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PULMONARY FUNCTION ANALYSIS IN RELATION TO STATIC POSTURE, THORACIC SPINE MOBILITY AND PHYSICAL ACTIVITY LEVEL IN PATIENTS WITH CYSTIC FIBROSIS

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ABSTRACT

Cystic fibrosis (CF) is a multisystemic disease, mainly affecting the pulmonary system. In order to optimize respiratory work, postural changes and progressive physical exercise limitation may appear over time. **Objective:** To identify whether there is a relationship between pulmonary function and static posture, thoracic spine mobility and physical activity level in CF patients. Cross-sectional study with a consecutive sample of 44 children and adolescents. The following evaluations were carried out: anamnesis, physical activity level, measurement of the angle and mobility of the thoracic spine, spirometry and pulmonary plethysmography. **Results:** Positive and significant correlation between: (1) flexion angle with forced vital capacity (FVC), forced expiratory volume in one second (FEV_1) and forced expiratory flow ($FEF_{25-75\%}$) (in liters); (2) extension mobility with FVC, FEV_1, $FEF_{25-75\%}$ (%pred); (3) total thoracic spine mobility with all pulmonary variables except Tiffeneau Index and plethysmographic variables. Negative and significant correlation was found between the extension angle with FVC, FEV_1 and $FEF_{25-75\%}$ (% predicted). The level of physical activity was negatively correlated with Residual volume (RV_L) (%pred). **Conclusion:** CF patients do not appear to have a significant impairment of static posture; however, thoracic spine mobility seems to correlate with worsening lung function.

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INTRODUCTION

Cystic fibrosis (CF) is a hereditary, progressive and multisystem disease, causing several changes in the human body due to the involvement of exocrine glands (Penafortes, 2012; Barker, 2014 and Radtke, 2015). However, pulmonary impairment is the main determinant of CF-related morbidity and mortality (Penafortes, 2012 and Barker, 2014). When ventilation is compromised, posture changes may occur to allow chest muscles to assist Breathing (Barker, 2014). In addition, patients with CF show signs of air trapping, thus causing pulmonary obstruction. Since the chest region has two primary functions, which are ventilation and postural control, changes in lung anatomy can cause mechanical changes in the bone structure of the rib cage. Thus, patients with CF tend to have a more kyphotic posture, as demonstrated by Okuro, *et al* (2012), who found a prevalence of postural kyphosis in 61.9% of children and adolescents with CF. Faced with such postural changes, it is also postulated that there are changes in the thoracic mobility of these patients, contributing to the impairment of lung function (McIlwaine, 2014). Patients with CF frequently experience progressive limitations to physical exercise; this is a multifactorial issue, derived from the various systemic effects generated by the underlying disease[11]. Such limitations notwithstanding, adherence to a regular exercise program is associated with better prognosis and increased survival (García, 2011 and Radtke, 2015). Few studies have addressed the association between pulmonary impairment and musculoskeletal alterations in patients with CF, or assessed whether level of physical activity can influence these parameters. Within this context, the objective of this study is to verify whether pulmonary function is associated with static posture, thoracic spine mobility, and level of physical activity in children and adolescents with CF.

MATERIALS AND METHODS

This is a cross-sectional study with a correlational design (Gaya, 2008). The sample of this study was consecutive and its size was defined by sample calculation using the Gpower 3.1 software, admitting a two-tailed test, a moderate correlation expectation ($r = 0.6$), large size effect ($f = 0.5$), an $\alpha = 0.05$ and a power of 80%, resulting in a minimum sample of 43 patients. The study included patients between six and 18 years old, of both sexes, diagnosed with CF, according to the Brazilian CF Diagnosis and Treatment Guidelines[30], followed up at the Children's Pulmonology Clinic of Hospital de Clínicas de Porto Alegre (HCPA), who performed more than one spirometry, with clinical stability at the time of the evaluations, defined as absence of disease exacerbation. Patients with heart disease were excluded from the study; patients with neuromuscular or bucomaxillofacial disorders; previous diagnoses of spinal deformities such as scoliosis; or any other clinical condition that prevented them from performing the evaluations were also excluded. This study was approved by the Research Ethics Committee of HCPA (No. 16-0062) and all participants signed the Consent Form and the parents, the Free and Informed Consent Form. Patients participated in the following evaluations: (a) anamnesis, (b) IPAQ (International Physical Activity Questionnaire), (c) static evaluation of curvature and mobility of the thoracic spine, both in the sagittal plane, (d) spirometry and (e) pulmonary plethysmography. The anamnesis, whose objective was to obtain information to characterize the sample, such as body mass, height and age, was completed by the evaluator with the answers provided by the legal guardian or the patient himself. The IPAQ is a valid and reproducible instrument (Matsudo, 2001), which allows estimating the time spent per week in different dimensions of physical activity, being answered by the patient if he could read and interpret the questions, or with the help of those responsible.

