

Rehabilitation in Connective Tissue Diseases

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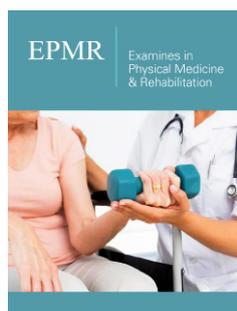
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Abstract

Connective Tissue Disease (CTD) is a heterogeneous group of chronic inflammatory conditions characterized by involvement of conjunctive tissue and a common autoimmune signature. These diseases include systemic lupus erythematosus, polymyositis, dermatomyositis, systemic sclerosis, Sjogren's syndrome, mixed CTDs, and undifferentiated CTDs. Cutaneous, articular, muscular, pulmonary and cardiovascular manifestations are among the most frequent during these diseases and often lead to a functional and physical disability that sometimes severely affects the quality of life of these patients. Rehabilitation and physical therapy play a crucial role in the management of patients with CTDs but despite their proven benefits, they are often neglected and represent a challenge for clinicians. This overview focuses on the rehabilitation of cutaneous, musculoskeletal and cardio-pulmonary involvement of CTDs.

Keywords: Connective tissue disease; Rehabilitation; Physical therapy; Physiotherapy

Introduction

Connective Tissue Disorders (CTDs) are defined as a group of acquired diseases resulting from persistent immune mediated inflammation [1]. The immune dysregulation results in autoreactive T Cells or autoantibodies generation, which can attack any organ of the body leading to a wide range of signs and symptoms [2]. The classic autoimmune CTDs include Systemic Lupus Erythematosus (SLE), Juvenile Dermatomyositis/Polymyositis (JDM/PM), Systemic Sclerosis (SSc), Sjogren's Syndrome (SS), Undifferentiated CTD (UCTD) and overlap syndromes e.g. mixed CTD (MCTD) [3,4]. They typically present insidiously, and many patients experience years of general malaise, pain and fatigue before a diagnosis is made and treatment begins [1-4]. Although early diagnosis and treatment is essential in reducing morbidity and mortality, cutaneous signs, joint pain, muscle weakness and cardiopulmonary signs, which represent the most common presenting features of the disease, may compromise functional outcome and represent a source of disability [2,4].

Because of the chronic nature of these diseases and the frequency of relapses, the drugs are most often insufficient to maintain a complete remission [5]. Cutaneous-tendon, joint, pulmonary and cardiovascular complications are often the cause of significant motor and functional impairment during the course of CTD [3,5,6]. Thus the place of physical therapy and rehabilitation is crucial in the management of patients with these diseases [5-8]. Despite its importance and its proven benefits, rehabilitation is often neglected in the management of CTDs [5], and because of the specificities of these diseases, it still represents a challenge for clinicians [8]. This manuscript focuses on the management of cutaneous, musculoskeletal and cardio-pulmonary symptoms and complications of CTDs. Other forms of supportive care and systemic therapy will not be covered by this review. Manifestations of CTDs and their rehabilitation-based treatment are divided by organ system.

Skin

Cutaneous involvement is the most common manifestation of CTDs, affecting about 73% of patients [9,10], can lead to a great deal of functional impairment, and may have a significant impact on the quality of life [10-12]. It includes thickening of the skin, hidebound skin, hyperpigmentation, retractions due to skin thickening, Raynaud's phenomenon, skin rash (malar rash, Heliotrope rash, nodules), photosensitivity, and alopecia [9,10,13,14]. When the involvement of the skin and/or fascia surrounds joints, it can cause decreased range of motion, significantly restricting a patient's ability to perform activities of daily living and contributing to prolonged periods of immobility [10-12].

Rehabilitation management includes connective tissue massage which is a manual technique used to treat altered connective tissues, in order to increase local bloodstream and relax involved tissue by stretching, Kabat method which is a neurorehabilitation technique that uses spiral and diagonal movement patterns in conjunction with stretch, resistance and other proprioceptive facilitation techniques to reinforce neuromuscular recruitment, Kinesiotherapy exercises for mouth opening and jaw lateralization, and specific exercises to increase mimic muscles motility (face stretching exercises), and to recover motions of temporo-mandibular joints [5-8,12,15].

Joints

Commonly affected joints are the wrists, shoulders, ankles, and hips, with the distal joints often affected first. Involvement is typically symmetric and can be progressive, involving more areas of the skin and joints over time [16-18]. Cutaneous and musculoskeletal involvement [12,19] and tendinopathy [20] are often associated with joint complications in CTD aggravating their outcome (tendon contractions, joint deformities, and destructions). In these complex forms, the functional disability and the limitation of the sectors of articular mobility are often very marked [12,19,20]. Joint rehabilitation aims mainly to improve or at least preserve the existing range of motion in order to allow normal function [21]. Massage therapy techniques including pressure and kneading are recommended for stiff joints. Passive, active-assisted and active mobilization are utilized according to pain intensity and muscle strength [5-8,15]. Posture for the hips, knees, ankles, shoulders, elbows and wrists are recommended at the end of rehabilitation sessions in order to help preserve the obtained progress. It is essential that patients learn self-rehabilitation exercises and perform them regularly supervised by the healthcare provider at first then at home [21-24].

