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Mini Review

Effects of Pulmonary Rehabilitation in Patients with Idiopathic Pulmonary Fibrosis: Integrative Review

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Abstract: Idiopathic Pulmonary Fibrosis (IPF) is a chronic lung condition characterized by progressive scarring of lung tissue, primarily affecting individuals aged 60 to 70, with a higher prevalence among men and smokers. It carries a poor prognosis, with an average life expectancy of up to 5 years. Pulmonary Rehabilitation (PR) is a comprehensive intervention involving detailed patient assessments and tailored therapies aimed at improving physical and emotional well-being, promoting long-term health-promoting behaviors. To assess the effectiveness of PR in IPF patients, an integrative review study was conducted, focusing on articles published from 2010 to 2020. Results showed improvements in exercise tolerance, functional capacity, and quality of life. While the Six-Minute Walk Test (6MWT) produced mixed results, most patients demonstrated gains. Interventions ranged from physical training alone to combined exercise and educational support programs, lasting 6 to 12 weeks, conducted 2-3 times weekly. Despite noted improvements during the intervention, sustaining gains remains a challenge, highlighting the need for further research to refine PR protocols tailored to IPF patients' specific needs.

Keywords: Idiopathic Pulmonary Fibrosis; Pulmonary Rehabilitation; Exercise Therapy.

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1. Introduction

Idiopathic Pulmonary Fibrosis (IPF) is defined as an inherent form of chronic fibrosing interstitial pneumonia, associated with the histological pattern of Usual Interstitial Pneumonia (UIP), exclusively affecting the lungs. IPF tends to manifest around the ages of 60 to 70 and is more prevalent in men and smokers, expressing a poor clinical prognosis with an average survival of up to 5 years [1]. Interstitial lung diseases are rare, with IPF being the most prevalent and having a high degree of lethality. Current studies show a high global incidence ranging from 4-10 deaths per 100,000 population in ten countries across Europe, North America, Asia, and Oceania. In Brazil, there has been an increase from 0.65 deaths per 100,000 in 1996 to 1.21 deaths per 100,000 in 2010 [2].

Patients who present with unexplained chronic cough, exertional dyspnea, bibasilar crackles, and/or digital clubbing should be referred for investigation, especially when multisystem diseases have been ruled out. Clinical presentation, along with lung imaging obtained through volumetric chest scanning, plays a significant role in the diagnosis of IPF [3]. Due to the complexity of the disease, patients with IPF experience a reduced quality of life due to dyspnea, severe fatigue, and exercise intolerance. These factors are evident in the Six-Minute Walk Test (6MWT), the St. George's Respiratory Questionnaire specific to IPF (SGRQ-I), and the modified Medical Research Council dyspnea score [4].

Pulmonary Rehabilitation (PR) is characterized by comprehensive intervention, starting with a detailed patient assessment followed by tailored therapies involving ed-

ucation, behavioral change, and physical training designed to improve both physical and mental conditions, with the aim of encouraging long-term adoption of behaviors that promote health restoration [5]. Given the clinical complexity of IPF, a multidisciplinary approach is essential to address symptoms and support daily needs, ultimately enhancing the quality of life for these individuals. This is particularly important since it is directly related to the role of the physiotherapist [6].

It is believed that the role of physiotherapy in addressing the functional kinetic condition, severely affected by the disease, could help define the profiles of patients suitable for PR and the implications of this intervention. This would provide a better understanding of the effectiveness of respiratory, motor, and functional aspects, potentially offering improved prognostic insight for the implementation of PR in IPF. Consequently, the objective of this study is to identify the effects of PR programs on patients with IPF concerning functional capacity (FC), activities of daily living (ADLs), anxiety, depression, and quality of life (QoL).

2. Methodology

We conducted an integrative review study, characterized as a broad methodological approach that includes the analysis of research providing a basis for clinical practice [7]. This study was carried out from March to May 2020 in the city of Fortaleza, Ceará, Brazil. We searched for original articles related to the topic in databases including the Latin American and Caribbean Center on Health Sciences Information (BIREME), Scientific Electronic Library Online (SciELO), U.S. National Library of Medicine (PubMed), and the Physiotherapy Evidence Database (PEDro). Articles were sorted within the established period and criteria.

We included original articles published in Portuguese and English that addressed the proposed theme within the specified time frame. The chosen time frame focused on the most recent studies demonstrating the effects of pulmonary rehabilitation in patients with idiopathic pulmonary fibrosis. Exclusion criteria included literature reviews, case reports, articles not freely available in the search databases, duplicates, and articles not in the specified languages for this study.

We used the terms "Idiopathic Pulmonary Fibrosis, Exercise Therapy, Pulmonary Rehabilitation" individually and in combination, along with their English counterparts "Idiopathic Pulmonary Fibrosis, Exercise Therapy, Pulmonary Rehabilitation," combined using the Boolean operator "and". The initial search was performed using these descriptors, followed by a preliminary review of titles and abstracts to filter materials. Subsequently, full texts were analyzed. At the end of this process, studies that met the inclusion criteria and exhibited good methodological quality were included in the study. The articles were independently collected by four researchers. After the search was completed, the articles were read separately, and then one of the authors conducted a filtering process, counting duplicates and selecting eligible articles according to the established criteria.

