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Anthropometric and musculoskeletal assessment of patients with Marfan syndrome

Avaliação antropométrica e musculoesquelética de pacientes com síndrome de Marfan*

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Abstract

Background: Marfan syndrome (MS) is an autosomic dominant condition of the connective tissue that involves the ocular, cardiovascular and musculoskeletal systems. MS is caused by mutations in the fibrillin-1 gene, leading to joint ligaments flaccidity, joint hypermobility and an overgrowth of the long bones. **Objectives:** The aim of the present study was to assess anthropometry, musculoskeletal alterations and the prevalence of physical therapy treatments among patients with MS. **Methods:** Twenty-six patients were included in this study [17 females (age: 13.23±2.77 years; body mass 51.5±24-68 Kg; height 1.70±1.40-1.81 m; arm span: 1.73±0.12 m) and 9 males (age: 14.44±2.18; body mass: 61.0±42-72 Kg; height: 1.83±1.66-1.97 m; arm span: 1.93±0.13 m)]. Anthropometric measurements and musculoskeletal abnormalities were determined in a standardized fashion: pectus and scoliosis were assessed through radiography and angulation (â) of the scoliosis curve using the Cobb method; arachnodactyly was assessed through the thumb sign and Walker-Murdoch test and dolichostenomelia was assessed by arm span in relation to height. Patients also responded to a questionnaire addressing participation in physical therapy. **Results:** In comparison to values estimated for the Brazilian population, mass and height were greater among the patients with MS (females: p=0.001 e p<0.0005 e males p=0.019 e p=0.0001, respectively). The following musculoskeletal abnormalities were found: pectus in 3 patients (11%), pectus and scoliosis in 19 (73%), dolichostenomelia in 11 (42%) and arachnodactyly in 21 (80%). Eleven patients (42%) with MS had previously undergone physical therapy. **Conclusions:** Patients with MS exhibit altered musculoskeleto and anthropometry and have infrequent physical therapy treatment.

Keywords: Marfan Syndrome; anthropometry; musculoskeletal system; connective tissue; physical therapy.

Resumo

Contextualização: A Síndrome de Marfan (SM) é uma doença autossômica dominante do tecido conjuntivo que envolve os sistemas ocular, cardiovascular e musculoesquelético, causada por mutações no gene da fibrilina1, gerando flacidez nos ligamentos articulares, favorecendo a hipermobilidade articular e redução na contenção do crescimento ósseo. **Objetivos:** Avaliar as medidas antropométricas, alterações musculoesqueléticas e a frequência do tratamento fisioterapêutico nos pacientes com SM. **Métodos:** Participaram deste estudo 26 pacientes, sendo 17 do gênero feminino, com idade de 13,23±2,77 anos, massa corpórea de 51,5±24-68 Kg, altura de 1,70±1,40-1,81 m e envergadura de 1,73±0,12 cm, e nove do gênero masculino, com idade de 14,44±2,18, massa corpórea de 61,0±42-72 Kg, altura de 1,83±1,66-1,97 m e envergadura de 1,93±0,13. Foram obtidas medidas antropométricas, alterações ME de forma padronizada, sendo o *pectus* e a escoliose, por avaliação radiológica, e a angulação (â) da curva escoliótica, pelo método de Cobb; a aracnodactilia, pelo sinal do polegar e teste de Walker-Murdoch, e a dolicostenomelia, pela envergadura em relação à altura. Os pacientes responderam a um questionário quanto à participação em tratamento de fisioterapia. **Resultados:** Quando comparados com a estimativa brasileira, a massa corpórea e a altura apresentaram valores maiores no gênero feminino (p=0,001 e p<0,0005) e masculino (p=0,019 e p=0,0001). Das alterações musculoesqueléticas, encontrou-se *pectus* em 3 (11%), *pectus* e escoliose em 19 (73%), dolicostenomelia em 11 (42%) e aracnodactilia em 21(80%). Onze (42%) pacientes com SM já haviam realizado tratamento de fisioterapia. **Conclusões:** As alterações antropométricas e musculoesqueléticas estão presentes na SM, e o tratamento fisioterapêutico é pouco frequente.

Palavras-chave: Síndrome de Marfan; antropometria; sistema musculoesquelético; tecido conjuntivo; fisioterapia.

