Exercise training in idiopathic pulmonary fibrosis

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Exercise training in idiopathic pulmonary fibrosis


Idiopathic pulmonary fibrosis (IPF) is a chronic, progressive and ultimately fatal interstitial lung disease (ILD) occurring primarily in older adults.[1] The mortality rate in the United States in 2003 was 61.2 deaths per 1,000,000 in men and 54.5 per 1,000,000 in women, which is the most common cause of death in progressive lung disease (60% of deaths).[1] The etiology is usually unknown although several risk factors have been suggested, and typically it has a poor prognosis, with an estimated median survival range between 2 and 5 years from the time of diagnosis.[1–3] IPF is characterized by progressive pulmonary restriction, ventilatory insufficiency, dyspnea impaired gas exchange and hypoxemia, and diminished exercise capacity.[1,3,4] Patients with IPF are more breathless and tend to be less physically active to avoid such symptoms.[5,6] All these manifestations have a significant negative impact on functional capacity and health-related quality of life (HRQOL) in IPF patients.[3–6] Long-term effective treatment, apart from lung transplantation, is still limited for most IPF patients despite some encouraging recent findings with pharmacotherapy.[1,7–9] According to the latest American Thoracic Society/European Respiratory Society (ATS/ERS) guidelines, pulmonary rehabilitation has only a “weak” level of recommendation for IPF patients based on the GRADE methodology for evidence quality.[1] The aim of this article was to comprehensively review the evidence of the wide impact of exercise training programs on patients with IPF and to address important research directions that should be focused on in the future.

Pathophysiology of IPF

Anatomically IPF manifests over several years by structural scar tissue within the lungs and pulmonary remodeling. This results in thickening of the alveolar walls and a characteristic appearance of honeycombing on high-resolution computed tomography.[3,10] In addition, parenchymal damage, interstitial fibrosis, collapse and apposition of alveolar walls, resulting in obliteration of alveolar lumina and distortion of normal lung architecture, have all been documented.[3,10]

Resting pulmonary function

Standard spirometry reveals decreased measures of forced vital capacity (FVC) and forced expiratory volume in 1 s (FEV₁), while the ratio of FEV₁/FVC remains normal (or increased), consistent with restrictive physiology as a consequence of reduced pulmonary compliance. This restrictive pattern is confirmed by reduced total lung capacity on lung volume body plethysmography measurements. [3,4] Gas exchange is impaired in IPF which
can be demonstrated by reduced diffusion capacity for carbon monoxide, although maximal voluntary ventilation is usually normal.[3,4] In general, as the disease progresses, lung compliance decreases and lung volumes fall.[4] The resting arterial blood gas is usually normal or may reveal mild hypoxemia; however, breathing pattern is often rapid and shallow as the disease progresses.[4]

**Exercise capacity and limitation**

Exercise intolerance is a cardinal feature of IPF, associated with severe exertional dyspnea and fatigue and poor QOL.[11–13] Patients with ILD and IPF typically exhibit reduced maximal or peak aerobic capacity (VO
subpeak), peak work rate and submaximal exercise endurance compared with age- and sex-matched normal subjects.[12]

Limitations to exercise capacity usually are multifactorial including abnormal pulmonary gas exchange, inefficient breathing mechanics, exercise induced hypoxemia, circulatory impairments and respiratory and skeletal muscle dysfunction.[4,11,12] A hallmark clinical sign among IPF patients is a decline in arterial O
sub2 pressure and arterial O
sub2 saturation (SaO
sub2) in response to exercise, which is mainly related to abnormalities in pulmonary gas exchange due to alveolar ventilation-perfusion (VA/Q) mismatching, oxygen diffusion limitation and low mixed venous oxygen content.[11,12] Ventilatory pattern is also abnormal in most IPF patients; however, breathing reserve in most patients is kept at normal levels.[11,12] Part of the elevated ventilatory drive in IPF during exercise relates to the increased dead space (VD) ventilation.[11,12] Although dyspnea is a predominant symptom in IPF patients, leg pain, chest discomfort and fatigue are common reasons for exercise test termination as well.[11,12] Several reports have suggested quadriceps weakness and peripheral muscle dysfunction in IPF patients.[14–16] Inflammation, hypoxemia, impaired oxidative capacity and high oxidative stress were proposed as contributing mechanisms in these studies.[14–16] In addition, patients can also be often bothered by a dry cough which interferes with daily activities. The onset of symptoms is slow, but symptoms become progressively worse over time.[3]

