127 - EVALUATION OF THE RESPIRATORY MUSCLE STRENGTH AND THE PEAK FLOW IN FIBROCYSTIC TEENAGERS AND CHILDREN

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1 INTRODUCTION

Cystic Fibrosis (CF) is caused by the dysfunction of the cystic fibrosis transmembrane regulator (CFTR), a chlorine-rich canal established in the epithelial cells. This way, the mutations which affect the respiratory tract, gastrointestinal, hepatobiliary and reproductive systems, as well as the sweat glands take place. Most of the patients with CF do not resist the respiratory failure due the chronic respiratory infection (HALL; SPARKS; ARIS, 2010).

It is a hereditary disease with high incidence (1/2,500 live born children) among Caucasian population. It presents an autosomal recessive pattern of inheritance set by a mutation in the Cystic fibrosis transmembrane regulator gene (RIBEIRO-PAES et al., 2009). The average age these patients have survived has increased. In 1950, the patients died in average when they were about 2 years old. Nowadays, such outcome happens when they are about 37 years old. This survival is initially linked to the degree of pulmonary dysfunction and the nutritional status (HALL; SPARKS; ARIS, 2010).

The progression of the pulmonary disease in CF decreases the ability practicing physical activities. Protein-energy malnutrition (PEM) and the metabolic changes generated by pulmonary inflammation lead to the reduction of muscle mass and bone mineral density (CHAVES et al., 2007). When FC is chronic it results in premature death. The fact that the patients develop a chronic pulmonary disease because of the progressive damage to pulmonary function and nutrition problems related to pancreatic insufficiency is very frequent. The nutritional deficit present in these patients comes from the malabsorption of nutrients they have consumed. Furthermore, the children who have CF spend more energy than the healthy children, so they need to eat more food, more frequently and using nutrition additive (GABATZ; RITTER, 2007).

Respiratory muscle strength (RMS) is reduced in people with CF which contributes for the development of fatigue in the respiratory muscles, dyspnea and difficulty in weaning from mechanical ventilation. The respiratory muscle fatigue can be a consequence of an imbalance between the load and the capacity of exercising respiratory muscles. The capacity of doing respiratory exercises is reduced because there is a reduction in the efficiency of respiratory muscles as a result from malnutrition (DUNNINK et al., 2009).

For those patients it is essential a multidisciplinary approach to improve survival and to provide a better health quality. Physiotherapy must be done starting from the diagnosis moment, with the goal of preventing and delay chronic pulmonary infections which are possible to happen, as well as to provide a higher aerobic capacity and respiratory muscle endurance. The nutritional treatment on the other hand aims to reduce malabsorption and balance the energetic absorption, which is deficient on these patients. Along with other professionals, daily tracking is absolutely important so the people who suffer from CF can reduce complications and manifestations presented by the illness. Faced with such respiratory ailments, this study aims to evaluate if body posture variation (Fowler 45º, sedestation 90º and or orthostasis) results in changes in the respiratory muscle strength and peak flow.

2 MATERIALS AND METHODS

This study is characterized as being a transversal, prospective study (HULLEY et al., 2008). This research was conducted with children and teenagers aged from 6 to 18 years old, from both genders (male and female) and different races. All of them were admitted at Hospital das Clinicas in Porto Alegre (HCPA). The ones responsible for the patients who are participating of the study signed an informed consent (IC), were also obtained from the patient acceptance.

The criteria used for including individuals in the study where to be a patient diagnosed with CF, established according to Cystic Fibrosis Foundation Consensus Panel criteria (ROSENSTEIN; CUTTING, quoted by DALCIN et al., 2007) and admitted at HCPA – Pediatric Pulmonology Service. The patients should show clinical stability, defined as the absence of findings in clinical exacerbation. The exclusion criterias incluiam presence of comorbities associated to CF that could limit the study. The research project was submitted to UNISC-CEP/UNISC Ethics Committee (Protocol number 2452/09) and it has been examined and approved according to ethical criteria for researches using human beings (Resolution 196/96 of CNS).

Firstly, were placed on the evaluation form, vital signs (Heart Rate (HR) and peripheral oxygen saturation (SpO2) using pulse oximetry (Nonin Medical®, Inc), respiratory rate (RR) and axillary temperature (AT), as well as the anthropometric characteristics.

