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Influence of thoracic spine postural disorders on cardiorespiratory parameters in children and adolescents with cystic fibrosis

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Abstract

Objectives: To assess the impact of increased thoracic kyphosis on pulmonary function and functional capacity in children and adolescents with cystic fibrosis (CF) and to verify the influence of disease severity, age and nutritional status on this deformity.

Method: This was a cross-sectional, analytical study conducted at a university hospital. It included CF patients with confirmed diagnosis and without pulmonary exacerbation. The sample was submitted to postural assessment, spirometry (FEV_1, FVC and FEV_1/FVC) and 6-minute walk test distance (6-MWT distance). Data were analyzed using the Mann Whitney test, Spearman correlation and logistic regression.

Results: Forty-two patients were enrolled, 61.9% presented increase of thoracic kyphosis. There was no difference in values of FEV_1, FVC, FEV_1/FVC and 6-MWT distance between the groups with or without thoracic kyphosis ($p = 0.407$; $p = 0.756$; $p = 0.415$; $p = 0.294$). In the group without alteration, patients with more disease severity had a mean FEV_1 of 74.1±21.9% and FVC of 79.8±18.7% while in those of lesser severity higher values were found (95.6±12.2% and 97.6±13.2%, respectively) ($p = 0.027$ and $p = 0.027$). The presence of kyphosis was correlated with age ($p = 0.048$) but not with severity ($p = 0.151$) and body mass index ($p = 0.088$).

Conclusions: There was a high prevalence of increased thoracic kyphosis in children and adolescents with CF. The deformity did not affect pulmonary function and functional capacity and there was no relationship with disease severity. Regardless of posture, worsening of disease severity determined worsening of pulmonary function.

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Introduction

The increase in life expectancy in cystic fibrosis (CF) involves the development of secondary complications related to the musculoskeletal system.[1] The occurrence of changes in body posture that can harm functions related to the cardiopulmonary system is among them.[2]

This is because postural abnormalities have a strong influence on the respiratory effort, as this determines a permanent stimulus on the musculoskeletal support, potentially aggressive in age groups in development.[3,4]

The trunk, which involves muscles and the thoracic spine, has a dual relationship with postural support and ventilation. This dual function requires that it regulates both postural needs and the needs of the respiratory system.[5]

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In CF, pulmonary disease exerts a positive pressure, repeated over and over the thoracic framework, resulting in a thoracic kyphosis. This deformity is the result of the obstructive process and the constant coughing episodes caused by the hypersecretion characteristic of the disease. Its presence may contribute to declining respiratory function.[6]

It is recommended that interventions be performed in an attempt to prevent and/or minimize emerging postural abnormalities. Early treatment is crucial in order to minimize the impairment of respiratory function. The pre-puberty phase (8-12 years) is described as the best period to begin this closer attention, because the growth phase is the most suitable for any intervention.[6]

Therefore, the objective of this study was to assess the impact of increased thoracic kyphosis on respiratory function and on the functional capacity of children and adolescents with CF and to assess the influence of disease severity, age and nutritional aspects on the presence of this deformity.

Method

A cross-sectional analytical study was carried out at the Cystic Fibrosis Clinic of the Department of Pediatrics, Hospital de Clinicas da Universidade Estadual de Campinas (UNICAMP), Brazil. Included subjects were children and adolescents from 7 to 19 years, followed in regular service, with a diagnosis of CF confirmed by clinical history, altered sweat test (chloride greater than 60 mmol/L) in at least two samples and molecular genetic study. The excluded patients were those dependent on oxygen and those in acute pulmonary exacerbation, according to the Cystic Fibrosis Clinical Score[7] and Cystic Fibrosis Foundation scores[8]. These are instruments routinely used in outpatient consultations.[9]

The CF outpatient service provides assistance to approximately 150 children and adolescents. All patients followed up by the service within the range defined by the study group were invited to participate in the study, which was performed from January to December, 2011.

The sample was submitted to postural assessment through the New York test (NYT),[10,11] examination of pulmonary function by spirometry and the 6-minute walk test (6MWT). Height and weight anthropometric data, as well as the computation of the body mass index (BMI) were also recorded.

