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Individualized home-based exercise program for idiopathic pulmonary arterial hypertension patients: a preliminary study

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SOUHRN

Úvod: Je prokázáno, že rehabilitační cvičení je přínosnou formou nefarmakologické léčby pacientů se stabilní plicní arteriální hypertenzí (PAH). Většina studií hodnotila i výsledky programů nabízených v nemocnicích a prováděných pod odborným dohledem. Je třeba zajistit lepší přístup pacientů k této významné formě intervence; současně je nutno zaměřit výzkum i na dlouhodobé účinky programů cvičení prováděného samotnými pacienty v domácím prostředí. Cílem naší studie bylo zhodnotit adherenci, bezpečnost i účinek 12týdenního individualizovaného programu cvičení v domácím prostředí pro pacienty s idiopatickou PAH.
Metody: Náš projekt byla prospektivní pilotní nekontrolovaná intervenční studie. Pro účast v ní bylo z lotyšského registru PAH vybráno šest pacientů s idiopatickou PAH, potvrzenou pravostrannou srdeční katetrizací. Následně byl vypracován 12týdenní cvičební program přizpůsobený každému pacientovi podle jeho funkčního stavu a domácího prostředí. Program byl zaměřen na posilování, dechová, aerobní cvičení i techniky relaxace nervového systému i svalstva, monitorování sebekontroly, každotýdenní kontrolu pacienta telefonem a osobní návštěvy v ordinaci, kde fyzioterapeut mohl upravovat režim i sledovat jeho dodržování pacientem; současně se dbalo o maximální bezpečnost pacientů. Primárními sledovanými parametry z hlediska účinku programu byly zátěžová kapacita, způsob dýchání a kvalita života, z hlediska adherence to byly dny s cvičením a z hlediska bezpečnosti jakákoli příhoda jako „varovaní" během cvičení, neúplné zotavení a zhoršení symptomů plicní hypertenze.
Výsledky: Výsledky prokázaly poměrně vysokou míru adherence k předepsanému cvičebnímu režimu (průměr 92,5 %). V průběhu programu nebyly zaznamenány žádné nežádoucí příhody. Výsledky prokázaly význam zajištění optimálních dovedností jak z hlediska objektivních sledovaných parametrů, tak subjektivních symptomů. Výsledky šestiminutového testu chůze (6-minute walking test, 6MWT) prokázaly, že vypracovaný program významně zlepšuje zátěžovou kapacitu (průměrné zlepšení 39 ± 17,5 m). U čtyř účastníků (66,7 %) byl pozorován minimální klinicky významný rozdíl (minumum clinically important difference, MCID) ve vzdálenosti překonané pacienty s PAH při 6MWT (25–33 m). Statisticky významné zlepšení ve výchylkách pohybu hrudníku potvrzují změny ve způsobu dýchání naznačující větší zapojení bránice při dýchání po absolvování programu. Výsledky neprokázaly významné zlepšení v žádné z domén z dotazníku SF-36. Nicméně polovina účastníků studie dosáhla po absolvování programu hodnoty MCID (11 %) podle příslušných stupnic fyzického zdraví.
Závěr: Předběžné výsledky této studie prokázaly, že vypracovaný individualizovaný program pro cvičení v domácích podmínkách je bezpečný, snadno jej lze provozovat a umožňuje postupně zvyšovat intenzitu zátěže; současně zlepšuje fyzický funkční stav klinicky stabilizovaných pacientů s idiopatickou PAH. Hypotéza této studie podporuje představu o nutnosti provádět další výzkum formou randomizovaných kontrolovaných studií s ověřováním dosud získaných výsledků.

ABSTRACT

Background: Exercise-based rehabilitation has been proved as a beneficial additional non-pharmacological treatment in patients with stable pulmonary arterial hypertension (PAH). Majority of studies include hospital-based supervised programs. To improve patient accessibility to this important intervention and long-term effect the research on home-based programs is warranted. The purpose of our study was to evaluate the adherence, safety, training effects of 12-week individualized home-based exercise program in patients with idiopathic PAH.