Respiratory Muscle Strength

The RMS was evaluated by digital manometer (MDV 300 Globalmed®). This is a simple technique to apply; low cost, non-invasive and it can be used to quantify the respiratory muscle strength in healthy individuals and in those who have disorders from different origins. To evaluate the maximum inspiratory pressure (MIP), the patients have done expiration up to the residual volume (RV) level and later, occluded against a valve and performed the maximum inspiratory effort for about three seconds, up to the total lung capacity (TLC). To evaluate the maximum expiratory pressure (MEP), the patients had to breath in up to their total lung capacity (TLC) and they had to do a maximum expiratory effort up to the RV (BRUNETTO; ALVES, 2003). Was done three measurements of the RMS in each evaluated position (Fowler 45o, Sedestation (sitting in a 90o position) and Upright Positions). It is noteworthy that was evaluated the stability of vital signs during the body posture variations was always evaluated.

Peak Flow

Peak Flow (Peak Flow Meter®), is an important equipment to evaluate the maximum peak expiratory flow of the patient. Such a device can measure the peak expiratory flow, being it easy to apply and reproduce. Three measurements were done where the expiratory flow was forced starting from an inspiratory positions with the head set in a neutral position. There were performed three measurements of the peak flow in the fowler 45o, sedestation (sitting in a 90o position) and upright position.

Data Analysis

The data were distributed according to the median and analyzed using the software Statistical Package for Social Sciences (SPSS, version 18.0). To compare the RMS values found in the patients with their respective foretold values, as well as

to evaluate the MIP, MEP and Peak Flow behavior in the Fowler 45o, sedestation and orthostasis positions were performed Wilcoxon and Friedman non-parametric tests ($p<0.05$).

3 RESULTS

The sample of this study was built of six (06) caucasian children of an average age of 13 years old, four (04) out of six children were female and two (02) of them were male. The average for the Body Mass Index (BMI) was 18.5 Kg/m2, which did not have relevant statistical significance among the values which were found and their respective foretold value (Table 1). Due to the small sample size it was not possible to evaluate the homogeneity of anthropometric characteristics among the individuals.

Table 1 – Anthropometric data and gender analysis of cystic fibrosis patients

Variables	Average (IIQ)
Male gender, n (%)	2 (33,3)
Age (years)	13,0 (9,5 – 15,0)
Weight (Kg)	42,0 (26,3 – 62,7)
Height (cm)	148,0 (130,0 – 168,0)
BMI (Kg/m^2)	18,5 (15,4 – 21,6)

Where: M: Male Gender; BMI: Body Mass Index
IIQ: Interquartile interval.

The average time for diagnosing cystic fibrosis had a median of 20.3 months after birth, only one (1) of the patients born from preterm labor and the others were born from term labor. No one of the patients presented previous pathology, four (04) patients who participated of the sample had cough symptoms, one (01) patient had cough and dyspnea and another one had only dyspnea. Two (02) patients had cystic fibrosis cases in their families (Table 2).

Table 2 - Clinical characteristics analysis of cystic fibrosis patients

Patient	Kind of Delivery	Ethnicity	Age it was diagnosed (month)	Manifestation	Previous pathologies	CF cases in the family
V.S.	Term	Caucasian	60	Cough	No	No
A.C.	Term	Caucasian	6	Cough	No	No
L.M.	Term	Caucasian	3	Cough	No	Sister
M.G.	Term	Caucasian	6	Dyspnea	No	No
L.S.	Term	Caucasian	7	Cough	No	No
J.S.	Preterm	Caucasian	7	Cough and Dyspnea	No	Sister

Where: CF: Cystic Fibrosis

It is noteworthy that the change in body posture was implemented in the tests only at the moment the vital signs of the evaluated patient would return to their base level. When comparing MIP and MEP values of the fibrocystic patients against foretold values, there were no changes in (MIP = 67. 00 cmH2O 69.18 cmH2O) and (MEP = 86.00 cmH20 90.60 cmH2O) respectively ($p = 0.917$) (WILSON et al. 1984). The peak flow in turn showed a statistically reduced in fibrocystic children and teenagers in relation to what was foretold (Peak Flow = 230.00 l/min 365.00 l/min respectively) ($p = 0,046$) (Chart 1).