NYT is an objective method of assessing posture that includes 13 body segments. It has a scoring system that allows quantitative analysis and the rating of the assessed postural disorder. Each body segment is scored according to the suitability of their alignment (5 points - normal pattern; 3 - moderate change; and 1 - severe). At the end of the assessment the points of all segments are added up, and the individual's posture is classified as normal (56-65 points), moderate (40-55) or severe (1-39).[11] This study specifically examined the position of the thoracic spine in lateral view, which determines the degree of curvature of this segment (normal, with a slight increase or a high degree of kyphosis). This change was also classified as normal, moderate or severe, according to the NYT, and patients were grouped according to presence (NYT moderate or severe) or absence (normal NYT) of increased thoracic kyphosis. They were labeled as group with thoracic kyphosis and group without it, respectively. For data analysis, this variable was considered as qualitative.

Pulmonary function was assessed by spirometry (Medgraphics CPFS/D spirometer) in the Pulmonary Function Laboratory, School of Medical Sciences, UNICAMP, following regulations by the American Thoracic Society (ATS, 1995).[12] The percentage of the predicted value of the variables of forced expiratory volume in one second (FEV_1) and forced vital capacity (FVC) was considered. A cutoff point of 80% was considered to divide the sample below the predicted value (less than 80%) and above it (80%).

After a 15 minutes rest, the 6MWT was performed to assess the functional capacity of patients with CF. 6MWT was also performed following ATS' recommendations (ATS, 2002).[13] The distance covered (DC) was recorded. The predicted distance for each participant was calculated according to Prienitz et al.[14] For data analysis, patients were grouped according to test performance and DC (DC: below and above the predicted).

The severity of cystic fibrosis was also considered using the Shwachman score (SS) modified by Doershuk.[15] The SS covers items related to general activity, physical examination, nutrition and radiological findings, with a range from 20 to 100 points. Patients were classified as severe (score below 40), moderate (between 40 and 55), medium (from 56 to 70), good (between 71 and 85) and excellent (from 86 to 100 points). For data analysis, the sample was divided into two groups: the groups of lesser severity (patients classified as good or excellent by SS) and of greater severity (classified as severe, moderate and medium),[15] and considered dichotomized.

The data were processed using the Statistical Package for Social Sciences 16.0 computer program (SPSS Inc., Chicago, IL, USA). The frequency of the variables for characterizing the sample was determined. The lack of data normality was verified by using the Kolmogorov-Smirnov and Shapiro-Wilk test. The comparison of the spirometric variables (FVC, FEV_1, FEV_1/FVC) and of DC in 6MWT between groups with and without thoracic kyphosis was made by using the nonparametric Mann-Whitney test for independent samples. The same test was used to compare the same variables in accordance with the severity of the groups. The Spearman correlation test

was used to determine the influence of thoracic kyphosis on spirometric parameters, BMI, age and severity. The relationship between the presence of kyphosis and the spirometric variables (FEV_1 and FVC) and functional (DC in 6MWT) was analyzed by univariate logistic regression, followed by multivariate, using the Forward Wald method. Thoracic kyphosis was considered as an independent variable, and FEV_1, FVC and DC in 6MWT were considered as dependent variables. The significance level adopted was 5%.

The project was approved by the Research Ethics Committee of the School of Medical Sciences, UNICAMP (n. 172/2010). All study participants had a term of informed consent signed by parents or guardians.

Results

The study included 42 children and adolescents from 7 to 19 years, mean age 12.47±3.43 years. Patients had a minimum of 12 kg and a maximum of 64.4 kg (mean 30.82±14.71 kg) and a mean height of 1.32±0.23 meters.

Table 1 presents the characteristics of the sample concerning demographics, gender, age, disease severity, postural and thoracic spine pattern, spirometry and DC in 6MWT.

The relation between the presence of alterations in the thoracic spine, according to the NYT classification, and the variables FEV_1, FVC, FEV_1/FVC and DC in 6MWT was analyzed. The mean values of FEV_1 in the group without increased thoracic kyphosis (n = 16) was 84.8±20.4 (percentage compared to the predicted), and in the group with increased thoracic kyphosis (n = 26) it was an average of 78.8±20.7. This difference was not statistically significant (p = 0.407). Regarding the values of FVC, the groups with and without increased thoracic kyphosis had averages of 88.7±18.1 and 85.5±18.4, respectively (p = 0.756).

The mean values of FEV_1/FVC for the group without thoracic kyphosis was 95.1±10.6 (percent relative to the predicted value), and in the group with thoracic kyphosis it increased to 91.7±13.7. This difference was not statistically significant (p = 0.415). The mean DC in 6MWT in groups without and with increased thoracic kyphosis was 597.6±93.2 and 564.2±67.6 meters, respectively (p = 0.294).

Tables 2 and 3 refer to the distribution of values of FEV_1, FVC, FEV_1/FVC and DC in 6MWT compared to SS levels of disease severity, present in the groups with and without alterations of the thoracic spine.

