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ORIGINAL RESEARCH

Respiratory muscle strength and Modified Shuttle Walk Test performance in schoolers with cystic fibrosis

Força muscular respiratória e desempenho no Modified Shuttle Walk Test em escolares com fibrose cística

Fuerza muscular respiratoria y rendimiento en el Modified Shuttle Walk Test en escolares con fibrosis quística

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ABSTRACT | Modified Shuttle Walk Test (MSWT) is a potentially maximal exercise test that, together with the assessment of respiratory muscle strength (RMS), reflects the respiratory condition and exercise capacity of schoolchildren with cystic fibrosis (CF). This study aimed to investigate the relationship between the RMS and the performance in the MSWT by schoolchildren with CF and to compare the data obtained with the values predicted in the literature. This is a cross-sectional observational study that included schoolchildren with CF. Anthropometric evaluation, spirometry and RMS evaluation were performed, using the maximal inspiratory (MIP) and expiratory (MEP) pressures (Globalmed MVD300® manovacuometer) (ATS/ERS) (2002). Two MSWT were performed, with an interval of 30 minutes between them. The distribution of the data by the Shapiro-Wilk test was applied and paired t-test was used to compare the values of the evaluations with those predicted, as well as for comparison between genders. Pearson test was used for correlation between MIP and MEP and the performance in the MSWT. Significance of 5% was accepted. 28 children (9.9±1.9 years) participated; 57.14% showed MIP below the predicted (15 children) and 53.57% showed MEP below the predicted (16 children). The mean performance was 730.4±266.1m, which is lower than the values predicted in the literature. No relationship between performance and RMS was observed. A moderate correlation was observed between MIP and MEP values (r=0.58, p=0.01). No relationship between the RMS and the MSWT performance was observed in schoolchildren with CF of this study. The RMS and the performance in the MSWT were below the predicted in the literature.

Keywords | Cystic Fibrosis; Child; Respiratory Muscles; Exercise Test.

RESUMO | O *Modified Shuttle Walk Test* (MSWT) é um teste de exercício potencialmente máximo que, associado à avaliação da força muscular respiratória (FMR), reflete a condição respiratória e a capacidade de exercício de escolares com fibrose cística (FC). O objetivo desta pesquisa foi investigar a relação entre FMR e distância percorrida (DP) no MSWT realizado por escolares com FC e comparar os dados obtidos com valores preditos na literatura. Trata-se de um estudo observacional transversal que incluiu escolares com FC. Realizou-se avaliação antropométrica, espirometria e FMR, utilizando as pressões inspiratória máxima ($PI_{máx}$) e expiratória máxima ($PE_{máx}$) por meio da

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manovacuometria. Dois MSWT foram realizados, com intervalo de 30 minutos entre eles. Verificou-se a distribuição dos dados pelo teste de Shapiro-Wilk e aplicou-se teste t pareado para comparação entre valores das avaliações e predito, bem como para comparação entre gêneros. Aplicou-se teste de Pearson para correlação entre $PI_{máx}$ e $PE_{máx}$ e DP no MSWT. Aceitou-se significância de 5%. Participaram 28 crianças (9,9±1,9 anos) destas, 57,14% apresentaram $PI_{máx}$ abaixo do predito (15 crianças) e 53,57% da $PE_{máx}$ (16 crianças). A média da DP foi 730,4±266,1m, abaixo do predito na literatura. Não houve relação entre DP e FMR. Identificou-se correlação moderada entre valores de $PI_{máx}$ e $PE_{máx}$ (r=0,58 e p=0,01). Não houve relação entre FMR e desempenho no MSWT nos escolares com FC estudados. A FMR, bem como o DP no MSWT, apresentou-se abaixo do predito na literatura.

Descritores | Fibrose Cística; Criança; Músculos Respiratórios; Teste de Esforço.

