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the high prevalence of alteration in atrioventricular–transvalvular flow, mainly with moderate severity, the electrocardiographic monitoring demonstrated that there were no serious arrhythmias or signs of myocardial ischemia in the MS group. This is the first report of such a finding and may suggest future modifications regarding the limitation criteria for physical activity in this population. The routine use of β -blockers in patients with MS is strongly recommended, especially for individuals with structural alterations in the aorta (31). This recommendation was heeded in the majority of subjects in the MS group, and there was a depressed chronotropic response to effort during the spirometric maneuver. Despite the greater prevalence of atrial arrhythmia, this population did not differ significantly from the CG on the SE, particularly with respect to the prevalence of atrial or VT, thereby suggesting the safety of this maneuver in patients with MS.

The moderate correlation between AS/H and LVMI and between AS/H and aortic diameter demonstrates an association between clinical staging, cardiovascular performance and aortic diameter, the former of which are functional characteristics and the latter is a biotype feature. However, the association between axillary to xiphoid perimeter ratio and FEV₁/FVC demonstrated an association between rib cage dysfunction and PF.

This study has limitations that should be addressed. One is related to the lack of a comprehensive assessment of PF, including plethysmography and broncodilatation, which could have furnished additional information regarding the impairment of the pulmonary parenchyma. However, despite the small number of patients in the subgroup analysis, the isolated comparison between individuals with no thoracic cage deformity seemed sufficient to infer the impairment of the pulmonary parenchyma. The lack of a height measurement from a sitting position did not allow for secondary comparisons of PF. However, considering the total population evaluated and the population restrictions imposed by other equations for predictive values, the decision was made to employ the equation proposed by Knudson et al. (26), as it is the broadest and most widely used in the international literature. Finally, despite the fact that the study was conducted on a young population, which may limit the frequency of signs of severe MS, the clinical characteristics such as height and body composition did not undergo any major changes, considering the medium age of the population. Furthermore, these findings may provide earlier and more detailed information on this population.

In conclusion, PF is reduced in patients with MS, and associated deformities in the thoracic cage contribute to this reduction in PF. Despite the evident structural alterations in the cardiovascular system in these patients, exertion during the SE appears to be safe in this population. On the basis of the results of this study, a future investigation is intended following up PF over time to determine whether there are further alterations with the aggravation of the disease. Moreover, further diagnostic studies on both younger and older patients could provide information that favors a complete understanding of the clinical staging, which would lead to improvements in the treatment of patients with MS.

Conflicts of interest

The authors have no conflicts of interest to disclose.

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Ethics approval

This study was approved by the Ethics Committee of the Universidade Federal de São Paulo (No. IRB-0381/05; <http://www.unifesp.br/reitoria/orgaos/comites/etica/>).

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Abstract

Background: Marfan syndrome (MS) is a dominant autosomal connective tissue disease that impacts multiple systems, such as the musculoskeletal, ocular, cardiovascular, pulmonary, tegumentary and neurological. This mutation may produce impairment in the connective tissue which causes modifications in the vascular viscoelastic properties, tegumentary elasticity, bone calcification matrix and pulmonary parenchyma. **Purpose:** To evaluate pulmonary function test (PFT) in patients with MS and relate it to clinical evaluation aspects, especially possible thoracic cage abnormalities (TCA), and the occurrence of cardiac arrhythmias during the spirometric exam (SE). **Method:** From a sample of 75 subjects, we evaluate 46 MS patients, 29 female and aged $20 \pm 0,51$ years, who underwent clinical, anthropometric, echocardiographic, radiographic and PF evaluation; 51 subjects (33 with MS) had their electrocardiography information evaluated during PFT. These individuals were matched and compared to a healthy control group (CG). **Results:** Forced vital capacity (FVC) and forced expiratory volume in the first second (FEV_1) in the patients with MS were significantly lower in comparison to the CG ($p=0.012$ and $p=0.0006$) and predicted values ($p=0.04$ and $p=0.003$). Subgroup analysis based on TCA revealed differences between patients with MS with two combined abnormalities (*pectus* + scoliosis) in comparison to both the CG ($p=0.012$ and $p=0.002$) and patients without abnormalities ($p=0.05$ and $p=0.006$). Some aspects from clinical evaluation (Arm span to height ratio, m and axillary to xiphoid perimetry ratio, cm), cardiovascular behavior (Left ventricular mass Index-LVMI, g/m^2 and Aortic Diameter- Ao , mm) and PF ($FEV_1/FVC\%$) has demonstrated a moderate correlation. There were no differences regarding the occurrence of arrhythmia during exertion on the SE. **Conclusion:** PF is reduced in patients with MS, and deformities in the thoracic cage appear to contribute to this reduction. Some aspects clinical, cardiovascular and PF are associated in MS. Despite the apparent structural alterations in the cardiovascular system in this young population, exertion during the spirometric exam appears to be safe in relation to electrocardiography modifications.

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Tabela 6- Comparação das características da Função Pulmonar nos grupos: Controle, SM e Subgrupos de pacientes com Síndrome de Marfan

Grupos	n	(%)	CVF (L)			VEF1 (L)			VEF1/CVF (%)		
			Observado Média ± DP	Predito Média ± DP	% Predito Média ± DP	Observado Média ± DP	Predito Média ± DP	% Predito Média ± DP	Observado Média ± DP	Predito Média ± DP	% Predito Média ± DP
Controle	29		4,5 ± 0,89	4,47 ± 0,88	101,6 ± 11,38	3,9 ± 0,7	3,92 ± 0,65	99,72 ± 10,74	86,98 ± 5,6	87,08 ± 2,24	99,93 ± 6,66
SM	46		3,85 ± 1,19 *	4,36 ± 1,11 **	88,39 ± 16,79 *	3,19 ± 0,9 *	3,78 ± 0,97 **	85,18 ± 15,96 *	84,02 ± 7,87	86,52 ± 1,94 **	97,08 ± 9,09
Caixa Torácica											
Sem alterações	9	19,57	3,81 ± 0,98 ***	3,81 ± 0,7	98,85 ± 14,78	3,11 ± 0,69 ***	3,33 ± 0,61	93,47 ± 12,75	83,04 ± 7,06	87,23 ± 2,09	95,16 ± 7,25
Com Pectus	8	17,39	4,14 ± 1,21	4,92 ± 1,38	85,02 ± 15,14 ***	3,36 ± 0,79	4,2 ± 1,13	82,2 ± 17,22 ***	82,45 ± 12,36	85,6 ± 1,99	96,14 ± 13
Com Pectus + Escoliose	29	63,04	3,78 ± 1,26 ***	4,38 ± 1,11	86,07 ± 17 ***	3,17 ± 1,02 ***	3,81 ± 0,98 **	83,42 ± 16,15 ***	84,76 ± 6,74	86,54 ± 1,85	97,92 ± 7,34

Legenda: CVF= Capacidade vital forçada, VEF₁= Volume expiratório forçado no primeiro segundo e VEF₁/CVF= Relação do volume expiratório forçado no primeiro segundo pela capacidade vital forçada. Dados categóricos representados em Número (N^o) de pacientes (% do total). Dados paramétricos contínuos representados em Média ± Desvio padrão; *Teste t não pareado*. p≤0,05. * SM vs Grupo controle; ** Valores observados de SM vs valores previstos *** Subgrupos de SM vs Grupo controle.