time spent on respiratory therapy over a 3-month period. We used the two-part 'one representative-week' questionnaire (6, 7) for time spent on PA. Based on the structure of that questionnaire, we created a questionnaire for CPT with a distinction between physiotherapy that was performed on its own or with a physiotherapist, including the nature of each therapy. In addition, the time spent on IT and MM was also assessed. For PA, activities at school, within a club, personal trainer and activities executed on its own respectively within the family were addressed. The questionnaire was not age-dependent, for each part the child and his/her parents had to fill in a weekly chart detailing the time spent on these activities each day.

To note that in our cohort the basic principle of physiotherapy is autogenic drainage, and that our patients have at a minimum of once a week supervised one-to-one therapy session with a professional physiotherapist.

Lung function measurements

Pulmonary function testing was conducted according to American Thoracic Society (ATS)/European Respiratory Society (ERS) – guidelines (8) simultaneously with the distribution of the questionnaire. The severity of FEV1% predicted results were classified according to the ATS/ERS guidelines (9).

Statistical analysis

This study used SPSS 15.0 (SPSS Inc., Chicago, Illinois, US) for Windows® (linearity analysis) for analysis.

Results

Of the 40 enrolled patients, 22 participated (13 boys and 9 girls), aged between 7 and 15 (mean age, 11 years). Twenty patients (91%) had mild [forced expiratory volume in one second (FEV1) > 70%], and 2 (9%) patients had moderate disease (FEV1 60–69%). The patient's demographics from visit 1 are shown in the Table 1.

Table 1. Study population

Number of patients	22	
Sex	13 boys	
Mean age (minimum – maximum)	11 (7–15 years) (standard deviation ± 2.27)	
Lung disease severity (9)	Patients	Boys
Mild (FEV1 > 70%)	20	12
Moderate (FEV1 60%–69%)	2	1
Moderately severe (FEV1 50%–59%)	0	
Severe (FEV1 35%–49%)	0	
Very severe (FEV1 < 35%)	0	

An average time of 19.46 h per week [standard deviation (SD) ± 7.53, 8.00–35.25 h] was spent on respiratory therapy; with 9.70 h for PA (50%, SD ± 2.03), 5.92 h for CPT (30.58%, SD ± 0.73), 2.97 h for IT (15.10%, SD ± 0.34) and 0.87 h for MM (4.32%, SD ± 0.37) (Fig. 1).

Boys spent 18.5 h a week on respiratory therapy (19.14 for girls, $P = 0.91$), 8.17 h on CPT (9.4 for girls, $P = 0.52$) and 10.33 h on PA (9.33 for girls, $P = 0.067$); there was no statistical significance between the sexes.

Patients performed on their own 69.79% of time for CPT (4.13 h a week), with one patient admitting not to do any CPT unescorted. All patients (100%) had supervised CPT by a physiotherapist, with time spent under supervision of 30.21% (1.79 h a week). All patients (100%) were performing autogenic drainage, 91% were using the Flutter® system (F. Uhlmann-Eyraud SA, Meyrin, Switzerland), 36% were using the PEP Mask® system (PARI Medical Holding, Starnberg, Germany) and 23% were using the Percussionaire® system (a device for intra-pulmonary percussive ventilation,

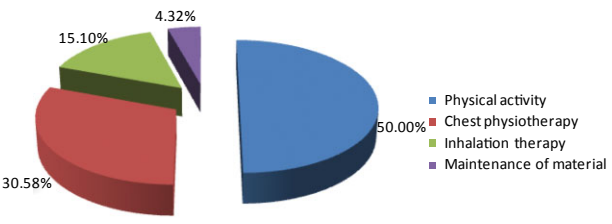

Figure 1. Repartition of activities in % of respiratory therapy.