The quantification of the thoracic curvature in the static posture and the evaluation of the thoracic spine mobility was performed with the Flexicurve, a valid and reproducible instrument (Oliveira, 2012), which consists of an 80 cm long flexible ruler (Trident, Brazil). To quantify the thoracic curvature in the static posture, the protocol described by Oliveira *et al* (2012) in which the patient remains standing, in his usual posture, with straight knees, parallel and bare feet, bare back, while spinous processes of vertebrae C7, T1, T12, L1, L5 and S1 were marked. Then, the ruler was molded into the spine, carefully removed and placed on a paper, transcribing the curvature contour and identifying the previously marked spinous processes. For the evaluation of thoracic spine mobility, the protocol described by Burton (Burton, 2015) was adapted, and the spinous processes of T1, T6 and T12 in the flexed and extended positions were palpated and marked. For the evaluation of the flexed position the patient remained seated, with the feet supported and performed the maximum flexion of the thoracic spine. For the evaluation of the extended position the patient was positioned in the prone position, upper limbs supported on the stretcher and a maximum extension of the thoracic spine was requested without moving the hip. In each of these positions the Flexicurve was molded into the spine and later transcribed on paper. To obtain the flexion mobility, the angular value obtained in the flexion position was subtracted from the angular value of the neutral posture (the neutral position was considered the curvature of the thoracic spine) and, to obtain the extension mobility, the angular value obtained in the extended position was subtracted of the angular value of the neutral posture. Spirometric values were obtained from the patient's online medical record. The examinations were performed with Jaeger v4.31a equipment (Jaeger, Würzburg, Germany) using the technical acceptability criteria of the Brazilian Pulmonary Function Consensus[1]. Data on forced expiratory volume in one second (FEV_1), forced vital capacity (FVC), Tiffeneau index (FEV_1/FVC) and mean forced expiratory flow ($FEF_{25-75\%}$) were obtained. Values were expressed as a percentage of predicted for gender, age and height (Sociedade Brasileira de Pneumologia e Tisiologia, 2002). Spirometry values after bronchodilator use were used for this study, as it results in higher fidelity of pulmonary function (Costa, 2004). Plethysmography examination data were collected from the online medical records of 29 patients. Residual volume (RV) and effective airway resistance (REf) values were obtained. Data were expressed as absolute values and % predicted for gender, age and height (Pereira, 2002).

In the IPAQ-short version, the data were classified according to IPAQ's own guidance (Lee, 2017), which divides into: sedentary, insufficiently active, active, very active. For Flexicurve data analysis regarding static posture, the paper curvature contour was digitized and analyzed using BIOMEC FLEX© 3.0 software (www.ufrgs.br/biomec/materials.html), providing the kyphosis angle of the thoracic spine, from trigonometric relations (Oliveira, 2012). To obtain the angular values of thoracic spine mobility, the thoracic curvature contour, in flexion and extension positions, was digitized and analyzed using Matlab® 7.5 *software*, with a routine specially developed for this study. Body mass, height and age data were used to determine the “body mass index for age” (BMI/A), using the percentile according to the World Health Organization recommendation. Nutritional indicators were obtained using the WHO Anthroplus 2007 program. Nutritional status was determined according to the Cystic Fibrosis Foundation Pediatric Nutrition Consensus Report. Malnutrition corresponded to the percentile below 10; nutritional risk at the 10 to 25 percentile; and eutrophy above the 25 percentile (Borowitz, 2002). In the analysis by age group, we considered children, those patients who were up to 12 years old and adolescents, those who were among 12 and 18 years old, according to Law 8.069 of the Child and Adolescent Statute (Brasil, 1990). The data obtained were initially analyzed from mean, standard deviation and frequency table. Data normality was assessed using the Shapiro-Wilk test. Pearson's correlation test or Spearman's correlation test was used to verify the correlation between the variables. Statistical analysis were performed using the SPSS 20.0 software, and a significance level of 0.05 was adopted.