**The rationale for exercise training in IPF**

IPF is a chronic progressive lung disease that manifests in dyspnea and desaturation during physical effort, exercise intolerance and poor QOL.[1] Among other chronic lung diseases such as chronic obstructive pulmonary disease (COPD) patients, there is strong evidence for improvement following exercise interventions in exercise and functional capacities, level of dyspnea and HRQOL.[13,17–19] Despite the fact that IPF and COPD have a different pathophysiology, they share similar exercise and functional limitations in daily activities, dyspnea and poor QOL.[5,20] Furthermore, the American College of Sports Medicine stated in a position paper that exercise and physical activity are effective evidence-based strategies for limiting the progression of chronic diseases and disability in older adults.[21] Recently, the ATS/ERS documented that pulmonary rehabilitation is recommended for chronic lung diseases other than COPD, including ILD.[13] Furthermore, our team and others have shown significant short-term improvements following supervised exercise training among IPF patients.[13,20,22–24] These enhancements may have a significant clinical impact in helping IPF patients cope with functional activities of daily life and improve HRQOL.[22] In addition, IPF patients often present with comorbidities such as coronary arterial disease, chronic inflammation [1] and skeletal muscle dysfunction.[13] Exercise training has been established as an effective treatment for prevention and rehabilitation of those pathophysiology in many well-designed large studies.[21,25] Finally, exercise capacity outcomes such as 6-min walk distance (6MWD) and VO
subpeak are strong prognostic predictors for mortality in IPF[26–29] and are improved following exercise training programs with IPF patients.[16,22,23,30–38] It is possible that improvement in these outcomes following participation in exercise programs may also have some beneficial effect on prognosis in IPF; however, this has not yet been demonstrated.

**Effects of exercise training in IPF**

Exercise training has not been studied as extensively in IPF as in COPD patients.[5,13,20,24,39] However, a growing body of evidence has shown encouraging results with some health benefits following participation in these programs.[13,20,24,40] In the past few years, several retrospective, prospective and randomized controlled trials (RCTs) have demonstrated significant improvements in some outcomes after short-term exercise programs among IPF patients.[5,13,20,24,39] Table 1 summarizes the exercise-training studies among patients with IPF.

In general, studies showed that exercise training interventions are safe and effective treatment for ILD and IPF patients.[40] The exercise programs were undertaken for 4–12 weeks with 2–3 weekly exercise sessions of 30–60 min. All training programs included aerobic endurance modality such as walking and cycling, whereas some programs also combined resistance and flexibility training, respiratory muscle training and breathing exercises.[16,22,23,30–38,41]

**Exercise tolerance**

Exercise intolerance is a cardinal manifestation of IPF associated with shortness of breath, poor QOL and mortality.[11–13,26,27] The majority of studies reported significant improvement in 6MWD following short-term exercise training interventions ranging from 15 to 81 m (Figure 1).[16,22,23,30–38] However, only few were conducted as RCTs [16,22,23,37] and the level of recommendation for pulmonary rehabilitation in the latest ATS/ERS guidelines is still weak.[1]

The minimal clinical important difference (MCID) for 6MWD was established as 24–45 m among patients with IPF [42]; most studies reported an improvement above this
Figure 1. Improvements in 6-min walking distance across exercise training studies in patients with idiopathic pulmonary fibrosis.
MCID: Minimal clinical important difference; Δ6MWD: Pre- to postdifference of 6-min walking distance. ▲ Randomized controlled trials with mean difference of improvement between the exercise and the control groups. ● Retrospective study. ■ Prospective noncontrolled study.

Table 1. Exercise training studies in idiopathic pulmonary fibrosis patients.