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Introduction

Marfan Syndrome (MS) is an autosomic dominant disease of the connective tissue caused by mutations in the fibrilin-1-gene of the chromosome 15[1] that involves several systems such as skeletal, ocular, cardiovascular, pulmonary, integumentary and neurological.

MS incidence is approximately 1/9.800 individuals and its prevalence is from 4 to 6/10.000 individuals without racial or ethnic preference. Thirty percent of the MS cases do not have an associated family history, meaning that they are resultant of isolated cases of new mutations[2-4]. The life expectancy of individuals with the condition is around 32 years and is primarily determined by the severity of the cardiovascular involvement. However, with the optimization of clinical treatments such as the use of beta blockers and elective surgery, the survival can increase up to 72 years, although patients require life long care from different health professionals including physical therapist[5].

According to Nosology of Ghent[6], given its clinical and phenotypic variability, the MS diagnosis is based on a clinical criteria that includes primordial manifestations in the cardiovascular, ocular, skeletal, neurological and other systems in addition to family history[7].

The genetic abnormalities present on the MS cause abnormalities in the connective tissue that is present in all systems. The connective tissue is responsible for the viscoelastic property of blood vessels, tension and elasticity of the skin and the matrix for bone calcification[2,3]. The alteration in fibrilin leads to flaccidity in the joint ligaments favouring joint hypermobility and consequently, reduction in the containment of bone growth compromising the musculoskeletal system[1]. The primary skeletal manifestations of the syndrome consist of increased stature, chest wall abnormalities and scoliosis with the later being present in approximately 62%[7] of patients and *pectus excavatum* and *pectus carinatum* present in 60% of patients[8]. Arachnodactyly, identified by long and slender fingers, and dolichostenomelia, characterized by disproportion between the relationship of the arm span and height, are also common characteristics of MS[4,7].

Considering that MS is a multisystem condition[9] with variation in the severity of phenotypes related to the disease and that the signs are age-dependent, MS patients need care of a specialized multidisciplinary team. Physical therapist are an indispensable professional of this multidisciplinary team working on the prevention and treatment of limitations associated with MS.

The increase of life expectancy in this population[5] associated with some risk factors predisposes individuals with MS to some important orthopedic manifestations. Thus, physical therapist are able to identify early disorders of the musculoskeletal system through functional kinetic evaluations and provide appropriate treatment and improved of quality of life of these patients. The present study aims to evaluate anthropometric carachteristics and musculoskeletal abnormalities in patients with MS and to analyze the frequency of physical therapy treatment in these patients.

Methods

The study was a cross-sectional observational design. Twenty six patients with a genetic-clinical diagnosis of MS based on the Ghent criteria[6] were evaluated. The research was carried out at the public multidisciplinary clinic of Marfan syndrome at the hospital of the Universidade Federal de São Paulo (UNIFESP), São Paulo, SP, Brazil from February 2006 to February 2008. All patients with MS who were aged between 6 and 19 years old and had radiography of the spine, chest and sternum of the last six months were include in the research. The study was approved by the Ethics in Research Committee of the UNIFESP, protocol nº 0381/05. An informed consent was signed by the parent or guardian of the children and adolescents before participation in the study.

Clinical evaluation

Patients answered to a standardized questionnaire with information related to personal and family history and demographic characteristics. After the interview a physical examination was conducted. The chest was evaluated in the sagittal and frontal plane in order to identify deformities of the anterior chest wall. Following the chest examination, the anthropometric measures were conducted and complementary exams analyzed.

Anthropometric evaluation

A conventional mechanical scale platform (Filizola®, São Paulo, SP, Brazil), with maximum capacity of 140 kg was used to measure body mass. Participants were asked to remove their shoes and remain with the least amount of clothes possible. Body mass was measured with patients standing at the center of the scale in orthostatic position with arms along the body. Height was measured using a stadiometer (model E120P, Tonelli, Siderópolis, SC, Brazil), with a field of use from 0.80 to 2.25 m. Arm span was assessed using a measuring tape of fiberglass and with a length of 200 cm and accuracy of 0.1 mm. This measure was conducted with patients in the orthostatic position, leaning against a wall with arms in abduction. The measurement was performed from the extremity of the middle

finger of one hand to another. A positive dolichostenomelia was determined as a value of arm span/height index of less or equal to 1.05[4,7] (Figure 1).

