

Aerobic exercise training in Kartagener's syndrome: case report

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Kartagener's syndrome is a rare, autosomal recessive inherited disease, which is characterized by a triad of chronic sinusitis, bronchiectasis, and situs inversus. In this report, we aimed to represent the effect of aerobic exercise training in addition to chest physiotherapy in an outpatient with Kartagener's syndrome. An 18-year-old female diagnosed with Kartagener's syndrome applied with the complaints of productive cough and dyspnea with exertion and attended pulmonary rehabilitation program comprising exercise training in addition to standard treatment. Aerobic exercise training was performed three times weekly at 80% of the peak heart rate, for 8 weeks as supervised sessions. Respiratory physiotherapy and postural exercises were taught to the patient to be performed at home each day of the week. Before and

after pulmonary rehabilitation program, incremental shuttle walk test (ISWT) was performed; dyspnea and fatigue were assessed using Borg Scale. Pulmonary function test was evaluated using spirometer. The patient wore activity monitor for 7 consecutive days before and after training. After pulmonary rehabilitation program, 90-m increase in ISWT was observed. We recommend exercise training in patients with Kartagener's syndrome to increase exercise capacity. Further randomized control trial is needed to clarify the effects.

Keywords: Bronchiectasis, Exercise, Pulmonary rehabilitation, Respiratory disease


INTRODUCTION

Primary ciliary dyskinesia is a rare autosomal recessive inherited disease with the congenital abnormality of the primary cilia. Situs inversus totalis (complete mirror-image reversal of the thoracic and abdominal organs) is present in 50% of patients with primary ciliary dyskinesia (Leigh et al., 2009). The triad of situs inversus, bronchiectasis, and chronic sinusitis became known as Kartagener's syndrome. The dysfunction of ciliary motility leading to impaired mucociliary clearance. Accumulated secretions on the epithelial surface and following persistent or recurrent bacterial infections result in bronchiectasis. Respiratory physiotherapy involving the combination of airway clearance techniques and exercise is recommended to alleviate the sputum clearance and may suppress the further deteriorative impacts of diseases. There is no random-

ized study in the treatment of primary ciliary dyskinesia/Kartagener's syndrome. As the aim is to correct or maintain the normal lung function for all chronic respiratory diseases, all recommendations are derived from cystic fibrosis guidelines, even though it follows a different pathophysiology (Barbato et al., 2009).

Pulmonary rehabilitation in addition to regular chest physiotherapy was shown to improve exercise tolerance, and health-related quality of life in patients with noncystic fibrosis bronchiectasis and this benefit was sustained at 12 weeks post-end of pulmonary rehabilitation (Mandal et al., 2012). Pifferi et al. (2013) reported that up to date proper therapeutical approach in primary ciliary dyskinesia includes the use of respiratory physiotherapy and regular exercise to favor airways clearance and antibiotics administration to control acute airway infections.

In this report, we aimed to represent a case of Kartagener's syn-

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drome with showing the effects of aerobic exercise training in addition to chest physiotherapy in an outpatient with Kartagener's syndrome.

MATERIALS AND METHODS

Case history

An 18-year-old female Kartagener's syndrome diagnosed patient presented to our outpatient department with the complaints of chronic productive cough and progressive dyspnea with exertion.

Clinical history and demographic information of the patient

The patient had a past medical history of chronic sinusitis, bronchiectasis, and situs inversus totalis. She was born to consanguineous parents. She consulted to our department with the complaints of a productive cough with yellow sputum and dyspnea on exertion. She had no history of smoking. She was prescribed an inhaler (salmeterol and fluticasone propionate) to use twice daily by her doctor. The signed informed consent was obtained from the patient for the use of the information obtained in the assessment and the treatment process and for the publication of this case report.

Examination

Her anthropometric features were: body weight, 48 kg; height, 157 cm, and body mass index, 19.47 kg/m². Her vital signs were: heart rate (HR), 96 beats/min; respiratory rate, 20 breaths/min; oxygen saturation (SpO₂), 91%; blood pressure 110/80 mmHg. Auscultation of the chest revealed coarse crackles and diminished breath sounds over bilateral lung fields.