<table>
<thead>
<tr>
<th>Study</th>
<th>Number of IPF patients</th>
<th>Research mode</th>
<th>Exercise Program’s type</th>
<th>Training modality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jastrzebski et al. [41]</td>
<td>13</td>
<td>Prospective</td>
<td>Twice weekly 4-week inpatient + 4 week home-based</td>
<td>Leg cycling and respiratory muscle training</td>
</tr>
<tr>
<td>Holland et al. [37]</td>
<td>34</td>
<td>RCT</td>
<td>Twice weekly 8-week outpatient</td>
<td>Walking + cycling and resistance training for lower body</td>
</tr>
<tr>
<td>Nishiyama et al. [23]</td>
<td>28</td>
<td>RCT</td>
<td>Twice weekly 10-week outpatient</td>
<td>Walking + cycling and resistance exercises for peripheral muscles</td>
</tr>
<tr>
<td>Ferreira et al. [34]</td>
<td>50</td>
<td>Retrospective</td>
<td>2–3 sessions a week, 6–8 weeks outpatient</td>
<td>Walking + cycling, resistance and respiratory muscle training</td>
</tr>
<tr>
<td>Ozalevi et al. [31]</td>
<td>17</td>
<td>Prospective</td>
<td>Daily home-based sessions 12 weeks</td>
<td>Walking, functional resistance training and breathing exercise</td>
</tr>
<tr>
<td>Swigris et al. [32]</td>
<td>14</td>
<td>Prospective</td>
<td>2–3 sessions a-week, 6–8 weeks outpatient</td>
<td>Walking, cycling and resistance training</td>
</tr>
<tr>
<td>Kozu et al. [30]</td>
<td>65</td>
<td>Prospective</td>
<td>2 sessions a week 8 weeks outpatient/home-based</td>
<td>Leg cycling and strength training walking and resistance training</td>
</tr>
<tr>
<td>Rammaert et al. [33]</td>
<td>13</td>
<td>Prospective</td>
<td>Daily home-based, 8 weeks</td>
<td>Leg cycling</td>
</tr>
<tr>
<td>Huppmann et al. [35]</td>
<td>202</td>
<td>Retrospective</td>
<td>Inpatient 5 days/week, 4 weeks</td>
<td>Walking, cycling and resistance training</td>
</tr>
<tr>
<td>Arizono et al. [36]</td>
<td>48</td>
<td>Prospective</td>
<td>2 sessions a week, outpatient, 10 weeks</td>
<td>Leg cycling, resistance training and respiratory muscle training</td>
</tr>
<tr>
<td>Ryerson et al. [38]</td>
<td>22</td>
<td>Prospective</td>
<td>2 sessions a week, outpatient, 6–8 weeks</td>
<td>Walking, leg cycling and sitting elliptical</td>
</tr>
<tr>
<td>Jackson et al. [16]</td>
<td>21</td>
<td>RCT</td>
<td>2 sessions a week, outpatient, 12 weeks</td>
<td>Walking, leg cycling, resistance and flexibility</td>
</tr>
<tr>
<td>Vainshelboim et al. [22]</td>
<td>34</td>
<td>RCT</td>
<td>2 sessions a week, outpatient, 12 weeks</td>
<td>Walking, leg cycling, stairs climbing, resistance and flexibility + breathing exercise</td>
</tr>
</tbody>
</table>

RCT: Randomized controlled trial.
MCID threshold as presenting a clinically meaningful effect of exercise training interventions.[16,22,23,31–38] Few studies have also assessed exercise tolerance with metabolic variables using cardiopulmonary exercise testing.[16,22,36,37] While Arizono et al. [36] and Vainshelboim et al. [22] demonstrated significant improvements in peak work rate, VO2peak and anaerobic threshold, Jackson et al. [16] showed only maintenance of steady state VO2 and Holland et al. [37] failed to show any significant improvement in cardiopulmonary variables despite a significant enhancement of 35 m in the 6MWD test.[37] This inconsistency should be addressed in future large group studies in order to understand the physiological mechanisms that underline adaptation to training in IPF.[16,22,36,37]