RESUMEN | El Modified Shuttle Walk Test (MSWT) es una prueba de ejercicio potencialmente máxima que, asociada a la evaluación de la fuerza muscular respiratoria (FMR), refleja la condición respiratoria y la capacidad de ejercicio de escolares con fibrosis quística (FQ). El objetivo de esta investigación fue investigar la relación entre FMR y distancia recorrida (DP) en el MSWT realizado por escolares con FQ y comparar los datos obtenidos con valores predichos en la literatura. Se trata de un estudio observacional transversal que incluyó a los escolares con FQ. Se realizó una evaluación antropométrica, espirometría y FMR, utilizando las presiones inspiratoria máxima ($PI_{máx}$) y espiratoria máxima ($PE_{máx}$) por medio de la manovacuometría. Se realizaron dos MSWT, con un intervalo de 30 minutos entre ellos. Se verificó la distribución de los datos por la prueba de Shapiro-Wilk y se aplicó una prueba t pareada para la comparación entre los valores de las evaluaciones y el predicado, así como para la comparación entre los géneros. Se aplicó una prueba de Pearson para la correlación entre $PI_{máx}$ y $PE_{máx}$ y DP en el MSWT. Se aceptó una significación del 5%. Participaron del estudio 28 niños (9,9±1,9 años); 57,14% presentaron $PI_{máx}$ por debajo del pronóstico (15 niños) y 53,57% por debajo del pronóstico para $PE_{máx}$ (16 niños). El promedio de la DP fue 730,4±266,1m, por debajo del predicho en la literatura. No hubo relación entre DP y FMR. Se identificó una correlación moderada entre valores de $PI_{máx}$ y $PE_{máx}$ (r=0,58 y p=0,01). No hubo relación entre FMR y desempeño en el MSWT en los escolares con FC. La FMR, así como la DP en el MSWT, se presentó abajo del predicho en la literatura.

Palabras clave | Fibrosis Quística; Niño; Músculos Respiratorios; Prueba de Esfuerzo.

INTRODUCTION

Cystic fibrosis (CF) is an autosomal recessive multisystem genetic disorder. Pulmonary and digestive alterations are among the clinical manifestations evidenced[1].

Alterations in the digestive system favor malnutrition, which leads to a poor development of body, indirectly affecting the muscular system[1,2]. Respiratory tract involvement is due to electrolyte changes that make the secretions thicker, increasing the susceptibility to infections and persistent inflammatory responses[2]. These changes favor the reduction in respiratory muscle strength (RMS), a factor that may contribute to fatigue and exercise intolerance[3].

The assessment of RMS can be performed using a digital manovacuometer, a non-invasive instrument that is easy to use and has a high success rate in children and adolescents[4].

Testing the exercise tolerance and cardiorespiratory fitness of children with CF is necessary to measure the impact of the disease on the patient's life[5]. Once the typical changes caused by the disease can reduce the exercise functional capacity by 95 % [6]. The literature reports that the Modified Shuttle Walk Test (MSWT) is highly recommended for clinical practice, as a valid, reliable and effective stress test, and can be safely performed in patients with CF[7].

Therefore, considering the importance of these resources when assessing CF patients, this study aimed to verify whether RMS is associated with MSWT performance or not; and to compare the values obtained in children and adolescents with CF with those predicted in the literature. The hypothesis was that RMS influences the physical performance of the population studied.

METHODOLOGY

This is a cross-sectional quantitative study that included students aged between 6 and 15 years, from a

CF reference center and a university extension program. The participants' CF diagnosis was confirmed by the sweat and genetic tests[1], and the mutation was obtained by analyzing the medical records. The study was approved by the Research Ethics Committee of the children's hospital (CAAE: 36493314.8000.5361), respecting the principles of ethics in research with human beings present in the Resolution 466/12 of the Brazilian National Health Council.

All participants had the Informed Consent Form and the Minor Assent Form signed by their parents/guardians.

Children instructed, without acute respiratory disease at data collection, whose clinical stability was confirmed by the Cystic Fibrosis Clinical Score[12] and the Cystic Fibrosis Foundation Score [8]were included. Disease severity was classified according to the Shwachman-–Doeurshuk score (SS)[9].

Data were collected in two stages. In the first stage, the anthropometric data and the assessment of RMS were obtained by measuring the maximum inspiratory pressure (MIP) and the maximum expiratory pressure (MEP). In the second stage, the patient underwent spirometry and two assessments of MSWT, with a 30-minute interval between them.

A digital scale, Wiso® Ultra Slim W903, was used for measuring body mass (kg), according to the criteria established by the World Health Organization [10]. Height was measured (cm) using a Sanny® portable stadiometer. The body mass index (BMI) of each child was calculated using the equation: body mass (kg)/height^2 (m). The reference values used followed the classification available from Programa Nacional Telessaúde Brasil Redes of the Ministry of Health website.

