Exercise capacity and maximum inspiratory pressure in patients with chronic respiratory failure due to Kyphoscoliosis in Turkey

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Abstract

Aim: Kyphoscoliosis is one of restrictive disorders in which respiratory muscle weakness develops. The aim of the study was to verify if exercise capacity and inspiratory muscle strength could be related with body composition, arterial blood gases (ABG), pulmonary functions, systolic pulmonary arterial pressure (PAP), psychological status, quality of life, dyspnea, and these could be indicators for exercise capacity in patients with chronic respiratory failure due to kyphoscoliosis.

Methods: The data of thirty patients using long-term noninvasive mechanical ventilation (NIMV) were used for this retrospective study conducted in Ankara, Turkey. Respiratory muscle strength was assessed using maximal inspiratory, expiratory pressure (MIP, MEP); exercise capacity using incremental shuttle walk test (ISWT); dyspnea using Medical Research Council (MRC) scale; quality of life using St. George's Respiratory Questionnaire (SGRQ); psychological status using Hospital Anxiety Depression (HAD) score; systolic PAP using echocardiagraphy. ABG, body, fat-free mass index (BMI, FFMI) were also recorded.

Results: The median value of MIP was 44, MEP was 83 cmH_2O. MIP was found correlated with ISWT (r=0.385, p=0.043), MEP (r=0.817, p<0.001), arterial oxygen pressure (r=0.642, p<0.001), smoking (r= -0.723, p<0.001) MRC (r= -0.671, p<0.001), sPAP (r= -0.428, p=0.026). MEP was correlated with smoking (r= -0.792, p<0.001), MRC (r= -0.489, p=0.008). ISWT was correlated with SGRQ (r= -0.531, p=0.003), depression, MRC (r=0.640, r=0.690, p<0.001, respectively), MIP (r=0.385, p=0.043).

Conclusion: Patients with chronic hypercapnia due to kyphoscoliosis could have low inspiratory muscle strength. Patients with pulmonary hypertension, hypoxemia, limited exercise capacity, especially severe dyspnea and history of smoking should be evaluated for respiratory muscle weakness. Additionally, patients with limited exercise capacity could have worse quality of life, more severe dyspnea and depression.

***Keywords*:** exercise capacity, kyphoscoliosis, long-term noninvasive mechanical ventilation, maximal expiratory pressure, maximal inspiratory pressure, respiratory muscle weakness.

Introduction

Kyphoscoliosis is one of the chest wall disorders in which respiratory failure develops, which results in reduced chest wall and lung compliance. The main pathology seems to be in changes to the mechanics of the rib cage and compliance. The severity of lung restriction has been found related to the severity of the deformity. The most important and progressive symptom in these patients is exercise dyspnea, which results in reduced exercise tolerance. As a result, deterioration in daily life activities and health-related quality of life (HRQOL) are frequently seen (1,2). One of prognostic factors in patients with kyphoscoliosis is hypercapnic respiratory failure. It has been shown that blood gas values, exercise capacity, HRQOL, survival, and also maximum inspiratory pressure (MIP) are likely to be improved by using NIMV and long-term oxygen therapy (LTOT) in these patients (3-8).

Maximal inspiratory pressure (MIP) and maximal expiratory pressure (MEP) are widely used tests for evaluating respiratory muscle strength, which are noninvasive and easy to perform. Furthermore, they have been shown to be useful in diagnosing and following up patients with various pulmonary and cardiac disorders (9,10).

The aim of the study was to verify if exercise capacity and inspiratory muscle strength could be related with body composition, arterial blood gas (ABG) analysis, pulmonary functions, systolic pulmonary pressure, psychological status, quality of life, and sensation of dyspnea, and whether these parameters could be indicators of exercise capacity in patients with chronic respiratory failure due to kyphoscoliosis.

Methods

The data of thirty patients who were diagnosed as having kyphoscoliosis were investigated and recorded for use in this retrospective cross-sectional study conducted in Ankara, Turkey. Approvals were obtained from local institutional review board before the parameters were recorded. Patients who were under age of 18, non-smokers, have uncontrolled diabetes mellitus, systemic hypertension and not able to perform walk test were not included into study. All patients were using NIMV and LTOT for about 6-8 hours per night for a minimum of three months. Respiratory muscle strength was assessed using maximal inspiratory, expiratory pressure (MIP, MEP); exercise capacity using the incremental shuttle walk test (ISWT); dyspnea using the Medical Research Council (MRC) scale; quality of life using the St. George's Respiratory Questionnaire (SGRQ); psychological status using the Hospital Anxiety and Depression (HAD) score; and systolic pulmonary pressure (PAP) using echocardiagraphy. Arterial blood gas (ABG), body mass index (BMI), and the fat-free mass index (FFMI) were also recorded.