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Methods: This was a prospective pilot uncontrolled interventional study. Six patients with iPAH confirmed by right-heart catheterization from the Latvian PAH registry were selected. A 12-week exercise program adaptable for each patient's functional state and home environment was created. The program included muscle strength training, respiratory, aerobic exercise and neuro-muscular relaxation techniques, self-control monitoring, weekly phone control and on-site re-assessment by physiotherapist were parts of the program to ensure both individualized adjustments and proper execution, and to maximize clinical safety. The primary outcome measures for training effects were exercise capacity, breathing pattern and quality of life, for adherence days of performance, for safety any event of "alarm sign" indicators during exercising, incomplete recovery, worsening of PH symptoms.

Results: The results showed a rather high degree of adherence to the prescribed exercise regimen (in average 92.5%). No adverse events were observed during the course of the program. The results proved the importance of ensuring optimal self-control skills both for objective measures and subjective symptoms. The 6-minute walking test (6MWT) results show that the developed program significantly improves exercise capacity (mean improvement 39 ± 17.5 m). In four participants (66.7%) the minimum clinically important difference (MCID) for 6MWT distance in PAH patients was observed (25–33 m). Significant improvement in chest excursions confirm changes in breathing pattern suggesting better engagement of diaphragm during breathing after the program. Results did not show significant improvements in either SF-36 survey domain. However, half of the participants reached MCID (11%) after the program at the physical health subscales.

Conclusion: The results of this preliminary study prove that the created individualized home-based exercise program is safe, easily followed and allows progression in exercise intensity and improves physical functional state in clinically stable iPAH patients. This study hypothesis supports the need for RCT to continue research and approve the results.

Keywords:
Exercise training
Home exercise program
Pulmonary arterial hypertension

Introduction

Pulmonary arterial hypertension (PAH) is a severe disease characterized by increase in pulmonary artery pressure and pulmonary vascular resistance which promote right ventricular failure – the leading cause of death in PAH.[1] The pathogenetic mechanisms are complex and only partially understood.[2] The most common symptoms on presentation are progressive dyspnea, fatigue, dizziness and episodes of syncope, which substantially limit patient's physical capacity and overall quality of life.[3]

At this moment there are fourteen PAH specific therapies available, that mostly act as vasodilators, on four PAH relevant molecular pathways. However, the key pathogenetic features of disease, such as adverse vascular remodeling or functionality of the right ventricle are not targeted by these drugs.[4]

Since PAH remains incurable, life-debilitating disease with high mortality despite recent advances in disease-targeted pharmacotherapy, other treatment options to improve quality of life for these patients are actively sought.

Guidelines on Diagnosis and Treatment of Pulmonary Hypertension published by European Society of Cardiologists in 2015, recommend (strength of recommendation, IIb) that exercise training programs should only be implemented in PAH centers experienced in both PAH patient care and rehabilitation of compromised patients as the available data on positive impact of exercise and long-term effects are highly limited.[3]

Recent studies on the impact of exercise training in patients with PAH suggest that exercise training is beneficial and safe non-pharmacological approach that should be included in treatment strategies of PAH. In most of them, patients underwent supervised training in hospitals or rehabilitation centers beforehand, and continued the prescribed exercise-training program at home in contact with the specialists to guarantee patient safety and compliance.[5]

The purpose of our study was to evaluate the adherence, safety, training effects of 12-week individualized home-based exercise program in patients with iPAH. As the first preliminary study on home-based exercise program for Latvian iPAH outpatients, the program's protocol was based on a low-intensity home-based training in conditions of self- and telemonitoring, and the study population included patients who had been stable for at least three months and were receiving optimized medical therapy.

Methods

Study population and design

This was a prospective pilot uncontrolled interventional study. Six patients with iPAH confirmed by right-heart catheterization from the Latvian PAH registry were selected. The inclusion criteria: age between 18 and 80 years, stable and on optimized medical therapy for at least 3 months before entering the study. Exclusion criteria consisted of the following: NYHA functional class IV; unstable condition; inability to visit the university clinic for on-site assessments.

The study was approved by the Riga Stradins University Ethics Committee and the patients signed written informed consent prior to participation. The patient enrollment was done in two steps, at first according to the registry data screening and afterwards the selection criteria were evaluated by a cardiologist (Fig. 1).

