

Case Report

Pulmonary rehabilitation in pulmonary alveolar proteinosis: a case report

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ABSTRACT

Pulmonary alveolar proteinosis is a rare lung disease in which surfactant accumulates in the alveoli causing impairment of gas exchange, pulmonary circulation, restrictive lung functions, respiratory and muscular dysfunction. Pulmonary rehabilitation is a core aspect in the management of patients with chronic respiratory diseases. However so far there are no documented studies on the effects of pulmonary rehabilitation in pulmonary alveolar proteinosis. Here authors report the case of a 20 year old female diagnosed with pulmonary alveolar proteinosis and having considerable activity limitations. Pulmonary rehabilitation program of 24 weeks was designed for her after a thorough assessment. The 6-minute walk distance (6 MWD), forced vital capacity (FVC), diffusion capacity for carbon monoxide (DLCO), exercise induced desaturation (SPO₂) and QOL were evaluated using the SF-36 questionnaire. All parameters were recorded pre, post and at a follow up at 6 months after pulmonary rehabilitation. The 6MWD, FVC, DLCO, exercise induced desaturation and QOL improved considerably after 24 weeks of PR. At a follow up at 6 months all measures expect the FVC remained maintained. The authors thus conclude that Pulmonary Rehabilitation is effective in improving the exercise capacity, lung functions and quality of life in pulmonary alveolar proteinosis.

Keywords: Exercise capacity, Pulmonary alveolar proteinosis, Pulmonary rehabilitation, Quality of life

INTRODUCTION

Pulmonary alveolar proteinosis (PAP) is an ultra-rare disease in which surfactant accumulates in the alveoli due to impaired surfactant production and/or metabolism.¹ PAP can present with gradual increasing dyspnea, dry cough, fatigue, muscle weakness and functional limitations which cause restrictions of daily physical activity, anxiety and depression. The quality of life (QOL) deteriorates considerably. Exercise limitation is mainly because of impairment of gas exchange, pulmonary circulation, restrictive lung functions, respiratory and muscular dysfunction.

Whole lung lavage (WLL) continues to be the current standard of care for PAP. This approach allows physical

removal of proteinaceous material and local GM-CSF antibodies. The mean duration of response is 15 months, and 15% of patients need lavage every 6 months for maintenance of symptoms.¹

The ATS/ERS defines Pulmonary rehabilitation (PR) as a comprehensive intervention based on a thorough patient assessment followed by patient-tailored therapies that include, but are not limited to, exercise training, education, and behavior change, designed to improve the physical and psychological condition of people with chronic respiratory disease and to promote the long term adherence to health enhancing behaviors.² There is ample evidence that PR benefits the large population with COPD.^{2,3} To the best of our knowledge there is no documented study on the effects of PR in PAP. The aim of this case report is to highlight the effectiveness of

pulmonary rehabilitation that authors found on exercise capacity, lung functions and quality of life in a patient with Pulmonary Alveolar Proteinosis.

CASE REPORT

This case report is made in accordance with the CARE guidelines. A 20-year-old female, student by profession presented with a long- standing history (4-5years) of dry cough and progressive dyspnea on exertion. She gave a history of pulmonary tuberculosis in 2013. She was diagnosed as a case of pulmonary alveolar proteinosis in 2015. Chest radiographs demonstrated bilateral alveolar infiltrates. CT scan of thorax showed diffuse ground glass appearance bilaterally with interlobular septal thickening. Anti GM-CSF antibody testing on bronchoalveolar lavage (BAL) fluid confirmed autoimmune PAP.

Patient had undergone four sessions of BAL in an interval of 6 months each. The patient did not have comorbidities or any form of addictions. No form of rehabilitation was incorporated in her treatment earlier.

She came to the physiotherapy department with the chief complaints of breathlessness on walking for 2-3 minutes, climbing the stairs, doing ADLs like bathing, dry cough which increased during activities and fatigue almost throughout the day. She did not go to college, had limited social interactions with her friends and no indulgence in any form of hobby or sport.