RESULTS

The sample consisted of 44 patients with CF (61.4% female), with a mean age of 12.7±3.3 years (Table I). Regarding the specific characteristics of CF, 27.3% were from the homozygous DeltaF508 mutation, 34.1% had bacterial colonization of *Staphylococcus aureus* and 15.9% had colonization of *Staphylococcus aureus* and *Pseudomonas aeruginosa*. Regarding the physical activity level, 77.3% were classified as active or very active. As for the nutritional classification, 29.5% of patients presenting nutritional risk classification and 6.8% with malnutrition.

Pulmonary, postural and thoracic spine mobility variables are described in Table 2. In the total sample, the flexion angle of the thoracic spine was positively correlated with FVC, FEV_1 and $FEF_{25-75\%}$ (L), the extension angle was negatively correlated with the same pulmonary variables FVC, FEV_1 and $FEF_{25-75\%}$, in % predicted. The extension mobility of the thoracic spine was correlated with the same previous variables, but positively. The total mobility of the thoracic spine was correlated with all pulmonary variables analyzed, except for the Tiffeneau Index and plethysmographic variables.

function of the diaphragm (torso stabilization and respiratory control) to respiratory control alone (Hodges, 2001). Still, the prolonged adoption of a flexed position for coughing gradually exerts abnormal pressure on the spine, contributing to a kyphotic posture (Henderson, 1994 and Massery, 2005). In a Systematic Review study (Lee, 2017) that investigated postural alignment in children and adults with obstructive respiratory disease, including 12 studies with CF, the increase in the kyphotic angle of the thoracic spine ranged from 3% to 62%. Furthermore, the angle was always greater than in the control group.

Table 1. Sample description

	Mean±SD	Mininum-Maximum
Age (years)	12,7±3,3	6-17
Body Mass (kg)	44,0±12,7	19,8-63,8
Stature (m)	1,52±0,18	1,15-1,81
BMI (kg/m²)	18,5±2,4	13,9-23,4

BMI= Body Mass Index

Table 2. Description of the sample regarding the spirometric and plethysmographic pulmonary variables, postural and thoracic spine mobility, for the total sample and separately by age group (children and adolescents)

	Total Sample (n=44)	Children (n=13)	Adolescents (n=31)
	Mean±SD	Mean±SD	Mean±SD
FVC (L)	2,9±1,1	1,8±0,5	3,4±0,9
FVC (% pred)	98,7±18,3	99,0±15,5	98,5±19,6
FEV_1 (L)	2,3±0,9	1,5±0,4	2,7±0,8
FEV_1 (% pred)	87,9±20,5	89,5±16,6	87,2±22,1
Tiffeneau Index (% pred)	67,6±39,8	95,3±8,3	86,3±18,8
$FEF_{25-75\%}$ (L)	1,9±1,3	1,2±0,6	2,1±1,4
$FEF_{25-75\%}$ (% pred)	67,6±39,8	70,3±33,0	66,5±42,7
RV_L (obs)*	2,0±0,6	1,7±0,6[a]	2,2±0,6[b]
RV_L (%pred)*	182,0±68,0	187,4±104,2[a]	196,7±53,1[b]
R eff (obs)*	0,4±0,2	0,6±0,2[a]	0,4±0,2[b]
Kyphosis Angle (º)	39,1±9,7	34,8±9,2	41,0±9,5
Flexion Angle (º)	59,6±7,6	54,6±5,3	61,7±7,5
Extension Angle (º)	21,0±10,8	18,8±8,9	22,0±11,5
Flexion Mobility (º)	20,8±8,2	20,9±5,4	20,7±9,3
Extension Mobility (º)	18,3±10,4	16,7±11,9	19,0±9,8
Total Mobility (º)	38,6±11,3	35,8±12,3	39,7±10,8

FVC = forced vital capacity; FEV_1 = forced expiratory volume in one second; $FEF_{25-75\%}$ = forced expiratory flow between 25-75% of vital capacity; RV = residual volume; R eff = effective airway resistance; L = liters; % pred =% of predicted; (obs) = observed; *sample of 29 subjects; [a] sample of 9 subjects; [b] sample of 20 subjects.

