Functional assessment of scleroderma patients

ABSTRACT

It is known that functional capacity of patients with rheumatic diseases is reduced. However, there are few studies that evaluate functional capacity in patients with scleroderma, a rare disease with vascular, skeletal muscle and visceral manifestations. **Objective:** To evaluate functional capacity of scleroderma female patients. **Method:** Thirteen patients without pulmonary impairment and 13 controls, all sedentary and paired by age and body mass index (BMI), undertook a treadmill exercise test. **Results:** Oxygen consumption (VO₂ max) of patients was statistically lower than control group (p=0.0395), as well as the percentage of predicted VO₂ max (p=0.0383) and maximal attained exercise intensity (p=0.0395). Time between ventilatory thresholds was also lower in scleroderma group (p=0.0271). **Conclusion:** Scleroderma patients, even without pulmonary disease, present a lower ability for a moderate to intense physical effort when compared with sedentary women of the same age and BMI.

INTRODUCTION

Scleroderma is a rare disease, which incidence is higher in women than in men, ranging from 3:1 to 8:1. Some studies show an incidence of 19 cases per million per year, and a prevalence of 19 to 75 cases per year per a hundred thousand inhabitants (KLIPPEL, et al., 1997; SILMAN, et al., 1996; SAMARA, 2004). The disease is characterized by skin thickening, Raynaud’s phenomenon and other vascular abnormalities, as well as skeletal muscle manifestations and visceral involvement, mainly in the gastrointestinal tract, lungs, heart and liver. (SUBCOMMITTEE FOR SCLERODERMA CRITERIA OF THE AMERICAN RHEUMATISM ASSOCIATION DIAGNOSTIC AND THERAPEUTIC CRITERIA COMMITTEE, 1980).

One of the initial symptoms of scleroderma are non-specific skeletal muscle complaints, like arthralgias and mialgias (KLIPPEL, 1997). The most common problem as disease advances is muscular atrophy and weakness
Disuse and decondition caused by fibrotic skin contractures and malnutrition may be the main causes of these symptoms, but muscular fibrosis associated to serum creatine kinasis elevation can also occur (KLIPPEL, 1997).

There are few studies that evaluate the functional capacity of scleroderma patients. Non-invasive exercise assessment with gas analysis and cardiac monitoring allows a detailed examination of exercise impairment that correlates well with invasive assessment. The measurements it provides serve as objective, reproducible indices of exercise capacity that can be applied to the management of various clinical problems (NEUBERG et al., 1988).

Some scleroderma patients are not able to complete a maximal test, interrupting it because of fatigue or weakness. Anaerobic threshold and functional capacity are reduced compared to predicted values for healthy individuals (SUDDUTH et al., 1993; BLOM-BÜLOW et al., 1983; MORELLI et al., 2000).

Scleroderma is a chronic disease with a great impact in patient's quality of life. In studies published until this moment, the maximal effort test has been conducted in a bicycle ergometer, which does not use to be an activity present in the daily routine of the patients. Therefore, our objective was to evaluate functional capacity using the treadmill. This assessment will contribute to a better understanding about the functional capacity of these patients, and allow prescription of safe and efficient exercise programs.

**PATIENTS AND METHODS**

This study has been approved by University of Sao Paulo Medical School Ethics Committee, protocol number 813/03.

For this study, 13 scleroderma outpatients from the Rheumatology Service of Clinicas Hospital from University of Sao Paulo Medical School were recruited (Scleroderma Group). Thirteen employees from the same hospital also took part of the study (Control Group).

Scleroderma Group (SG) had 13 female patients with diagnostic of Scleroderma (SUBCOMMITTEE FOR SCLERODERMA CRITERIA OF THE
AMERICAN RHEUMATISM ASSOCIATION DIAGNOSTIC AND THERAPEUTIC CRITERIA COMMITTEE (1980), ages above 20 years old, sedentary for at least 6 months, without pulmonary involvement (forced vital capacity (FVC) higher than 75%, diffusion lung capacity of oxygen (DLCO) higher than 75%, pulmonary artery systolic pressure (PASP) lower than 40mmHg, without the following symptoms: cough, dispnea and thoracic pain, without renal insufficiency, badly-absorption syndrome and malnutrition, uncontrolled hypertension and arrhythmia, without diagnostic of fibromialgia and anemia (hemoglobin lower or equal to 11 mg/dl).