Her expectations from physiotherapy were to achieve improved muscle strength and endurance, limit her breathlessness and anxiety such that she would be able to do all her ADLs independently and live a near normal social life.

On general examination, she was tachypnoeic RR 29/min, HR 79 bpm, SpO2 89-90% on room air, BP 110/80 mm Hg and BMI 23.8 kg/m². On observation she had overuse of accessory muscles for breathing and inverse I: E ratio. On auscultation there were crackles bilaterally and the air entry was found to be reduced at all levels.

The 6 minute walk distance (6MWD), Forced vital capacity (FVC) and diffusion capacity for carbon monoxide (DLCO) were measured in view of a restrictive lung defect. Eight domains of QOL were evaluated using the SF-36 questionnaire.⁴

A single point ‘Health change’ perception was also recorded. During the session, heart rate and SpO2 were monitored continuously.

The optimal exercise prescription for people with PAP is not yet known. Previous studies of pulmonary rehabilitation in with chronic respiratory disease other than COPD have used exercise prescriptions similar to those commonly employed in COPD.^{2,5}

The detail components of the PR program are summarized in Table 1.

Table 1: Pulmonary rehabilitation program.

Durartion	24 weeks of supervised exercise sessions.
Frequency	3 times a week.
Time	60-90 minutes per session.
Intensity	80% of the 6-MWT speed and increased each week by 0.25-0.5 km per hour, target rate of perceived exertion during training was modified Borg scale range of 4-6. 1-2% incline was added weekly. Muscular strengthening consisted of one set of 10 repetitions (60-70% of one-repetition maximum) in each major muscle group.
Type	Endurance training (treadmill, stationary bikes, arm ergometry) and resistance training (light weights, resistance bands) for major muscle groups on alternate sessions. Breathing training consisted of breathing techniques (controlled and diaphragmatic breathing), pacing and energy conservation. Inspiratory muscle training (IMT) and Incentive Spirometry. Patient education included , nutrition, panic control and relaxation techniques, as well as psychosocial support to take up a vocational course etc.

All parameters were recorded pre, post and at a follow-up at 6 months after pulmonary rehabilitation (Table 2 and 3).

Table 2: Outcome measures.

Outcome measures	Pre-intervention	Post-intervention	Follow up after 6 month
6MWD (m)	375	410	390
FVC (L/min)	1.63	1.69	1.59
DLCO (%)	33	42	39
SPO2 (during exertion)	94-83%	97-91%	96-88%

This case study demonstrates that pulmonary rehabilitation has beneficial effects in PAP. The parameters are summarized in Table 2. Authors found an increase in FVC at the end of 6 months of PR. This improvement however was not maintained at the 6 months follow- up. Improvements in the exercise capacity (6MWD), the diffusion capacity (DLCO) and exercise induced desaturation (SpO2) were also found at the end of the PR. These improvements remained maintained and did not return to baseline at 6 months follow-up. Most importantly the quality of life of the patient improved considerably (Table 3). At the end of rehabilitation program the patient reported improved confidence, took up a vocational course and reported better social interactions with family and friends.

Table 3: SF36 scores % change from post PR readings.

Domains	Pre PR (%)	Post PR (%)	% change from pre PR readings	Follow up after 6 months (%)	% change from post PR readings
Physical functioning	15	65	+50.00	70	+15.00
Role limitations due to physical health	0	50	+50.00	75	+25.00
Role limitations due to emotional problems	0	100	+100.00	100	±00.00
Energy/Fatigue	25	50	+25.00	55	+5.00
Emotional well-being	32	60	+28.00	60	±00.00
Social functioning	38	75	+37.00	63	-12.00
Pain	45	78	+33.00	68	-10.00
General health	45	55	+10.00	58	+3.00
Health change	50	100	+50.00	100	±00.00