Body mass index (BMI) was used to diagnose the nutritional status of individuals [body mass (kg)/height (m^2)]. The thoracic expansion was calculated using a measuring tape on the patient's chest wall. The measures were performed in the axillary area and in the distal third of sternum (xiphoid area), with the patient in the orthostatic position and in maximum inspiration[10].

A flexible measuring tape was positioned in the smallest curvature located between the ribs and the iliac crest to measure the waist perimeter. To measure hip perimeter, the measuring tape was positioned on the greatest gluteal protuberance area. An index measure of the relationship between waist and hip circumference (RC/Q) was obtained by dividing the perimeters of the waist (cm) by hip (cm). A relationship between waist and hip circumference was considered inadequate if ≥0.95 for men and ≥0.80 for women[4,5].

The Steinberg thumb sign test was used to confirm the presence of arachnodactyly[1,3,11]. The test consisted of requesting the patient to perform an adduction of the thumb and flexion of fingers. The test was considered positive when the distal phalanx of the thumb surpassed the palmar area. The Walker-Murdoch wrist test was also used. In this test, patients were requested to hold the wrist with the contralateral hand. The test was considered positive when the little finger and thumb overlapped while taking hold of the wrist[1,11] (Figure 2).

Population anthropometric data

Anthropometric data and population body mass and height were obtained from published data by the Brazilian government, through the Ministério do Planejamento, Orçamento e Gestão (MPOG) and Instituto Brasileiro de Geografia e Estatística (IBGE)[12].

Complementary exams

Radiography of spine, chest and sternum

Panoramic spine x-rays from T1 to the sacrum were performed in lateral and postero-anterior positions. Patients that used spine orthoses were asked to remove it for a period of four to six hours before x-rays. Angulation measures of the scoliotic curve were performed between T1-T12 vertebrae using the Cobb method[13].

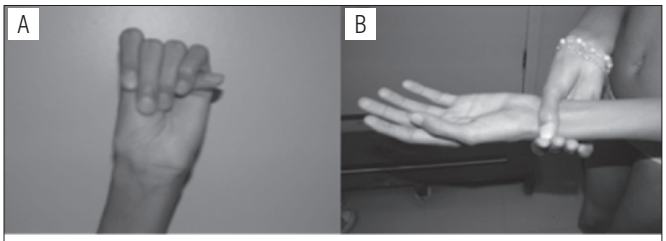
The anterior chest wall analysis was conducted in an objective way through the evaluation of the sternum on the lateral x-ray. Chest wall deformities were classified as: *pectus carinatum* - when a protrusion of sternum was observed and *pectus excavatum* - when a depression of sternum was observed[14,15].

Echocardiogram

Echocardiogram exams from the previous six months were analyzed. All echocardiograms were conducted at our hospital service, in uni and bidimensional modes, with the study of continuous wave Doppler, pulsed and color flow mapping, in an equipment with transducers of 5.0 and 2.5 MHz (*model SD800, Philips Healthcare, Andover, MA, United States*). Cardiac function was analyzed based on: 1) aortic diameter (mm), 2) left atrium (mm), 3) interventricular septum (mm), 4) left ventricular posterior wall (mm) and 5) left ventricular diastolic diameter (mm) in addition to research of valvular regurgitations[16].

Statistical analysis

Data was analyzed using the *GraphPadPrism* software (*GraphPad Software, Inc. CA, USA*). Data normality was evaluated using K-S distance test. Parametric continuous data were presented as mean and standard deviation and the non parametric as medians and minimum and maximum values. The anthropometric data obtained in this study were compared with the medians of the population published by IBGE using the *WMW test* (*Wilcoxon-Mann-Whitney*). Categorical data were presented as absolute frequency (n) and relative frequency (%) and compared between independent groups using a qui-squared test. For all analyses α was set as 5%.



A) Steinberg thumb signal test B) Walker-Murdoch wrist test.

Figure 1. Female patient, 12 years old.

Figure 2. Male, 19 years old with dolicostenomelia.

