International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Effectiveness of Pulmonary Rehabilitation in a Patient with Idiopathic Pulmonary Fibrosis: A Case Study

Isha Shripad[1], Razia Nagarwala[2], Ashok K. Shyam[3], Parag K. Sancheti[4]

[1]MPT, [2]MPT, HOD and Professor,
Sancheti Institute College of Physiotherapy, Pune, Maharashtra, India.
[3]MS.Ortho, Research Officer, [4]MS.Ortho, Chairman,
Sancheti Institute for Orthopedics and Rehabilitation, Pune, Maharashtra, India.

Corresponding Author: Razia Nagarwala

ABSTRACT

Interstitial lung disease (ILD) is an umbrella term which consists of variety of disorders. ILDs are characterized by impaired gas exchange function and restricted pattern on lung function physiology testing with associated radiological findings. Lung damage occurring in most ILDs is progressive and irreversible Idiopathic Pulmonary Fibrosis (IPF) is one of the commonest ILD encountered in practice. The symptoms experienced by the patients in most interstitial lung disease are cough, breathlessness or shortness of breath, limited functional capacity. Here, we identified a case study of Idiopathic Pulmonary Fibrosis for which a tailor-made pulmonary rehabilitation program was introduced. The 8 week pulmonary rehabilitation program showed significant improvement in the aerobic capacity and quality of life of the patient.

Keywords: interstitial lung disease, idiopathic pulmonary fibrosis, pulmonary rehabilitation

INTRODUCTION

Interstitial Pulmonary Fibrosis (IPF) is defined as a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, limited to the lungs, and associated with changes in the histopathology and radiologic pattern bilateral, patchy, peripheral, and basilar predominant disease with reticulonodular infiltrates, often with honeycomb and cystic changes of Usual Interstitial Pneumonia (UIP). [1] IPF is diagnosed by- a] excluding other known causes of interstitial lung disease (ILD) e.g., domestic and occupational environmental exposures, connective tissue disease, and drug toxicity, b] the presence of a UIP pattern on high-resolution computed tomography (HRCT) in patients not subjected to surgical lung biopsy, c] HRCT and surgical lung biopsy pattern. A gradual lung function worsening over years is seen in most of the patients while few remain stable. The patients with IPF demonstrate increase in respiratory symptoms with decline in PFT, progressive fibrosis on HRCT, acute respiratory decline or death. These patients with IPF may have co-morbid conditions including pulmonary arterial hypertension, gastro-oesophageal reflux disease, obstructive sleep apnoea, obesity, and emphysema. The impact of these conditions on the outcome of patients with IPF is unclear. [2]

Case Description

A 29 year old male, operating designer (desk job) by occupation, visited OPD with dry cough, progressive dyspnea on exertion from grade 1 to grade 3 (mMRC scale) with no known allergies and negative family history. He was diagnosed as a case of clinical schizophrenia in 2007 and is on regular medications (Rexipra, Olizet) since then. He was asymptomatic till, when in 2012, patient started experiencing shortness of breath on exertion. Many clinics were visited for the same complaints and symptomatic relief was found. Standard chest X-ray investigation was done in 2012 which did not show any significant change. Over the years, he started experiencing increased episodes of breathlessness but all these complaints were correlated to his clinical psychological condition and were not addressed appropriately for a very long time. Again in June 2018, he experienced progressive increase in breathlessness and limitation in carrying out his activities of daily living and was started with oral steroids and home oxygen therapy (2 litres O_2 at night). Investigations were done following these episodes which consisted of Chest X-rays, CT Chest scan (plain and contrast), CT Thorax, USG, body plethysmography, DLCO (diffusing capacity of the lungs for carbon monoxide), blood investigations and 6MWT. HRCT and X-ray showed Usual Interstitial Pattern. CT thorax showed signs of diffuse bronchiolitis, bronchiectatic changes in inferior lingual and bilateral lower lobes, paraseptal emphysematous changes in bilateral upper and lower zones and excessive deposition of fat in right pleuropericardial space causing displacement of heart towards left. CT chest showed the presence of right sided Morgagni hernia with omental fat, defect measuring 1.5*4.2 cms was noted in anterior aspect of righthemidiaphragm with omental fat herniation. Herniated fat noted in anterior mediastinal and right pericardial region causing sub segmental atelectasis of underlying lung. Pulmonary function test report showed severe restriction, small airway obstruction with modest improvement in small and large airway function after inhalation of bronchodilator. DLCO single breath test showed 1.39% (predicted 10.72%). Body plethysmography report showed Total Lung Capacity of 2.39 which was 37.6% of predicted.