The RMS was assessed using the Globalmed MVD300® digital manovacuometer, which has a 2mm diameter orifice to prevent glottal closure during the MIP and to reduce the use of buccal muscles during the MEP. After orientations, the evaluator used verbal stimuli to perform the patient's measurements. During the maneuver, the child remained seated, with the feet on the floor, and used nose clip. The measurements were performed according to the norms of the American Thoracic Society (ATS)[11]. To obtain the MIP, the child was instructed to exhale close to the residual volume, then to perform a maximal inspiration up to about the total lung capacity (TLC). The MEP was measured from an inspiration close to the TLC, followed by a maximal expiration up to about the residual volume. At least three and at most seven maneuvers were performed for each of the MIP and MEP measurements, and the best value obtained was recorded. An interval of 30 seconds occurred after each maneuver. Between the measurement of MIP and MEP, a three-minute interval occurred to avoid the patient's fatigue [12]. The data obtained were compared with the data proposed in the reference equations by Rosa et al.[13] for children aged between six and ten years. From 11 years old on, the equation proposed was that by Domenech et al.[14].

To characterize the sample, spirometry was performed using the pneumatograph Jaeger Master Scope IOS - Germany. At least three acceptable and two reproducible curves were obtained, with at least one-minute interval between maneuvers, according to the standardization by the ATS[15]. Measurements were used in percentage and absolute values, according to Polgar[16] and Knudson et al.[17] After spirometry, the functional capacity was assessed using MSWT, according to the protocol proposed by Bradley et al.[18]. The MSWT is an incremental shuttle walk test composed of 15 levels, whose speed is determined by a beep. If the patient reported being very tired and unable to continue the test or if there was a loss of rhythm of the audio signal, the test would be stopped, which did not happen. The best performance was considered for analysis.

Descriptive and frequency statistics were used to present the results. Initially, the Shapiro-Wilk test was used for assessment of the normality of data. The comparison between the values of RMS variables measured and of the performance in the MSWT and the corresponding values predicted was made using the paired t-test for the total of participants, as well as for the comparison between the genders. The Pearson correlation test was used to analyze the correlation between MIP and MEP and the performance in the MSWT. The significance value set for this study was $p<0.05$.

RESULTS

The sample of this study was composed of 28 children and adolescents with CF, whose mean age was 9.9 ± 1.9 years. The data for sample characterization are shown in Table 1. Regarding the genotype, 53.57% of participants were ΔF508 heterozygotes, 28.57% ΔF508 homozygotes, and 17.85% carried other mutations.

Table 1. Distribution and frequency of data of sample characterization

Parameters	Mean±SD	(minimum – maximum)
Age (years old)	9.9±1.9	(6 – 14)
BMI (Kg/m^2)	15.7±1.9	(13.4 – 19.7)
Height (cm)	132.2±28.8	(116 – 168)
Weight (kg)	30.5±8.6	(20.6 – 54.3)
Shwachman score (scores)	83.4±2.7	(50 – 100)
FEV_1	66.7±22.2	(21.8 – 105.6)
FVC (% predicted)	80.1±18.2	(44.3 – 116.6)
FEV_1/FVC (%)	87.4±14.4	(54.0 – 113.0)
FEF_{25-75} (%)	51.1±28.7	(7.40 – 101.7)
PEF (%)	65.5±21.6	(22.5 - 108.3)

SD: standard deviation; BMI: body mass index; FEV_1: forced expiratory volume in the first second; FVC: forced vital capacity; FEV_1/FVC: ratio between forced expiratory volume in the first second and forced vital capacity; FEF_{25-75}: forced expiratory flow at 25-75% of the FVC; PEF: peak expiratory flow; %: percentage of value predicted.

Regarding the RMS, 57.14% of the total sample showed a measured value of MIP (ten boys and six girls) below that predicted, and 53.57% of the participants obtained lower MEP values (ten boys and five girls). The performance in the MSWT of all participants was below the expected. In the comparison between the absolute values of the parameters of RMS and of performance in the MSWT measured and the corresponding values predicted for each gender, a difference was observed between boys and girls regarding the values measured and predicted (Table 2).

Table 2. Comparison between the mean of absolute values of RMS and performance in the MSWT obtained by the participants and the mean of the values predicted for each parameter

Parameters	Total n=28 (Mean±SD)	Boys n=16 (Mean±SD)	Girls n=12 (Mean±SD)
MEP measured (cmH_2O)	77.1±22.0	80.4±22.4	72.8±21.4
MEP predicted equation (cmH_2O)	83.9±27.7	89.6±30.0	76.3±23.4
MEP measured (cmH_2O)	−66.0±26.0	−62.6±28.4	−70.7±22.9
MEP predicted equation (cmH_2O)	−66.9±18.1	−70.6±20.1	−61.9±14.5
Performance MSWT – measurement (m)	730.4±266.1	805.0±280.6	630.8±218.1
MSWT performance predicted (m)	1018.6±131.1*	1103.7 ± 90.6 *	905.2±80.1*

SD: standard deviation; MEP: maximum expiratory pressure; MIP: maximal inspiratory pressure; MSWT: Modified Shuttle Walk Test; cmH_2O: centimeter of water; m: meters; *Paired t test: $p<0.05$, difference is observed between the mean of the value predicted and the mean of the value measured in the participants.

