The short and long term role and effectiveness of physiotherapy in children with Cystic Fibrosis

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Abstract

Hypothesis and purpose: Physiotherapy is one of the main interventions that contribute to quality of life improvement for these patients. The purpose of this work is to asses the physiotherapy efficiency in infants. *Method:* The study was conducted in the Paediatric Clinic II and in Romanian National Cystic Fibrosis Centre Timisoara and evaluations were made at the "Politehnica" University of Timisoara. Study group included 15 patients (10 male, 5 female), aged between 2 months and 3 years. Long-term clinical evaluation and evaluation during exacerbations included: general clinical condition and nutritional status evaluation, cough character, presence of dyspnoea and pulmonary physical signs (wheezing, rales). Patients also benefit of bacteriological examination of sputum or hipofaringian aspirate, radiological examination and quantification of changes severity with "Norman and Chrispin" score, CT and pulse oximetry. Physiotherapy treatment was established according to the diagnosis time and the clinical condition of patients. *Results:* At discharge, 80% of the patients had good overall condition, including those infected with persistent Staphylococcus aureus. Nutritional status improved in all patients and coughing disappeared in 46% of the cases. Bacterial culture remained positive in 40% of the patients. Oxygen saturation became normal after physiotherapy, even in patients with persistent infection and Norman and Chrispin score improved and sterilization in patients with Staphylococcus aureus was successful in 42% of cases. *Conclusions:* The results of this study highlight the importance of short term physiotherapy in improving clinical and nutritional status and the importance of clearance techniques in staphylococcal infection sterilization and Chrispin-Norman score improving. Assessment of the long term (one year) physiotherapy effectiveness, showed the importance of physiotherapy in reducing the number of exacerbations and therefore the number of readmissions, especially for the uninfected subjects.

***Key words**: Physiotherapy, cystic fibrosis, quality of life*

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Rezumat

Ipoteza şi scopul studiului: Fizioterapia este una dintre principalele intervenţii care contribuie la îmbunătăţirea calităţii vieţii la pacienţii cu Fibroza Chistică. *Scopul* acestei lucrări este evaluarea eficientei fizioterapiei la sugari. *Metodă:* Studiul a fost efectuat în cadrul Clinicii II Pediatrie şi a Centrului de Mucoviscidoză Timişoara, iar evaluările s-au desfăşurat în cadrul Universităţii „Politehnica" din Timişoara. Lotul de studiu a fost format din 15 copii (10 de sex masculin, 5 de sex feminin), cu vârsta cuprinsă între 2 luni şi 3 ani. Evaluarea clinică a bolnavului în timpul exacerbărilor şi pe termen lung a inclus: starea clinică generală şi statusului nutriţional, caracterul tusei, prezenţa dispneei, semne fizice pulmonare. Pacienţii au beneficiat, de asemenea, de examen bacteriologic din spută sau aspirat hipofaringian, examinarea radiologică şi cuantificarea severităţii modificărilor cu scorul "Norman Chrispin", computer tomografie şi pulsoximetrie. Tratamentul de fizioterapie a fost stabilit în funcţie de momentul diagnosticului şi de starea clinică a pacienţilor. *Rezultate:* Evoluţia clinică a bolnavilor pe termen scurt a fost în general favorabilă. La externare 80% dintre pacienţi au avut stare generală bună, inclusiv cei infectaţi cu Staphylococcus aureus persistent (n = 3). Statusul nutriţional s-a ameliorat la toţi bolnavii şi tusea a disparut la 46% dintre pacienţi. Examenul bacteriologic a rămas pozitiv la 40% din cazuri, iar saturaţia de oxigen a înregistrat valori normale după fizioterapie, chiar şi la pacienţii cu infecţia persistentă şi cu scor Norman Chrispin modificat. La cei cu infecţie cu Stafilococ auriu, s-a reuşit sterilizarea la 42% din cazuri. *Concluzii:* Rezultatele acestui studiu evidenţiază importanţa fizioterapiei pe termen scurt în ameliorarea stării clinice şi nutriţionale, precum şi importanţa tehnicilor de clearance în sterilizarea infecţiei stafilococice şi îmbunătăţirea scorului Norman şi Chrispin. Evaluarea eficienţei fizioterapiei pe termen lung (pe perioadă de un an), efectuată după un program riguros, a evidenţiat importanţa fizioterapiei în reducerea numărului de exacerbări a suferinţei respiratorii şi implicit a numărului de reinternări, mai ales la cei neinfectaţi.