METHODOLOGY

The patient did not get the clearance for the surgical management of hernia owing to the lung condition and was decided to be managed conservatively. In functional assessment, resting HR- 93 bpm, RR- 26/min, SpO_2- 93% and BP- 130/90 mmHg. The tests done were 6 Minute Walk Test, [3] Lower limb endurance using 1-minute sit-to-stand test, [4,5] Maximum Inspiratory and Expiratory Pressures using MicroRPM™ device, [6] and A Tool to Assess Quality of life in Idiopathic Pulmonary Fibrosis (ATAQ-IPF) questionnaire. [7] In 6MWT, patient walked 100-meter distance. After 1st minute, patient became tachypnoeic and tachycardic and stopped the test. Patient covered 16.98% of his predicted 6MWD [(561.022 – (2.507*age in yrs) + (1.505* weight in kgs) – (0.055* height in cm)] [8] and did not complete the test successfully. Post-test: HR- 121 bpm, RR- 32/min, SpO2–78%and BP- 150/100 mmHg with recovery occurring in 30secs to HR- 95 bpm, RR- 26/min, BP- 140/90 mmHg and SpO_2- 92%. During 1 minute sit-to-stand test, patient performed 22 sit ups with heart rate 102 beats/min and $SpO_2$77%. Post 30 seconds and 1 minute of the test, the recovery occurred with heart rate going down to 95 beats/min and 86 beats/min and SpO_2 rising to 85% and 93% respectively. The patient demonstrated good lower limb strength and endurance. The MIP and MEP values were 89 (82.32%) and 103 (77.85%) cmH_2O respectively using age predicted formula [MIP = 120 - (0.41*age) and MEP = 174 – (0.83*age)]. [9] ATAQ-IPF questionnaire is a 13 section questionnaire dealing with different aspects of life. Higher the score, poorer is quality of life. Maximum and minimum scores are 370 and 74 respectively. Patient scored 258 out of 370.

The focus of pulmonary rehabilitation program was on improving aerobic capacity of the patient, strength of the respiratory muscles, peripheral muscles and quality of life. The intervention duration was 8 weeks, 6 days a week, 20 minutes per session twice a day. The program was structured as follows: diaphragmatic breathing (2-3 minutes), Anulom-Vilom pranayama (inhale through one nostril while the other is kept closed, then open the closed nostril and exhale through it keeping the other closed, repeat same with other nostril, this completes one cycle; 3 sets of 10 cycles), Suryanamaskar (Patient performed SN in a rapid manner so that all 12 postures were completed in 2 minutes; 3 cycles using 2 litres of O_2), breathing control (1-2 minutes), ventilator muscle training using Respironics™ Threshold IMT (Inspiratory Muscle Trainer) at 30% of MIP (10-15 repetitions/2 times a day). The progression was made after 4 weeks which included Anulom-Vilom pranayama (3 sets of 15 cycles), Suryanamaskar (5 cycles without hold) using 2 litres of O_2 and Threshold IMT at 40% of MIP (10-15 repetitions/2 times a day). After 6 weeks, O_2 usage was stopped while performing Suryanamaskar with same repetition. Post 8 weeks, readings were taken for 6MWT, MIP, MEP and ATAQ-IPF questionnaire to see the effect of pulmonary rehabilitation.

RESULTS

After 8 weeks of pulmonary rehabilitation, patient's resting heart was 82 beats/min, respiratory rate 14/min and SpO_2 97% at room air. Patient successfully completed the 6 MWT. Patient walked a distance of 256 meters (43.47% of predicted 6MWD) with 2 rest periods of 32 seconds and 46 seconds respectively; showed significant improvement in the walking distance by 156 metres (26.4%) with MCID. [10] MIP and MEP values showed improvement by 116 (107.29%) and 139 (92.70%) cmH_2O with a significant difference of 24.97% and 14.85% respectively. [11] In 1 minute sit-to-stand test, patient performed 27 sit -ups which showed significant improvement achieving MCID. [12] On ATAQ-IPF questionnaire, patient scored 215 out of 370 with improvement of 43 points (16% change).