The variables collected for the research included heart rate (HR), respiratory rate (RR), peripheral oxygen saturation (SpO_2), systolic and diastolic blood pressure, as well as the degree of dyspnea measured using the Borg scale. These measurements were taken during the six-minute walk test (6MWT). Additionally, quality of life tests included the Short Form Health Survey 36 (SF-36), Maximal Inspiratory Pressure (MIP), Maximal Expiratory Pressure (MEP), and the 1 Repetition Maximum test (1RM). These variables were collected from articles that contained all of them or separately, depending on the variable studied in each literature source.

3. Results

A total of 475 articles were identified in the literature through the initial search using the combination of the descriptors "Idiopathic Pulmonary Fibrosis (IPF)" and "Exercise

Therapy (ET)/Pulmonary Rehabilitation (PR)." After applying available filters in some of the databases, following the predefined study criteria, and subsequently reviewing the titles and/or abstracts, 33 complete articles remained, which were potentially relevant for further evaluation. The complete reading of these articles was conducted by four co-authors to obtain additional details, and a consensus was reached to exclude 17 articles as they were duplicates, 2 articles as they were guidelines, and 3 articles as they were projects, leaving 11 articles for the final analysis.

The analysis reveals that the studies were conducted in 6 different countries, primarily consisting of randomized clinical trials (RCTs). The number of patients varied from 10 to 21 in the RCT groups. A prospective study and one RCT assessed the effects of PR in various interstitial lung diseases (ILDs), with a total of 99 and 142 patients, respectively. The Pilot Study, on the other hand, evaluated PR in IPF and COPD with a total of 77 patients. Results indicated improvements in exercise tolerance, functional capacity, quality of life (QoL), and the Idiopathic Pulmonary Fibrosis-specific St. George's Respiratory Questionnaire (SGRQ-1). These improvements were associated with enhanced leg strength and reduced sensation of dyspnea, which showed positive trends in most of the literature. The Six-Minute Walk Test (6MWT) did not yield unanimous results but generally exhibited positive markers. Even variables that showed the most significant improvements did not maintain their benefits in assessments conducted six months after the intervention.

The protocols used in the intervention groups showed consistency across all studies, with minor variations. They all included structured physical training (PT), covering components such as warm-up, stretching, aerobic and resistance exercises, respiratory exercises, with or without health education. Some studies also involved medical supervision, nutritionists, psychologists, social workers, and other services offered in PR centers. The interventions ranged from 6 to 12 weeks, with sessions held two to three times per week, aiming to assess the effects of PT, whether included in PR protocols or as standalone PT, often with the assistance of other healthcare professionals. The objectives ranged from assessing dyspnea to evaluating functional capacity, and the data are summarized in Table 1.

Table 1: Characteristics of the studies included in the review.

Reference	Country	N	Time	Protocol	Results
[8]	Israel	GC=15 GI= 17	12 weeks	TF-1	↑ exercise tolerance, CF, leg strength, QoL
[9]	Israel	GI=15 GC=17	12 weeks	TF-2	↑ 6MWT, mMrc, and SGrQ-1 in exercise but not with echo in resting cardiography
[10]	Japan	FPI= 45 DPOC= 45	8 weeks	TF-3 + TE	↓ dyspnoea, ↑ muscle strength, ↑ 6MWT and ↓ ADL, effects not sustained on IPF after 6 months
[11]	USA	FPI: 21 DPOC: 56	6 weeks	TF-2 + TE	↓ Fatigue ↓ Anxiety, depression ↑ SGRQ-1 ↑ Mean TC6. No ≠ Between IPF and COPD TC6=Did not change
[12]	USA	GI=11 GC= 10	12 weeks	TF-1 + AM+ TE	O2= ↑ 0.577 during exercise MIP= ↑ p/0.05 Dyspnea= ↓ 0.281

[4]	Australia	Total = 142 FPI=61 Asbestosis=22 CTD-ILD =23 Others=36	8 weeks	TF-3 + TE	TC6, CDRQ, SGRQ-1 > in FPI and Asbestosis < in CTD-ILD, Not maintained after 6 months
[13]	USA	GI=11 GC=10	12 weeks	TF-2 + Palestras	SGRQ-I= ± 9 ± 22 ↑ IPAQ= 14,428 ± 8,884, Not maintained after RP BDI= there was no change
[14]	Brazil	Total = 99 FPI = 47 Emphysema = 27 Others= 25	12 weeks	TF-1 + AM + TE	↑ TC6 ± 72m ↑ SF-36 = CF-22, AF-10, AS-15, AM-8 and Vitality-10
[15]	USA	GI=10 GC=11	12 weeks	Video game exercise	6MWT (- 22 ± 56 m vs- 60 ± 111 m), respectively, SGRQ-1 (3 ± 9 vs 1 ± 11)
[16]	Israel	GI=16 GC=18	12 weeks	TF-1+ TE	30s test - chair ±3 p=0.01 SGRQ-1 ±6 p =0.037
[17]	Israel	GI=14 GC=14	12 weeks	TF-1	↑ Physical Activity≠ ↑ Body composition. Unmaintained Effects