All patients were under the care of a cardiologist, the medical therapy was continued and remained unchanged during the exercise intervention. The exercise program was carried out by an experienced physiotherapist.

Exercise program

A 12-week exercise program adaptable for each patient's functional state and home environment was cre-

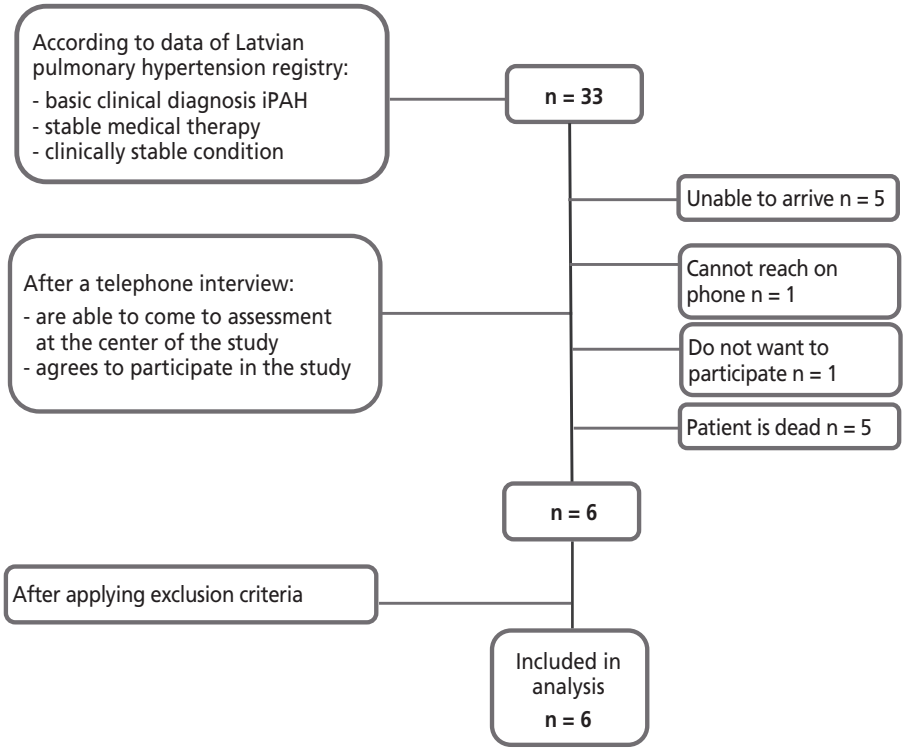


Fig. 1 – Enrollment of participants, describing number of patients screened, excluded (and reasons for exclusion). iPAH – idiopathic pulmonary arterial hypertension.

ated. The program included muscle strength training, respiratory, aerobic exercise, and neuro-muscular relaxation techniques (Table 1).

Self-control monitoring (HR, SpO_2, Borg scale, subjective clinical symptoms), weekly phone control and on-site re-assessment by physiotherapist were parts of the program to ensure both individualized adjustments and proper execution, and to maximize clinical safety. For the patient's convenience and improvement of compliance a visual material was created, which consisted of detailed program description, exercise plan, Borg's scale and information on the "alarm signs" which indicate the need to stop or not to start exercising. Each participant was provided with a paper format diary and pulse oximeter (Accurate FS20A).

Table 1 – The exercise program description

Monday	Aerobic excersise Breathing and relaxation
Tuesday	Resistance exercise (upper body) Breathing and relaxation
Wednesday	REST
Thursday	Aerobic excercise Breathing and relaxation
Friday	Resistance exercise (lower body) Breathing and relaxation
Saturday	Aerobic excercise Breathing and relaxation
Sunday	REST

"Alarm signs", which indicate the need to stop exercising, were determined as peak heart rate of more than 120 bpm, decrease in SpO_2 to 85%; perceived exertion as very hard (>6 on Borg scale) and subjective symptoms of exercise intolerance (severe dyspnea or fatigue, dizziness, pain etc.).[6,7]

The program included individual educational and motivational elements provided by a physiotherapist at on-site visits and weekly phone-controls. Education included both information about the program and its completion, overall benefits and possible adverse events of daily exercise, options to better manage activities of daily life, as well as self-management strategies for coping with exacerbation of disease symptoms.