Table 1: Pre- and Post- intervention parameters

Parameters	Pre-intervention	Post-intervention	% change
6 minute walk distance (6MWD)	100 meters	256 meters	60.93%
Maximum inspiratory pressure (MIP)	89 cmH_2O	116 cmH_2O	23.27%
Maximum expiratory pressure (MEP)	103 cmH_2O	139 cmH_2O	25.89%
1 minute sit-to-stand	22 sit-ups	27 sit-ups	18.51%

Table 2: ATAQ-IPF questionnaire dimensions

Dimensions	Patient pre-score	Patient post score
Cough	24/30	6/30
Shortness of breath	25/30	6/30
Planning and analyzing	25/25	25/25
Sleep	9/35	11/35
Mortality	20/25	16/25
Energy level	15/25	12/25
Mental and emotional well being	27/35	25/35
Ability to participate in social activities	22/25	21/25
Finances	24/30	24/30
Independence	22/25	22/25
Sexuality	5/25	5/25
Relationships with others	21/30	21/30
Therapies for IPF	19/30	21/30
Total score	258/370	215/370

DISCUSSION

The choices of exercises made for this patient were based on his clinical condition and were tailor made to meet his requirements. The results showed improvement in the aerobic capacity, ventilatory muscle strength and quality of life significantly.

MIP is the strength of the inspiratory muscles, primarily the diaphragm and allows assessment of respiratory muscles by a quick and non-invasive method, which is highly dependent on patient effort. [10] Respiratory muscle weakness is found to be an independent predictor of all cause mortality including other measures of pulmonary function test as well. [13] Suryanamaskar (SN) was performed sequentially in synchrony with breathing. Patient performed SN in a rapid manner, also called as Fast Suryanamaskar (FSN). Suryanamaskar has been found to be an effective modality in improving the patients. SN training is shown to improve both expiratory and inspiratory muscles strength. The different postures of SN involve isotonic contraction and chest wall expansion which may be improving strength of the intercostal muscles. This is in agreement with a study performed by Ananda et al which found significant increase in MIP with FSN than SSN. FSN had shown a significant effect on MIP and a statistically insignificant but appreciable increase in MEP. This can be attributed to the type of breathing and pattern of SN practiced. It is plausible that the slow and controlled inspiration and expiration, as practiced in SSN, contributed to significant increase in both pressures, whereas FSN resulted in increase in MIP alone as it was being performed rapidly and did not have controlled expiration phase. [14] Positive effects on levels of breathlessness and patients' ability to cope with breathlessness and satisfaction with the management of breathlessness and anxiety were found after 3 months of treatment. Changes in depression were evident from 8 weeks while quality of life indicators (fatigue, breathlessness mastery and emotional function) improved significantly from the first month of treatment suggesting more stable breathlessness experience with IMT.

One of the most important yogic practices which can produce different physiological responses in healthy individuals is breathing techniques. This art of prolongation and control of breath helps in reshaping breathing habits and bring awareness of conscious breathing. [15] During pranayama, the respiratory muscles are stretched to their full extent and the respiratory apparatus was able to work to their maximal capacity; the use of diaphragmatic and abdominal muscles more efficiently during the Anulom-Vilom. The 6 MWT is a submaximal, moderate-to-vigorous intensity testing which may be useful in the classification of aerobic fitness, which is associated with various health outcomes. The predictive value of the 6 MWT for estimating VO_2 max depends on the patient characteristics. It has important implications as it is a noninvasive and simple-to-use determinant of maximal aerobic power. [16] Increased aerobic capacity in this patient could be attributed to improved respiratory muscle function. Pranayama helps to reduce the relative load on the muscles and increase maximal sustained ventilatory capacity, lung volumes and vital capacity by its slow breathing; slow and deep breathing technique which is commonly used to decrease the work of breathing and accessory muscle activity; to increase the efficiency of breathing; and to improve the ventilation by its slow breath rate and more tidal volume. Additive effect is seen in terms of increased 6MWD after practice of this specific breathing technique. [17]

ATAQ-IPF questionnaire is a disease specific instrument to health related quality of life in patients with idiopathic pulmonary fibrosis (IPF). It is a multidimensional questionnaire which comprises of different domains of psychometric characteristics. [7] With improvement in all other functions, patient has reported improved quality of life with ATAQ-IPF questionnaire. This is in agreement with a review done by Dowman et al [18] which included 9 studies, 5 were included in the meta-analysis. Pulmonary rehabilitation improved the 6 minute walk distance significantly. Quality of life has improved following pulmonary

rehabilitation for all participants on a variety of reasons.

CONCLUSION

Pulmonary rehabilitation incorporating spiritual and physical domains of aerobic fitness, respiratory muscle strength and quality of life has been found to improve aerobic capacity and QOL in a patient with IPF.