Thirteen women, employees from Clinicas Hospital from University of São Paulo Medical School, were invited to join CG, their ages were above 20 years old, sedentary for at least 6 months, and without diagnostic of any chronic disease or chronic use of any kind of medicine.

Both groups were paired by age and body mass index (BMI).

All patients from SG were evaluated at zero moment, before functional assessment. Pre-test evaluation included pulmonary function testing (to determine forced vital capacity (FVC) and diffusion lung capacity of oxygen (DLCO)); echocardiogram (to determine pulmonary artery systolic pressure (PASP)); hemogram (to determine hemoglobin concentration (Hb)); consult to each patient's registers to check for possible exclusion factors, disease type (diffuse or limited) and disease duration; medical appointment at the Rheumatology Service from Clinicas Hospital from University of Sao Paulo Medical School, where clinical assessment was performed and also questioned for the level of physical activity of each patient.

We excluded patients with FVC or DLCO lower than 75% predicted, PASP higher or equal to 40mmHg, Hb lower or equal to 11mg/dl, patients with symptoms like cough, dispnea and thoracic pain, uncontrolled hypertension or arrhythmia, and also fibromialgic and malnourished patients.

CG was also evaluated at zero moment. They had a clinical evaluation at the Rheumatology Service from Clinicas Hospital from University of São Paulo Medical School, where the presence of any possible exclusion factors was verified and also questioned for the level of physical activity of each participant.
Both groups performed a maximal ergospirometric test in treadmill (Inbramed Millenium, RS, Brazil), following BRUCE (1973) protocol, with direct determination of the maximal oxygen consumption (VO₂ max) and maximal heart rate checking. Metabolic data were analyzed by metabolic analysis system Aerosport-teem100 (Ann Arbor, USA). The software we used for this procedure was Micromed ErgoPC Elite 3.2 (Brásilia, Brazil). The chosen protocol has fixed increments in speed and incline at every three minutes. Anaerobic threshold and respiratory compensation point were determined by the method of ventilatory equivalents.

We also analyzed the percentage of predicted VO₂ max, metabolic equivalent of oxygen consumption (MET) at maximal effort and time between ventilatory thresholds.

Statistical analysis was performed with a software package (Instat™ v.2, GraphPad Software, San Diego, CA, USA). Comparisons between groups were performed by unpaired Student's T test. P values less than or equal to 0.05 were accepted as significant.

RESULTS

Both groups were similar regarding age, BMI and race, as shown in table 1. The patients presented a mean of 8.46 years of disease and the limited form was predominant.

<table>
<thead>
<tr>
<th><em>Table 1</em> - Demographic data</th>
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<tbody>
<tr>
<td><strong>Scleroderma Group</strong></td>
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<tr>
<td>(n=13)</td>
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<tr>
<td>mean ± SD</td>
</tr>
<tr>
<td>Age (years)</td>
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<tr>
<td>BMI (kg/m²)</td>
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<tr>
<td>Race</td>
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<td>Disease duration (years)</td>
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<td>Disease form</td>
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VO₂ max of SG (mean 19.82 ml/kg/min) was significantly lower (p=0.0395) than the one achieved by CG (mean 23.72 ml/kg/min). The same result was observed with statistical significance (p=0.0383) about the attained percentage of predicted VO₂, SG reached 53.69%, and CG 61.79% (table 2).

Statistical analysis did not reveal a significant difference when we compared VO₂ of SG and CG in both ventilatory thresholds (p=0.6021 for the anaerobic threshold and p=0.3387 for the respiratory compensation point). However, when we compared the time of testing between those thresholds, SG presented a lower time interval between anaerobic threshold and respiratory compensation point than CG (p=0.0271) (table 2).

Regarding MET, CG reached a significantly higher effort than SG (p=0.0395), and kept the test for a longer time space (p=0.0203) (table 2).