Strength training

Six to seven resistance exercises scheduled two times a week, each done for one set, 5 to 10 repetitions. Muscle strength training was accomplished using person's own body weight and after 8th week, the use of additional weight (small dumbbells [500–1 000 grams] or water bottles; resistance bands) was strongly encouraged. The exercises were adapted individually and clearly explained for each patient.

Aerobic exercise

The aerobic workload was adjusted individually, taking in account 6-minute walk test (6MWT) results (both distance and reaction of HR, BP, SpO_2, perceived exertion [Borg scale], complaints) and the daily amount of physical activities of each patient. It consisted of 20 to 40 minutes of cyclic activity, three times a week. It was initially planned to adjust the intensity according to heart rate (HR), but during

the 6MWT on primary evaluation, increasing the intensity did not increase HR, and most importantly lowered SpO_2. For this reason, criteria used in dosing intensity of aerobic exercise were based on rating of perceived exertion as 5–6 points on Borg scale (hard) and sustained SpO_2 at starting level or decrease of no more than 5% from the initial.

The type of aerobic exercise was selected individually, taking in account patient's conditioning, living environment, daily habits and preference. The standard choice was walking, but other options such as walking while sitting on a chair, marching on spot and cycling were provided.

Breathing exercises and relaxation techniques

Breathing and relaxation exercises were done five times a week, in five to ten minute sessions. Respiratory exercises mainly aimed to improve chest expansion, breathing rhythm and emphasized diaphragmatic breathing. Relaxation methods included elements of progressive neuromuscular relaxation and body awareness.[8,9]

Outcome measures

Training effects

The primary outcome measures for training effects were exercise capacity assessed using 6MWT, breathing pattern evaluation using chest excursions and health related quality of life by SF-36 survey at baseline and 12 weeks.

6MWT was performed according to guidelines[10] together with SpO_2, HR, BP monitoring.

Chest excursions were recorded at level of processus xiphoideus and 10th rib by measuring difference in chest circumference between maximal inspiration and expiration.[11] Two lower levels of chest measurements were chosen to indirectly estimate involvement of diaphragm in the breathing pattern.

The SF-36v2 survey measures eight physical and mental health areas, as well as physical and mental component summary scores.[12] For norm-based scoring, every area has the same range (0–100), mean (50) and standard deviation. The subscale and component summary scores usually range between 20 and 70. If an individual respondent's scores are less than 45 or if the group's mean is less than 47, it is considered to be lower, compared to the healthy population.[13] The scores were calculated with SF-36 software.

Adherence

Adherence to the home program was assessed using patient diaries, where the patients had to write down daily records on completion of intended exercises on each day of the program. The total length of program was 84 days (12 weeks), where a total of 60 days was devoted to exercising and 24 days were rest days (Table 1). Performance on each exercise day was accepted as "full", if both types of exercises were completed, "partial" – one type of exercise completed, and "non" if no exercises were performed. Reasons of non-adherence were analyzed both from diaries and phone-control data.

Safety

Analysis of daily records in the patient's diaries, weekly phone-control data and on-site reassessment results at 4 weeks were used to detect any adverse effects: "alarm sign" indicators during exercising, incomplete recovery, worsening of PH symptoms.

Statistical analysis

Data are presented as mean ± standard deviation (SD) and for individuals as absolute values. Intragroup comparison between baseline and 12 weeks was done using Wilcoxon signed-rank test for all outcome variables. A value of $p < 0.05$ was considered statistically significant. All analyses were carried out with the SPSS v. 23.0 software program.

Results

Enrollment

Six PAH patients were enrolled in the study (Fig. 1) and all of them completed it. Five of the patients were women and the patients were aged from 56 to 79 years (mean 68 ± 7.6). PAH diagnosis in participants had been confirmed 3.9 ± 3.5 years ago (Table 2).

