

1. STRESZCZENIE W JĘZYKU ANGIELSKIM

Pulmonary arterial hypertension (PAH), due to complex etiopathogenesis and non-specific symptoms, is a serious diagnostic and therapeutic problem. Delayed diagnosis, reduced quality of life, decrease in exercise capacity, and progressive impairment of the functional state or right ventricular failure lead to rapid clinical deterioration and death of patients. Despite the specific treatment of PAH optimized in recent years, no significant impact on physical performance and survival time was achieved. It is believed that people with pulmonary hypertension constitute a group of patients in which the impairment of exercise tolerance and prognosis are comparable, if not worse, than in patients with left ventricular heart failure (HF).

In recent years, few experimental and clinical studies have been conducted showing the beneficial effects of regular physical training on the course of PAH. Actually conducted studies focus on developing an optimal rehabilitation model being a compromise between the effectiveness, costs and availability of therapy for this group of patients. This study is an important link in the therapeutic management strategy, contributing to the broadening of knowledge about training in PAH.

The study was designed in order to assess the effectiveness and safety of a proprietary, six-month home cardiac rehabilitation program dedicated to PAH patients, based on endurance, general fitness, resistance, and improving the efficiency of the respiratory system training. The analysis of the effects of training after one year from the start of exercises allows to fill the gaps in long-term data in the world literature, which is a significant limitation of the previous reports, assessing health behaviors and the impact of training on the functional parameters of patients after the implementation of the rehabilitation program.

The conducted clinical trial was of prospective, interventional nature, 109 patients (46 patients with pulmonary arterial hypertension and 63 patients with left ventricular heart failure with a reduced ejection fraction) were enrolled. The control group consisted of 25 healthy people without cardiovascular diseases. Patients who agreed to participate in a home cardiac rehabilitation were supervised by a therapeutic team and performed a structured rehabilitation program for a period of six months. After completing the intervention phase, the group of patients with PAH was observed for the next six months, allowing the long-term effects of the therapy to be assessed.

In order to prevent skipping training sessions (minimum 120 sessions in a 24-week period), thoughtful, comprehensive support of exercising persons was used (education, telemonitoring, self-monitoring diary, accelerometry, blood pressure and heart rate measurements, assessment of fatigue and dyspnea). The support model turned out to be highly effective (compliance rate ~ 91%) not only in relation to patients in the PAH group, but also in relation to HF, indicating the correct selection of measures. It was established that the ability to self-control supported by the use of various activity monitors and telephone supervision over the course of training constituted a necessary link in the rehabilitation program performed at home and seemed to be an important component in encouraging exercise.

After the intervention period, the PAH group experienced a significant increase in the average distance covered in the six-minute walk test (6MWD) by 71.4 m, which indicates comparable effectiveness to intensive rehabilitation in the hospital model. The prevalent part of exercising individuals (75%) exceeded the minimal clinically important difference (MCID) established for this group of patients. The six-month observation period, during which the majority of participants did not undertake training, allowed the observation of a stabilization of the achieved improvement in 6MWD at the level of 57 m. Moreover, there was a significant reduction (exceeding MCID) in reported dyspnea at rest after the intervention period and further improvement during the observation period. Systematic, controlled physical activity of patients and a long period of intervention also led to a significant increase in the level of resting SaO₂, which strengthens the value of the above analyses. This effect did not persist after exercises were discontinued in the next six months of observation, which suggests a relationship between systematic physical activity and resting blood oxygenation. In the HF group, there was also significant improvement in 6MWD (37.9 m), but without reaching the MCID for this population of patients. The observation made supports the notion, that despite the fundamental importance of researches on heart failure in a pioneering approach to pulmonary hypertension rehabilitation, the current design of the rehabilitation program seems to be more focused on the problems of PAH patients.

Despite the lack of a direct impact of resistance training, a statistically significant improvement in handgrip strength was achieved in the PAH group. Unfortunately, the effect was not sustained and fluctuated around the baseline values one year after the beginning of intervention. This observation suggests that central adaptations are responsible for the improvement of muscle strength in the absence of direct skeletal muscle resistance training. In the HF group, the increase in peripheral muscle strength was minimal, confirming the greater effectiveness of the developed exercise program in the PAH group.

Performed rehabilitation program also led to an increase in the maximal inspiratory pressure (PI_{max}) and expiratory pressure (PE_{max}) after the intervention period in both groups and a further increase in both parameters after the observation period in the PAH group. These data indicate that an inseparable element of the rehabilitation program should be resistance training of the inspiratory muscles along with exercises that restore the proper functioning of the respiratory system and mobilize the diaphragm and chest.

There was a significant improvement in health-related quality of life (HRQoL) of patients in the PAH group, as expressed by the SF-36 questionnaire (subscales: PF, RP, VT, RE, PCS). It was observed that after stopping exercise, the quality of life gradually decreased, and although it was higher than before starting the intervention (12 months earlier), no statistical significance was noted in relation to the pre-intervention values. It was also shown that the change in HRQoL (Δ PCS before and after the intervention) in the PAH group was not dependent on the degree of improvement in physical endurance parameters, peripheral muscle or respiratory muscle strength. However, in patients with HF, the quality of life improved together with a decrease in resting dyspnea and fatigue.

In this study, only mild cases of excessive fatigue or a single sudden increase in heart rate during training were reported. The obtained data confirm the view that physical training in PAH is not associated with any serious adverse events, reinforcing the belief that there is a need to popularize rehabilitation specific for pulmonary arterial hypertension in this population of patients.

Summarizing, developed for the needs of PAH patients, safe, a relatively simple and inexpensive six-month rehabilitation program allows the improvement of functional parameters, quality of life and physical endurance of patients with pulmonary arterial hypertension and patients with left ventricular heart failure with reduced ejection fraction. In addition, it has been proven that the assumptions made regarding the training method, the duration of rehabilitation process, the frequency of training sessions and the intensity of individual exercises, show greater effectiveness in the treatment of patients with PAH. The fact, that the beneficial effects of rehabilitation persist for six months after cessation of the exercises, indicates the purposefulness of implementing proprietary rehabilitation program in the group of patients with pulmonary arterial hypertension.