Abbreviations. RCTs: Randomized Clinical Trials; PR: Pulmonary Rehabilitation; IG: Intervention Group; CG: Control Group; IPAQ: International Physical Activity Questionnaire; BDI: Baseline Dyspnea Index; FC: Functional Capacity; QL: Quality of Life; AM: Multidisciplinary Assistance; TE: Educational Training (stimulation and conservation of energy); TF: Physical Training (1 = warm-up, stretching, aerobic, resistance, breathing exercises; 2 = aerobic, resistance, flexibility, breathing; 3 = stretching, strength and breathing exercises).

4. Discussion

Pulmonary Rehabilitation (PR) is included in the treatment protocols for Idiopathic Pulmonary Fibrosis (IPF) as recommended by ATS (2002). However, its effects and limitations are still under development. It is known that the PR protocols used for IPF patients are essentially the same as those originally designed for Chronic Obstructive Pulmonary Disease (COPD) patients. This is primarily due to the commonality of exercise limitations and disability among these patients. Nevertheless, the differences in the pathophysiology of these diseases raise questions about the effects, the appropriate timing for initiation, and the relationship with the degree of impairment, especially considering that many IPF patients are elderly and often have other comorbidities.

It is of considerable importance to assess the physical conditions of patients facing this condition. As evidenced by the analysis of the studies, physical debilitation, combined with the limited treatment options available, directly impacts the emotional well-being and significantly reduces the quality of life for these patients. The selected studies demonstrate a range of effects that can benefit IPF patients. Improvements in leg strength, functional capacity, and reduced sensation of dyspnea during exercise can provide a minimum level of independence in performing activities of daily living (ADLs). This, in turn, may positively impact the patients' quality of life, as indicated by the SF-36 and SGRQ-1 tests. The increase in the distance covered in the Six-Minute Walk

Test (6MWT), assessed before and after the intervention, as well as improved exercise tolerance, were generally observed to be beneficial. Although some variables did not show significant improvements, they also did not worsen, maintaining baseline values in the intervention groups while declining in the control groups.

The effects of PR in COPD patients have been extensively studied, and two of the comparative studies involving COPD and IPF patients showed gains. However, only the COPD group maintained these gains after discontinuation, while the IPF group saw declines in all variables. The possibility that these effects may be more pronounced in COPD patients may be due to the distinct characteristics of the diseases and the fact that PR programs for chronic lung diseases are primarily based on studies in COPD patients.

In this context, it is evident that there is a need to develop PT protocols specifically tailored to IPF patients. This includes educational programs focusing on specific energy conservation techniques, appropriate duration, and considering the disease stage, which is directly related to the effectiveness of the training. Another important factor observed in the studies is the significance of multidisciplinary care, an integral part of PR protocols. Patients who receive both physical training and health education show superior results compared to studies that include only physical training.

In Jackson's study [12], the effects of PR were positive for Maximal Inspiratory Pressure (MIP), even though there were no effects on Maximal Expiratory Pressure (MEP) and respiratory mechanics. While some variables did not show improvements when comparing the intervention and control groups post-intervention, the baseline values in the intervention group were maintained, whereas there was a degradation in the control group. In a disease like IPF with numerous associated comorbidities, maintaining functional values while awaiting transplantation can be considered a positive outcome.

All studies indicate two or more variables with a positive effect. The primary concern in IPF is to maintain the quality of life for these patients, as there are no curative drugs available, and not all patients are eligible for transplantation. Additional therapies can provide some relief in terms of physical and emotional conditions. This underscores the need for a specific approach to IPF patients that addresses the unique aspects of this condition and emphasizes the importance of continuous PR intervention, as gains are lost with discontinuation. Due to the etiological profile of IPF, there is limited availability of studies, with most of them focused on pharmacotherapeutic interactions. The research in this area is limited, and the data are imprecise, making it difficult to allocate these patients. Therefore, there is a need for more IPF-specific protocols and further research in this area.

5. Conclusion

Pulmonary Rehabilitation (PR) undoubtedly leads to improvements in the functional capacity of patients with Idiopathic Pulmonary Fibrosis (IPF), enhancing their autonomy in performing activities of daily living (ADLs) and reducing symptoms of anxiety and depression, ultimately leading to a better quality of life (QoL). However, it is a consensus that these gains are not maintained in the long term, and the reasons for this phenomenon have yet to be fully determined. It is believed to be related to factors inherent to the disease profile. There is a clear need for further research regarding PR protocols for IPF patients since there is no consensus on the most effective approach for these individuals. Defining these protocols is essential to guide the application of therapy, both for patients eligible for transplantation to maintain preoperative physical condition and for patients for whom transplantation is not a viable option. In the latter case, the goal is to mitigate the physical, emotional, and social impairments caused by the disease, ultimately improving their QoL.

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