Adherence

Full performance of prescribed exercises in 57 and more of 60 days (>95%) was demonstrated by three participants. One patient full performance demonstrated in 54 of 60 days (adherence 90%) and one additional day

Table 2 – Baseline characteristics of the participants

Age (years)	68 ± 7,6
Gender (n)	
Women	5
Men	1
NYHA class (n)	
I	1
II	3
III	2
PAH therapy (n)	
PDE5 inhibitor	4
ERA	2
Spironolactonum	5
Oxygen therapy	1
Comorbidities (n)	
Hypertension	2
Dislipidemia	3
Bronchial asthma	1
Coronary artery disease	1
Time since diagnosis (years)	3.9 ± 3.5
Cardiac catheterization	
mPAP (mmHg)	46.8 ± 13.1
PAWP (mmHg)	13.8 ± 4.7
PVR (WU)	7.9 ± 4.1
6MWT distance (m)	375.8 ± 91.5

Data are presented as n, or mean ± SD. 6MWT – 6-minute walking tests; ERA – endothelin receptor antagonist; mPAP – mean pulmonary arterial pressure; NYHA – New York Heart Association; PAH – pulmonary arterial hypertension; PAWP – pulmonary arterial wedge pressure; PDE – phosphodiesterase; PVR – pulmonary vascular resistance.

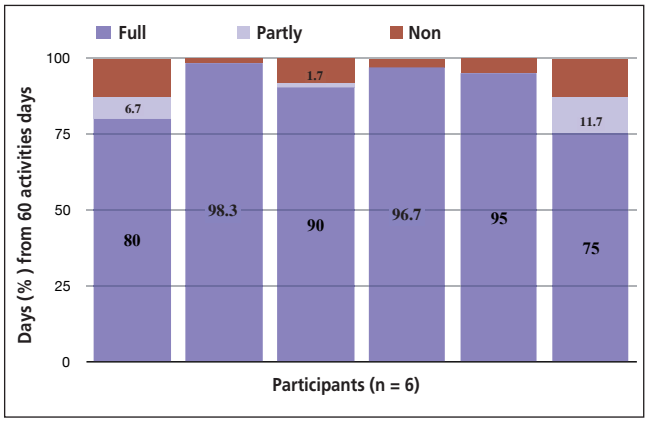


Fig. 2 – Exercise program adherence presented as performance (full [completion both prescribed exercises] [dark blue color], partly [completion of one kind from prescribed exercise] [light blue color], non [no kind of exercises was done] – in different colors) of intended exercises in all 60 program active days (expressed as days from 60 activity days [%]).

(1.6%) of partial performance. Remaining two patients demonstrated full performance in 48 days (80%) and 45 days (75%) (Fig. 2).

The main reasons for non-performance were associated with various errands that required leaving home (travelling, visiting friends, doctor's visits etc.) – recorded in five participants accounting for 1–5 days of non-performance. Two of the patients had health issues – one had back pain (2 days of non-performance) and one experienced fracture of the wrist after a fall on pavement, but on the next day of reduction, the patient resumed exercising. Two patients noted weather conditions that contributed to fatigue and served as reason for non-performance or partial performance of the exercise program (3 and 9 days). One of the patients had progressive peripheral edema due to not following doctor's orders regarding medication, which caused issues with full completion of exercises (reason for partial performance in 3 days).

Safety

None of participants was forced to interrupt exercising, no "alarm signs" or aggravation of clinical symptoms were observed. Data from the patient diaries represented normal HR recovery after exercising. All patients completed the entire 12-week program and no adverse events were recorded. During the phone or on-site visits, none of the patients noted that their symptoms had worsened in duration of this program. Only one of participants demonstrated insufficient self-monitoring skills.

