**RESEARCH PAPER**

# The Impact of a 12-Week Hybrid Pulmonary Rehabilitation Program on Quality of Life in Kartagener Syndrome – A Case Report

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**ABSTRACT**
A 32-year-old male presents with primary complaints of severe breathing and coughing. Following a chest examination and investigation, he was diagnosed with Kartagener syndrome. It is a rare inherited condition that falls under primary ciliary dyskinesia (PCD). It is characterized by chronic sinus infections, bronchiectasis, and a reversed orientation of internal organs (situs inversus). We implemented a structured pulmonary rehabilitation program, which included six weeks of inpatient rehabilitation followed by six weeks of telerehabilitation at home. In pulmonary rehabilitation, we focus on airway clearance techniques, including ACBT and Bottle PEP. The 6-Minute Walk Test (6MWT), Single Breath Count (SBC), Borg scale, and WHO Quality of Life questionnaire were used as outcome measures, and they were performed before and after rehabilitation. Results showed a significant 153-meter increase in walking distance, well beyond the minimal clinically important difference, as well as improved breathing capacity, less symptoms and improved quality of life. These findings highlight the potential of pulmonary rehabilitation to greatly improve daily function and overall well-being in people with Kartagener's syndrome.

**Keywords:** Pulmonary Rehabilitation (PR), Airway clearance therapy (ACT), Active Cycle of Breathing Techniques (ACBT), 6-Minute Walk Test (6MWT), WHO Quality of Life (WHO QOL), Single Breath Count (SBC)

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## INTRODUCTION

Kartagener's syndrome, also known as primary ciliary dyskinesia, is a rare autosomal recessive ciliopathic disorder. It is caused by congenital abnormalities of the primary cilia and shows extensive genetic heterogeneity. The disease is characterised by impaired ciliary motility. This leads to reduced mucociliary clearance and presents as a clinical triad of chronic sinusitis, bronchiectasis, and situs inversus. Bronchiectasis is a primary, heterogeneous disease that contributes to disease severity. It presents with a common triad of productive cough, dyspnoea and fatigue. These symptoms contribute to recurrent respiratory exacerbations and increased disease severity, and a poorer health-related quality of life (HRQOL). The ultrastructural genetic defect leads to recurrent chest, ear/nose/throat (ENT) and sinus infections. It also causes infertility [1, 2]. Pulmonary rehabilitation (PR) is an essential part of the management of bronchiectasis as per guidelines. Pulmonary rehabilitation is a structured program of exercise and education. It improves HRQOL and exercise capacity in people with bronchiectasis and reduces fatigue and dyspnoea. Airway clearance techniques are an important part of pulmonary rehabilitation in the management of bronchiectasis. These techniques facilitate sputum clearance, improve HRQOL and reduce the risk of acute exacerbations [3] Telerehabilitation is a substitute for conventional hospital-based pulmonary rehabilitation (PR) and resolves transportation problems and maintains the benefits of traditional care. Patients can obtain healthcare from the comfort of their own homes by using digital platforms and remote monitoring, which improves adherence and lowers expenses. Cardiorespiratory disorders are among the conditions for which telerehabilitation has shown potential to improve patient outcomes and reduce healthcare disparities [4].In this case report, we aimed to present a case of Kartagener's syndrome by emphasising the effects of pulmonary rehabilitation in improving QOL in a patient with Kartagener's syndrome.

**Case Presentation:**

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A 32-year-old male comes with chief complaints of breathlessness and cough. Over ten years, he also reported frequent bouts of nasal congestion, nasal pain, and productive cough. Since then, he has gone to the doctor several times for the same issue and has also taken medication. Over the past five years, he has observed periodic flare-ups of coughing with copious amounts of purulent sputum. He received treatment for the same symptoms, but there was no significant clinical

improvement after completion of 6 months of medication. Three months ago, he visited to ENT specialist who diagnosed him with chronic sinusitis and a nasal polyp. He was prescribed antibiotics and intranasal steroids for his condition. No one in his family had the same symptoms. Investigations at that time were done, but the results were not available. On physical examination, the patient was underweight with a body mass index (BMI) of 18.5 kg/m². His blood pressure (BP) was 116/74 mmHg, pulse rate (PR) 86 beats per minute, respiratory rate (RR) 22 breaths per minute, and temperature (T°) 37.5°C. Using 2 L/min of oxygen, his arterial oxygen saturation (SpO2) was 92% at room air. A respiratory system examination revealed coarse crackles on both basal lung fields and decreased chest expansion bilaterally. On cardiovascular examination, an apex beat was felt on the right fifth intercostal space along the midclavicular line, as also seen in Figure 1c. Heart sounds were best audible on the right side of his chest. Physical examination also revealed grade 2 digital clubbing, as seen in Fig. 1a. An evaluation of the neurological system revealed no abnormalities. Chest x-ray PA view showed evidence of blunting of the right costophrenic angle, heterogeneous opacity, and cardiac apex and aortic arch on the right side, suggesting dextrocardia Fig. 1c. HRCT thorax showed mild bronchiectasis changes in the bilateral lower lobes; mild right-sided pleural effusion was noted, as seen in Fig. 1b. We evaluated the functional capacity of the patient by using a 6-minute walk test. Additionally, we evaluated his pulmonary function using a single breath count, the Borg scale for dyspnea, and quality of life by using the WHO-QOL, which is below the normal range, as seen in Table No. 2

