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FASCIA SCIENCE AND CLINICAL APPLICATIONS: CROSS-SECTIONAL STUDY

Systemic sclerosis: Association between physical function, handgrip strength and pulmonary function

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ABSTRACT

Background: Systemic sclerosis (SSc) is a multisystem disease affecting the skin, respiratory system and skeletal muscles. In SSc patients, hand function disability is the major factor limiting daily activities.
Aim: To evaluate the association of physical function with handgrip strength and pulmonary function in SSc patients. A further aim was to assess the relationship between handgrip strength and pulmonary function in patients with SSc.
Method: A cross-sectional study in which 28 patients with SSc underwent isometric handgrip strength (IHGS) measurement and pulmonary function tests and completed the Health Assessment Questionnaire Disability Index (HAQ-DI) to measure physical function.
Results: The HAQ-DI scores were associated with the IHGS ($r_s = -0.599$, $P = 0.001$) and pulmonary function parameters, particularly the diffusion capacity for carbon monoxide (DLco; $r_s = -0.642$, $P = 0.0004$).
Conclusion: In patients with SSc, the degree of physical disability is associated with both hand grip strength and pulmonary function. However, there is no relationship between handgrip strength and pulmonary function in these patients.

1. Introduction

Systemic sclerosis (SSc) is an autoimmune and chronic connective tissue disease characterized by vascular dysfunction and progressive fibrosis of the skin and internal organs caused by an excess of collagen fibres (van den Hoogen et al., 2013). The incidence rates of the disease range from 0.6 to 122 cases per million people per year, and the prevalence rates range from 4 to 489 cases per million people; however, these numbers are likely underestimated (Chifflot et al., 2008). Although skin involvement is the most common manifestation of SSc, other systemic impairments are commonly observed, including cardiovascular, musculoskeletal, renal, pulmonary and gastrointestinal abnormalities.

Effective measurement of quality of life is essential for describing the disease's impacts and progress and the effects of therapy on patients' health (Bruce and Fries, 2003). Therefore, quality of life assessment instruments must be comprehensive, reliable, valid and responsive. In this context, the Health Assessment Questionnaire Disability Index (HAQ-DI) has shown good validity, reliability and sensitivity to changes in the overall health status of patients with SSc (Racine et al., 2016). In an evaluation of 205 patients with early diffuse SSc, Sultan et al. (2004) noted that an HAQ-DI score of less than 1.3 was predictive of at least a 20% improvement in clinical parameters (including health status and skin lesions) after one to two years of follow-up. In SSc, several factors can contribute to a worse quality of life and reduced physical capacity; muscle weakness and pulmonary dysfunction are among the factors with the greatest impact (Morrisroe et al., 2015; Paik et al., 2016).

Depending on the assessment method used, muscle weakness is found in up to 90% of patients with SSc and is associated with significant disability and important psychosocial and economic burdens (Ranque et al., 2007). A study showed that high serum creatine kinase (CK) in SSc increases mortality, especially for patients with diagnosed early diffuse disease, reduced forced vital capacity (FVC) and high HAQ-DI scores (Jung et al., 2014). Another study showed an association between the Medsger muscle severity

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score, which is a subjective assessment scale, and the HAQ-DI score in patients with muscle weakness caused by SSc (Paik et al., 2016). Studies using objective assessment tools, such as the one by Azevedo et al. (2009), also found muscle weakness in patients with SSc. Using isokinetic dynamometry, these authors noted that 100% of the patients in their study had low average peak torque values for the elbow flexor and extensor muscles.

A simple, inexpensive tool for assessing muscle strength is the measurement of handgrip strength, which has been defined as the sum of the strength of the flexor muscles against the palm of the hand (Mathiowetz et al., 1985). Because of its low cost, ease of use and portability, handgrip strength has been used to assess severity and therapeutic response for certain clinical conditions and to reflect the overall health status and physical activity level (Leong et al., 2015; Lima et al., 2016). In SSc, handgrip strength may be impaired due to several factors, such as musculoskeletal dysfunction, arthralgia and arthritis, muscle contractures in flexion, and rigidity of the hands and fists (Randone et al., 2008; Lima et al., 2015; Morrisroe et al., 2015). In SSc patients, hand function disability is the major factor limiting daily activities (Guillaume-Jugnot et al., 2016).

Another very common manifestation in SSc is pulmonary involvement, which occurs in more than 80% of patients and is considered the second most common visceral complication in this disease (Ferri et al., 2002). Interstitial lung disease and pulmonary arterial hypertension are the most common forms of pulmonary involvement. SSc patients with pulmonary disease present a worse prognosis than those without pulmonary involvement. In fact, pulmonary disease is currently the most common cause of death in SSc patients and has recently surpassed kidney disease as the main cause of mortality. The severity of the pulmonary involvement related to SSc is frequently quantified in routine clinical practice using pulmonary function tests (PFTs) (Antoniou et al., 2016).