Correlation between the performance in the MSWT and any variable of RMS was not observed. A moderate correlation was observed between MIP and MEP values ($r=0.58$ and $p<0.01$).

DISCUSSION

This study showed more than a half of the schoolchildren with CF assessed had MIP (57.14%) and MEP (53.57%) values below the values predicted, despite they had mild severity of the disease. The mean of performance in the MSWT was below the value estimated (730.4m±26m). No relationship was observed between performance in MSWT and RMS variable, but a moderate correlation was found between MIP and MEP values.

In the longitudinal study by Donadio et al.[19], in which 26 youths with CF were followed-up for 5 years , one found that, although MIP increased, MEP and performance in the 6-minute walk test (6MWT) remained stable. According to the authors, functional capacity apparently does not occur together with the expected decline in lung function over time, while inspiratory muscle strength increases as the disease progresses. These findings corroborate the results of this study, in which there was no correlation between the RMS and the performance in the MSWT.

Contrarily, Saglam et al.[20] found a moderate correlation between the MSWT and the MIP ($r=0.517$ and $p=0.01$) and between MEP ($r=0.552$ and $p<0.01$), as well as a correlation between the 6MWT and the MIP ($r=0.377$ and $p = 0.01$) and with the MEP ($r=0.343$ and $p=0.02$) in a study that evaluated 50 fibrocystic patients between 7 and 25 years of age.

In the case of adolescents and children with CF, the values of RMS are divergent, and there may be both weakness and increased strength of the respiratory muscles [21]. This is because, in CF, the nutritional status and oxygen uptake into muscle tissue greatly interfere. It results in the inability or weakness of the overall musculature, including the respiratory muscles. Contrarily, when the integrity of the muscle is preserved, the RMS increases due to the ventilatory demand, in cases of greater resistance to airflow. This causes muscle fatigue in the long term[21,22].

Regarding the performance in the MSWT, all the participants of this study showed values below that predicted in the literature, according to the reference equation by Lanza et al.[23]. This result may be related to the decline in pulmonary function and the decreased performance when doing aerobic exercise[19]. However, the study by Coelho et al.[24] compared the performance of 28 children in the MSWT (14 with mild CF and mean age of 11.5 ± 2.5 years and 14 healthy children with mean age of 11.2 ± 1.8 years) and showed MSWT performance was similar between the two groups.

Concering the percentage predicted for MIP and MEP, most of the study participants showed values below the expectations. The study by Dassios et al.[25] evaluated the nutrition, pulmonary function, and respiratory muscle function of 37 fibrocystic patients aged between 7 and 34 years who did regular aerobic exercises. These active patients were compared with a control group of CF paired for age and sex, whose participants did not do regular exercises. MIP and MEP were significantly higher in the group of those who do exercise compared with the control group (92x63cmH_20 and 94x64cmH_2O, respectively). The authors concluded that patients with CF who do regular aerobic exercises maintain higher rates of RMS.

In the same line, Orenstein et al.[26] reported a significant increase in respiratory muscle resistance in CF patients after a supervised three-month conditioning program. In its turn, the study by Vendrusculo et al.[27] showed 34 children and adolescents with CF, aged between 8 and 16 years, with normal lung function, had increased inspiratory muscle strength and decreased inspiratory muscle resistance when compared with healthy individuals. These findings suggest changes in respiratory muscle appear to be clearly associated with pulmonary involvement, and the force is related to lung function parameters, whereas inspiratory muscle resistance has a higher correlation with airway resistance.

Considering that, a shortage of studies correlating RMS with MSWT performance in fibrocystic children and adolescents is identified. One verifies that the behavior of the patients' population studied show RMS values and performance in an exercise test below the estimated when compared with those of the literature. Further studies, with a larger sample size, are necessary to draw attention to these findings. One realizes the relevance of studying the behavior of the RMS in these patients as a therapeutic focus and parameter of evaluation of the disease progression, as well as its relationship with the exercise capacity in this population.

CONCLUSION

In this study exclusively evaluating children and adolescents with CF, no relationship was found between the RMS and the performance in the MSWT. Although our initial hypothesis was not confirmed, our data indicate CF patients' values are below that predicted in the literature for the parameters RMS and physical performance assessed by the MSWT. Multidisciplinary care is known to be essential in this population, and, as the literature shows, nutritional status and regular physical exercise can influence the RMS measurements and the physical performance.

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