The level of physical activity was negatively correlated with RV_L (%pred). The thoracic kyphosis angle and the flexion mobility of the thoracic spine did not present a significant correlation with any of the pulmonary variables (Table 3). When analyzed in isolation only the children group (Table 4), the thoracic kyphosis angle showed a positive correlation with FVC (% pred) and the flexion angle with $FEF_{25-75\%}$ (L and % pred). The thoracic spine flexion mobility did not show any correlation with pulmonary variables. The thoracic spine extension angle was negatively correlated with FVC (L), FEV_1 (L and %pred) and FEF (L and %pred). Thoracic spine extension mobility correlated positively with FVC, FEV_1 and Tiffeneau Index (%pred); and the total mobility of the thoracic spine was positively correlated with FEV_1 (L and %pred), with $FEF_{25-75\%}$ (L e %pred) and negatively correlated with RV_L (%pred). The results for the adolescents group showed few correlations, being found only between the thoracic spine total mobility with FEV_1 (% pred) and $FEF_{25-75\%}$ (L and %pred). The other posture and thoracic spine mobility variables, as well as the physical activity level did not correlate with the pulmonary variables in this group (Table 5).

DISCUSSION

Several studies have investigated musculoskeletal repercussions in CF patients (Barker, 2014; Botton, 2003; Henderson, 1994; Okuro, 2012; Rawo, 2015; Schindel, 2015), hypothesizing that high levels of pulmonary hyperinflation might shift the dual

The findings of the present study corroborate this review; in the overall sample, the average thoracic kyphosis angle was 39.1°. When analysis was stratified by age groups, adolescents showed a slightly higher average kyphotic angle than children. In this sense, many studies have demonstrated that thoracic kyphosis increases with advancing age (Barker, 2014; Henderson, 1994; Okuro, 2012; Rawo, 2015). Nevertheless, the cause of this increase in thoracic kyphosis in these patients is uncertain and multifactorial; it may be associated with disease severity, osteoporosis, and chronic pain (Barker, 2014; Henderson, 1994 and Okuro, 2012). Despite this, in our study, the thoracic spine kyphosis angle correlated with FVC (% pred) only when analyzing the group of children. A possible explanation for this finding may be the fact that no stratification of disease severity levels was performed, no objective assessment of exercise capacity, and no analysis of vitamin D and corticosteroid levels. It bears noting that the literature lacks reference values for range of thoracic spine motion in children, both healthy and with CF. One of the few studies that have evaluated this outcome demonstrated that patients with CF had limited flexibility and muscle weakness in the torso, suggesting that this could promote a more flexed posture (Ross, 1987). In the present study, those patients with higher levels of pulmonary function reached wider angles of thoracic spine flexion; however, when range of flexion was evaluated, there was no correlation with any of the studied pulmonary variables. In other words, when considering a neutral position for analysis of range of motion, these variables did not correlate, probably

Table 3. Correlations between pulmonary and postural parameters, spine mobility, and IPAQ in the overall sample