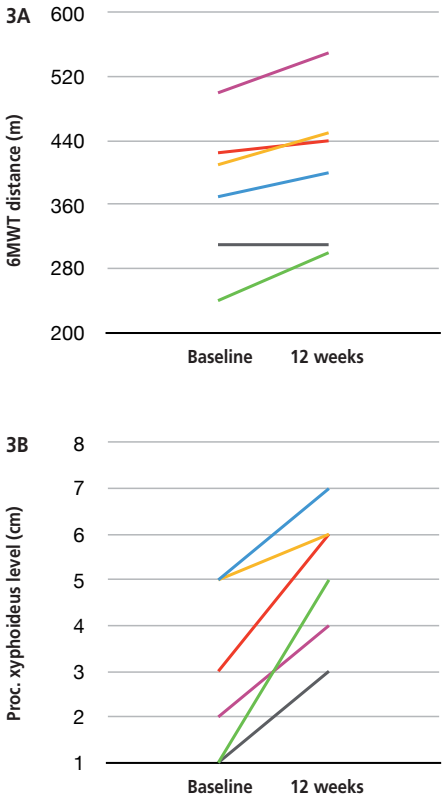
Training effect

6MWT distance increased for five of the patients. The total increase in distance ranges from 15 to 60 meters (mean 39 ± 17.5 m) (Fig. 3A, Table 3). In four participants (66.7%), the minimal clinically important difference for 6MWT distance in PAH patients was observed (approximately 33 m).[14] For one patient, 6MWT distance on final (12-week) assessment remained the same as at the baseline.

Table 3 – Change in 6MWT and chest excursions

Variable	Baseline	12 weeks	*p*-value
6MWT distance (m)	375.8 ± 91.5	408.3 ± 94.1	0.04*
Chest excursion (cm)			
Proc. xyphoideus	2.8 ± 1.8	5.0 ± 1.8	0.03*
10th rib	1.7 ± 0.8	5.3 ± 1.2	0.03*

Values are mean ± SD. Wilcoxon signed-rank was used to analyze changes between the baseline and at the end of 12 weeks in all outcome variables. *p <0.05 indicating a significant change. 6MWT – 6-min walk test.



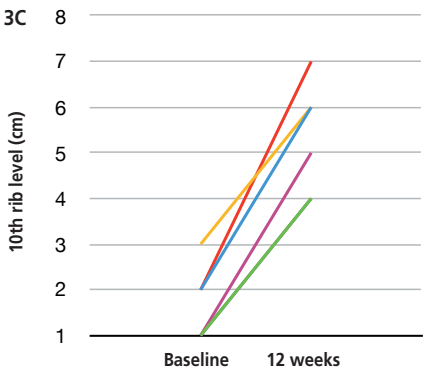


Fig. 3 – Individual changes in 6MWT and chest excursions at baseline and after 12 weeks. (A) 6MWT distance (m) for each participant (n = 6); (B) chest excursions at proc. xyphoideus (cm) for each participant (n = 6); (C) chest excursions at 10th rib level for each participant (n = 6). 6MWT – 6-min walk test.

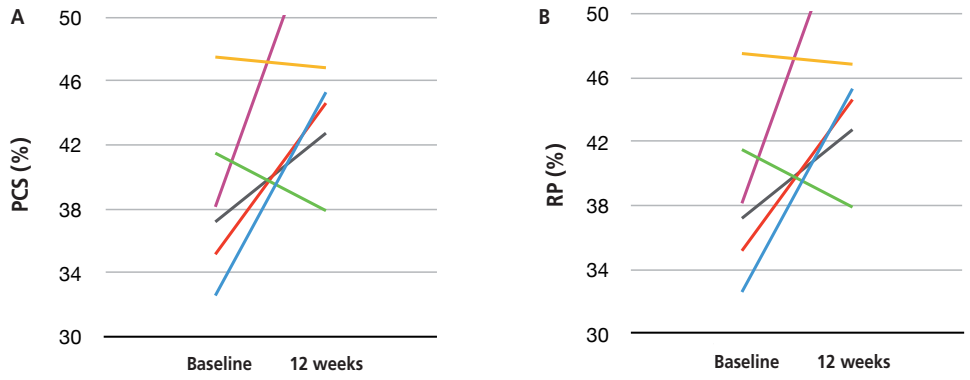


Fig. 4 – Individual changes in SF-36 domains RP and PCS at baseline and after 12 weeks. (A) PCS (%) for each participant (n = 6); (B) RP (%) for each participant (n = 6). PCS – physical component score; RP – physical role functioning.

Chest excursions increased in all participants (with mean increase 2.3 ± 1.1 cm at proc. xyphoideus level and 3.7 ± 0.8 at 10th rib level) and at 12 weeks demonstrated normal values in level of 10th rib (with mean increase 2.3 ± 1.1 cm at proc. xyphoideus level) (Fig. 3B, 3C, Table 3).