**Dyspnea**

Dyspnea is a predominant symptom of IPF and clinically significant due to its strong association with exercise intolerance, poor HRQOL and mortality.[1,6,30,43,44] Relief of dyspnea is therefore one of the important goals of IPF management.[43] Exercise training interventions in IPF have shown significant reduction in dyspnea sensation, although not all studies could demonstrate this benefit.[16,22,23,30–38] Evaluations of dyspnea were conducted using the Medical Research Council (MRC) dyspnea scale,[22,30–33,37,41] baseline dyspnea index,[16,23,41] Borg scale,[33,34,41] visual analogue scale [35] and University of California San Diego Shortness of Breath Questionnaire.[34,38] It is possible that the usage of different tools for dyspnea assessment and varied training programs could have an impact on the variability in these studies’ findings.[16,22,23,30–38]

**Quality of life**

QOL is impaired in IPF patients with severe symptoms and declined functional capacity.[5,45] Most HRQOL domains are poor,[5,45] and there are few therapies or other interventions that have shown efficacy to improve HRQOL in patients with IPF.[6] Swigris et al. [6] performed a systemic review of seven studies with 512 IPF patients that examined HRQOL.[6] The results of this review showed that IPF patients have significantly reduced HRQOL with a more pronounced repercussion in physical domains.[6] IPF patients suffer more breathlessness and tend to be less physically active in order to avoid such episodes which in turn influence their mobility and independence.[46] Usually they need more rest periods and longer recovery time after exercise.[46] In addition, IPF patients also report higher degrees of fatigue, exhaustion, anxiety, depression and fear.[5,45]

There is a large heterogeneity in the evaluation of HRQOL in response to exercise training in IPF studies. The Saint George’s Respiratory Questionnaire (SGRQ) and the Medical Outcome Short-Form (SF)-36 are the most common instruments that have been used, and recently were also validated with the MCID to assess improvement in HRQOL among IPF patients.[47–49] The MCID in IPF patients for the SGRQ was set to 5–8 points and to 2–4 points for the SF-36.[48] Several studies using SGRQ have shown significant improvement above the MCID (ranging from −5 to −11 points; a decrease in score denotes improvement) following supervised exercise training programs.[22,23,30,38,50] A few studies have also demonstrated enhancement above the MCID using SF-36 with a large distribution effect.[31,35]

Yorke et al. developed an IPF-specific version of SGRQ.[51] However, to the best of our knowledge, this new instrument is yet to be implemented in exercise-based studies and its utility should be examined in future exercise training trials. The chronic respiratory disease questionnaire (CRDQ) has been used in one of the landmark randomized controlled studies of exercise training in ILD.[37] The CRDQ showed significant improvement in QOL, dyspnea and fatigue domains among ILD and IPF patients,[37] although to the best of our knowledge no MCID for CRDQ in IPF was established. Most studies showed a beneficial effect of exercise training in improving HRQOL in IPF patients,[22,23,30,31,33,35,37,38,50] although a few have reported no enhancement,[16,32,34,36] possibly due to differences in patient characteristics, training program modalities and nonspecific QOL instruments for IPF.

Dowman et al. [40] recently published a Cochrane systematic review on pulmonary rehabilitation for ILD and subgroup of IPF. Their meta-analysis included five studies (86 participants who undertook pulmonary rehabilitation and 82 control participants).[40] The results for IPF patients showed significant improvements in 6MWD (36 meters, 95% CI 16 to 55), peak aerobic capacity (1.5 ml/kg/min, 95% CI 0.5–2.4), reduced dyspnea (−0.7 units, 95% CI −1.1 to −0.3) and improved QOL, (0.59 units, 95% CI 0.1–1). [40] These findings are consistent with the previous Cochrane review demonstrating clinical benefits of exercise training for ILD and IPF.[24]

**Skeletal muscle strength and endurance**

Only a few studies on exercise training in IPF patients have looked at the strength and endurance of peripheral muscles.[22,36] Arizono et al. showed significant improvement of hand grip and quadriceps strength using a dynamometer following 10 weeks of a pulmonary rehabilitation program.[36] Our group demonstrated significant improvement in a 30-sec-chair-stand-test (representing the functional strength-endurance capacity of lower limb muscles) after a 12-week exercise program.[22] Future studies need to address the muscular fitness component more extensively among IPF patients, due to its strong association with functional capacity and activity of daily living.[21,52]