	Kyphosis angle		Flexion angle		Extension angle		Flexion ROM		Extension ROM		Total ROM		IPAQ	
	r/rho	p	r/rho	p	r/rho	p	r/rho	p	r/rho	p	r/rho	p	r/rho	p
FVC (L)	0.153[a]	0.321	0.357[a]	0.017*	-0.098[a]	0.526	0.104[a]	0.502	0.217[a]	0.156	0.337[a]	0.025*	-0.038[b]	0.806
FVC (% pred.)	0.050[a]	0.746	0.061[a]	0.694	-0.344[a]	0.022*	0.037[a]	0.813	0.397[a]	0.008*	0.373[a]	0.013*	-0.108[b]	0.484
FEV_1 (L)	0.147[a]	0.341	0.369[a]	0.014*	-0.147[a]	0.342	0.119[a]	0.441	0.269[a]	0.078	0.391[a]	0.009*	-0.052[b]	0.737
FEV_1 (% pred.)	0.004[a]	0.980	0.111[a]	0.473	-0.375[a]	0.012*	0.103[a]	0.505	0.404[a]	0.007*	0.436[a]	0.003*	-0.083[b]	0.594
Tiffeneau index (% pred.)	0.075[b]	0.629	0.053[b]	0.732	-0.188[b]	0.222	-0.070[b]	0.653	0.286[b]	0.060	0.202[b]	0.188	0.047[b]	0.762
$FEF_{25-75\%}$ (L)	0.060[b]	0.698	0.346[b]	0.021*	-0.158[b]	0.306	0.186[b]	0.228	0.239[b]	0.119	0.386[b]	0.010*	0.038[b]	0.806
$FEF_{25-75\%}$ (% pred.)	-0.020[b]	0.898	0.199[b]	0.196	-0.303[b]	0.046*	0.129[b]	0.406	0.329[b]	0.029*	0.404[b]	0.007*	-0.005[b]	0.977
RV_L (obs)	0.053[a]	0.839	-0.140[a]	0.591	0.031[a]	0.907	-0.259[a]	0.315	0.009[a]	0.972	-0.129[a]	0.621	-0.168[b]	0.519
RV_L (% pred.)	-0.045[a]	0.865	-0.241[a]	0.351	0.210[a]	0.418	-0.246[a]	0.340	-0.277[a]	0.281	-0.391[a]	0.121	-0.496[b]	0.043*
R_{eff} (obs)	-0.052[a]	0.844	-0.213[a]	0.413	0.172[a]	0.509	-0.199[a]	0.443	-0.240[a]	0.354	-0.331[a]	0.195	0.067[b]	0.797

[a] Pearson correlation; [b] Spearman correlation; * Significant correlation; FVC = forced vital capacity; FEV_1 = forced expiratory volume in the first second; Tiffeneau index = FEV_1/FVC; $FEF_{25-75\%}$ = forced expiratory flow between 25-75% of vital capacity; RV = residual volume; R_{eff} = airway effective resistance; L = liters; % pred. = % of predicted; (obs) = observed; ROM = range of motion.

Table 4. Correlations between pulmonary and postural parameters, spine mobility, and IPAQ in children

	Kyphosis angle		Flexion angle		Extension angle		Flexion ROM		Extension ROM		Total ROM		IPAQ	
	r/rho	p	r/rho	p	r/rho	p	r/rho	p	r/rho	p	r/rho	p	r/rho	p
FVC (L)	0.093[a]	0.762	0.279[a]	0.357	-0.565[a]	0.044*	0.056[a]	0.857	0.462[a]	0.112	0.530[a]	0.062	0.308[b]	0.306
FVC (% pred.)	0.567[a]	0.043*	0.301[a]	0.318	-0.525[a]	0.066	-0.397[a]	0.179	0.790[a]	0.001*	0.511[a]	0.074	0.206[b]	0.499
FEV_1 (L)	0.050[a]	0.870	0.407[a]	0.167	-0.622[a]	0.023*	0.167[a]	0.586	0.497[a]	0.084	0.628[a]	0.022*	0.337[b]	0.259
FEV_1 (% pred.)	0.323[a]	0.282	0.479[a]	0.098	-0.581[a]	0.037*	-0.045[a]	0.883	0.711[a]	0.006*	0.629[a]	0.021*	0.329[b]	0.273
Tiffeneau index (% pred.)	0.110[b]	0.720	0.613[b]	0.026	-0.264[b]	0.383	0.363[b]	0.223	0.711[b]	0.006*	0.629[b]	0.021	0.082[b]	0.789
$FEF_{25-75\%}$ (L)	0.027[b]	0.929	0.553[b]	0.050*	-0.577[b]	0.039*	0.357[b]	0.231	0.357[b]	0.231	0.473[b]	0.103*	0.278[b]	0.358
$FEF_{25-75\%}$ (% pred.)	0.297[b]	0.325	0.674[b]	0.012*	-0.615[b]	0.025*	0.121[b]	0.694	0.440[b]	0.133	0.582[b]	0.037*	0.125[b]	0.683
RV_L (obs)	-0.673[a]	0.327	-0.436[a]	0.564	0.306[a]	0.694	-0.012[a]	0.988	-0.415[a]	0.585	-0.500[a]	0.500	0.458[b]	0.542
RV_L (% pred.)	-0.868[a]	0.132	-0.582[a]	0.418	0.771[a]	0.229	-0.035[a]	0.965	-0.814[a]	0.186	-0.987[a]	0.013*	0.867[b]	0.133
R_{eff} (obs)	-0.635[a]	0.365	-0.651[a]	0.349	0.230[a]	0.770	-0.245[a]	0.755	-0.348[a]	0.652	-0.547[a]	0.453	0.639[b]	0.361

