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CROSS-SECTIONAL STUDY

Relationship between functional capacity, joint mobility and pulmonary function in patients with systemic sclerosis

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Summary *Background:* In systemic sclerosis (SS), pulmonary involvement is currently the leading cause of mortality. Joint impairments limit the range of motion (ROM), which may reduce the functional capacity of these patients.
Aim: To assess the correlation between the functional capacity, joints mobility, and pulmonary function parameters in adults with SS.
Method: This was a cross-sectional study including ten SS patients who underwent goniometry, spirometry, carbon monoxide diffusing capacity (DLco) assessment, and the 6-min walk distance (6 MWD).
Results: Significant correlations were found between the 6 MWD and the tibiotarsal plantarflexion ROM ($r = 0.65$; $P < 0.01$), tibiotarsal dorsiflexion ROM ($r = 0.64$; $P < 0.01$), and hip adduction ROM (0.52; $P < 0.05$). Significant correlation was also observed between the 6 MWD and DLco ($r = 0.61$; $P < 0.01$).
Conclusions: Although the 6 MWD can be influenced by cardiovascular and pulmonary impairments in SS, our results suggest that the musculoskeletal dysfunction play an important role in the functional capacity of these patients.

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Introduction

Systemic sclerosis (SS), a generalized form of scleroderma, is a chronic inflammatory disease of the connective tissue characterized by extensive fibrosis and abnormalities of small vessels and microvasculature (Chizzolini et al., 2011). It is a rare disease with a prevalence ranging between 7 and 489 cases per million (Chifflot et al., 2008). It is predominant among females (3—8:1), with no predilection for race, and usually starts between the third and sixth decades of life (Barnabe et al., 2012). Although cutaneous involvement is the most evident, the disease can strike multiple organs and systems, such as the lungs, the musculoskeletal system, the kidneys, the heart, and the gastrointestinal tract, which can worsen the prognosis (Tani et al., 2013).

Currently, pulmonary involvement is the leading cause of death in SS. The lungs are affected in 70%—90% of patients, with interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH) being the most frequent manifestations and causing major clinical repercussions (Coaccioli et al., 2009; Gohari Moghadam et al., 2011). In pulmonary function tests, the first observed finding is usually a reduction in the diffusing capacity for carbon monoxide (DLco). This index is currently regarded as the main predictor of pulmonary deterioration in patients with SS (Lopes et al., 2011). Another feature is the reduction of forced vital capacity (FVC), which occurs in 40%—75% of patients (Solomon et al., 2013).

Joint involvement is observed in 46%—97% of subjects with SS (Baron et al., 1982; Avouac et al., 2012). It may cause joint pain, tenosynovitis, and polyarthritis, resulting in fibrosis around the tendons and other periarticular structures (Avouac et al., 2012). As the disease progresses, joint contractures may arise because of cutaneous thickening, shortening of the tendon, and intra-articular changes (Randone et al., 2008; Avouac et al., 2010, 2012). The involvement of the finger joints is common, along with compromises of the large joints such as the elbows, knees, and ankles (Randone et al., 2008; Iagnocco et al., 2012). Goniometry is a simple and widely used objective measure of joint range of motion (ROM) (Sabari et al., 1998; Sacco et al., 2007). Despite its low cost and high intra- and inter-rater reliability (Rothstein et al., 1983; Brosseau et al., 1997, 2001), goniometry has not previously been used in SS.

Because it is a multisystemic disease, several factors can potentially limit exercise in patients with SS. Previous studies have described abnormalities in gas exchange at rest and during exercise in patients with SS (Schwaiblmair et al., 1996). More recently, Dumitrescu et al. (2010), using the cardiopulmonary exercise test, observed that pulmonary vasculopathy and left ventricular dysfunction limit exercise capacity in these patients from an early stage. Another simpler and more widely used test to evaluate functional capacity is the 6-min walk test (6 MWT). In patients with SS, some studies have shown the importance of the 6 MWT in assessing the severity of the interstitial and pulmonary vasculature involvement (Villalba et al., 2007; Coaccioli et al., 2009; Deuschle et al., 2011), and the impact of the left ventricular diastolic dysfunction (Akdogan et al., 2011). However, since several investigators have found weak or moderate correlations between cardiopulmonary parameters and the 6 MWT, it is speculated that other disorders may also limit the 6-min walk distance (6 MWD) in patients with SS (Garin et al., 2009; Schoindre et al., 2009; Holland and Goh, 2012). Because of its multisystemic nature, the effects of other systems (including the osteoarticular system) on the 6 MWT results of patients with SS remain unclear (de Oliveira et al., 2007; Garin et al., 2009).

In SS, joint deformities lead to limitation of movement and consequent restriction of mobility, which potentially reduces functional capacity and directly affects the quality of life of these patients (Nguyen et al., 2011). As this has not been objectively demonstrated, we hypothesized that the joint impairments influence the ROM and influences the 6 MWT performance in patients with SS. Thus, our primary objective was to correlate joint ROM with functional capacity in these patients and, secondarily, to evaluate the association between pulmonary function variables and functional capacity.

Methods

Patients

A cross-sectional study was conducted between April and August 2013. Eighteen consecutive SS patients who presented to the Department of Rheumatology in the Federal Hospital of Bonsucesso in the city of Rio de Janeiro, Brazil, were evaluated. Adult patients (18 years of age or older) who had a diagnosis of SS by a rheumatologist according to the American College of Rheumatology/European League Against Rheumatism criteria (van den Hoogen et al., 2013) were included (Table 1). The following exclusion criteria were applied: respiratory infection in the four weeks preceding the study, peripheral oxygen saturation (SpO_2) at rest $<$ 90%; history of orthopedic surgery on the trunk or lower limbs; and difficulty walking. Individuals took part after being briefed on the objective of the study and giving their prior consent, in accordance with local ethical guidelines and the Declaration of Helsinki (1964). The protocol was approved by the Research Ethics Committee of the Augusto Motta University Center (Number 300.826/2013).

Measurements

ROM was always measured by the same examiner using a universal goniometer (Carci, Inc., São Paulo, Brazil). The participants were asked to wear suitable attire so that clothing did not affect the measurement results. The anatomical points were marked with self-adhesive labels (Fig. 1). The goniometer's axis was placed parallel to the joint axis, leaving the fixed arm of the apparatus on the proximal portion of the joint and the movable arm on the distal part of the joint to monitor joint dislocation from the beginning to end of the movement. The value on the protractor was recorded as the ROM of the involved joint. In this study, the ROM angles of the following joints were measured (Marques, 2003; Sacco et al., 2007; Santos et al., 2011): hip joint (flexion and extension) = an angle that includes the vertex of the greater trochanter of the femur