Results

Anthropometric and musculoskeletal characteristics

Twenty six patients with MS, aged 13.65±2.61yrs and 17 female, were evaluated. Anthropometric variables were divided according to gender as the differences between the studied variables were more accentuated in males (Table 1).

In comparison with the population data from IBGE[12] it was observed that children and adolescents with MS had higher values in relation to height and weight to those of the general population (Table 2).

Regarding orthopedic abnormalities, *pectus* and scoliosis were observed in 19 (73%) patients, *pectus* alone in 3 (11%) and no chest wall abnormalities in 4 (15%) patients. The mean degree of scoliosis was 20.76±9.83 analyzed by the Cobb's angle. Only one patient with greater severity of MS used orthoses during the evaluation which might have minimized the mean degree of scoliosis in the group. The *pectus* type most commonly presented was the *carinatum* in 16 (61%) patients. Dolichostenomelia was observed in 11 (42%) and arachnodactyly in 80% of patients. Questionnaires reveled that 11 (42%) patients had already received physical therapy for MS associated musculoskeletal disorders for a mean period of one year over the three preceding years.

Clinical characteristics

The presence of family history of Marfan syndrome was reported by the majority of patients n=15 (60%). The main medication used was beta blockers, in 15 patients (60%).

Cardiovascular and echocardiographic characteristics are presented on Table 3. The analysis revealed the presence of some echocardiographic alterations. Twelve (46%) patients had minimal mitral regurgitation, two (8%) had mild regurgitation and one (4%) had moderate regurgitation. Regarding the aortic diameter, only one (4%) patient presented with minimal ectasia and 8 (31%) with mild ectasia. In total only 2 (8%) patients had normal echocardiogram.

Discussion

The results of the present study demonstrated that patients with MS have anthropometric and musculoskeletal alterations. These characteristics may be detected by a physiotherapeutic evaluation which would allow an early detection, prevention and treatment.

The compromise of the connective tissue may lead to changes in the musculoskeletal system because the elastic and collagen fibers are responsible for the elasticity and resistance

Table 1. Anthropometric characteristics of patients with Marfan syndrome.

Anthropometric characteristics	Patients with MS (n=26)	
	Female (n=17)	Male (n=9)
	Mean±SD	Mean±SD
Age (years)	13.23±2.77	14.44±2.18
Body mass (Kg)	49.20±12.00	56.61±9.53
Height (m)	1.68±0.10	1.82±0.09
BMI (Kg/m2)	17.05±3.17	17.53±2.13
Arm span (m)	1.73±0.12	1.93±0.13
W/HR (cm)	0.75±0.13	0.85±0.4
Axillary perimetry (cm)	74.21±8.61	80.5±7.48
Xyphoid perimetry (cm)	68.71±6.40	78.77±3.59

MS=Marfan syndrome; BMI=body mass index; W/HR=waist to hip ratio; continuous parametric dada expressed as mean±standard deviation.

Table 3. Cardiovascular and echocardiographic variables of patients with Marfan syndrome.

Cardiovascular and echocardiographic characteristics	MS (n=26) Mean±SD
HR min (bpm)	57.7±7.76
SAP (mmHg)	103.8±12
DAP (mmHg)	70.38±11
SpO_2 (%)	97.42±1.5
Diameter of aorta (mm)	32.07±5.86
Diameter of left atrium (mm)	27.96±3.85
Diastolic diameter of left ventricle (mm)	44.7±6.02
Ejection fraction (%)	0.66±0.056

MS=Marfan syndrome; HR=heart rate; SAP=systolic arterial pressure; DAP=diastolic arterial pressures; SpO2=peripheral oxygen saturation; continuous dada expressed as mean±standard deviation.

Table 2. Anthropometric characteristics of patients with Marfan syndrome versus estimates from the Brazilian Institute of Geography and Statistics (Instituto Brasileiro de Geografia e Estatística) (IBGE, 2002-2003)[17].

