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RESPIRATORY FUNCTIONS AND EXER CISE CAPACITY IN ASYMPTOMATIC SWYER JAMES SYNDROME: REPORT OF TWO CASES

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SUMMARY

Aiming to determine the early functional changes and response to exercise test in two patients with Mac Leod Swyer syndrome. After performing the basal lung function tests, the subjects exercised until maximal level on a cycle ergometer. Maximal oxygen uptake (VO_2max) was determined from expired gas measurements during test. In conclusion, respiratory function abnormalities in both obstructive and restrictive manner may be seen in even in asymptomatic Swyer James syndrome. Although lung function tests are affected, VO_2max during exercise may be normal.

Key words: exercise testing, maximal oxygen uptake, spirometry, Swyer-James syndrome

ÖZET

Asemptomatik Swyer James Sendromunda Solunum Fonksiyonları ve Egzersiz Kapasitesi: İki Olgu Sunumu

Swyer James sendromu bir veya birden fazla lob veya tüm akciğerin hiperlusensi ve lezyonun olduğu tarafta hiler ve arteryel damarlanmada azalma ile karakterize bir tablodur.
Bu çalışmada Swyer James sendromu tesbit ettiğimiz iki hastada erken solunum fonksiyon test değişiklikleri ve egzersiz test cevaplarını ölçmeyi planladık. Bazal fonksiyon testlerini yaptıktan sonra , hastalar bisiklet ergometresinde maksimal seviyeye kadar egzersiz yaptılar. Maksimal oksijen alımı (VO_2max) test esnasında expire edilen gaz ölçümlerinden belirlendi. Sonuç olarak asemptomatik Swyer James sendromlu 2 hastada solunum fonksiyon test bozuklukları görülmekle beraber egzersiz esnasında VO_2 max değerinin normal olabileceği kanaatine varıldı.

Anahtar kelimeler: egzersiz test, maksimal oksijen alımı, Swyer-James sendromu

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INTRODUCTION

Swyer-James syndrome (SJS) is a radiologic entity: hyperlucency of one or several lobes, or even of one lung, scarce hilar shadow on the same side as the abnormal transradiency, and barely visible arterial network on the abnormal side of the thorax [1,2]. Distal bronchiectases and abnormal distal bronchi suggest that this disease may be due to bronchiolitis in childhood[3]. Obstructive type pulmonary disorder, assessed by pulmonary function tests, has been described in this syndrome[1,3].

The exercise responses in patients with lung diseases may differ from normal. There are many kinds of lung disease and, in different patients, the structural and functional severity of the disease may range from the barely discernible to the very severe[4]. As SJS is a very rare disorder, data on this syndrome are very limited and to our knowledge, none of previous studies investigated functional changes and cardiopulmonary exercise test responses in patients with this syndrome. Our aim was to determine the early functional lung status and exercise capacity in two patients with Swyer James syndrome.

CASE PRESENTATION

Two patients (17-year-woman, 173cm, 65kg; 25-year-man, 167cm, 55kg), diagnosed Swyer James syndrome were included to the study. Both of them were asymptomatic and diagnosed accidentally. Female patient described frequent respiratory tract infections during childhood. She was asymptomatic for at least two years before admitting to us. She had unilateral lung hyperlucency at chest X ray (Figure 1) and computerized tomography (CT) revealed hyperlucency of left lung with concomittant bronchiectatic changes (Figure 2). Lung scintigraphy showed the perfusion lost of left lung (Figure 3). Male patient diagnosed during the military service. He had no respiratory complaints and completed the military serve without a medical problem. His tomography also showed unilateral hyperlucency concomittant with bronchieactasis (Figure 4). Both patients had sedentary lifestyle and were not participating any regular sportive activities. Prior to exercise testing, patients read and signed a consent form that was approved by the University's Policy and Review Commitee on Human Research. This study was conducted in accordance with Helsinki Doctrine on Human Experimentation.