Respiratory muscle strength

Respiratory muscle strength was evaluated by measuring the MIP and MEP using a Micro-RPM respiratory pressure meter (Care Fusion, Hoechberg, Germany). MIP and MEP were measured with the subject in a sitting position by the same physiotherapist, in accordance with the recommendations of the American Thoracic Society and European Respiratory Society (ATS-ERS) (11). MIP was measured starting from residual volume and MEP was measured starting from total lung capacity. Tests were repeated a minimum three times, and the best value was recorded.

Other measurements

Spirometry was performed to determine forced vital capacity (FVC), forced expiratory volume in one second (FEV1), and FEV1/FVC using a spirometer (AS-507, Minato Medical Science, Tokyo, Japan), in accordance with the ATS-ERS guidelines (11). FVC and FEV1 were measured three times and the greatest value was recorded. Exercise capacity was assessed using the ISWT. The patients walked around a 10 m course. The speed at which the patient walked was dictated by an audio signal and increased by 0-17 m/s each

minute. The test continued until the subject was not able to continue or keep up with the required pace. The distance walked was recorded. The ISWT was conducted by the same physiotherapist (12). Blood gas analysis was performed at rest with room air (without nasal oxygen) using a COBAS B 121 (ROCHE, West Roxbury, USA) before the exercise test. Echocardiagraphy was performed using an Epiq 7 ultrasound machine (Philips, Amsterdam, Holland). Body mass and the fat-free mass index (FFMI) were measured using bioelectrical impedance (BIA model TBF-300; Tanita Corporation, Tokyo, Japan).Body mass index (BMI) and FFMI were calculated based on the formula where body weight / fat-free mass in kilograms were divided by height in meters squared.

Statistical analysis

Data were evaluated using the Statistical Package for Social Science for Windows version 18.0 and by analyzing descriptive statistics. The Shapiro-Wilk's test was used to determine the normality of distribution of the data. Correlations between parameters were assessed with Pearson's correlational coefficient. Correlation coefficients reflected poor (0.26-0.49), moderate (0.50-0.69), and high relationship ($\geq$0.70). A p-value $\leq$0.05 was considered as statistically significant in all cases.

Results

Thirty patients were enrolled in this study. Each of them was diagnosed as having chronic respiratory failure due to kyphoscoliosis. The overall mean age of the patients was 53±14 years. Ten patients were females. The mean FEV1 value was 36%±13%, whereas mean FVC was 39%±19%. Clinical characteristics of the patients are presented in Table 1.

Table 1. Clinical characteristics of the patients

	Mean±SD	Median	Minumum : maximum
Age (year)	53±13	54	26:76
Smoking (pack.year)	10±14	2	0:50
MRC	3.3±1	3	2:5
FEV1%	36±13	34	13:63
FVC %	39±19	37	15:85
MIP (cm H_2O)	48±23	44	17:94
MEP (cm H_2O)	83±42	83	21:152
PH	7.37±0.02	7.37	7.35:7.46
PO_2 (mm Hg)	58±8	57	45:76
PCO_2 (mm Hg)	50±6	51	40:64
SPAP (mm Hg)	40±10	37	26:65
BMI (kg/m^2)	26±5	26	17:40
FFMI (kg/m^2)	19±2	19	16:24
Anxiety score	9.8±1.5	9	7:12
Depression score	9.5±1.7	9	6:14
ISWT (meter)	222±129	220	30:530
SGRQ total	61±16	59	29:100

MRC: Medical Research Council, FEV1: forced expiratory volume, FVC: forced vital capacity, MIP: maximum inspiratory pressure, MEP: maximum expiratory pressure, PO_2: partial oxygen pressure, PCO_2: partial oxygen pressure, Spap: systolic pulmonary arterial pressure, BMI: body mass index, FFMI: fat-free mass index, ISWT: incremental shuttle walk test, SGRQ: St. George's respiratory questionnaire.

The median value of MIP was 44 cmH_2O and MEP was 83 cmH_2O. Eight patients had an MIP value under 30 cmH_2O, whereas four patients were over 80 cmH_2O. MIP was found positively correlated with ISWT (r=0.385, p=0.043), MEP (r=0.817, p<0.001), and arterial oxygen pressure (r=0.642, p<0.001) and negatively with cigarette smoking (r=-0.723, p<0.001), MRC scale (r=-0.671, p<0.001), and systolic PAP (r=-0.428, p=0.026). There was no correlation between MIP and age, BMI, FFMI, SGRQ, HAD score, and p CO2 values. MEP was correlated with cigarette smoking (r=-0.792, p<0.001), and negatively with the MRC scale (r=-0.489, p=0.008). FEV1 and FVC predicted were only correlated with age (p=0.009, r=0.478, p=0.013, r=0.452, respectively). ISWT was negatively correlated with SGRQ (r= -0.531, p=0.003), depression, and MRC scores (r=0.640, r=0.690, p<0.001 respectively), and positively with MIP value (r=0.385, p=0.043).