Proprioceptive rehabilitation is indicated to increase joint stability and enhance the patient's balance. Exercises are performed with joints at charge if possible, they can be specific to a joint or a through a complex motion of the limb enhancing the coordination between agonist and antagonist muscle groups [19,23,24]. Occupational therapy can help to increase significantly the upper limbs function and reduce functional dependence for daily activities through exercises that focus on the fine gripping gestures and wrist and fingers postures [5-8,19,23,24]. Orthotics for hands and wrists are indicated to decrease pain, to preserve

function and to reduce deformities. They can be used to maintain the joints in resting position to temporarily put to rest inflamed joints or to compensate for a lack in one direction of the range of motion. A regular evaluation based on their tolerance, degree of pain reduction and efficiency to reduce malalignment should be performed and decide whether they have to be renewed or not [21-24].

Technical assistance gadgets are very helpful for accomplishing daily activities. For the upper limbs they reduce pain and joint stress by widening grip making thin objects easier to hold and extending the reach, they are prescribed keeping in mind the patient's environment and individual expectations. For the lower limbs they provide much needed help to make walking less energy consuming and much safer by reducing the risk of falls [19,21-24].

Muscles

The muscle weakness mainly affects the shoulder and pelvic girdle muscles but can involve neck, respiratory and pharyngeal muscles [25-28]. Muscle involvement in CTD may be of the type of myalgia, localized myositis, or most often diffuse myositis. The long-term evolution is towards fibrosis of muscle fibers and amyotrophy [25-30]. Muscle strengthening must be adapted to the patient's general condition and the extent of joint damage [19]. It can be applied for all patients as soon as they manifest local or global muscle weakness. The following modalities for muscle strengthening have proven to be efficient:

- A. Analytic or integrated in a global training program.
- B. Isometric, dynamic and isokinetic methods.
- C. Mild to intense training (50 to 80% of the maximal contraction) [7,8,19].

In fact dynamic muscle strengthening is well tolerated and does not expose to disease reactivation or to a faster joint destruction. However, mechanical solicitation of severely damaged joints should remain prudent in the absence of sufficient knowledge to support it especially on the long term. That is why when a joint presents major destruction or an inflammatory thrust it is recommended to manage the surrounding muscles according to the following modalities:

- A. Isometric condition.
- B. Against light to mild resistance.
- C. In discharge if the joint is bearing.
- D. Respecting the pain threshold.
- E. Avoiding intense and heavy load bearing, which seem to accelerate damage progression [6-8,15,19].

Electrostimulation techniques are not recommended alone, they should rather be considered as adjuvant methods indicated to maintain or restore muscle strength in some groups. However it should be noted that they include an increased risk of cutaneous burns increased by skin fragility in patients treated with corticosteroids [6-8,15,19].

Cardiovascular and Pulmonary Systems

Literature suggests that lung disease in CTD's is related to pleural effusion, acute/chronic pneumonitis, pulmonary embolism, pulmonary hypertension or diffuse interstitial lung disease [31-33], while the cardiovascular system can suffer from pericarditis, myocarditis, Libman Sacks endocarditis, arterial and venous thrombosis, hypertension, and premature atherosclerosis [31,32,34-36]. Most cardiopulmonary issues related to CTDs are related to decreased aerobic capacity and performance status from a lack of physical activity. Inactivity can cause profound muscle atrophy, as well as a decline in cardiac performance, with increased resting heart rate, decreased stroke volume, and decreased maximum oxygen consumption (VO₂ max) [31-33].

Ischemic strokes and arterial occlusion of the limbs may result in focal or more generalized motor deficits depending on the location and severity of the vascular and neurological involvement [34-38]. Persons with CTDs may have impaired function of respiratory muscles, with the muscles of expiration more affected than those of inspiration. As pulmonary decline hinders patients' ability to participate in physical and occupational therapy, addressing these impairments is essential to prevent the patient from further physical decompensation [7,39-41]. Regular practice of physical aerobic activities enhance cardiopulmonary endurance and are recommended for all CTD's patients [39-41]. These activities consist in walking, cycling, swimming and Rhythmic dance. They have not shown to be particularly deleterious on the disease activation nor on pain neither joint destruction [5-8]. Their modalities and intensity should be however adapted to the patient's general, cardiovascular and articular status, they should have a low joint stress to be performed in discharge if the lower limb's joints are severely affected. Aerobic activities should be temporarily restricted during a disease reactivation phase [39-41].

Group physical activity supervised by a kinesio therapist have proved to be more efficient on the general wellbeing and joint mobility than an individual self-program [40,41]. The rehabilitation of motor deficits caused by neurovascular involvement in CTD is nothing special compared to the rehabilitation of deficits secondary to classical strokes. Balneotherapy is recommended as an adjuvant to other techniques and it allows mainly to have a physical inactivity while discharging the suffering joints, the included exercises can be active or passive and are performed in warm water (32° to 35 °C). It is indicated outside severe inflammatory periods, with special attention to immunodepression conditions in relation to long corticosteroid treatment and cutaneous lesions. They have a beneficial effect on pain and muscle stiffness, increasing joint range of motion and increasing the patient's self-confidence by allowing him to achieve exercises that were otherwise impossible [39-41]. A combination of exercises in a swimming pool and weakly group exercises are more recommended than group exercises alone [7,39].

Conclusion

Medications are usually insufficient to control CTD, and patients often find themselves with functional and/or physical disabilities affecting their quality of life. Physical therapy and

rehabilitation have proven benefits on the course of cutaneous, rheumatic, pulmonary, and cardiovascular complications of these diseases. Health professionals need to be more and more familiar with specificities and indications of these supportive therapies to improve outcomes for their patients followed for CTD.

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