Anthropometric characteristics	Subjects					
	Female (n=17)			Male (n=9)		
	MS	IBGE	p	MS	IBGE	p
	Median±Min-Max	Median±Min-Max		Median±Min-Max	Median±Min-Max	
Body mass (Kg)	51.5±24-68	41.46±30.7-54.4	0.001	61.0±42-72	46.96±32.4-58.5	0.0192
Height (m)	1.70±1.40-1.81	1.49±1.39-1.58	<0.0005	1.83±1.66-1.97	1.53±1.41-1.66	<0.0001

MS=Marfan syndrome; IBGE=Instituto Brasileiro de Geografia e Estatística; continuous dada expressed as median±min-max; WMW test (Wilcoxon-Mann-Whitney).

of ligaments[17,18]. Histopathological assessments have demonstrated fragmentation of fibrilin in muscular biopsy of patients with MS which would lead to the known musculoskeletal manifestations[19].

The present study demonstrated that patients with MS have increased stature when compared to those observed in the data from the Family Budgets Research of IBGE[12]. The lack of containment generated by ligament laxity favors an excessive growth and consequently decreased BMI to levels inferior to what is recommended by WHO. Some studies have already reported some limitations of BMI because it can not reflect the corporal composition appropriately, specially when considering the relationship between percentages of fat, muscles and bones and differences between gender, ethnicity and degree of regular activity[20,21].

The ratio between the measures of RC/Q, that characterize the central distribution of fat, have been used to identify risk to develop metabolic and cardiovascular complications[22]. Patients with MS have lower values than those recommended by WHO, which is certainly influenced by the phenotype of the syndrome[18,23].

Among the typical clinical signs of MS, arachnodactyly, identified by long and slender fingers, is a common characteristic[24]. Dolichostenomelia a common characteristic of MS, identified by elongated and thin extremities[25], was described for the first time in 1896. These characteristics were present in this study with arachnodactyly being extremely frequent and dolichostenomelia being not frequent. Dolichostenomelia may vary with age, during growth and development[4,7]. Other important clinical signs, such as ocular, cutaneous, pulmonary alterations, espondylolisthesis, lower elbow extension, *pes planus* and arched palate, were not evaluated in the present population, as specific equipments are required. However, the diagnosis was confirmed by genetic exam.

Regarding the chest wall alterations, one of the most common manifestations of MS is the *pectus*, characterized by a disproportion on the growth of cartilaginous plates of the sternum and the costochondral cartilages[14,15]. The presence of *pectus carinatum* was high in this population which may lead to the conclusion that the progression of *pectus carinatum* may occur during the growth phase and may become worse mainly in patients with genetic predisposition[26]. Respiratory disorders related to *pectus carinatum* will depend on the degree and on the association with other thoracic deformities such as kyphosis and scoliosis[26-28]. That's why it is important to identify early the musculoskeletal alterations in order to prevent future complications.

In this study, a high frequency of scoliosis was observed in patients with MS. Scoliosis seems to form as a consequence of the speed of growth peak that occurs earlier and for longer time in this population. This abnormal pattern of growth also leads to a disparate growth of limbs when compared to the general population also favoring the appearance of deformities on the spine[18].

Scoliosis diagnosis is extremely complex as patients don't commonly present with pain making the acceptance and adhesion to the treatment difficult. However, an early postural evaluation of patients with MS may identify the presence of this deformity before the symptoms appear. In the most severe cases of patients with moderate and severe scoliosis, cardiorespiratory complications by limitation of chest wall expansibility due to spine deviation may be observed. Cobb's angle is decisive for treatment indication. Patients with MS with no structured scoliosis show good evolution with conservative treatment, including physical therapy with or without the associated use of orthoses. Thus, the early diagnosis of scoliosis becomes important in patients with MS seeking to correct or to avoid scoliosis progression.

Physical therapy treatments were performed for patients who showed musculoskeletal disorders, however, because this is a multisystem disease, several areas of physical therapy may contribute to the prevention and treatment of the disorder. Some studies[17,30] report the need for physical therapy in the conservative treatment of some skeletal manifestations and they describe that the respiratory physical therapy, in the pre and postoperative of patient with MS contributes to the success in respiratory weaning, reducing respiratory disorders and consequently complications[30]. Furthermore, during adetailed physical and functional evaluation, physical therapist can identify clinical manifestations that are relevant to MS, contributing to a multiprofessional approach for the early diagnosis of MS and treatment of its complications.

Conclusion

Increased height and the presence of *pectus carinatum* and arachnodactyly are the anthropometric and musculoskeletal changes more frequently encountered in individuals with MS. Physical therapeutic treatments are not frequent.

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