Figur 1: PA x ray of the female patient

Figure 2: CT of the female patient

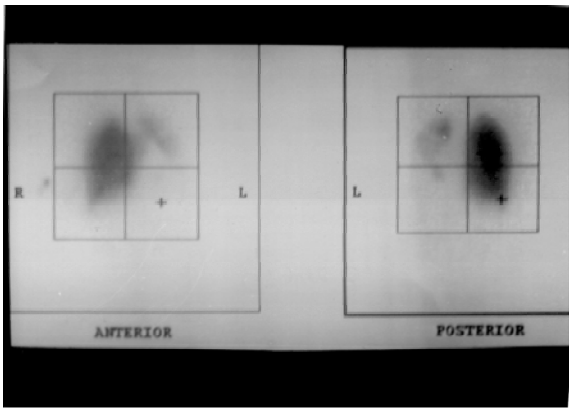

Figure 3: Scintigraphy of the female patient

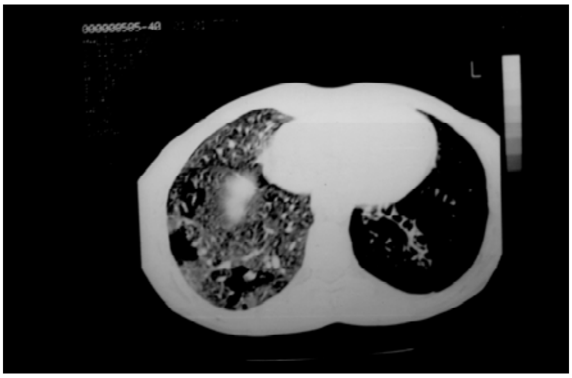

Figure 4: CT of the male patient

The maximal oxygen uptake (VO_{2max}) was measured during a maximal exercise testing (MXT) in order to establish their functional capacity. Each patient first underwent a comprehensive physical examination which included a 12 lead electrocardiogram (ECG) recording and blood pressure measurement at rest. They performed pulmonary function tests before exercise test. Subjects had a light breakfast 2 hours before exercise and abstained from strenuous exercise.
Lung function tests met the criteria of American Thoracic Society[5]. Dynamic and static volume and flow parameters detected by computer supported spirometer (Sensormedics Vmax 29c, Anaheim, CA, US). Acquired results were corrected for BTPS and compared with predicted normal values. European Coal & Steel criteria concerning age, height, gender and race as calculating predictive values was used [6].
The patients exercised on an electronically braked cycle ergometer (ergometrics 800, ergoline, Germany) at an initial work-rate of 20 Watts, which was increased by 20 Watts, at the end of each step lasting for 3-minute. The subjects were acclimated to the cycle ergometer with a 2-min warm up period (0 W). The pedal rate was maintained constant at 60 rpm throughout the test. The test was stopped at maximal exercise level. It was considered maximal if the patients achieved any two of the three test criteria. The criteria for reaching included a respiratory exchange ratio of 1.10 or higher, a maximal heart rate within ± 10 beats min^{-1} of age-predicted maximum and plateau of oxygen consumption with increasing work load. Maximal heart rate (HRmax) was calculated by subtracting subject's age from 220 (HRmax = 220-age). Heart rate was monitored electrocardiographically. Blood pressure was measured every three minutes using standard cuff manometry (ERKA, Germany). The VO2max was determined from expired gas measurements during test. Expired gases (O_2 and CO_2) were analyzed breath-by-breath on a SensorMedics Vmax 29c Metabolic Measurement Cart. Calculating predictive values was used to compare values of minute ventilation[7], VO_{2max}, VO_{2AT} and O_2 pulse[8-10].
Exercise performance and the cardio-respiratory measurements of two patients during maximal level of exercise testing can be seen at Table 1 and 2, respectively. All cardiopulmonary parameters at maximal level of exercise were within normal limits. For female and male patient, VO_{2max} values were measured as 26.9 ml/kg/min (81% of predicted) and 39.6 ml/kg/min (95% of predicted), respectively. Pulmonary function test parameters of two patients are depicted at Table 3 and 4, respectively. Obstructive respiratory insufficiency were detected in both patients. Degree of obstruction was more severe in female patient. Obstruction was more prominent at small airway level in both patients. FEF %25-75 values were 1.15 l/sec (28% of predicted) for female and 1.78 l/sec (37% of predicted) for male patient.

Table I: Aerobic capacity and the cardio-pulmonary measurements of female patient during maximal level of exercise testing

Parameters	Measured	Predicted	%
Aerobic capacity			
VO_{2max} (mL/kg/min)	26.9	33.1	81
VO_{2max} (L/min)	1.748	2.148	81
VO_{2AT} (L/min)	1.134	>40%	53
VO_{2AT} (mL/kg/min)	17.4	>40%	53
Cardio-pulmonary			
HRmax, (bpm)	183	203	90
O_2 pulse, (mL O_2/beat)	8.5	10.6	80
V_{Emax}, (L)	59.4	67.5	88

HRmax: Maximal heart rate, VEmax: Maximal minute ventilation, VO2max: Maximal Oxygen uptake, VO2AT : Oxygen uptake at the anaerobic threshold,

Table II: Aerobic capacity and the cardi o-pulmonary measurements of male patient during maximal level of exercise testing.