<table>
<thead>
<tr>
<th>Table 2 - Results</th>
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<tbody>
<tr>
<td><strong>Scleroderma Group</strong></td>
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<tr>
<td>Mean</td>
</tr>
<tr>
<td>HR max (bpm)</td>
</tr>
<tr>
<td>VO₂ max (ml/kg/min) *</td>
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<td>VO₂ AT (ml/kg/min)</td>
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<td>VO₂ RCP (ml/kg/min)</td>
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<td>% predicted VO₂ max *</td>
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<td>MET *</td>
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<td>Time between thresholds (s)</td>
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*statistically significant difference (p ≤ 0.05)

**DISCUSSION**

To our knowledge, this is the first study that used the treadmill to evaluate the functional capacity of scleroderma patients.

Although our patients did not present any pulmonary impairment, even though, the results indicated that patients present a reduced functional capacity compared to healthy controls.

It is known that heavy housework, such as scrubbing floors and lifting or moving heavy furniture is an activity that requires between 4 and 10 METs.
Strenuous sports, such as swimming, singles tennis, football, basketball and skiing are activities that usually demand more than 10 METs (EAGLE et al., 2002). Our study revealed that SG is able to perform a 5.66 MET task, and CG a 6.77 MET task. Therefore, both groups are impaired in some degree to develop some physical activities, but SG, according to our initial hypothesis, would be more impaired than CG.

There are evidences in the literature that some degree of pulmonary involvement may contribute to exercise intolerance (SUDDUTH et. al, 1993; MORELLI et al., 2000). We speculate that there can be other factors involved in this mechanism, such as peripheral deconditioning or circulatory impairment. On the other hand, the lack of pulmonary involvement associated with exercise intolerance could be a predictor of a future or a silent pulmonary disease.

It is worthy to note that, in order to avoid confounding variables, we excluded the possible interference of age and body mass in functional capacity by pairing both groups, and also compared sedentary patients with sedentary controls, but even in these conditions SG had a significantly lower VO\textsubscript{2} max.

Our results confirm the previous ones that used a bicycle ergometer and compared scleroderma patients with predicted values. They and also found a VO\textsubscript{2} max significantly lower than expected for healthy individuals (SUDDUTH et. al, 1993; BLOM-BÜLOW et. al.,1983; MORELLI et al., 2000).

Previous studies regarding functional assessment in scleroderma patients used a bicycle ergometer (SUDDUTH et al., 1993; BLOM-BÜLOW et al., 1983) and found a VO\textsubscript{2} lower than 40% predicted and a mean VO\textsubscript{2} of 51% predicted, respectively. Our study's patients presented a mean VO\textsubscript{2} of 53.69% predicted for healthy individuals, which is above the values found by both previous studies.

Research comparing effort tests in treadmill and cycle ergometer conclude that the evaluated maximal oxygen uptake tends to be 10 to 20% higher in treadmill (BUCHFUHRER et al., 1983; MYERS et al., 1991; HAMBRECHT, et al., 1992). This fact invalidates direct comparison of our results with the ones found by previous studies, once all of them used a cycle ergometer.
Some authors believe that exercise intolerance in scleroderma patients is mainly related to circulatory mechanisms, but sedentarism, that is a cause of peripheral fatigue, may also be responsible for that in some degree (MORELLI et al., 2000, SUDDUTH et al., 1993). However, other studies considered that the lack of physical conditioning, including diminished articular and skin mobility could also contribute to the exercise limitation presented by patients (SUDDUTH et al., 1993).

Besides the differences in VO\textsubscript{2} in the ventilatory thresholds of both SG and CG were not statistically significant, the significant lower time between AT and RCP in SG indicate that SG has a certain degree of exercise impairment, related to the aerobic metabolism. Unfortunately, it was not possible to determine if this impairment is due to disuse and decondition or to the vascular complications caused by scleroderma.

The patients of this study have a mean age lower than the general incidence of scleroderma, which occurs in the 5th and 6th decades of life (KLIPPEL, et. al., 1997). Evidences show that VO\textsubscript{2} declines 10% per decade in men and women regardless of activity level (HAWKINS & WISWELL, 2003), and a low cardio respiratory fitness challenges quality of life (BLAIR et al., 1996). Hence, a low VO\textsubscript{2} at this age may represent an even lower VO\textsubscript{2} in the future, and that can prevent patients from their work, recreational and daily living activities.

In conclusion, the results of this study suggest that scleroderma patients present a lower functional capacity compared to healthy sedentary controls. Thus, exercise testing with respiratory gas exchange must be used to prescribe safe physical exercise programs in order to increase cardio respiratory fitness in this population.

REFERENCES


