Main Components of Exercise Rehabilitation Training for PAH

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Physical activity is one of the most important modifiable factors in our lifestyle and may significantly impact the prevention and treatment of cardiovascular diseases. Exercise may modulate a variety of molecular mechanisms related to proliferation, apoptosis, oxidative stress, inflammation, thrombosis, proteolysis, and vasodilatation. Without a doubt, exercise training (ET) is necessary in order to reverse the physical impairment that accompanies pulmonary arterial hypertension (PAH) and to maximize the benefits of pharmacotherapy without apparent clinical risk.

Keywords: PAH ; pulmonary arterial hypertension ; exercise training

1. Introduction

Pulmonary arterial hypertension (PAH) is an incurable [1][2] pathophysiological [1] condition associated with non-specific symptoms, including general weakness, shortness of breath on exertion, decreased muscle strength and endurance, leg fatigue, and peripheral edema, significantly affecting functional performance. In addition, patients may experience chest pain, coughing, palpitations, dizziness, and syncope, leading to reduced activity in daily living, significantly reduced physical performance, and a deterioration in the quality of life [3][4][5]. Reported complaints initially occur during exercise and later, along with the progression of the disease, at rest [5]. Thus, the main goal of PAH treatment is to ensure an acceptable level of quality of life as early as possible in the course of the disease [1], reduce symptom progression, improve functional class [6] and, if possible, improve prognosis [1][6][7][8], which remains poor [2][9]. This poses difficulties due to the non-specific symptoms significantly delaying the diagnosis of PAH [10]. Reported exercise intolerance may be initially ignored by patients due to reduced daily physical activity, poor commonly understood physical fitness, and as a result, a reduction of adequate cardiovascular and respiratory system efficiency [5]. A relationship between physical activity and the severity of PAH has been found [11]. Diagnosed patients severely reduce their physical activity, and more than half of them limit their normal activities of daily living [12]. Significantly, reduced daily physical activity in people with PAH has been shown to be associated with more severe symptoms reported by patients [13].

Numerous clinical studies have been carried out in order to enhance the knowledge and understanding of the pathophysiology of PAH, and yet this incurable condition, characterized by a high mortality rate, still requires a better transfer of new scientific knowledge to inform medical interventions [14][15]. Advanced specific therapies for pulmonary hypertension (PH), which resulted in clinical improvement and improved prognosis in PAH patients, still had little effect on physical performance. Exercise limitations in PH, caused by a variety of physiological mechanisms, have a profound effect on morbidity and mortality [16]. These limitations are the result of complex interactions between the circulatory and respiratory systems and the musculoskeletal system [17].

In recent years, the perceived importance of physical activity and exercise in patients with PAH has changed significantly [17][18], and exercise training (ET) dedicated to PAH (as adjuvant therapy in pharmacological treatment), which results in measurable improvement in patients' health, is beginning to attract increasing interest among clinicians [19].

It is widely recognized that inequality in access to healthcare is a global concern [20]. Moreover, the availability of specialized rehabilitation programs for PAH patients and reimbursement of costs is largely limited to a small number of countries [15][19]. Therefore, the current research focuses on the development of an optimal rehabilitation model that is a compromise between the effectiveness, cost, and availability of therapy [18].

2. Main Components of the Exercise Training

Physical activity is one of the most important modifiable factors in our lifestyle and may significantly impact the prevention and treatment of cardiovascular diseases [21]. Exercise may modulate a variety of molecular mechanisms related to proliferation, apoptosis, oxidative stress, inflammation, thrombosis, proteolysis, and vasodilatation [19][22][23]. Without a doubt, ET is necessary in order to reverse the physical impairment that accompanies PAH and to maximize the benefits of pharmacotherapy [18] without apparent clinical risk [24]. Even a slight increase in normal activity levels can lead to beneficial effects on health, and individually adjusted ET can meet many potential therapeutic goals [24][25].

Unfortunately, despite the consensus in the relevant literature on the positive impact of the studied physical training programs and the recently made significant advances in understanding the pathobiology of PAH, the mechanisms underlying functional improvement in patients undergoing ET and its impact on long-term outcomes are currently not fully understood [19][22][24][25][26].

There is also insufficient data on what forms, frequencies, and intensities of exercise are required to obtain the most beneficial results [23][24][26][27][28]. The available literature shows that in addition to daily activity, regular endurance (or aerobic) training, strength training, and respiratory muscle training ought to be done [3][21][29][30][31]. Aerobic, resistance, and inspiratory muscle training, supported by individually adapted general conditioning training (also known as general fitness), stretching, body awareness training, neuromuscular relaxation techniques, positional relaxation, fascial therapy, etc., provide significant functional and physiological improvement [29][30].