DISCUSSION

The rationale for PR remains the same in non-COPD as for COPD.⁶ Aerobic exercises are known to strengthen peripheral and respiratory muscles, increase the rate and depth of respiration, reduce the airway resistance and motivate the subject to take deep inspiration and fill all air passages. The training induces an increase in aerobic enzyme levels and oxidative capacity of respiratory musculature contributing to enhanced ventilator muscle function.^{2,7} These findings were supported by Moazami M, et al, who stated that, aerobic exercise causes a significant increase in FVC due to increase in the strength and endurance of the respiratory muscles. Breathing exercises promotes a more efficient breathing pattern and improvement in ventilation.

Inspiratory muscle training (IMT) represents one component of PR. The rationale behind IMT is that enhancing respiratory muscle function can potentially reduce the severity of breathlessness and improve exercise tolerance.³ In line with the present study Chatham et al, studied the effect of IMT in a 43 year old person with deteriorating lung function for 8 weeks and found improvement in FVC from 0.85 to 1.21.⁹

Incentive spirometry increases the transpulmonary pressure through slow deep maximal inspiration thus increasing tidal volume and decreasing respiratory frequency.¹⁰ These effects facilitate increase in FVC.

An increase of 35 meters in the 6 MWD was noted. PR improves aerobic capacity, cardiac conditioning, quadriceps force thus making it fatigue resistant, and enhances the efficiency of skeletal muscle function at the cellular levels.^{2,11,12} All the above said effects translate to improvements in exercise capacity. Similar to the findings of this study Salhi et al, studied the effects of PR in RLD and found that the 6MWD improved significantly after 12 weeks and further improvements were noted after 24 weeks of PR in patients with RLD.¹²

In healthy subjects gaseous exchange improves with exercise in response to the increase in pulmonary blood flow, increased cardiac output, enlarged pulmonary capillary network, lung expansion, increased pulmonary hematocrit and homogeneous distribution of erythrocytes among capillaries.^{6,13} This helps to increase the SpO₂ and DLCO. In his study Hanson et al, reported DLCO improved by 9% after nine weeks of training.¹⁴ On the contrary Reuschlein et al, showed that five months of training failed to increase DLCO measured at rest or during exercise.¹⁵ Decreased diffusing capacity in PAP is accounted for by filling of the alveolar space with the proteinaceous fluid. With limited evidence available to justify the increase in the DLCO merely on exercise we attribute the improved diffusing capacity to the combined effects of WLL and exercise.

Improvements in the QOL after PR are noted in Table 3. The reliability of each subscale using this scoring system is between 0.93 (physical functioning) and 0.78 (general health and pain).^{4,16} The scores of all 8 domains of the SF-36 questionnaire changed positively over the course of the study. The highest percentage change (+100) was in the area of role -emotional. The general perception of health change also showed an improvement after PR. At follow up after 6 months, the improvements in 6 out of 8 domains remained consistent. PR has psychosocial benefits that help patients understand their disease and may mitigate anxiety and depression.¹¹ Supporting the present study findings several other studies have shown improvements in QOL after PR.^{3,5,11,12}

The duration of rehabilitation plays an important role. A longer PR program may assist with maintenance of benefits.⁶ Supporting the findings of the present study were the findings of Salhi and colleagues who identified greater benefits among their subjects, who completed 6 months of training.¹²

The outcome of 24 weeks of PR provides support to the use of PR alongside WLL which is currently the standard treatment for PAP. The results however cannot be

generalized and further research on a larger sample size may be considered. Despite robust evidence for the benefits of PR in chronic respiratory diseases, this field continues to evolve. Important issues still to be addressed include the ideal format and content for PR, methods to extend duration of benefits and how PR could best be tailored to the complex needs of people with pulmonary alveolar proteinosis.

CONCLUSION

This case study concludes that pulmonary rehabilitation is effective in improving the exercise capacity, lung functions and quality of life in Pulmonary Alveolar Proteinosis.

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