Parameters	Measured	Predicted	%
Aerobic capacity			
VO_{2max} (mL/kg/min)	39.6	41.5	95
VO_{2max} (L/min)	2.178	2.284	95
VO_{2AT} (L/min)	1.744	>40%	76
VO_{2AT} (mL/kg/min)	31.7	>40%	76
Cardio-pulmonary			
HRmax, (bpm)	185	195	95
O_2 pulse, (mL O_2/beat)	11.8	11.7	100
V_{Emax}, (L)	72.9	88.3	83

HR_{max}: Maximal heart rate, V_{Emax}: Maximal minute ventilation, VO_{2max}: Maximal Oxygen uptake, VO_{2AT} : Oxygen uptake at the anaerobic threshold,

Table III: Pulmonary volume and capacity of female patient during resting

Variable	Measured	Predicted	%
FVC, L	2.83	3.86	73
FEV_1, L	1.73	3.71	47
FEV_1/FVC %	61	86	71
VC, L	2.99	3.86	77
FEF %25-75(L/sec)	1.15	4.09	28
PEFR(L/sec)	2.46	7.21	34

FVC: Forced vital capacity, FEV1: Forced expiratory volume at one second, FEV1/FVC: Tiffeneau index, VC: vital capacity, FEF25-75: Forced expiratory flow 25-75, PEFR:Peak expiratory flow rate.

Table IV: Pulmonary volume and capacity of male patient during resting

Variable	Measured	Predicted	%
FVC, L	3.20	4.45	72
FEV1, L	2.33	3.84	61
FEV1/FVC %	73	83	88
VC, L	3.20	4.65	69
FEF %25-75(L/sec)	1.78	4.81	37
PEFR(L/sec)	5.40	9.16	59

FVC: Forced vital capacity, FEV1: Forced expiratory volume at one second, FEV1/FVC: Tiffeneau index, VC: vital capacity, FEF25-75: Forced expiratory flow 25-75, PEFR:Peak expiratory flow rate.

DISCUSSION

Swyer-James syndrome (SJS) is one of the manifestations of postinfectious obliterative bronchiolitis. The involved lung or portion of the lung does not grow normally and is slightly smaller than the opposite lung. The lung is expected to grow by progressive alveolarization for a child's first 2-8 years. Thereafter, lung growth is related to hyperexpansion of existing alveoli. SJS is a postinfectious syndrome in which diminished vascularity, arrest of progressive growth and alveolarization of the lung, and resultant hypoplasia occur. Multifocal areas of air trapping may be seen. The pulmonary parenchymal pattern is similar to obliterative bronchiolitis[11-14].

Bronchoalveolar lavage studies of two patients with Swyer-James syndrome suggested that there is an ongoing inflammation which is characterised an increase of neutrophils[15]. Involved lung of patients with SJS is small. Compensatory overexpansion of the contralateral lung, peripheral bronchi and bronchioles "pruned" secondary to obliterative bronchiolitis, a mosaic pattern of hyperlucency on CT, and small vessels and vascular occlusions in the abnormal areas can be observed[16,17]. Both of our patients showed unilateral hyperlucency concomittant with bronchiectasis. We observed compensatory hyperinflation of uninvolved lung. Pulmonary scintigraphy in Swyer James syndrome reveals unilateral abscence of pulmonary perfusion[18]. We also observed this finding in our patients. Patients with Swyer James syndrome may show respiratory function abnormalities[3]. We found obstructive respiratory insufficiency in lung function tests in both patients. This functional impairment has not seem to affect the patients daily life and has not cause any complaint. Many patients with pulmonary disease can have concomitant cardiovascular disease, deconditioning, or anxiety/ depression that might also reduce exercise performance. And a low aerobic capacity is an important indicator for co-morbidity[19]. Exercise testing can provide important clinical data about these and other possible conditions that might cause dyspnea and/or limit exercise performance in these patients. It is preferable to measure minute ventilation and its components, frequency of respiration and tidal volume, as well as expired gases, O_2 and CO_2[20] VO_{2max} is accepted as the most accurate measure of cardiopulmonary physical fitness. It is an integrated evaluation of the state of the oxygen transport system, and includes the functions of the pulmonary, cardiovascular and muscular systems. In adults, during a maximal exercise testing VO_2 reaches a plateau at VO_2 and no further increase is observed with further increases in the rate of work.

From the results of this study it is concluded that respiratory function abnormalities in both obstructive and restrictive manner may be seen in even in asymptomatic Swyer James syndrome. Although lung function tests are affected, aerobic capacity measured as VO_{2max} during exercise may be normal. We suggest that studies focusing on the long term follow up with spirometry and cardiopulmonary exercise testing in patients with this syndrome would be beneficial.

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