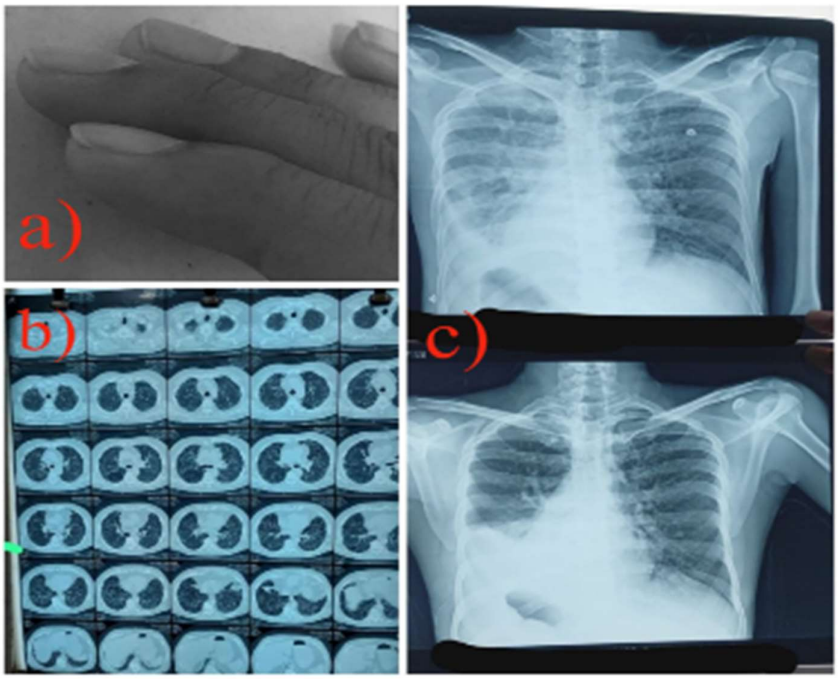


**Figure 1: a) Digital Clubbing, b) HRCT, and c) Pre-post Chest X-ray Presentations**

**Discussion :**

Patients with KS have reduced airway clearance capacity and lower aerobic capacity than healthy subjects, and repetitive episodes of infection ultimately lead to reduced overall quality of life. To manifest these complaints, we enrolled patients in pulmonary rehabilitation in a mix of settings, including hospital or conventional outpatient programs, home-based or telerehabilitation, with the exercise structure focused on mild- to moderate-intensity endurance and resistance exercise prescription, for a typical duration of 12 weeks [2].

During the 12 weeks of pulmonary rehabilitation, the patient underwent a hybrid approach. The first phase, involving six weeks of inpatient department (IPD) rehabilitation, was conducted at a hospital. Following this initial phase, the patient transitioned to six weeks of telerehabilitation, a remote approach using technology to continue the rehabilitation in the

comfort of the patient's home [3]. This phase likely included scheduled virtual sessions with therapists, guided exercise programs, and the use of monitoring tools to track progress and maintain engagement, as shown in Table No. 1.

**Table 1: Physiotherapy intervention**

| Sr.No | Intervention objectives | Clinical intervention | Frequency |
|---|---|---|---|
| 1. | Patient Education | 1. Understanding the aetiology of the condition and prognosis, including uncertainty<br>2. Airway clearance therapy – alternative options for techniques, frequency of review, and advice on cleaning of devices<br>3. Management anxiety, depression | On the first day of the session |
| 2. | To remove the secretion | 1. Active cycle of breathing techniques<br>2. Bottle PEP | Every 3 hours, 10 repetitions.<br>15 min |
| 3. | To improve lung capacity | 1. Diaphragmatic breathing exercise<br>2. Thoracic expansion ex<br>3. Incentive spirometer | Every 3 hours, 10 repetitions. |
| 4. | To reduce breathlessness | 1. Dyspnoea relieving position<br>2. Breathing exercise<br>3. Purse lip breathing | Every 3 hours, 10 repetitions. |
| 5. | To improve aerobic capacity | 1. Hall ambulation<br>2. Sit to stand<br>3. Stepping<br>4. Leg ergometer | F – 3 times/day<br>I – RPE 3-4/10<br>T- 10-15 min<br>T- stepping, walking, |
| 6. | To improve strength | 1. UL & LL strengthening ex. | F – 3 times/day<br>I - 40-80% of 1RM<br>T- 10-15 Repetition<br>T- stepping, walking, |

