Case Report

A Combined Outpatient and Home-Based Rehabilitation Program for a Patient with an Open Atrial Septal Defect (ASD) and Severe Pulmonary Hypertension: A Case Report

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ABSTRACT

Objectives: This paper presents a rehabilitation intervention for improving functional capacity and physical activity level in a patient with an open atrial septal defect (ASD) with severe pulmonary hypertension contraindicated to ASD closure.

Study design: Case report.

Setting: Hasan Sadikin General Hospital, Bandung, West Java, Indonesia.

Subjects: A-37 year old female, an elementary school teacher, complained of shortness of breath when doing daily activities, walking, and speaking for three years. She was planned for ASD closure but could not be done because of severe pulmonary hypertension. The right heart catheterization showed pulmonary hypertension (mean Pap: 70 mmHg). In addition, the CT-Scan showed cardiomegaly with pulmonary hypertension and pulmonary fibrosis.

Methods: A combined outpatient and home-based comprehensive rehabilitation program including cardiopulmonary endurance exercise with a treadmill, breathing exercises (controlled breathing and deep breathing), and sustained maximum inspiration technique were prescribed to reduce fatigue and dyspnea on exertion.

Results: Eight weeks after following the program, there were improvements in cardiorespiratory fitness (from 2.92 to 5.27 METs), maximum inspiratory volume (from 1,500 to 2,500 ml), the Modified Medical Research Council (MMRC) scale (from 2 to 1), and the Fatigue Severity Scale (FSS) (from 71 % to 45 %).

Conclusions: In a limited inpatient rehabilitation service country like Indonesia, a combined outpatient and home-based rehabilitation program of exercises benefits the functional capacity of a patient with an open ASD with severe pulmonary hypertension and mild pulmonary fibrosis.

Keywords: atrial septal defect, dyspnea, exercise, fatigue, pulmonary hypertension

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Introduction

The second most common congenital heart disease (CHD) is atrial septal defects (ASDs).[1] It accounts for roughly 13% of CHD cases, with a female to male ratio of 2:1.[1,2] Al though most ASDs are benign, left to right shunting can occasionally overwhelm the right heart, resulting in RHF and arrhythmias.[1] In the context of severe shunting and symptoms of RHF, shunt closure is recommended.[1] Closure of ASD is not recommended if irreversible pulmonary arterial hypertension (PAH) develops due to the risk of right ventricular decompensation and death following the operation.[1,3]

Pulmonary hypertension (PH) is a chronic illness that worsens over time. Dyspnea frequently appears gradually in the early stages of PH, delaying diagnosis.[4] Physical activity was thought to negatively affect PH patients since it increased the risk of disease progression, right ventricular decompensation, and sudden cardiac death. As a result, people with PH were advised to limit their physical activity, which worsened their mobility and exercise tolerance.[4] Despite improved medical treatment, most individuals with PH, continue to experience symptoms, decreased exercise capacity, poor quality of life (QoL), and disease progression.[5] In most cases, medication will not be able to completely stop or reverse right ventricular dysfunction, nor will it be able to normalize pulmonary vascular resistance.[5] Exercise training is helpful in various illnesses, including cardiac and pulmonary diseases, and is one of the most essential, safe, and cost-effective therapy alternatives.[5]

Long-term endurance exercise, according to research, increases right ventricle size and improves early diastolic right ventricular function and left ventricular stiffness.[5] As a result, moderate physical activity has been recommended to avoid various cardiovascular problems.[5] Exercise training for patients with left heart failure was given a 1A recommen-

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dation in the most recent guidelines. It has been shown to improve QoL and exercise capacity, as well as lower the risk of heart failure-related hospitalization and morbidity events.[6] However, excessive physical activity, particularly in untrained individuals, can increase the risk of myocardial infarction.[5]

There have been no exercise guidelines for patients with PH. However, in addition to pharmacological therapy, recent guidelines advocate a supervised and thoroughly monitored exercise and respiratory training program in specialized clinics for stable PH patients (class II, level of evidence B).[5] Exercise training for patients with PH, on the other hand, is only frequently reimbursed by insurance programs or government financing in a few high-income countries[5], including phase 2 cardiac rehabilitation in Indonesia.