When dealing with the evaluation of RMS behavior in the different body postures adopted, it was found that there was a statistically significant variation of PLmax among the groups that were studied ($p = 0.03$). It must be observed that the significance was obtained when changing the body posture from Fowler 45o to the orthostatic position ($p = 0.009$) (Chart 2). And yet, the orthostatic position was the one responsible for generating the greater inspiratory muscle strength.

Chart 1 – Peak expiratory flow CF patients analyzed.

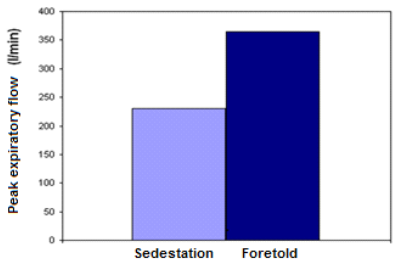
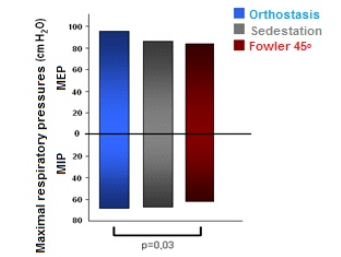


Chart 2 – Maximal respiratory CF patients analyzed.



Where: MIP: Maximum inspiratory pressure
MEP: Maximum expiratory pressure

It is observed that the MEP ($p = 0.055$) as well as the Peak Flow ($p = 0.154$) showed no variations between the body postures that were analyzed.

4 DISCUSSION

Cystic Fibrosis is an inherited autosomal recessive disease characterized as being a chronic suppurative lung disease, pancreatic insufficiency, multifocal biliary cirrhosis, male infertility and great electrolytic loss on sweat (MATTAR et al., 2010).

It was possible to observe, in this study, that the Caucasian Ethnicity prevailed. This information is consistent with

Ziegler et al. (2007) and Dalcin et al. (2007) who found in their studies that the caucasoid patients highly prevailed among others.

With the results obtained meet with what Rosa et al. (2008) observed that the mucoviscidosis is a chronic, progressive disease which can affect several organs and body systems. This disease is common in the white race and, it equally reaches both genders. However, according to Morrow et al. (2008), although the disease prevails in caucasoid people, in his study he investigated the lung function changes in a population diagnosed with CF in South Africa for a period of 8 years, reporting significant improvements of the lung function in this fibrocystic population.

In our study, most patients had adequate nutritional status for their age group. Only two of the patients had differences in their BMI, one of them faced overweight risk and the other excess weight. It is possible that the BMI within or close to the normal values found in our study may be assigned to the fact that these patients receive systemic and multidisciplinary monitoring starting from the moment the disease is diagnosed. In Ziegler's et al. (2007) study, from the 46 patients who participated in the research, 26 (63.4%) were in a adequate nutritional status, 6 (14.6%) were in a nutritional risk zone and 9 (22%) were malnourished and it was not possible to identify a significant association between BMI and the age of the patients, the age when CF was diagnosed, clinical score, radiological score, lung function, respiratory pressures and the Six Minute Walk Test (WT6). According to the study of Zieger et al. (2007) the nutritional approach is an important aspect for succeeding in the treatment of fibrocystic patients. It is considered an independent prognostic factor in the sickness outcome, and the regular monitoring of the nutritional status aims to detect and treat malnutrition in an early state.

In our study, the patients had a median of 20.3 months in the CF diagnosis, and that Ziegler et al. (2007) said that, in his study, there was an average of 9 years for CF diagnosis. For Rosa et al. (2008) advances in diagnosis and therapeutical strategies developed in the last 30 years have significantly increased life expectancy of patients diagnosed with Cystic Fibrosis.

According to Chaves et al. (2007), in Cystic Fibrosis, the cellular hypoxia may be caused by decreasing the alveolar ventilation and/or alterations in the ventilation/perfusion interaction what decreases the alveolar oxygen levels. Besides this, the mucus retention facilitates the presence of infection in the respiratory tract and harms lung function leading to respiratory symptoms like cough and dyspnea.