**Physical activity**

Physical inactivity is identified by the World Health Organization as the fourth leading risk factor for global mortality.[53] Approximately 31% of adults are estimated to be physically inactive, contributing to 6% of all deaths.[53,54] In
2008, physical inactivity alone caused 5.3 million premature deaths worldwide.[55] IPF manifests in severe signs and symptoms especially during physical exertion which is, in turn, associated with inactivity in order to avoid these episodes.[3,5] Inactivity among chronic respiratory disease patients is associated with poorer outcomes including higher mortality risk.[13] Nonetheless, the effect of participating in supervised exercise training or pulmonary rehabilitation programs on physical activity was not studied extensively among patients with IPF. Only two prospective studies examined the effect of short-term pulmonary rehabilitation programs on physical activity levels in IPF, with some conflicting results in the follow-up outcomes.[38,50] Both studies reported short-term improvement following the interventions; however, at 6-month follow-up Gaunaud et al. [50] in a RCT showed deterioration in physical activity levels assessed by the International Physical Activity Questionnaire. Both studies have significant limitations in the accuracy of assessing physical activity by using subjective self-report questionnaires rather than more objective electronic devices. In addition, it is also possible that some bias effect exists in Ryerson’s results due to the lack of a control group.[38]

Wallaert et al. [56] using accelerometers for step counting demonstrated a 65% lower daily life physical activity level in patients with fibrotic idiopathic interstitial pneumonia compared to healthy sedentary controls.[56] Moreover, this study also showed that among fibrotic interstitial pulmonary patients, physical activity <3287 steps/day was associated with poorer prognosis and an almost three times higher risk for death (hazard ratio = 2.72).[56] Recently, Nakayama et al. [57] showed that disease severity as measured by blood biomarkers, the extent of honeycombing on high-resolution computed tomography, 6MWD and MRC dyspnea scale scores were associated with physical activity levels among 31 stable IPF patients.[57] More studies using accurate, objective, electronic instruments are warranted to ascertain the short- and long-term effects of participating in supervised exercise programs on physical activity levels in IPF patients.

**Predictors of benefit and response to exercise program**

Several authors working with rehabilitation of IPF patients attempted to identify predictors of benefits from exercise training programs.[30,58] It seems there is agreement with respect to the group of patients who benefit the most from these interventions.[30,58] Holland et al. [58] reported a greater improvement in mild disease severity among 25 IPF patients out of a group of 44 ILD patients based on FVC, level of desaturation and right ventricle systolic pressure.[58] This notion aligns with Kozu’s et al. [30] findings among 65 IPF patients, demonstrating greater improvement in patients with mild to moderate MRC dyspnea compared to severe and very severe levels.[30] In contrast, Ryerson et al. [38] showed higher prerehabilitation functional capacity (as measured by a higher 6MWD) was associated with a lower improvement rate of this outcome in ILD and IPF patients.[38] This inconsistency may be related to methodological limitations and bias due to lack of intention to treat analysis for noncompleters and nonsurvivors in the latter study.[38] In addition, a large variation in response and adaptation to exercise training exist among IPF patients (Figure 1), and future exercise interventions should explore this issue more deeply.

**Methodology, program modalities and training types**

There is a large variability in exercise program modalities, research methodology and training types in IPF studies (Table 1). Although most studies conducted supervised outpatient-based programs,[16,22,23,30,32,34,36–38] few conducted home-based,[31,33,41] inpatient [35,41] or combined [30,41] programs. Most studies were prospective cohorts [16,22,23,30–33,36–38,41] and several were RCTs, [16,22,23,37] while few were retrospective.[34,35] In the majority of these studies, training programs combined aerobic exercise (walking or cycling or both) with resistance and flexibility exercises for peripheral skeletal muscles.[16,22,23,30–32,34–37] Some programs also included respiratory muscle training or breathing exercises.[22,31,34,36,41]

Despite the fact that all these studies have reported some improvement in IPF patients following the exercise training period, inconsistency still exists with respect to the magnitude of improvement and different measures that have been used across studies.[16,22,23,30–38,41] Furthermore, most of these studies adopted the established COPD guidelines of exercise training in the pulmonary rehabilitation program. These guidelines might be less appropriate for IPF due to the different pathophysiological mechanisms of the exercise limitations, and therefore might consequently not deliver optimal exercise stimuli and adaption to training.[13,20] Recommended training variables such as frequency, intensity, type and time are unknown yet and also needs further investigation in large randomized controlled studies to optimize the exercise training programs for IPF.