According to the guidelines, for patients with severe PH who are stable on medical therapy, an inpatient and outpatient program should be used.[5] The majority of rehabilitation programs and services are provided at outpatient clinics in our setting; there are no rehabilitation beds or wards for inpatient treatment. As a result, the goal of presenting this case is to show that employing current guidelines at a limited health/rehabilitation care institution can successfully rehabilitate a patient with an open ASD and severe PH, which causes limitations in daily activities and restrictions in working as a school teacher.

Case presentation

A-37 year old female, an elementary school teacher, came to the rehabilitation clinic with a chief complaint of shortness of breath while doing daily activities such as sweeping and mopping, which started three years ago after giving birth. The symptoms disappeared with resting. However, the symptoms restricted her work as a teacher. She could not teach because of shortness of breath when speaking for more than 10 minutes.

The oxygen saturation was sometimes below 90% if she developed shortness of breath. Later, she also felt easily tired when doing daily activities, walking more than 150 meters, and speaking for more than 10 minutes, and became sedentary because of shortness of breath and fatigue. An atrial septal defect (ASD) closure was planned but could not be done because of severe pulmonary hypertension. For about 10 minutes, the oxygen saturation was reduced during the talk. The maximal inspiratory volume was 1,500 ml with chest expansion at axillary, nipple, and xiphisternum levels of 2 cm, respectively. The trunk alignment was straight (normal posture). Her body weight was 43 kg, her body height was 146 cm, and body impedance analysis revealed 21% visceral fat, 13.7 kg of skeletal muscle mass, and 56.6% of total body water.

The spirometry test was requested and showed mild restrictive. The echocardiography revealed dilated right atrium and ventricle, diastolic dysfunction, and reduced right ventricular contractility with a high probability of pulmonary hypertension. The right heart catheterization showed secundum ASD with balance shunt; pulmonary hypertension (mean pulmonary arterial pressure of 70 mmHg); low flow, high resistance, and nonreactive oxygen test. The CT scan of the thorax with contrast showed: cardiomegaly with pulmonary hypertension; minimal fibrosis spread over the lateral segment of the middle lobe, the anterobasal segment of the inferior lobe of the right lung, and almost the entire segment of the left lung; minimal left pleural thickening; minimal ground-glass opacity in the superior and anterobasal segments of the left inferior lobe of the left lung, compatible with idiopathic pulmonary fibrosis (IPF).

Using the Modified Medical Research Council (MMRS) Scale, the dyspnea scale was 2 (moderate dyspnea). The cardiorespiratory fitness using the treadmill exercise stress test with the “Bruce protocol” showed low cardiorespiratory fitness (METs = 2.92) with desaturation during exercise and the Fatigue Severity Scale (FSS) was 45 (71.43%).

The patient was referred to a rehabilitation physician by a cardiologist at the outpatient clinic. Under the supervision of a rehabilitation physician, the patient got oxygen supplementation 2-3 liters/minutes via nasal cannula while performing cardiopulmonary endurance exercise, which consisted of a 20-minutes session of low-intensity conditioning treadmill exercise with warming-up and cooling-down activities for 5 minutes, 3 days per week. Exercise duration was increased by 5 minutes every 2 weeks as tolerated. The oxygen saturation was maintained at above 88%.

In addition, the patient was trained to perform a home program of sustained maximum inspiration (SMI) using an incentive spirometer for two sessions per day. Each session consisted of 3 sets of 10 repetitions, rest 2-3 minutes between

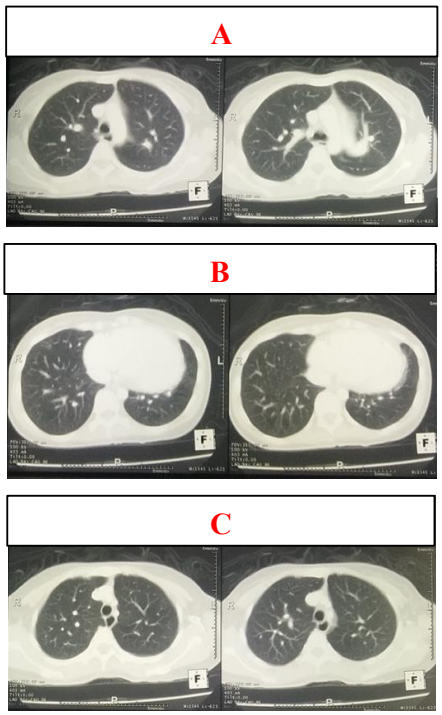

Figure 1. CT scan of thorax with the contrast of this patient showing (A) pulmonary hypertension; (B) cardiomegaly; (C) bilateral pulmonary fibrosis.