The mean values of SF-36 survey did not show a significant difference between baseline and at 12 weeks in either domain (Table 4). However, three patients reached improvement of more than 11% in domains of physical role functioning (RP), bodily pain (BP) and summary physical component score (PCS) (minimal clinical important difference) (Fig. 4).

Discussion

This was the first pilot study to evaluate the adherence, safety and training effects of 12 week individualized home-based exercise program for patients with iPAH in Latvia. The results of this study demonstrate that the individually adjusted home-based exercise program with close self- and telemonitoring was safe and significantly improved functional outcomes.

Table 4 – Change in SF-36 results

SF-36 domain	Baseline	12 weeks	*p*-value
PF	42.6 ± 6.6	42.5 ± 8.2	0.91
RP	41.1 ± 7.2	50.0 ± 6.1	0.08
BP	42.2 ± 10.0	54.6 ± 12.2	0.12
GH	41.9 ± 7.5	42.7 ± 8.1	0.50
VT	53.1 ± 9.3	51.6 ± 6.7	0.59
SF	49.8 ± 4.2	49.0 ± 9.3	0.96
RE	48.6 ± 8.1	51.0 ± 4.8	0.47
MH	53.9 ± 5.8	54.35 ± 1.4	0.92
PSC	38.7 ± 5.3	45.8 ± 6.5	0.12
MSC	55.7 ± 6.5	53.7 ± 3.7	0.75

Values are mean ± SD. Wilcoxon signed-rank was used to analyze changes between the baseline and at the end of 12 weeks in all outcome variables. * p <0.05 indicating a significant change. BP – bodily pain; GH – general health; MCS – mental component score; MH – mental health; PCS – physical component score; PF – physical functioning; RE – emotional role functioning; RP – physical role functioning; SF – social functioning; VT – vitality.

Adherence and safety

Patient compliance is one of the most important factors affecting the efficacy of home-adapted exercise programs. The results of our study show a rather high degree of adherence to the prescribed exercise regimen (in average 92.5%). It is, however, slightly lower in comparison to similar studies.[15,16] Our program was completely adapted to home environment under close self and telemonitoring. The participants appreciated weekly communication with physiotherapist during phone-controls, which both encouraged to maintain motivation and gave confidence on proper exercise execution.[17] Based on previous research on long-term exercise adherence in cardiac patients,[18] we could assume that self-monitoring using records in diaries (exercise performance, SpO_2, HR, Borg scale and subjective symptoms) improves both adherence and safety.

It is known, that self-monitoring of clinical stability indicators such as BP or blood glucose help to improve long-term health care results and coping ability.[19–21] Recently published studies on PAH patients have proved importance of developing the patient's own skills to manage daily life, supporting patients to take control of their disease to gain better health related quality of life.[22–24] Our study results emphasize patient education, high quality, and user friendly technical equipment as requirements to ensure optimal self-control and self-monitoring.

No adverse events were observed during the course of the program. In agreement with previous research,[15,25] our study supports the notion that exercise training is not associated with any serious adverse events, if done over close monitoring of objective and subjective parameters. Our study proves the importance of ensuring optimal self-control skills both for objective measures and subjective symptoms.

As the program was developed as completely home-based, in future it should be supplemented with technologies for remote, real-time data monitoring and storage (SpO_2, HR, physical activity level with accelometer) that may enhance safety. Use of telemedicine technologies has been approved in cardiac rehabilitation,[26,27] and newly available technologies can be considered as an opportu-

nity that may promote compliance, safety, and effectiveness of home-based exercise interventions in PAH patients.

Improvement in functional status and health related QoL

The 6MWD results show that the developed program significantly improves exercise capacity. The improvements in 6MWT distance seen in other studies range from 17 m to 96 m (in our study 39 ± 17.5 m).[28] Comparing the results with those of other studies by using the 6MWT distance is difficult because the treatment effect depends on the study design, exercise protocol, and patient selection criteria. In majority of them in-patient exercise programs or in-patient program followed by home-based exercises were studied. Compared to the results of home--based pulmonary rehabilitation program and home--based walking regimen,[15,29] rather similar improvement in 6MWT distance was observed (33.3 ± 25.1 m and 40 ± 13 m, respectively).