In SSc, the evaluation of handgrip strength is extremely relevant because its impairment is closely related to morbidity. We hypothesized that both muscle dysfunction of the hands and fists and pulmonary dysfunction, two of the most common abnormalities in SSc, have negative impacts on the physical capacity of these patients. Thus, the objective of the present study was to assess the association of physical function (determined with the HAQ-DI) with handgrip strength and pulmonary function in SSc patients. A further aim was to assess the relationship between handgrip strength and pulmonary function in this patient population.

2. Methods

2.1. Patients

Between October 2015 and August 2016, a cross-sectional study was performed with 37 SSc patients who were followed at Pedro Ernesto University Hospital of the State University of Rio de Janeiro, in the city of Rio de Janeiro, Brazil. The disease diagnosis was based on clinical characteristics and was confirmed using the American College of Rheumatology/European League Against Rheumatism recommendations (van den Hoogen et al., 2013). Patients of both genders aged ≥18 years were included in the study. The exclusion criteria were a diagnosis of SSc with other collagen diseases (except Sjogren's syndrome), confirmed by a rheumatologist; a history of smoking ≥ 10 pack-years; a history of a previous pulmonary disease not related to SSc (including asthma and tuberculosis); an inability to perform the PFTs; and respiratory infection within the last month before recruitment. The protocol followed the Helsinki Declaration principles and was approved by the Research Ethics Committee of the Augusto Motta University Centre under number CAAE-[47112015.0.0000] 5235. All patients signed an informed consent form prior to their enrolment in the study.

2.2. Measurements

The HAQ-DI was used to assess the patients' physical function levels. This questionnaire includes questions related to the fine movements of the upper extremities and the motor activities of the lower limbs (Bruce and Fries, 2003). The assessment consists of 20 questions divided into eight categories that represent a comprehensive set of functional activities performed over the past seven days, including the ability to dress, stand, eat, walk, perform personal hygiene, reach or grab something and perform usual activities. Responses are given on a scale from zero (no disability) to three (complete handicap). Each category contains at least two questions addressing a specific component.

Isometric handgrip strength (IHGS) was measured with a maximum isometric test of the upper dominant limb using SH5001 equipment (Saehan Corporation, Korea). The participants were positioned according to the American Society of Hand Therapists recommendations (Crosby et al., 1994): seated in upright position with the knee angle of flexion at 90°, the shoulder positioned in neutral adduction and rotation, and the elbow flexed at 90°, with the forearm in half pronation and the fist in a neutral position. The arm was kept suspended in the air with the hand positioned on the dynamometer, which was supported by the evaluator. A 60-s rest time was established between tests, and the highest value of three tests was used for the analysis.

The PFTs included spirometry, static lung volume measurements using whole-body plethysmography, assessment of the diffusion capacity for carbon monoxide (DLco), and measurement of respiratory muscle strength. All these tests were conducted using HDpft 3000 equipment (nSpire Health, Inc., Longmont, CO, USA) and were standardized and interpreted as recommended by the American Thoracic Society (Miller et al., 2005). National equations were used to calculate the predicted values for each participant (Neder et al., 1999a, 1999b, 1999c; Pereira et al., 2007).

2.3. Data analysis

The data analysis was conducted using SAS 6.11 software (SAS Institute, Inc., Cary, NC, USA). When the data distribution was not Gaussian (Shapiro-Wilk test), the results were expressed as medians and interquartile ranges or frequencies (percentages). The Spearman correlation coefficient (r_s) was used to evaluate the associations between the physical function, handgrip strength and pulmonary function variables. We considered a correlation coefficient ≤0.29 weak; those between 0.30 and 0.49 were considered moderate, and those ≥0.50 were considered strong (Cohen et al., 2003). Statistical significance was set at $P < 0.05$.

3. Results

Among the 37 patients who were initially recruited, nine were excluded for the following reasons: four for having SSc plus other collagen diseases, three for an inability to perform the PFTs and two for respiratory infection in the four weeks before recruitment. Thus, the sample included 26 women and two men with a median age of 51.2 (44.2–60.2) years. Eighteen subjects had the limited form of the disease, and ten had the diffuse form of the disease. The mean duration of the disease since the appearance of the first symptom, excluding Raynaud's phenomenon, to the date of inclusion in the study was 14 (6.50–19) years. Six subjects were ex-smokers who smoked less than ten pack-years. Diabetes and hypertension were reported by three and eight individuals, respectively.

HAQ-DI scores ≥1.0 were observed for 46.4% of the patients.