In the group without thoracic kyphosis, patients with more severe CF had a mean FEV_1 of 74.1±21.9%, while the group of lesser severity showed higher values (95.6±12.2%). This difference was statistically significant (p = 0.027). In FVC, the group without thoracic kyphosis and classified as less severe had a mean of 97.6±13.2% of the predicted, and the more severe group, 79.8±18.7% (p = 0.027). The FEV_1/FVC ratio in the group without increased thoracic kyphosis and less severe had a mean of 98.3±8.6, whereas in the more severe group the mean was 91.9±12.1 (p = 0.141). In the group with increased thoracic kyphosis, the group of lesser severity had a FEV_1/FVC score ratio of 93.7±14.1, while the group of greater severity, an average 87.2±12.5 (p = 0.120). These data are presented in Table 2.

There was no difference in DC in 6MWT and the level of severity of disease (lesser and greater severity) in the groups

Table 1 - Sample characterization concerning anthropometric data, sex, age, disease severity, postural and thoracic spine pattern, spirometry and distance covered in the 6-minute walk test

Variable	n	%
Sex		
Male	19	45.2
Female	23	54.8
Age (years)		
Under or equal to 12 years	20	47.6
Over 12 years	22	52.4
BMI		
< percentile 5	12	28.6
Between percentile 5 and 50	23	54.8
Between percentile 50 and 85	7	16.7
Above percentile 85	0	0
Severity score		
Excellent	4	9.5
Good	22	52.4
Medium	13	30.0
Moderate	3	7.1
Severe	0	0
Global posture		
Normal	8	19.0
Moderate	26	61.9
Severe	8	19.0
Thoracic spine		
Without thoracic kyphosis	16	38.1
With thoracic kyphosis	26	61.9
FEV_1		
Below the predicted	19	45.2
Normal/above the predicted	23	54.8
FVC		
Below the predicted	13	31.0
Normal/above the predicted	29	69.0
FEV_1/FVC		
Below the predicted	8	19.0
Normal/above the predicted	34	81.0
DC in 6MWT		
Below the predicted	23	54.8
Normal/above the predicted	19	45.2

BMI = body mass index; DC in 6MWT = distance covered in the 6-minute walk test; FEV_1 = forced expiratory volume in the first second; FVC = forced vital capacity.

without and with thoracic kyphosis (639.8±105.2 *versus* 555.5±59.1 meters, p = 0.093; and 559.7±64.3 *versus* 574.5±78.0 meters, p = 0.868, respectively) (Table 3).

There was a correlation between thoracic kyphosis and age (r = 0.308, p = 0.048). BMI (r = 0.088, p = 0.581) and disease severity (r = -0.151, p = 0.339) did not correlate with the deformity.

After conducting univariate logistic regression, it was found that there was a harmful association between DC in 6MWT and children with kyphosis, as well as with FEV_1 (odds ratio > 1.00). Patients with abnormal chest showed DC below predicted and FEV_1 below 80% (Table 4). In this study, a patient with CF and kyphosis showed a 0.972 chance having FEV_1 below the predicted and 0.2513 of reaching a lower DC in 6MWT compared to a CF without kyphosis. For FVC, the result was -0.0225. In the application of multivariate logistic regression, none of the variables remained in the model.

Table 2 - Distribution of forced expiratory volume in the first second and forced vital capacity values in relation to cystic fibrosis in groups with and without thoracic kyphosis

Variable	n	Mean	SD	Minimum	Maximum	p
FEV_1						
Without thoracic kyphosis						
Lesser severity	8	95.6	12.2	80.0	115.0	
Greater severity	8	74.1	21.9	38.0	112.0	0.027
With thoracic kyphosis						
Lesser severity	18	80.3	22.5	32.0	112.0	
Greater severity	8	75,3	16.9	41.0	94.0	0.487
FVC						
Without thoracic kyphosis						
Lesser severity	8	97.6	13.2	81.0	118.0	
Greater severity	8	79.8	18.7	52.0	115.0	0.027
With thoracic kyphosis						
Lesser severity	18	85.2	19.6	53.0	118.0	
Greater severity	8	86.2	16.5	63.0	116.0	0.846
FEV_1/FVC						
Without thoracic kyphosis						
Lesser severity	8	98.3	8.6	83.3	106.9	
Greater severity	8	91.9	12.1	73.1	104.5	0.141
With thoracic kyphosis						
Lesser severity	18	93.7	14.1	60.4	112.0	
Greater severity	8	87.2	12.5	65.1	102.9	0.120

FEV_1 = forced expiratory volume in the first second; FVC = forced vital capacity; p = non-parametric Mann-Whitney test probability; SD = standard deviation. Values given in percent in relation to the predicted.