[a] Pearson correlation; [b] Spearman correlation; * Significant correlation; FVC = forced vital capacity; FEV_1 = forced expiratory volume in the first second; Tiffeneau index = FEV_1/FVC; $FEF_{25-75\%}$ = forced expiratory flow between 25-75% of vital capacity; RV = residual volume; R_{eff} = airway effective resistance; L = liters; % pred. = % of predicted; (obs) = observed; ROM = range of motion.

Table 5. Correlations between pulmonary and postural parameters, spine mobility, and IPAQ in adolescents

	Kyphosis angle		Flexion angle		Extension angle		Flexion ROM		Extension ROM		Total ROM		IPAQ	
	r/rho	p	r/rho	p	r/rho	p	r/rho	p	r/rho	p	r/rho	p	r/rho	p
FVC (L)	-0.098[a]	0.600	0.080[a]	0.670	-0.202[a]	0.276	0.161[a]	0.386	0.143[a]	0.442	0.272[a]	0.139	-0.193[b]	0.299
FVC (% pred.)	-0.100[a]	0.592	0.021[a]	0.909	-0.303[a]	0.098	0.117[a]	0.532	0.260[a]	0.158	0.339[a]	0.062	-0.216[b]	0.244
FEV_1 (L)	-0.056[a]	0.766	0.123[a]	0.509	-0.232[a]	0.209	0.154[a]	0.407	0.220[a]	0.235	0.335[a]	0.066	-0.204[b]	0.270
FEV_1 (% pred.)	-0.068[a]	0.716	0.076[a]	0.684	-0.323[a]	0.076	0.129[a]	0.489	0.315[a]	0.084	0.399[a]	0.026*	-0.177[b]	0.340
Tiffeneau index (% pred.)	0.174[b]	0.349	-0.030[b]	0.874	-0.090[b]	0.632	-0.169[b]	0.363	0.330[b]	0.070	0.130[b]	0.487	0.048[b]	0.799
$FEF_{25-75\%}$ (L)	0.014[b]	0.941	0.254[b]	0.168	-0.127[b]	0.495	0.188[b]	0.311	0.234[b]	0.205	0.360[b]	0.047*	-0.017[b]	0.926
$FEF_{25-75\%}$ (% pred.)	-0.006[b]	0.973	0.188[b]	0.312	-0.194[b]	0.295	0.154[b]	0.409	0.302[b]	0.099	0.388[b]	0.031*	-0.064[b]	0.733
RV_L (obs)	0.041[a]	0.895	-0.135[a]	0.659	0.008[a]	0.980	-0.257[a]	0.398	0.031[a]	0.920	-0.107[a]	0.728	0.079[b]	0.797
RV_L (% pred.)	0.138[a]	0.652	-0.155[a]	0.613	-0.072[a]	0.816	-0.445[a]	0.128	0.236[a]	0.437	-0.032[a]	0.916	0.029[b]	0.924
R_{eff} (obs)	0.035[a]	0.910	-0.138[a]	0.654	0.163[a]	0.595	-0.250[a]	0.409	-0.182[a]	0.551	-0.283[a]	0.349	0.168[b]	0.583