Besides the symptoms found in patients from the sample, it was also possible to notice the hereditary traces from the disease because CF cases were discovered in the family of two (02) patients. This information meets what Rosa et al. (2008) which reports that, in reports from the last 70 years, CF has been recognized as the most prominent inherited disease, potentially lethal.

For Ribeiro - Paes et al (2009), CF is a hereditary disease of high incidence (1/ 2.500 live born) among Caucasian population, reporting even the lung is, normally, the most affected organ, with a tendency to recurrent infections, loss in gas exchange and cardiopulmonary functions failure. This way, the knowledge about the effects the force of gravity has on cardiovascular function and the negative impacts physiopathological status has on lung cardiopulmonary function offer foundation for the clinical application of body posture as the main therapeutical intervention to optimize oxygen transportation, this being the main non-invasive physical therapy intervention for the non-invasive forms, pharmacological and mechanic support treatments can be postponed, reduced or yet avoided (FROWNFELTER; DEAN, 2004).

In our study we found that the MIP and MEP from fibrocystic people are under the foreseen value, not that there was any significant difference. For Dunnink et al. (2009), the available studies about respiratory muscle strength in fibrocystic patients had conflicting results, showing decreased or normal values for the RMS. Hyperinflation and malnutrition seem to be the main factors for weakness of respiratory muscles.

According to the author, it is difficult to understand the studies about the RMS in fibrocystic patients because they differ from each other in various characteristics as: age, nutritional status and the amount of respiratory dysfunction. Also, the different methods and ways of measuring used to evaluate the RMS of these patients.

This study found that the patients had a peak flow lower than the foretold and, this fact may be justified by the hypersecretion these patients had. For Camargo and Queiroz (2002), as much as the results about peak flow have showed statistically significant values, these are lower than the ones gotten through conventional spirometry. Therefore, these results are weak for clinical relevance because the alterations associated to peak flow reflect, mainly, proximal airway involvement, which can be observed in the furthest stage of the disease.

According Camargos and Queiroz (2002), peak flow measures do not openly express neither the operating level of involvement nor the magnitude of clinic-radiological involvement and, that said, they do not constitute a reasonable alternative, not even as an acceptable marker to evaluate the pulmonary function in this disease (CAMARGOS; QUEIROZ, 2002).

When talking about RMS, the present study revealed that the orthostatic position must have a greater value in relation to the other positions. It was possible to notice this because according to Frownfelter and Dean (2004), the orthostatic position maximizes volumes and pulmonary capacity. In addition, the functional residual capacity (FRC) is higher in orthostat than it is in a sitting and standing position, which in turn, FRC is higher than the supine position in about 50%. The results obtained in this research showed that the FMS had variations from Fowler 45º to orthostatic position. According to Frownfelter and Dean (2004), when a patient is in a vertical position, the diameter if his/her airways, increases slightly. In the orthostatic position, the gravitational gradient is in its maximum position, the Antero-posterior dimension of the chest walls is higher, and heart and lungs compression is minimal, therefore, in the orthostatic position, the shorted position of diaphragmatic fibers increase the neural command for breathing.

We recognized that the RMS in fibrocystic patients is decreased. However, when the systemic risk is monitored in these patients since the moment they born, it becomes possible to offer them a better quality of life because it is essential for these children and teenagers to have a childhood filled with life.

5 FINAL CONSIDERATIONS

When conducting this study, we determined the impact of disease severity on the respiratory function of individuals with cystic fibrosis analyzed. The limitations this study had were limited amount to sorting children and teenagers suitable to the inclusion and exclusion criteria of this study, this resulted in a reduced sample size. It is necessary to continue with this study, having a more significant sample of people diagnosed with Cystic Fibrosis thus it will be possible to get more comprehensive results. Cystic Fibrosis is a systemic disease and a serious social problem, and it is desirable to continue this research, as well as other ones, which can contribute for a longer survival of fibrocystic patients and also offer quality to the years of life.