Table 3 - Distribution of distance covered in the 6-minute walk teste values in relation to cystic fibrosis severity in groups with and without thoracic kyphosis

DC in 6MWT	n	Mean	SD	Minimum	Maximum	p
Without thoracic kyphosis						
Lesser severity	8	639.8	105.2	486.0	772.8	
Greater severity	8	555.5	59.1	429.6	630.0	0.093
With thoracic kyphosis						
Lesser severity	18	559.7	64.3	415.2	643.0	
Greater severity	8	574.5	78.0	502.8	733.3	0.868

DC in 6MWT = distance covered in the 6-minute walk test; p = non-parametric Mann-Whitney test probability; SD = standard deviation. Values given in meters.

Table 4 - Distribution of frequencies and raw odds ratio in relation to spirometric variables (forced vital capacity and forced expiratory volume in the first second) and distance covered in the 6-minute walking test

Variable	n	With kyphosis n (%)	Without kyphosis n (%)	OR	95%CI	p*
FEV_1						
Above 80%	23	14 (60.8)	9 (39.2)	1	0.3148-3.8582	0.8792
Below 80%	19	12 (63.1)	7 (36.9)	1.1020		
FVC						
Above 80%	29	18 (62)	11 (38)	1	0.2546-3.7558	0.9739
Below 80%	13	8 (61.5)	5 (38.5)	0.9778		
DC in 6MWT						
Above/equal to the predicted	22	13 (60)	9 (40)	1	0.3678-4.4949	0.6939
Below the predicted	20	13 (65)	7 (35)	1.2857		

% = relative frequency; 95%CI = 95% confidence interval; DC in 6MWT = distance covered in the 6-minute walk test; FEV_1 = forced expiratory volume in the first second; FVC = forced vital capacity; n = absolute frequency; OR = odds ratio.
* Univariate logistic regression.

Discussion

The close relationship between breathing and posture is a consensus in the management of CF. Postural disorders are considered secondary to pulmonary disease. In this context, this study sought to assess the impact of increased thoracic kyphosis in pulmonary function parameters (spirometry) and functional capacity (6MWT) of children and adolescents with CF.

The identification of the high prevalence of thoracic kyphosis in the studied population of children and adolescents with CF corroborates findings and conclusions that the literature presents. The pathophysiological process and the progression of pulmonary disease are pointed as having the major responsibility for the development of alterations in body posture. This is because the disease progression with pulmonary hypersecretion leads to hyperinflation and frequent coughing episodes. These events have repercussions on the musculoskeletal system through important muscle shortenings. The shortenings are able to cause postural changes that can alter the respiratory mechanics.[2]

This relationship between posture and respiratory mechanics has been the focus of discussion by various authors.[2,4,16-20] According to them, air trapping is caused by recurrent inflammation, bronchial hypersecretion and decreased elastic recoil of the airways. These factors result in a remodeling of this structure, followed by chronic infection, with a consequent reduction of airway diameter and loss of lung elasticity. The hyperinflated thorax elevates the upper thorax and flattens the diaphragmatic domes. Such compensations raise the thoracic volume and the negativity of the pleural pressure, increasing transpulmonary pressure and airway diameter. The inspiratory muscles gradually adapt to the new position of the bone structure, which will lead to a shortening of the fibers and a reduction of these muscles' ability to generate strength.[2,16] In individuals with poor nutritional status, the development of spinal deformities seems to be enhanced.

Given this pathophysiological process and its intimate connection with the rib cage, the posture alteration most commonly seen in CF is increased thoracic kyphosis.[4,17-21] This alteration was found in the present study, with a prevalence (62%) similar to that found in the study by Parasa & Maffulli.[18] A lower incidence of around 15.1% had been published in the 1980s.[22]

The high incidence of musculoskeletal complications in CF has been associated with increasing age and decreasing pulmonary function.[4,18,23] Specifically on the posture, it is suggested that the presence of postural alterations has a correlation with reductions in lung function and in the capacity for physical exercise.[5,21,24] However, this was not observed in the current result, in which increased thoracic kyphosis was not related to spirometric data nor to 6MWT performance. The findings of Logvinoff & Erkilla[4,17] corroborate those presented here, because the kyphosis was also prevalent and had no correlation with the spirometric variables.