ACKNOWLEDGEMENT

The authors of the present study thank the patient for the co-operation and consent for the conduction of the study.

REFERENCES

1. Hunninghake GW, Lynch DA, Galvin JR, Gross BH, Mu N, Schwartz DA, King Jr TE, Lynch III JP, Hegele R, Waldron J, Colby TV. Radiologic findings are strongly associated with a pathologic diagnosis of usual interstitial pneumonia. Chest. 2003 Oct 1;124(4):1215-23.
2. Raghu G, Collard HR, Egan JJ, Martinez FJ, Behr J, Brown KK, Colby TV, Cordier JF, Flaherty KR, Lasky JA, Lynch DA. An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management. American journal of respiratory and critical care medicine. 2011 Mar 15;183(6):788-824.
3. Guidelines of 6 minute six walk test, American journal of respiratory and critical care medicine 2002
4. Miranda S, Machado A, Melro H, Oliveira A, Valente C, Marques AS. Is the 1-minute sit-to-stand test related to respiratory muscle strength in patients with COPD? Conference: ERS International Congress 2017 abstracts
5. Diener MH, Golding LA, Diener D. Validity and reliability of a one-minute half sit-up test of abdominal strength and endurance. Research in Sports Medicine: An International Journal. 1995 Aug 1;6(2):105-19.
6. ATS/ERS Statement on Respiratory Muscle Testing, American journal of respiratory and critical care medicine 2002
7. Swigris JJ, Wilson SR, Green KE, Sprunger DB, Brown KK, Wamboldt FS. Development of the ATAQ-IPF: a tool to assess quality of life in IPF. Health and quality of life outcomes. 2010 Dec;8(1):77.
8. Ramanathan RP, Chandrashekharan B. Reference equations for 6-minute walk test in healthy Indian subjects (25-80 years). Lung India: official organ of Indian Chest Society. 2014 Jan;31(1):35.
9. Evans JA, Whitelaw WA. The assessment of maximal respiratory mouth pressures in adults. Respiratory care. 2009 Oct 1;54(10):1348-59.
10. Nathan SD, Du Bois RM, Albera C, Bradford WZ, Costabel U, Kartashow A, Noble PW, Sahn SA, Valeyre D, Weycker D, King Jr TE. Validation of test performance characteristics and minimal clinically important difference of the 6 minute walk test in patients with idiopathic pulmonary fibrosis. Respiratory medicine. 2015 ul 1;109(7): 914-22.
11. Veena Kiran Nambiar, Savita Ravindra, Maximal respiratory pressures and their correlates in normal Indian adult population, International journal of physiotherapy and research, 2015; 3(4):1188-96
12. Vaidya T, de Bisschop C, Beaumont M, Ouksel H, Jean V, Dessables F, Chambellan A. Is the 1-minute sit-to-stand test a good tool for the evaluation of the impact of pulmonary rehabilitation? Determination of the minimal important difference in COPD. International journal of chronic obstructive pulmonary disease. 2016; 11:2609.
13. Van der Palen J, Rea TD, Manolio TA, Lumley T, Newman AB, Tracy RP, Enright PL, Psaty BM. Respiratory muscle strength and the risk of incident

cardiovascular events. Thorax. 2004 Dec 1;59(12):1063-7.
14. Bhavanani AB, Udupa K, Madanmohan PN. A comparative study of slow and fast suryanamaskar on physiological function. International journal of yoga. 2011 Jul;4(2):71.
15. Patil YR, Sawant RS. Effect of bastrika pranayama on pulmonary function. Int J Pharm. 2012;3:204–7.
16. Burr JF, Bredin SS, Faktor MD, Warburton DE. The 6-minute walk test as a predictor of objectively measured aerobic fitness in healthy working-aged adults. The Physician and sports medicine. 2011 May 1;39(2):133-9.
17. Pramanik T, Pudasaini B, Prajapati R. Immediate effect of a slow pace breathing exercise Bhramari pranayama on blood pressure and heart rate. Nepal Med Coll J. 2010;12:154–7.
18. Dowman L, Hill CJ, Holland AE. Pulmonary rehabilitation for interstitial lung disease. Cochrane Database of Systematic Reviews. 2014(10).

How to cite this article: Shripad I, Nagarwala R, Shyam AK et.al. Effectiveness of pulmonary rehabilitation in a patient with idiopathic pulmonary fibrosis: a case study. Int J Health Sci Res. 2019; 9(12):74-79.

******