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EVALUATION OF THE RESPIRATORY MUSCLE STRENGTH AND THE PEAK FLOW IN FIBROCYSTIC TEENAGERS AND CHILDREN

ABSTRACT

Introduction: Cystic Fibrosis (CF) is characterized as a chronic obstructive pulmonary disease, increasing the number of electrolytes in sweat and exocrine pancreatic failure. It is possible that the respiratory muscle strength (RMS) is decreased in this population and that selected body postures entail in a higher reduction of the RMS and peak flow. Goal: To watch if there is reduction of maximum inspiratory pressure (MIP), maximum expiratory pressure (MEP) and peak flow in fibrocystic patients under different body postures. Materials and Methods: An observational, descriptive and cross sectional study, built of six patients from both genders (male and female) and aged between 6 and 18 years old. It was done three different measurements of the RMS using a digital vacuum manometer (MVD 300 Globalmed®) and peak flow (Peak Flow Meter®) in the Fowler 450, sedestation (sitting in a 90o position) and orthostasis position, the vital signs were also evaluated during the postural assessment. Results: Comparing the MIP and MEP values against the predicted values there were not any variations (MIP = 67.00 and 69.18 cmH2O and MEP = 86.00 and 90.60 cmH2O) (p = 0.917). Orthostasis showed the highest MIP and MEP values (68.50 cmH2O and 95.50 cmH2O respectively). It is possible to highlight there was a variation of the MIP in Fowler 45o for orthostasis (p= 0.003), while MEP did not show any significant variation between the body postures. The peak flow, on the other hand, was statistically decreased in these patients (p = 0.046). Conclusions: The RMS of the fibrocystic patients analyzed did not change in relation to what was predicted, but the Fowler 45o for orthostatic showed a significant variation. Orthostasis resulted in the highest values of MIP and MEP. The peak flow showed an arway obstruction and did not vary changing the body postures.

KEY-WORDS: Cystic Fibrosis; different body postures; manovacuometry.

ÉVALUATION DE LA RÉSISTANCE DES MUSCLES RESPIRATOIRES ET DÉBIT DE POINTE DU FLUXE EXPIRÉ EN ENFANTS ET ADOLESCENTS FIBROKYSTIQUES

RÉSUMÉ

La fibrose kystique est caractérisée par la BPCO l'augmentation des électrolytes dans la sueur et l' insuffisance pancréatique exocrine. Il est possible que la force des muscles respiratoires (FMR) soit réduite dans cette population et que certaines postures impliquent une plus grande réduction de la FMR et du débit expiratoire de pointe (Peak Flow). Observer s' il y a une réduction du PImax et du PEmax et du Peak Flow chez les patients FK sous différentes postures. Une étude d'observation descriptive et transversale, composé de six patients des deux sexes et âgés entre 6 et 18 ans.Trois mesures sucessives on été

realisées par rapport à la FMR par manovacuometry numérique et le débit expiratoire de pointe dans des positions Fowler 45º, sedestation (assis à 90 degrés) et hypotension orthostatique, ont également été évalués les signes vitaux pendant les changements de posture. En comparant les valeurs du PImax et du PEmax obtenues avec les valeurs prédits il n'y a eu aucune variation (PImax=67.00 et 69.18 cmH2O et PEmax=86,00 et 90,60 cm H2O) (p=0,917). L'hypotension orthostatique a montré la plus grande valeur de PImax et PEmax (68,50 cm H2O et 95,50 cmH2O respectivement). Il est à remarquer qu'il y a eu une variation de PImax en Fowler à 45 ° pour la hypotension orthostatique (p=0,03), tandis que le PEmax n'a pas changé de façon significative entre les postures. Le débit expiratoire de pointe s'est montré statistiquement réduit chez ces patients (p=0,046). La FMR de patients fibrokystiques analisés n'a pas varié par rapport au prédit, mais la variation de Fowler 45° pour l' orthostatique a déterminée une variation significative. L'hypotension orthostatique a donné une plus grande hausse des valeurs obtenues de PImax et PEmax. Le Peak Flow exposé obstruction des voies aériennes haute d'une alternance de position haute et n'a pas changé.

MOTS-CLÉS: Fibrose kystique; postures différentes; manovacuometry.