***Cuvinte cheie**: fizioterapie, fibroză chistică, calitatea vieţii*

Funding Acknowledgement

This paper work was supported by a research grant from CNCSIS Romania, code TE_36, Contract No. 13/09.08.2010

Introduction

Cystic fibrosis (CF) is an inherited genetic disease, caused by a mutation in the CFTR gene. This gene has many complex functions, the most important being the chloride channel. ΔF508 mutation is the most common. Although most people have two functional alleles of the CFTR gene, only one is needed to prevent CF. CF develops when neither allele can produce a normal functional CFTR protein. Therefore, CF is considered autosomal recessive disease. CFTR gene is found on the q31.2 locus of chromosome 7. [2]

Patients with CF have normal lungs at birth; however most of them soon develop a pulmonary chronic illness that is a major cause of morbidity and mortality. There is considerable variation in terms of clinical presentation and lung function at a given age and rate of these functional variations can not be predicted. [2]

In CF lung disease begins in the peripheral airways, in an area particularly difficult to investigate in infants and young children and for this reason pulmonary function tests are difficult to execute, and the results are difficult to interpret. [3]

Hypothesis

Modern physiotherapy includes: airway clearance techniques, physical exercise, inhalatory therapy and is an integrant part of CF patient management. This interventions contribute to quality of life improvement for these patients. Along with other therapeutic tools, physiotherapy helps in bronchial secretion mobilization, improve dyspnea and exercise tolerance. [4]

The purpose of these work is the assesment of the physiotherapy eficiency in infants.

Material and method

The study was conducted in the Paediatric Clinic II and National CF Centre Timisoara and evaluations were made at "Politehnica" University of Timisoara.

The study group consisted of 15 patients (10 male, 5 female), aged between 2 months and 3 years (mean age: 9.6 months).

Long-term and during exacerbations clinical evaluation of the patient includes: general clinical condition and nutritional status, cough character, presence of dyspnoea and pulmonary physical signs (wheezing, rales etc.).

Patients also benefit of bacteriological examination of sputum or hipofaringian aspirated, radiological examination and changes severity quantification with "Norman and Chrispin" score (maximum possible score is 38), computer tomography in selected cases (Bhalla system - assess the severity of lesions by evaluating the degree of development of bronchiectasis), and pulse oximetry (assessment of the oxygen saturation). [5, 6]

Table I. Clinical and paraclinical aspects before treatment

Patients	General clinical state	Nutritional staus	Cough character	Pulmonary physical signs	Bacteriological exam	Pulmonary Xray (Norman – Chrispin score)	Pulse oximetry
1	good	dystrophy gr. II	rare, spastic	hyperinflation	staphylococcus aureus	2	96%
2	moderate	dystrophy gr II	rare, productive	rare bronchoalveolar rales	nonpathogenic flora	4	95%
3	moderate	dystrophy gr I	frequent, productive	bronchoalveolar rales, disseminated	staphylococcus aureus	6	95%
4	severe	dystrophy gr II/III	frequent, productive	bronchoalveolar rales, disseminated	staphylococcus aureus	8	94%
5	moderate	dystrophy gr I	rare, productive	bronchoalveolar rales, disseminated	nonpathogenic flora	2	97%
6	severe	dystrophy gr I	Intens, productive	bronchoalveolar rales, disseminated	Pseudomonas aeruginosa	10	92%
7	good	dystrophy gr I	rare, iritative	hyperinflation	nonpathogenic flora	2	97%
8	moderate	dystrophy gr. I	rare, spastic	hyperinflation	nonpathogenic flora	2	96%
9	severe	dystrophy gr.II	frequent, productive	bronchoalveolar rales, hyperinflation	staphylococcus aureus	8	92%
10	moderate	dystrophy gr. II	rare cough, spastic	hyperinflation	staphylococcus aureus	4	95%
11	good	normal	rare, iritative	hyperinflation	nonpathogenic flora	2	98%
12	severe	dystrophy gr. II	frequent, productive	bronchoalveolar rales, disseminated	staphylococcus aureus	12	86%
13	good	normal	rară, spastică	hyperinflation, wheezing	nonpathogenic flora	1	98%
14	severe	dystrophy gr. II	frequent, productive	bronchoalveolar rales, disseminated	staphylococcus aureus	8	92%
15	severe	dystrophy gr. III	frequent, productive	disseminated bronchoalveolar rales, wheezing	Pseudomonas	12	86%

For general condition assessment we examine the following clinical features: reactivity state, temperature, hydration state, skin coloration, respiratory rate, heart rate, shortness of breath, wheezing.In our patients, diagnosed in infancy, environmental factors that may have influenced the evolution of respiratory symptoms, especially infectious factors, had different influences from patient to patient.

Another important factor that definitely can influence the development of respiratory symptoms is the nutritional status of the patient.

We noticed that in our patients, infants who had grade II or III dystrophy, evolved with stronger clinical symptoms (productive cough, bronhoalveolar disseminated rales), while for those with normal weight, respiratory symptoms were mild to moderate. Paraclinical aspects before treatment are presented in Table I.