Pulmonary function assessment

Before incremental shuttle walk test (ISWT), pulmonary function was determined according to standard spirometry guideline (Miller et al., 2005). Forced vital capacity (FVC), forced expiratory volume at 1 sec (FEV₁) (expressed as % of predicted) and FEV₁/FVC were obtained.

Functional capacity assessment

The ISWT is an externally paced maximal exercise test that the walking speed is controlled using auditory signals and increases continuously until the participant can no longer continue (Cox et al., 2014). The distance completed by the patient was recorded throughout the test period. SpO₂ and HR were recorded before

and after the test using a portable pulse oximeter (Nonin Palmsat 2550A, Nonin Medical Inc., Plymouth, MN, USA). Before and after the test, blood pressure was measured. Modified Borg Scale was used to evaluate fatigue and dyspnea perception. The completed distance was expressed in meters and as ISWT% distance, taking the percentage of normal reference values (Probst et al., 2012).

Physical activity assessment

Physical activity level was determined objectively using SenseWear Armband (SWA) (SenseWear Armband Model MF-SW, BodyMedia, Pittsburgh, PA, USA). Our patient wore SWA for 7 consecutive days. She was informed to remove the device only when in contact with water, such as shower, swimming, etc., and then rewearing it after these kinds of activities. As described in the manual, the device was fitted by adjusting the band tightness relative to the arm of the patient, ensuring that the sensors were in contact with the arm, on the left arm over the triceps. She was asked to continue their daily life activities. The SWA was used, and its validity was tested in various chronic diseases such as chronic obstructive lung disease, cystic fibrosis, and cardiac patients (Cole et al., 2004; Cox et al., 2014; Hill et al., 2010).

Exercise training program

Exercise training consisted of a prescribed exercise program and included treadmill walking, with the intensity set to 80% of the peak HR achieved on the ISWT. Our patient participated aerobic exercise training three days of the week through 8 weeks. Monitoring of vital signs including SpO₂ and HR was ensured pre-, during, and postexercise training. Patient continued her standard treatment consisting of respiratory physiotherapy using oscillatory positive expiratory pressure device, postural exercises, and positions to reduce dyspnea to be performed at home.

RESULTS

Pulmonary function test parameters before and after exercise

Table 1. Pulmonary function test parameters before and after exercise training

Pulmonary function parameters	Before	After
FVC (%)	66.0	61.0
FEV ₁ (%)	51.0	48.0
FEV ₁ /FVC	69.3	66.0
PEF (%)	85.0	82.0
FEF _{25-75%} (%)	25.0	20.0

FVC, forced vital capacity; FEV₁, forced expiratory volume at 1 sec; PEF, peak expiratory flow; FEF_{25-75%}, forced expiratory flow at 25%–75% at vital capacity.

Table 2. Incremental shuttle walk test parameters before and after exercise training

Incremental shuttle walk test parameters	Before training		After training	
	Pretest	Posttest	Pretest	Posttest
HR (beats/min)	95	185	103	190
Maximum HR (beats/min)	185		190	
Maximum HR% (%)	91.58		94.05	
SpO ₂ (%)	95	95	96	87
Systolic blood pressure (mmHg)	90	170	110	160
Diastolic blood pressure (mmHg)	60	80	80	90
Dyspnea (Borg 0–10)	0	10	0.5	4
Quadriceps fatigue (Borg 0–10)	0	10	0.5	4
Fatigue (Borg 0–10)	0	10	1	4
ISWT distance (m)	540		630	
ISWT% distance (%)	52.94		61.76	

HR, heart rate; SpO₂, oxygen saturation; ISWT, incremental shuttle walk test.

training were shown in Table 1. The patient had evident airway obstruction according to the results before and after aerobic exercise training. The ISWT parameters were shown in Table 2. The parameters such as HR, SpO₂, blood pressure, and fatigue and dyspnea perception to ISWT before and after the exercise training were demonstrated in Table 2. The ISWT distance increased 90 m after training compared with the distance before aerobic exercise training. The physical activity assessment parameters were shown in Table 3.

DISCUSSION

Patients with primary ciliary dyskinesia manifest lower aerobic fitness than healthy subjects and their aerobic fitness correlate with pulmonary function and self-reported complaints of limitations in vigorous physical activity (Madsen et al., 2013).