EVALUACIÓN DE LA FUERZA MUSCULAR RESPIRATORIA Y DEL PICO DEL FLUJO EXPIRADO EN NIÑOS Y ADOLESCENTES FIBROCÍSTICOS

RESUMEN

Introducción: La fibrosis cística se caracteriza por enfermedad pulmonar obstructiva crónica, aumento de electrólitos en el sudor e insuficiencia pancreática exocrina. Es posible que la fuerza muscular respiratoria (FMR) esté reducida en esa población y que determinadas posturas impliquen en una mayor reducción de la FMR y del pico de flujo espiratorio (Peak Flow). Objetivo: Observar si hay reducción de la PImax y PEmax y del Peak Flow en pacientes fibrocísticos so distintas posturas. Materiales y Métodos: Estudio observacional descriptivo y transversal, compuesto por seis pacientes de ambos los géneros y edad entre 6 y 18 años. Fueron realizadas tres afericiones de la FMR a través de la manovacuometría digital (MVD 300 Globalmed®) y del pico de flujo espiratorio (Peak Flow Meter®) en las posiciones Fowler 45º, sed estación (sentado a 90º) y ortos tasis, siendo también evaluados las señales vitales durante las variaciones posturales. Resultados: Al comparar los valores de la PImax y de la PEmax obtenidos con valores predichos no hubo variación (PImax = 67,00 y 69,18 cmH2O y PEmax = 86,00 y 90,60 cmH2O) (p = 0,917). El ortos tasis presentó el mayor valor de PImax y PEmax (68,50 cmH2O y 95,50 cmH2O respectivamente). Se resalta que hubo variación de la PImax en Fowler 45 °para ortos tasis (p=0,03), mientras la PEmax no presentó variación significativa entre las posturas. El pico de flujo espiratorio por su vez, se ha presentado estadísticamente reducido en esos pacientes (p = 0,046). Conclusiones: La FMR de los fibrocísticos analizados no cambió en cuanto al predicho, pero la variación de Fowler 45° para ortos tasis determinó variación significante. El ortos tasis resultó en los mayores valores obtenidos de PImax y PEmax. El Peak Flow demostró obstrucción de las vías aéreas altas y no cambió con la mudanza postural.

PALABRAS CLAVE: Fibrosis cística, distintas posturas, manovacuometría.

AVALIAÇÃO DA FORÇA MUSCULAR RESPIRATÓRIA E DO PICO DE FLUXO EXPIRADO EM CRIANÇAS E ADOLESCENTES FIBROCÍSTICOS

RESUMO

Introdução: A fibrose cística caracteriza-se por doença pulmonar obstrutiva crônica, aumento de eletrólitos no suor e insuficiência pancreática exócrina. É possível que a força muscular respiratória (FMR) esteja reduzida nessa população e que determinadas posturas impliquem em maior redução da FMR e do pico de fluxo expiratório (Peak Flow). Objetivo: Observar se há redução da PImax e PEmax e do Peak Flow em pacientes fibrocísticos sob diferentes posturas. Materiais e Métodos: Estudo observacional descritivo e transversal, composto por seis pacientes de ambos os gêneros e idade entre 6 e 18 anos. Foram realizadas três aferições da FMR através da manovacuometria digital (MVD 300 Globalmed®) e do pico de fluxo expiratório (Peak Flow Meter®) nas posições Fowler 45º, sedestação (sentado a 90º) e ortostase, sendo também avaliados os sinais vitais durante as variações posturais. Resultados: Ao comparar os valores da PImax e da PEmax obtidos com valores preditos não houve variação (PImax = 67,00 e 69,18 cmH2O e PEmax = 86,00 e 90,60 cmH2O) (p = 0,917). A ortostase apresentou o maior valor de PImax e PEmax (68,50 cmH2O e 95,50 cmH2O respectivamente). Ressalta-se que houve variação da PImax em Fowler 45° para ortostase (p=0,03), enquanto a PEmax não apresentou variação significativa entre as posturas. O pico de fluxo expiratório por sua vez, apresentou-se estatisticamente reduzido nesses pacientes (p = 0,046). Conclusões: A FMR dos fibrocísticos analisados não variou em relação ao predito, porém a variação de Fowler 45° para ortostática determinou variação significante. A ortostase resultou nos maiores valores obtidos de PImax e PEmax. O Peak Flow demonstrou obstrução das vias aéreas altas e não variou com a mudança postural.

PALAVRAS-CHAVE: Fibrose cística; diferentes posturas; manovacuometria.