Physiotherapy treatment was established according to the diagnosis time and the clinical condition of patients. Therefore, we developed a short-term treatment plan focused on education of the mother and acute problems solutions, and a long-term one, spread over a period of one year from time of diagnosis.

The short-term therapeutic plan was carried out in cascade following acute problems identification and solving. Acute problems identified were: lobar or segmental atelectasis, linear and micro nodular opacities, low oxygen saturation, excessive bronchial secretions (Figure 1).

Therapy program aimed re-expansion of the affected lung lobes, treatment of pulmonary micro opacities, improving oxygen saturation and removing excessive bronchial secretions. Physiotherapy treatment consisted of: postural drainage, chest vibration, percussions and secretions suction.

Long-term therapeutic plan aimed implementation of physiotherapy schemes at home and included: postural drainage, chest vibration percussions and inhalatory therapy.

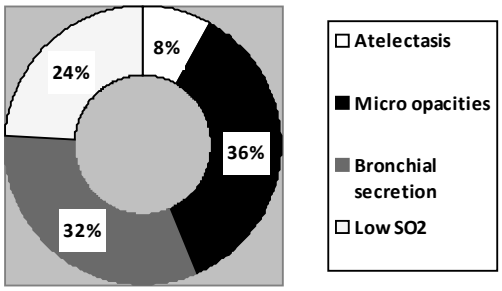

Figure 1. Items of respiratory illness exacerbations

Results

The short term clinical evolution of the patients was generally favourable, with a range of shades, depending on the initial status.

At hospital discharge, 12 of the patients (80%) had good overall condition, including those infected with persistent Staphylococcus aureus (n = 3). Nutritional status improved in all patients.

In case of Lung infection in CF, thick and sticky mucous secretions constituting the "primum movens", immune deficiency is not common in this condition. [7]

Coughing as a symptom characteristic of the disease, disappeared in 7 patients (46%) directly related to good clinical condition. In some patients infected with Staphylococcus aureus (n=2) and two patients infected with Pseudomonas aeruginosa (n=2), the cough became more rare, but it keeps the productive character.

In terms of paraclinical investigations, bacterial culture remained positive in six of the patients (40%). We note that in both patients infected with Pseudomonas aeruginosa, infection persisted, and they had a more pronounced clinical status than

other patients. Sterilization in patients infected with Staphylococcus aureus was successful in 42% of cases (n=3). Radiological aspects quantified by Chrispin-Norman score, were consistent with the clinical and bacterial infection, and in patients who were sterilized, radiological score became zero. For those infected with Staphylococcus aureus, score has improved, but remained at high levels in those with persistent Pseudomonas aeruginosa infection.

Oxygen saturation in our patients recorded normal values after physiotherapy, even in patients with persistent infection and modified Chrispin Norman score.

Long term evolution (one year term) in the context of physiotherapy application, was dependent of the initial results. Uninfected patients showed no exacerbation of the respiratory disease, with no need for subsequent readmissions.

Among patients with Staphylococcus aureus persistent infection, two patients had good clinical course following aerosol antibiotic treatment and intermittent oral treatment, the others two, with Chrispin-Norman score 4 and 6, had two episodes of exacerbation and needed readmission and intravenous antibiotic treatment.

Patients with persistent infection with Pseudomonas aeruginosa showed long term exacerbation, involving prolonged antibiotic treatment, 21 days intravenously and 14 days oraly.

The results can be seen in Table II.

Table II. Clinical and paraclinical aspects after physiotherapy

Patients	General clinical state	Nutritional staus	Cough character	Pulmonary physical signs	Bacteriological exam	Pulmonary Xray (Norman – Chrispin score)	Pulse oximetry
1	good	dystrophy gr. I	absent	absent	nonpathogenic flora	0	98%
2	good	dystrophy gr I	rare	absent	nonpathogenic flora	2	97%
3	good	normal	rare	absent	nonpathogenic flora	2	98%
4	good	dystrophy gr II	rare	bronchoalveolar rales	Stafilococcus aureus	4	97%
5	good	normal	absent	absent	nonpathogenic flora	0	98%
6	good	dystrophy gr I	rare, productiv	bronchoalveolar rales	Pseudomonas aeruginosa	6	96%
7	good	normal	absent	absent	nonpathogenic flora	0	99%
8	good	normal	absent	absent	nonpathogenic flora	0	98%
9	moderate improved	dystrophy gr.II	rare, productiv	bronchoalveolar rales, hyperinflation	Stafilococcus aureus	6	95%
10	good	dystrophy gr. I	rare	absent	nonpathogenic flora	2	97%
11	good	normal	absent	absent	nonpathogenic flora	0	98%
12	good	dystrophy gr. I	absent	absent	Stafilococcus aureus	8	94%
13	good	normal	absent	absent	nonpathogenic flora	0	98%
14	moderate improved	dystrophy gr. II	frequent, productive	bronchoalveolar rales	Stafilococcus aureus	4	95%
15	moderate improved	dystrophy gr. II	rare, productiv	rare bronchoalveolar rales	Pseudomonas aeruginosa	10	94%