Table 1. The results before and after eight weeks of following the comprehensive rehabilitation program

	Before	After
MMRC score	2 (moderate dyspnea)	1 (mild dyspnea)
FSS score (%)	45 (71.4)	30 (47.6)
Maximum inspiratory volume (ml)	1.500	2.500
Chest expansion (cm)	2/2/2	3/3/3
Cardiorespiratory fitness (METs)	2.92	5.27
Oxygen saturation at rest (%)	92-93	95-96

MMRC, Modified Medical Research Council; METs, metabolic equivalents; FSS, fatigue severity scale
Max FSS score is 63; the higher the score, the greater the fatigue severity.[7]
Oxygen saturation during treadmill exercise was 87-91%

sets. The progression was to increase inspiratory volume every week as tolerated, and the target was 2,500 ml. Moreover, the patient was educated about controlled breathing during activity, relaxation techniques, and energy conservation.

The patient could complete the exercise program as planned. However, oxygen saturation was initially low, sometimes falling below 88% during exercise training. Therefore, the patient planned to provide the program for 8 weeks from the beginning.

Eight weeks after the comprehensive rehabilitation program, dyspnea and fatigue decreased while the maximum inspiratory volume, chest expansion, and cardiorespiratory fitness increased, as shown in Table 1. The patient could do daily activities with minimal shortness of breath and fatigue and resumed working as a part-time teacher.

Discussion

We describe a middle-aged female patient whose closure of ASD was contraindicated because of severe PH and the risk of right ventricular decompensation and death after the operation. The patient was referred to a rehabilitation physician (physiatrist) to rehabilitate a case that was not common in our setting. Before planning a comprehensive rehabilitation program for this patient, we were aware of her physical condition. Her respiratory disturbance might be caused by PH and restrictive lung disease due to IPF. This comprehensive exercise program aimed to lessen her symptoms of shortness of breath and dyspnea on exertion. The program consisted of controlled breathing techniques, which are generally used to improve pulmonary function tests, reduce dyspnea, reduce work of breathing, improve ventilator muscle function, facilitate relaxation, and are indicated in patients with obstructive and restrictive pulmonary problems.[8,9]

A home program of SMI with an incentive spirometer seemed like a cost-effective breathing exercise technique. After 8 weeks of SMI, her maximum inspiratory volume reached the target of 2,500 ml. This supports the evidence that SMI can increase chest expansion and lung volume to improve cardiorespiratory fitness for daily activities.[10,11] The target inspiratory volume of 2,500 ml was based on age.[12]

As spirometry showed mild restrictive lung disease in this case, we believe that the most likely cause of oxygen desaturation during exertion was of cardiac origin, rather than pulmonary. Therefore, besides breathing exercises, a cardiore¬spiratory exercise training program was planned at the outpatient rehabilitation clinic to improve her cardiorespiratory endurance. We started with light intensity aerobic exercise for patient safety, appropriate for patients with severe chronic disease or very deconditioned individuals.[13] The exercise duration was increased as tolerated based on recommended dyspnea ratings of between 3 and 6 on the Borg 10 scale.[14] And the exercise program was conducted at outpatient rehabilitation clinics under the supervision of a physiatrist. According to guideline recommendations, frequency is at least 3-5 days per week, and duration is 20-60 minutes per day at low-to-moderate intensities as tolerated.[5] In this case, we chose a session of 20 minutes of treadmill walking exercise as her cardiorespiratory fitness was low, and treadmill walking is generally the preferred method because walking is a functional activity.[15] We are also concerned about oxygen desaturation during exercise and prescribed oxygen supplementation, which is indicated for patients with a PaO_2 ≤ 55 mm Hg or a SaO_2 ≤ 88% while breathing room air,[16] so that SaO_2 is maintained at > 88% as recommended. After 8 weeks of this program, the patient's respiratory functions and cardiorespiratory fitness improved. The patient was advised to continue using the SMI technique at home. The patient was satisfied with the outcome of this rehabilitation program. She could perform her daily activities as usual, and go back to work as a teacher.

Conclusions

An 8-week combined outpatient and home program of cardiopulmonary and breathing exercises provided a beneficial result in improving the functional capacity and resuming work of a patient with an open ASD with severe pulmonary hypertension who is not fit for operation but is expected to reduce the risk of the worsening of pulmonary hypertension.

Disclosure

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