Discussion

This study showed that cigarette smoking, increased systolic PAP, sensation of dyspnea, reduced exercise capacity, and arterial partial oxygen pressure were correlated with inspiratory muscle weakness in patients with kyphoscoliosis using long-term NIMV. The strongest correlation was found between inspiratory strength and history of smoking. Smoking and dyspnea were also related with expiratory muscle strength. FEV1 and FVC predicted were only correlated with age. Additionally, reduced exercise capacity was associated with deterioration in quality of life, dyspnea, and depressive status.

Kyphoscoliosis is one of the most common pulmonary restrictive lung diseases. The mechanism of dyspnea is multi-factorial; decreased compliance of the chest wall results in increased work in breathing. The patient's breathing pattern is usually rapid and shallow, but it causes alveolar hypoventilation (13). Reduced diffusion of oxygen across the alveolar membrane results in shunting of blood through non-aerial areas of the lungs and deterioration in the ventilation-perfusion ratio (14-16). As a result of these pathologies, hypoxemia and hypercapnia occur. The main treatment for hypoxemia and hypercapnia is LTOT and NIMV. However, a definite effect of NIMV has not been shown, it is supposed to increase respiratory drive secondary to improving nocturnal blood gases and reducing sleep fragmentation, resting respiratory muscles, and increasing chest wall and lung compliance by opening the at electatic areas and improving the ventilation-perfusion ratio (8). Another pathologic finding is respiratory muscle weakness. The strength of the diaphragm and other inspiratory muscles could be detected using MIP, and that of abdominal muscles and other expiratory muscles with MEP. Several studies demonstrated that MIP was improved in patients with kyphoscoliosis using NIMV (8,17).MIP values under 30 cmH_2O have been found related with respiratory failure, especially in patients with hypercapnia. Muscle weakness has not been seen over 80 cmH_2O for MIP and 100 cmH_2O for MEP values. In this study, the median values of MIP and MEP were above these cutoffs; eight patients had MIP values under 30cmH_2O, four patients were over 80 cmH_2O. MIP was correlated moderately with PO_2 but not correlated with PCO_2. A relation may have been found if the patients had not been using NIMV.The age and degree of chest wall deformity were shown as factors that contributed to abnormalities in gas exchange and ABG (17). Similar to our findings, in another study, PaO_2 values ranged from 41-64 mm Hg and the average value of $PaCO_2$ was 49.85 mm Hg in patients with kyphoscoliosis (8).

Reduced exercise capacity and exertional dyspnea are important symptoms that are often seen. It was shown that exercise capacity was not related with the severity and nature of the spinal deformity but related with respiratory muscle strength (18,19). Combined low MIP and MEP values reflect systemic muscle weakness. In our study, albeit poorly, inspiratory muscle strength was related with exercise capacity. In several studies, inspiratory

muscle strength and exercise capacity were shown to be related with the degree of dyspnea sensation (18,19). In our study, dyspnea as assessed using the MRC scale was moderately correlated with both MIP and MEP values. Another significant correlation in our study was found between MIP and systolic PAP. It has already been demonstrated that respiratory muscle strength, especially MIP, is low in patients with pulmonary hypertension (20,21). Smoking is another factor with which a relationship has been shown with respiratory muscle weakness (22). In our study, a significant and high correlation was found between cigarette smoking and expiratory and inspiratory muscle strength. It is compatible that smoking alone is likely to contribute to reducing muscle weakness (23).

Reduced exercise capacity and exertional dyspnea are also important factors that limit quality of life in chronic respiratory disorders. It has previously been shown that peripheral muscle strength, dyspnea, and exercise capacity are independent factors that affect quality of life in patients with kyphoscoliosis who are diagnosed as having chronic respiratory failure (24). In our study, exercise capacity was significantly related with quality of life, dyspnea, and depressive mood. It was not an unexpected result that patients with kyphoscoliosis who had depression and dyspnea had reduced exercise capacity and consequently reduced HRQL.

A limitation of this study included the lack of age- and sex-matched group that was not using NIMV. Lung volumes could have been performed and the Cobb angle could have been measured.

Conclusion

Patients with chronic hypercapnia who have kyphoscoliosis could have low inspiratory muscle strength regardless of using NIMV. Patients with a history of smoking, pulmonary hypertension, reduced exercise capacity, hypoxemia, and dyspnea could have more inspiratory muscle weakness with any FEV1, FVC values. Both inspiratory and expiratory muscle weakness may be seen in patients with more severe dyspnea and with a history of smoking. Additionally, patients with limited exercise capacity could have worse quality of life, more severe dyspnea, and depression.

Conflicts of interest: None declared.

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