**Long-term effect of exercise training**

Only few studies in IPF patients have conducted a follow-up reassessment after a completion of a short-term exercise program, and report some inconsistent results.[37,38,59] While Holland et al. [37] and Vainshelboim et al. [59] showed nonsustained outcomes of exercise capacity, dyspnea and QOL at 6 and 11 months, respectively, Ryerson et al. [38] in contrast demonstrated preserved 6MWD, QOL, depression and physical activity levels at 6 month follow-up after 6–8 weeks of a pulmonary rehabilitation program.[38] Again this inconsistency is possibly related to methodological weaknesses in the Ryerson’s study as discussed above.[38] Interestingly, in Vainshelboim’s [59] study, despite a deterioration in 6MWD, V02peak, peak power and anaerobic threshold at 11-month follow-up, leg strength and QOL improvements were still preserved.[59]
Possibly a detraining effect in patients with IPF behaves differently across physiological and perceived outcomes, as was previously reported in young athletes, the elderly and COPD patients.[17,60–62] This issue too, should be ascertained in future research and maintenance of improvement strategies are warranted for IPF patients.

**Issues for consideration in the exercise program**

To the best of our knowledge no specific exercise guidelines exist for IPF.[20] Thus, issues presented in this chapter are based on observational reports of the published exercise training studies in IPF,[16,22,23,30–38,41] the pulmonary rehabilitation documents of several leading respiratory organizations [13,17,18] and our clinical experience and opinion. In general, exercise training seems to be safe and beneficial for IPF patients;[40] however, several issues should be considered in the management of exercise training programs in IPF (Table 2).

Since patients with IPF tend to have accompanied comorbidities, such as coronary arterial disease, systemic and pulmonary hypertension, severe dyspnea and desaturation and arrhythmias during exercise,[1,3,4,12] it is important that baseline evaluation should be conducted. This may include a pulmonologist assessment, pulmonary function test and cardiopulmonary exercise testing to ensure the patient’s safety before entering an exercise program.[13,63] Furthermore, IPF patients present exercise intolerance due to multifactorial limitations, including dyspnea and desaturation induced by physical demand.[11,12] It would be valuable to assess directly the electrocardiographic, hemodynamic and respiratory and gas exchange responses to evaluate the physiological limitations and the lowest level of oxygen saturation during incremental cardiopulmonary exercise testing. This would be in order to optimize personal exercise prescriptions and perform appropriate adjustments such as oxygen supplementation during the exercise sessions.[13]

Oxygen supplementation seems to be necessary for hypoxicemic IPF patients during exercise, as was recommended by the ATS/ERS guidelines.[1] To the best of our knowledge, in IPF there is no cutoff point of desaturation for oxygen delivery, although a threshold of SpO$_2$ > 90% was recommended for appropriated oxygenation in ILD,[39] based on COPD data,[64] Vainshelboim et al. [22] and Holland et al. [37] reported a threshold of SpO$_2$ < 88% and <85%, respectively, for oxygen supplementation during their exercise training sessions. The latter thresholds are probably more practical since IPF patients desaturate to much lower values compared to COPD patients.[65]