[a] Pearson correlation; [b] Spearman correlation; * Significant correlation; FVC = forced vital capacity; FEV_1 = forced expiratory volume in the first second; Tiffeneau index = FEV_1/FVC; $FEF_{25-75\%}$ = forced expiratory flow between 25-75% of vital capacity; RV = residual volume; R_{eff} = airway effective resistance; L = liters; % pred. = % of predicted; (obs) = observed; ROM = range of motion.

because the patients in our sample already had some degree of thoracic kyphosis. When analyzing the thoracic extension angle, patients with higher angles (closer to the neutral position) had worse pulmonary function. Similar results were obtained on analysis of spine extension. Although issues of spine mobility have not been evaluated in the literature, studies that assessed postural or musculoskeletal outcomes in CF populations often provide suggestions for the improvement of respiratory mechanics. Okuro *et al*[19] and Massery[16] state that aerobic, thoracic mobility, and stretching exercises promote improvements in posture and chest wall compliance, preserving and optimizing pulmonary function. In this sense, the results of the present study highlight the importance of maintaining thoracic spine mobility in patients with CF. When analysis was stratified by age groups, correlations were significantly more present in children than in adolescents. In children, nearly all musculoskeletal variables (except range of thoracic spine flexion) were associated with some pulmonary parameter, while in the adolescent group, only total range of motion seemed to interfere significantly with pulmonary function. Possible explanations are that children are in a growth phase subject to anatomical changes; hence, impairments in pulmonary function may be more likely to lead to such alterations, as all structures are not yet stabilized. Other potential explanations include disease severity, type of bacterial colonization, adherence to treatment, and the relatively small number of children in relation to adolescents. There is a need for additional research to address these issues, as well as to evaluate the effect of early interventions on thoracic spine mobility and pulmonary capacity in these patients.

Isolated elevations of RV (and the RV/TLC ratio) may indicate slight airflow limitations, especially if the drop in RV is observed after bronchodilator administration. This may be the result of an extrapulmonary restrictive process or due to expiratory muscle weakness. In CF, there is an increase in RV/TLC ratio that expresses the air trapping component of hyperinflation (Barreto, 2002). The postural attitude of the insufflated thorax presupposes a series of spinal, shoulder girdle, and pelvic compensations. In adults with CF, spinal deformities are common because of the advanced progression of the disease (Tattersall, 2003). To date, only one study has evaluated the correlation between pulmonary function (using spirometry and plethysmography) and posture, but in adult CF patients[21]. In the present study, in the general sample, no correlations were found between the postural and pulmonary function variables, with a correlation only in the group of children when analyzing the total mobility of the thoracic spine. The lack of correlations can be attributed to the fact that only 29 patients were included, since the exam was not routinely performed in the service. Thus, further investigations are still needed to understand the relationship between pulmonary plethysmographic variables and postural variables, especially in the population of children and adolescents. Although the benefits of the regular practice of physical activity in this public were well consolidated, the present study showed a negative correlation between the level of physical activity and only one plethysmographic variable (RV_L) in the general sample. One explanation may be the fact that patients are constantly encouraged to perform physical activity at the center where they receive treatment and 77.3% of them have an "active or very active" level of physical activity. The study by Schindel *et al*[27] corroborates our results, as they did not find a significant difference in lung function (FEV_1) when providing guidelines for the practice of physical exercise for three months. According to Elkin *et al* (Elkin, 2000), even if these patients do not exhibit significant pulmonary repercussions, there is still a need for early multidisciplinary intervention, as studies with adult populations still show that postural issues correlate with pulmonary function, quality of life, and functional capacity (Barker, 2014 and Rovedder, 2014). Limitations of the study include the fact that the pulmonary plethysmography was performed in only 29 of the 44 studied patients, which may have contributed to Type II error. Longitudinal studies to evaluate the progression of thoracic spine alterations and the effect of therapeutic interventions are suggestions for future research. In this sample of patients with CF, there were no significant correlations between pulmonary function and the kyphotic angle of the thoracic spine or the level of physical activity. Thoracic spine mobility correlated with pulmonary parameters, especially in extension; participants with reduced thoracic spine mobility showed worse pulmonary function. When stratified by age, children with CF seemed to show greater correlations between musculoskeletal alterations and pulmonary function than adolescents.

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