The exercise was found to be a more potent stimulus for bronchodilation than by inhaled β_2 -agonists (Phillips et al., 1998). Considering the effects of exercise to enhance airway clearance and improve functional capacity in patients with bronchiectasis and chronic lung diseases, we were in favor of including aerobic exercise training for treatment program of our patient. This case report was the first report to demonstrate the effects of aerobic exercise training of a patient with Kartagener syndrome. Our patient exhibited increased exercise capacity and also showed decreased dyspnea and fatigue scores with exercise training, without presenting any sign of exacerbation.

The pulmonary function parameters of the patient after exercise training were similar to her baseline values. A 6-month super-

Table 3. Physical activity parameters before and after exercise training

Physical activity parameter	Before	After
Daily energy expenditure (kcal/day)	1,774	1,798
Daily physical activity duration (≥ 1.5 MET)	11.63 hr	8.11 hr
Lying down (hr/day)	9.28 hr	9.38 hr
Daily active energy expenditure (≥ 1.5 MET) (kcal/day)	1,051 kcal	912 kcal
Step counts (steps/day)	5,015	6,212
Sleep duration (hr/day)	7.88 hr	7.06 hr
MET	1.7	1.6
Distance (km/day)	2.9	4.3
Physical activity level index	1.4	1.5
Mean sedentary activity duration (< 1.5 MET)	12.06 hr	15.75 hr
Light physical activity duration (1.5–3 MET)	10.55 hr	6.56 hr
Moderate physical activity duration (3–6 MET)	1.25 hr	1.30 hr
Vigorous physical activity duration (6–9 MET)	7 min	14 min
Very vigorous physical activity duration (> 9 MET)	1 min	4 min

MET, metabolic equivalent.

vised training program was shown to improve lung function including FEV₁ and aerobic performance in patients with cystic fibrosis underlining the importance of physical activity (Kriemler et al., 2013). It was reported that the benefits of regular care plus exercise training might be influenced by the type and duration of the training program in patients with cystic fibrosis (Radtke et al., 2017). We did not observe such difference in our results probably due to relatively shorter time, eight weeks, of aerobic exercise training program.

The ISWT distance increased 90 m after training compared with the distance before aerobic exercise training. The 47.5 m was identified as the minimum clinically significant improvement for the ISWT in patients with chronic obstructive pulmonary disease (Singh et al., 2008). In the same study, 78.7 m was determined a higher level of perceived improvement. Our patient's result was higher than these.

We evaluated the physical activity level of a patient with a Kartagener Syndrome before and after aerobic exercise training. There is no evidence of physical activity level in this disease other than we provided. According to our findings, our patient showed a sedentary lifestyle, and her results were similar before and after exercise training. However, our patient achieved the required physical activity level, at least 150 min of aerobic activity or at least 75 min of vigorous aerobic activity, or the equivalent combination of both activities per week, which defined by World Health Organization's (WHO) recommendations for maintenance of cardiovascular health (World Health Organization, 2010). We believe exercise training may have a favorable role to adopt a relatively more active lifestyle.

Tudor-Locke and Bassett (2004) classified physical activity level < 5,000 steps/day as a sedentary lifestyle index; 5,000–7,499 steps/day as low active; 7,500–9,999 steps/day as somewhat active; and > or = 10,000 steps/day as active. The same study reported that 5,000–7,499 steps/day considered as low active which was inferred as typical of daily activity excluding sports or exercise (Tudor-Locke and Bassett, 2004). We can classify our patient as low active. The daily step count of our patient before training and after training was respectively 5,015 steps/day and 6,212 steps/day.

According to the findings, the aerobic exercise training program in addition to standard physiotherapy treatment for a patient with Kartagener syndrome was a success by improving exercise capacity, lowering perceived dyspnea and fatigue with exertion.

Respiratory muscle strength assessment and, if required, respiratory muscle strength training may be helpful for management of the disease. We were not able to provide this due to technical issues. However, respiratory training may have been confounding with the effects of individual aerobic exercise training. In this respect, evaluation and investigating the impact of respiratory training in this population may be considered.

This is the first case report of a Kartagener's syndrome patient who attended aerobic exercise training including the assessment of physical activity level. The functional capacity of patient increased after training. We recommend exercise training in patients with Kartagener syndrome to increase exercise capacity and relief the symptoms.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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