We started pulmonary rehabilitation with patient education, including education about the pathology and prognosis of the disease, and also explained the importance of exercise training in the disease. It involves a range of interventions to improve respiratory function and quality of life in individuals with chronic lung conditions. For people with bronchiectasis, airway clearance is essential to remove mucus and reduce the risk of infection. It includes Active Cycle of Breathing Techniques (ACBT), postural drainage, and the use of a bottle PEP device, which are commonly recommended in pulmonary rehabilitation to help manage bronchiectasis by clearing the airways, mobilising mucus, and reducing the risk of infection. The physiological rationale is that these techniques augment sputum clearance via a number of mechanisms: improvement in collateral ventilation and independence; increase in expiratory airflow velocity; reduction in airway resistance; use of gravity; changes in airway pressure; and generation of airway oscillations [3, 4].

**Figure 2: Patient Engaged in Telerehabilitation**

To improve ventilation, we started an incentive spirometer and breathing exercises, including diaphragmatic breathing, segmental breathing, thoracic expansion exercises, and positional breathing exercises. Breathing exercises can reduce the harmful physiological impacts of reduced lung function by improve the strength and endurance of the respiratory muscles, improve the mechanics of movement of the chest and abdominal wall, decreasing dynamic hyperinflation, and improving gas exchange [5, 6].

Due to the benefits of exercise to improve airway clearance and functional capacity in patients with bronchiectasis and chronic lung diseases [7, 8], we preferred to include aerobic exercise training in our patients' treatment program, like stair climbing, brisk walking and sit-to-stand with a set RPE scale of 3-4/10. Mostly in clinical practice, patients with bronchiectasis do not comply well with hospital-based pulmonary rehabilitation, primarily because of the inconvenience of transportation. A reliable home-based tele-rehabilitation program could emerge as the new standard for pulmonary rehabilitation in cases with bronchiectasis [9, 10]. According to a 2021 workshop on the future of pulmonary rehabilitation organised by the American Thoracic Society, telerehabilitation may help increase program accessibility and completion. The authors of the workshop noted that the terms "access," "uptake," and "completion" are crucial in describing the difficulties that pulmonary rehabilitation programs confront [11, 12].

Close monitoring in telerehabilitation is needed to monitor the safety of the patient and the success of the program. Real-time monitoring helps to detect any adverse response immediately and to ensure that the breathing exercises and aerobic exercises at home are within the recommended exercise intensity range of 60-80% of the maximum HR achieved during the 6MWT [10]. Continuous observation and feedback through remote methods ensure that patients perform exercises correctly and follow their rehabilitation plan safely from home, as shown in Fig. 2 [13, 14].

A study on tele-yoga and telerehabilitation for COPD patients showed improvements in 6-minute walk distance, anxiety, and depression. A meta-analysis further found that telerehabilitation is as effective and safe as centre-based programs for chronic respiratory diseases [15]. Evidence from a systematic review indicates that patient attendance and adherence to real-time telerehabilitation self-management programs are equal to or better than those observed with conventional in-person physiotherapy. After completing rehabilitation, the patient's well-being was improved, enabling him to return to work and participate actively in social activities, as reflected in the higher quality of life scores as seen in Table No. 2.

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**Table 2: Outcome Measure of Pre- and Post-rehabilitation**

| Sr. No. | Parameters | Prehabilitation (Baseline) | Post-rehabilitation (After 12 weeks) | Reference value |
|---|---|---|---|---|
| **1.** | **Spo2** | 92% at room air | 97% at room air | 100% |
| **2.** | **SBC** | 23 | 35 | 30-40 counts |
| **3.** | **BORG SCALE** | 6 | 2 | 0/10 |
| **4.** | **6 MWT** | 445m | 580m | 533 m |
| **5.** | **WHO QOL** | | | |
| | **D1** | 48% | 86% | 100% |
| | **D2** | 56% | 89% | 100% |
| | **D3** | 58% | 68% | 100% |
| | **D4** | 44% | 78% | 100% |

This is the first case report of a Kartagener's syndrome patient who attended Pulmonary Rehabilitation with mixed modes, including telerehabilitation. The overall result showed significant improvement in aerobic capacity, reduced breathlessness, and improved quality of life. In order to improve exercise capacity and alleviate symptoms, we recommend pulmonary rehabilitation for patients with Kartagener syndrome. (16)

**Conclusions:**

This case demonstrates that a hybrid pulmonary rehabilitation approach—combining inpatient care with telerehabilitation—can significantly improve functional capacity, respiratory symptoms, and quality of life in a patient with Kartagener's syndrome. Also, this case highlights the feasibility and effectiveness of telerehabilitation as a safe and accessible modality for improving functional outcomes and quality of life in patients with chronic respiratory conditions.

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