Discussions

CF is a chronic disease with a progressive evolution. Specific drug therapy combined with an early and properly applied physiotherapy plan, positively influence the disease course and reduce the exacerbations and hospitalizations number and increase life expectancy and quality of life. Our results agree with the literature observations. [8]

Given that all patients received targeted antibiotic treatment according to antibiogram, we believe that physiotherapy has contributed substantially to the favourable development of unsterilized patients, fact also highlighted in literature. [4]

Nutritional status improvement can be explained in two ways: on one hand improved the respiratory distress led to the resumption of proper nutrition and on the other hand we know that good respiratory condition leads to lower energy losses. [2]

Our results regarding Staphylococcus aureus sterilization are similar with the literature reports in the field, defining the extremely important role of physiotherapy in the management of CF, whose benefit is even greater if the diagnosis is made early. [2]

Our study is consistent with the literature data, and revealed at this age group Staphylococcus aureus infection is predominant. (Figure 2)

Patients with cystic fibrosis have periodic exacerbations of pulmonary infection.

Appropriate treatment consists of parenteral administration of antibiotics, intensified airway clearance, and bronchodilator administration. Pseudomonas aeruginosa may survive longer in airways filled with sputum and airway plug formation is one factor reducing the eradication of pathogens from CF airways by antibiotic therapy.

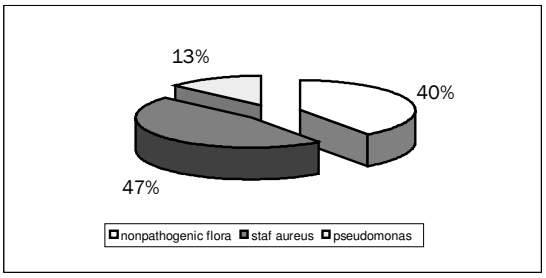

Figure 2. Distribution of cases according to the germ involved

We consider that the factors that influenced the disease in the context of a proper therapy were: lung status at diagnosis, bronchial tree with different germ colonisations (especially Staphylococcus and Pseudomonas aeruginosa), nutritional status, socio-cultural family and primary and secondary genetic factors (genetic modifiers).(Figure 3)

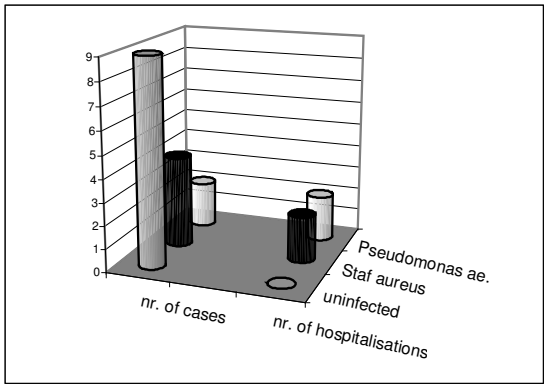

Figure 3. Correlation between infection status and hospitalisations number

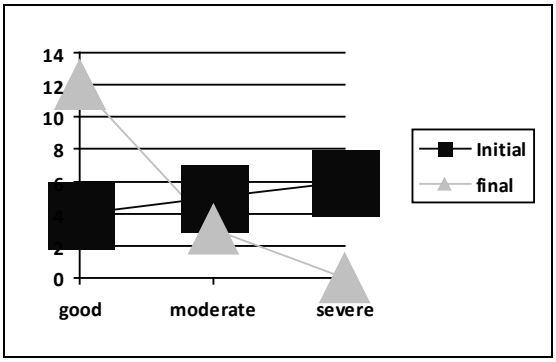

Figure 4. General clinical state before and after treatment

Conclusions

Education about the disease and its treatment starts at the diagnosis and is a never-ending process. The chronic infection and the inflammatory response is the reason for lung tissue damage, and the most logical objective must prevent or at least slow the progression of lung disease through physiotherapy.

The response to physiotherapy and antibiotic therapy was reflected by improved pulmonary function, decreased density of bacteria in sputum and improved well-being.

The effectiveness of the short term physiotherapy (during hospitalization) revealed overall the substantial benefits in improving clinical status and nutritional status, the important role of the clearance techniques in staphylococcal infection sterilization and improvement of Chrispin Norman radiological score. Assess of the long term physiotherapy effectiveness (one year), made after a rigorous therapy program, highlighted the importance of physiotherapy in reducing the number of exacerbations of respiratory illness and therefore the number of readmissions, especially for the uninfected patients.

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