In the analysis of improvement in exercise capacity, it should be taken into account that the training protocol in our study included low-intensity exercise program, but the expected results to improve exercise capacity were achieved. We can assume that combination of resistance, aerobic, breathing and relaxation exercises,[28,30] and secondary benefits (activity level, self-efficiency)[23,29] from the program's implementation and adaptation to patient everyday life contributed to the positive impact of the program on improvement of exercise capacity in the analyses of iPAH patients.

In four participants (66.7%) the minimum clinically important difference for 6MWT distance in PAH patients was observed (25–33 m).[14,31]

The results demonstrate that the improvement in 6MWT results can be achieved both in patients with relatively low (baseline 6MWT distance 240 m, improvement 60 m) and high exercise capacity (baseline 6MWT distance 500 m, improvement 50 m) that could be explained by an individualized approach to adapting the program to the functional state and needs of each patient.

Unlike in other published studies on exercise training for PAH patients, where intensity of the aerobic exercises was aimed as 50–85% of target heart rate,[15,28] we were forced to seek another way of dosing the exercise intensity (perceived exertion, HR and SpO_2)[6] as on the initial visit during 6MWT increasing the intensity did not increase HR and most importantly lowered SpO_2. The slower chronotropic response may be the result of autonomic imbalance,[32] and is associated with clinically worse outcomes[32,33] that can explain our findings in context of rather late diagnosis of illness and initiation of pathogenetic medical therapy in the participants of our study.

Significant improvement in chest excursions confirm changes in breathing pattern suggesting better engagement of diaphragm during breathing after the program. The program emphasized promoting the engagement of diaphragm and "slow breath" to improve breathing pattern[34] and inspiratory muscle strength[35,36] that contributes to better exercise capacity[6,37] as well as more adaptive stress response (autonomic balance)[34,38] as were mentioned by the participants practicing the breathing exercises almost every day to manage daily activities. Because neither the pulmonary function nor level of oxygen consumption was assessed after the exercise program in the present study, the directly defined effects remain unclear and further studies are required.

SF-36 survey results in all physical health domains at baseline approved lower HRQoL compared to the healthy population, but mental health domains were rather high (more than 47). Similar trend in mental health domains, as well as better ratings of quality of life in older PH patients (>55 years) and those who were retired was observed in Finland PH cohort.[39] In our opinion, the SF-36 questionnaire did not reveal mental health issues such as anxiety, uncertainty and their dynamics after completing the program. Simultaneously there is evidence on high prevalence of clinically significant anxiety in PH patients.[40]

Whilst most previous studies on exercise-based rehabilitation programs reported improvements in HRQoL, but in majority this improvement may not be clinically important,[25] our results did not show significant improvements in either SF-36 domain. However, half of participants reached minimal clinical important difference (11%) after the program at the physical health subscales.[14] The results of HRQoL in the participants were likely be affected by co-morbidities, non-specific musculoskeletal complaints (clinically important reduction in pain was observed in three patients as secondary gain from the program) and life events (one participant got arm fracture).

The study limitations

The present study was designed to evaluate feasibility of an individualized home-based exercise program, therefore there was no control group. The limitation is the small sample size and there may have been some selection bias towards the more motivated patients. This was the first trial on any exercise-program in Latvia, therefore participants were well aware of the assignment to an intervention study, which may also have favored the present results.

The program was designed to be adaptable to everyday life subsequently affecting physical activity habits of oneself, but we did not verify (due to financial limitations) this issue by objective assessment of daily activities by accelerometry.[41,42] Such changes in daily habits could contribute to promoting long-term benefits, therefore absence of long-term follow-up we could consider as the study limitation.

Conclusions

The results of this preliminary study prove that the created individualized home-based exercise program is safe, easily followed and allows progression in exercise intensity and improves physical functional state in clinically stable iPAH patients. This study hypothesis supports the need for RCT to continue research and approve the results.

Conflict of interest
The authors have no conflicts of interest to disclose.

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