Some articles point to the decline in pulmonary function through important postural impairments,[21] especially the increased thoracic kyphosis.[23] Massie et al.[23] showed that this type of change is associated with increased age and low pulmonary function, which is still controversial, since the study by Erkilla et al. did not find similar results. In it, thoracic deformity did not correlate with the severity of the disease or with pulmonary function, as it does not in the present study, although the prevalence of kyphosis

increased with age.[4]

The association between the severity of cystic fibrosis and the impairment of posture does not follow a standard in the literature, just as the relationship between a worse posture and a worse pulmonary function. Unlike the findings presented here, as well as the study of Erkilla et al.,[4] Walshaw & Tattersall[21] found a correlation between posture and the severity of CF in adults. Denton et al.[22] had found the same in 1981 with a sample of 91 children and adolescents, whose increased thoracic kyphosis was related to the severity of lung disease. The same behavior was published by Henderson & Specter,[20] which also found a correlation between the presence of kyphosis and age.

In the results obtained here, although, statistically, the influence of the severity of cystic fibrosis on pulmonary function of patients with increased thoracic kyphosis was not found, the second logistic regression analysis, even in the absence of this thoracic deformity, children with lesser severity showed higher values of FEV[1] and FVC compared to the more severe patients (95.6±12.2 *versus* 74.1±21.9, p = 0.027, and 97.6±13.2 *versus* 79.8±18,7, p = 0.027). Thus, irrespective of the posture, the worsening in disease severity is related to worsening pulmonary function. This probably is associated to the fact that clinical deterioration determines the progression of lung disease.

In contrast, the study by Tejero Garcia et al.[19] found, in a sample of 50 CF patients aged over 16 years, a correlation of disease severity with FEV_1, FVC and increased thoracic kyphosis, and the latter was also related to FEV_1. However, this study was a sample of spirometry values lower than those presented here, as well as with an older population. Moreover, disease severity was classified by the spirometry data only and not by the broad SS, as done in the current investigation. All these factors may have influenced the difference in results between studies. Another divergent finding in the study by this author is due to the correlation found between the severity of the disease and the DC in 6MWT, an event that did not happen here, since it was observed that the older the subject, the higher is the DC in 6MWT. There was no statistically significant difference in DC in 6MWT between groups of greater and lesser severity, in patients with and without thoracic kyphosis (p = 0.616 and p = 0.068, respectively).

According to literature, the development of thoracic kyphosis has many causes. Among the factors that contribute to this deformity are: reduction of muscle mass,[25] of bone mineral density,[26] excessive respiratory effort,[22] osteopenia[24] and chronic pain.[26]

Its presence is often reported in publications on adult cystic fibrosis populations with a high percentage identified already in the pubertal phase,[27] as is the profile of this study's sample. And, according to Elkin,[27] even if those individuals do not have repercussions on lung function yet, there is a need for an early multiprofessional intervention. Lannefors et al.[28] emphasize this preventive idea, considering that there is a high prevalence of increased thoracic kyphosis that can be reversed if treated properly. According to the author, the maintenance of postural alignment reduces the risk of low back pain and spinal complications, besides contributing to the preservation of physical function.[28]

Longitudinal studies show that the treatment of postural disorders can be successful through assessment methods and appropriate programs.[16,21] Exercises for thoracic mobility, muscle stretching and coordination activities to promote improved posture and chest wall compliance, resulting in the maintenance and optimization of pulmonary function. Patients with more severe pulmonary disease, lower aerobic capacity and more sedentary lifestyles deserve special attention because they are predisposed to lower bone mineral density, higher prevalence of vertebral fracture and development of major thoracic kyphosis increases.[19]

Although the logistic regression analysis was not statistically significant for any of the variables, the odds ratio value of the variables DC in 6MWT and FEV_1 show a tendency for the presence of kyphosis to be a risk factor for lower performance on the 6MWT and for lower spirometric value. A larger sample could sensitize statistical tests and contribute to the identification of new results.

The absence of a larger sample, characterized by patients more severely impaired and older, may have restricted the identification of results, as well as the application of a more sensitive instrument of postural assessment. Studies with longitudinal design to assess the evolution of thoracic deformity and the effect of therapeutic interventions, both in the musculoskeletal and respiratory systems, are suggestions for future supplementary investigations.

Conclusion

There was a high prevalence of increased thoracic kyphosis in children and adolescents with CF. This deformity did not alter pulmonary function and functional capacity and was not related to the severity of the disease. However, regardless of posture, the worsening in disease severity caused a greater decline in pulmonary function.

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