**Expert commentary and future research directions**

IPF is a chronic, progressive deadly lung disease usually presenting a poor prognosis.[1] We believe that it is important to address with large prospective trials whether rehabilitative exercise training can slow the progression of or change IPF’s clinical course. Especially, knowledge whether exercise interventions can reduce the rate of exacerbations and mortality among patients with IPF will be highly valuable. We are entering into a new era where IPF patients now have access to promising pharmacotherapy that has been recently approved by the Food and Drug Administration.[7–9] These drugs have shown benefits in slowing disease progression and increasing survival.[7–9] Patients’ lives can now be prolonged, meaning exercise and functional capacity and QOL have become even more important issues in order to maintain for longer the ability to cope with activities of daily life. Furthermore, in our opinion, it is valuable to deliver the rehabilitative exercise training as early as possible in IPF’s disease course and to explore the optimal training modality for this group of patients. This is in order to more effectively target IPFs compromises and to maximize the improvement and maintenance of outcomes. In addition, a combination of the new pharmacotherapy with rehabilitative exercise training

### Table 2. Issues for consideration in exercise program in idiopathic pulmonary fibrosis patients.

<table>
<thead>
<tr>
<th>Issue for consideration</th>
<th>Safety</th>
<th>Efficacy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baseline assessment using cardiopulmonary exercise testing</td>
<td>Arrhythmias, ischemia and abnormal hemodynamics</td>
<td>Direct measure of aerobic capacity</td>
</tr>
<tr>
<td>Desaturation during exercise</td>
<td>Monitoring of SpO$_2$ &gt;85–88%</td>
<td>Oxygen supplementation Interval training-resistance training with adequate time for resaturation between the exercise bouts</td>
</tr>
<tr>
<td>Cardiac signs:</td>
<td></td>
<td>Further evaluation searching for reasons</td>
</tr>
<tr>
<td>Arrhythmias:</td>
<td>Termination of exercise</td>
<td></td>
</tr>
<tr>
<td>Ventricular tachycardia, multifocal PVCs or triplets of PVCs, supraventricular tachycardia, heart block, new bundle branch block, or bradycardia.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypotensive response: drop in SBP &gt;10 mmHg from baseline</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypertensive response: SBP &gt;250 mmHg or DBP &gt;115 mmHg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Symptoms:</td>
<td>Termination of exercise</td>
<td>Further evaluation searching for reasons</td>
</tr>
<tr>
<td>Chest pain</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dizziness</td>
<td></td>
<td></td>
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<tr>
<td>Syncope or near syncope</td>
<td></td>
<td></td>
</tr>
<tr>
<td>DSB: Diastolic blood pressure; SBT: Systolic blood pressure; SpO$_2$: Oxygen saturation measured by pulse oximeter; PVC: Premature ventricular contraction.</td>
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</tr>
</tbody>
</table>
Exercise training in idiopathic pulmonary fibrosis

Key issues

- Idiopathic pulmonary fibrosis (IPF) is chronic deadly lung disease with few effective therapeutic options.
- IPF manifests in pulmonary restriction, dyspnea, hypoxemia, exercise intolerance and poor quality of life.
- Exercise training studies in IPF showed clinical improvement in the short-term; however the benefits are usually not sustained after exercise program cessation.
- Exercise training improves 6-min walking distance ranging from 15 m to 81 m in IPF.
- Exercise training may reduce dyspnea, increase muscular fitness and enhance quality of life in IPF.
- Participating in a supervised exercise program can increase home-based physical activity in IPF.
- It is possible that at early stages of IPF with mild to moderate disease severity, patients respond better to exercise training interventions compared to more advanced or severe disease conditions.
- Optimal training modalities for IPF patients are yet to be established.
- Meticulous evaluation before starting the program and consideration during exercise sessions are important.

References

Papers of special note have been highlighted as:

• of interest
•• of considerable interest


** Recent American Thoracic Society/European Respiratory Society position statement on pulmonary rehabilitation.


** This is an important review paper of pulmonary rehabilitation in idiopathic pulmonary fibrosis.


** This is an important randomized controlled study of pulmonary rehabilitation in idiopathic pulmonary fibrosis.


** This is an important randomized controlled study of pulmonary rehabilitation in idiopathic pulmonary fibrosis.


** This is an important systematic review paper of exercise training in interstitial lung disease patients.


** This is an important randomized controlled study of pulmonary rehabilitation in interstitial lung disease.


** This is a recent systematic review paper of pulmonary rehabilitation in interstitial lung disease.