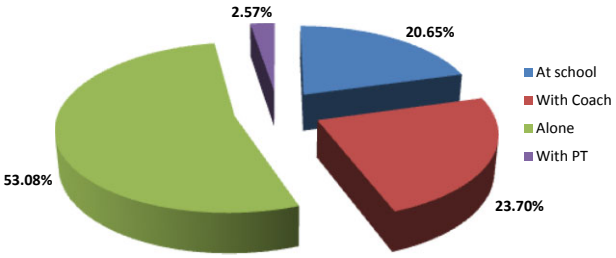

Figure 2. Repartition of supervised versus non-structured activities. PT, physiotherapist.

Percussionaire Corporation, Sandpoint, Idaho, USA (10)). One patient was not prescribed any IT.

Of the 9.70 h a week of PA, activities at school accounted for 2 h per week (20.65%), 2 h and 30 min within a club (23.71%), 15 min with a trainer (2.58%), and 5 h and 15 min (53.09%) in a non-structured way (Fig. 2). The most frequent activity was walking or kick-scootering (44%), football (33%) and ski or snowboarding (22%). Nine per cent did not perform any activities outside school hours. There was no correlation between gender and the level of activity, neither for the FEV1 results nor for the differing age groups.

Discussion

The CF patients from our paediatric cohort spend an average of nearly 20 h a week in respiratory therapy, within a wide range of between 8 h and almost 36 h a week.

Respiratory therapy is considered to be a keystone of the treatment. However, there is as well a lack of consensus of what exactly respiratory therapy comprises, as evidence about which technique might be the best. We assessed the amount of time spent on respiratory therapy including CPT, IT and MM during a 3-month period, as well as the time spent on PA. This was included as PA might be associated with a slower FEV1 decline and act as an independent predictor of survival, as recently shown (11). In addition, aerobic activity has been recently recommended as adjunctive activity for airway clearance and for its additional benefits to overall health (3). Exercise may improve mucus clearance by increasing respiratory flow and by increasing ventilation with probably easier expectoration due to alteration of physical sputum properties (12). In CF, exercise may have a direct positive impact on the ion-channel dysregulation. Its adrenergic and purinergic activity could act on the CFTR and ENaC function, improving ion transportation (13). Therefore, PA might well be seen as a part of the overall respiratory therapy; however, to date, there is no evidence to say if CPT in its own right, or PA, is more efficient (14). Hebestreit *et al.* showed that a 6-month training period had a longer lasting effect for over 1 year after the end of the programme (15). Worth noting, PA is also a method of socialisation and plays an important part in developing positive self-esteem in children.

Our analysis shows that the children spent a significant amount of time on therapy (an average of nearly 20 h a week), and this has to be recalled each time a new therapy is added to the already one in place. To rationalise and optimise the treatment and to decrease the burden of therapy, it is crucial to tailor the therapy to the patient. The time spent on PA within our cohort seems to be greater than in other CF populations (9.70 vs 8.52 h a week) (16). Therefore, to optimise that time, our patients wore a thoracic belt during PA to help them descend to lower lung volumes, thus increasing the quality of drainage. MMs, particularly cleaning including disinfection, is important as poorly maintained equipment may be a source of contamination (17); in our cohort, this takes about 52 min a week; however, we did not monitor the efficacy in performing microbiological swabs, nor did we assess if the maintenance was performed by the patient or his/her carers.

Our results regarding PA are in line with those of Selvadurai *et al.* as in our cohort, there was also no difference in time spent on therapy between the sexes (18). In addition, we found no differences between age groups and stage of disease. Adherence to CPT is considered to be poor (19), but as our study was limited, we did not assess it as part of the study protocol. Another important limitation is that the results are based on a recall method, which can vary in different age groups especially in adolescents.

In conclusion, CF is the paradigm of chronic disease, with treatment efficiency with regard to respiratory therapy mainly depending on the active collaboration of the patient. Respiratory therapy remains an important keystone of treatment, but as we have shown, it is time consuming. For paediatric patients, unfortunately, it is also an easy way of rebelling, particularly against the parents, and often becomes the subject of intra-familial conflict. Therefore, physicians treating CF patients have to take into consideration the burden of the treatment, to optimise the therapy. PA, as part of respiratory therapy, however, may be a way to reinforce the treatment: all the more if it is encouraged from young age and with the aim of maintaining social interactions.

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