2.1. Aerobic Training

The ways in which endurance training is carried out are generally consistent, despite differing in details [18]. Aerobic training is most often performed in the form of walking on a treadmill, walking outside or riding a bicycle ergometer (or both, which may increase the effectiveness of training) [18][19][30]. It is obvious that, despite the lack of studies that would test the effects of other popular exercise options, such as a recumbent stepper, cross trainer, upper body or horizontal ergometer, moderate to vigorous physical activity, regardless of the type of exercise, should positively affect physical performance [32]. Patients should also be encouraged to take walks outside of exercise sessions in order to minimize the daily reduction of physical activity [30].

Exercises can be carried out in a continuous or interval mode. In most of the rehabilitation programs developed for treating PAH, interval training models were used, which enables optimizing the efficiency of the training process [18].

Aerobic training should constitute a large part of the rehabilitation program. Currently, it is believed that aerobic exercise, performed 2–3 times a week, sufficiently improves patients' 6-minute walk distance (6MWD) and provides a slight increase in peak VO_2 value in cardiopulmonary exercise testing (CPET) [30]. However, it is likely that a higher frequency of aerobic exercise, performed 5 or more times a week, may increase physical fitness [28][30][33][34][35].

Initial sessions may consist of alternating same-duration exercises of moderate intensity followed by passive rest to avoid prolonged shortness of breath. As tolerance increases, the duration of intense exercise increases with it, eventually turning into a continuous phase of at least 30–45 min [30] with an intensity corresponding to approximately 60–80% of the HR (heart rate) achieved in an exercise test [25][28][35].

In addition to aerobic exercise, the inclusion of resistance exercise and inspiratory muscle training for 15 to 30 min as an element of a rehabilitation program has been associated with significant improvements in cardiopulmonary fitness and skeletal muscle profile [28][29][30][32].

2.2. Resistance Training

Resistance training may be an effective treatment option in patients suffering from PAH. Therefore, taking into account the way pulmonary hypertension affects skeletal muscle dysfunctions [3], some studies have utilized elements of resistance training as supplementary to endurance exercises [15][18][30]. In accordance with the recommendations of the American Heart Association, resistance training consisted mainly of low-level dumbbell training for large muscle groups [15][18]. Training loads should not exceed 50% of 1RM (one repetition maximum) for 10–15 repetitions per set. The Valsalva maneuver should also be avoided [25]. The available literature on PAH patients suggests that resistance exercises using body weight or with dumbbells weighing 500–1000 g for 15–30 min [29] sufficiently supplement aerobic training [25].

2.3. Breathing Training

Decreased strength and endurance of the inspiratory muscles are among the consequences (of high prognostic value) of the development of pulmonary arterial hypertension. This dysfunction results in increased dyspnea and fatigue during physical activity [18][30].

Breathing training may include breathing techniques, stretching exercises, strengthening the inspiratory muscles using resistance exercise devices, and activities such as yoga [29].

A common recommendation is that breathing training ought to consist of stretching, shaping and mobilizing the chest, re-educating the correct breathing patterns that enable ideomotor action, and strengthening the inspiratory muscles 5–7 days a week for up to 30 min [25][29]. Inspiratory muscle resistance training (as the main element of breathing training) may be performed with a small handheld device that blocks the patients' airflow until a predetermined "threshold" pressure of 30–40% PImax (maximum inspiratory pressure) is created to effectively improve respiratory muscle strength and endurance [30][36][37].

2.4. Education and Psychological Support

Patient and family education is one of the essential elements of the rehabilitation program for PAH patients. The aim of these procedures is to learn about one's own physical abilities [15], to be able to recognize one's own limitations (beyond which the exertion may become excessive or dangerous) [25] and to learn how to cope with difficult situations [15]. Selected mental training techniques that improve physical and cognitive functions are also used [25]. Utilizing cognitive-behavioral therapy may facilitate the recognition of negative behavioral and/or thought patterns that lead to a deterioration in the quality of life in the realm of feelings and emotions and may be helpful in developing mechanisms of "coping" with these disorders. The aim of psychological training is also to broaden the knowledge, awareness, and acceptance of the disease and to facilitate intrapersonal, interpersonal, and professional adaptation to the current clinical state [25].

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