



# Allergologia et immunopathologia

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## ORIGINAL ARTICLE

# Lung involvement in rheumatologic diseases in children

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Received 17 December 2010; accepted 14 February 2011

### KEYWORDS

Children;  
Juvenile arthritis;  
Juvenile dermatomyositis;  
Lung;  
Lupus;  
Rheumatic diseases

### Abstract

**Background:** Lung involvement in rheumatologic disease in children has been described with low frequency.

**Objective:** To describe the lung function test and the radiological findings in a group of paediatric patients with rheumatologic diseases.

**Methods:** Descriptive study. Pulmonary function was evaluated with spirometry, carbon monoxide diffusing capacity, blood arterial gas at rest and post exercise in addition to chest radiography and high resolution computed tomography were performed in children with rheumatologic disease.

**Results:** Of the fourteen patients studied: 11 were diagnosed with juvenile idiopathic arthritis (JIA), two with systemic lupus erythematosus (SLE), and one with juvenile dermatomyositis (JDM). Mean age: 13 years (range 7–18 years). Nine females, duration of disease  $4 \pm 2.8$  years (range 1–11 years). Four patients had any grade of abnormalities in one or more lung function tests and/or radiological test.

**Conclusion:** We recommend routine checks of pulmonary function in all patients with rheumatologic diseases even in the absence of respiratory symptoms.

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## Introduction

Lung function tests have been used for the diagnosis, monitoring and treatment evaluation of a number of chronic lung diseases in children, such as asthma, cystic fibrosis, and interstitial pneumonitis. For each of these methods, there are standard procedures and reference values adjusted to normal population by age, sex, and height.<sup>1–3</sup> Rheumatic diseases are a group of immunologically mediated

inflammatory disorders associated with a large variety of affected organs.<sup>3</sup>

There are few descriptions of lung injuries in children with rheumatic disease, which incidentally may be affecting all its components such as airways, vessels, parenchyma, pleura, and respiratory muscles.<sup>4–9</sup> Juvenile idiopathic arthritis, systemic lupus erythematosus, juvenile dermatomyositis, and systemic sclerosis are the most frequent rheumatic illnesses associated with lung compromise.<sup>1,2,4,5</sup>

This impairment can be related to the illness itself, drugs used in treatment, or secondary to infections between others.<sup>7,8</sup>

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In adults, rheumatoid arthritis has been associated with mild restrictive defects, diffuse interstitial lung diseases, pulmonary vasculitis, and bronchiolitis obliterans. These alterations have also been observed in juvenile idiopathic arthritis.<sup>10,11</sup>

In childhood, lung impairment can be found in active diseases even in the absence of abnormalities in chest radiographs or in lung function tests such as diffusing capacity, lung volumes, low forced ventilatory flows and saturation in exercise.<sup>9</sup>

## Objective

To describe the lung function test and radiological findings in children in treatment for at least one year from diagnosis of some rheumatic disease stable in the preceding six months.

## Methods

### Study population

Fourteen patients attending the Immunology Department of Exequiel Gonzalez Cortes Children's Hospital (Santiago, Chile) were enrolled: 11 diagnosed with juvenile idiopathic arthritis (JIA) of different clinical form (three systemic, two oligoarticular, two polyarticular, and four enthesitis-related arthritis) according to the International League Association for Rheumatology classification<sup>12</sup>; two diagnosed with systemic lupus erythematosus (SLE) according to American Rheumatology Academic criteria updated by Hochberg<sup>13</sup>; and one diagnosed with juvenile dermatomyositis (JDM) according to Pearson criteria modified by Bohan and Peter.<sup>3</sup> Age of onset and history of treatment were recorded. None had history of chronic lung disease before the onset of the rheumatic disease.

### Lung function

In all patients spirometry was obtained using a Med Graphics Breeze V 3.7 spirometer, and the best values of at least three technically well-performed flow-volume curves were used. Reference values of Gutierrez for Chilean populations of healthy children 4–18 years were applied<sup>14</sup> and range 80–120% for reference values were considered normal.

Carbon monoxide diffusing capacity of lung (DLCO) and DLCO/alveolar volume (VA) were determined using a standardised single-breath technique (Master Screen Body V 4.3 Jaeger). Reference values from Zapletal for populations of healthy children aged 4–18 years were used<sup>15</sup> and range 80–120% for reference values were considered normal.

Blood arterial gases at rest and exercise were performed in the subjects before and after a run on a treadmill for three minutes after reaching submaximal heart rate. Reference values of Graef were used.<sup>16</sup>

In two cases with signs of hypoventilation, strength of respiratory muscles was assessed by measuring maximum static respiratory mouth pressures with a tube mouthpiece attached to pressure gauge. The absolute peak pressure for inspiratory effort (Pi max) was recorded and compared with reference values of Szeinberg.<sup>17</sup> This test is used in

**Table 1** Characteristics of the study population.

Diagnosis	
JIA S	3
JIA O	2
JIA P	2
JIA E-A	4
SLE	2
JDM	1
Female	64.3%
Age years	13 (range 7–18)
Duration of disease years	4 ± 2.8

JIA = juvenile idiopathic arthritis; S = systemic; O = oligoarticular; P = polyarticular; E-A = enthesitis-related; SLE = systemic lupus erythematosus; JDM = juvenile dermatomyositis.

our country as part of the evaluation of lung involvement in children with neuromuscular diseases.

### Radiology test

Chest radiography and chest high resolution computed tomography (HRCT) were interpreted by two blinded radiologists.

The protocol was approved by the local Ethics Committee and written consent for participation in the study was obtained from their parents.

### Statistical analyses

All variables were presented as means ± SD. The results from the lung function test were expressed as a percentage of reference values.

Epi Info 6.0 and Stata 10.0 were used for data analysis. Percentages and Fisher exact test were used for qualitative nominal variables such as gender differences. T student test was applied for gauge variation with mean and standard deviation and average comparison.

## Results

Fourteen children were included (Table 1). Mean age was 13 years (range 7–18 years), nine females. Duration of disease was 4 ± 2.8 years (range 1–11 years). Among these 14 children: nine had normal results in all lung function and radiologic tests (diagnoses JIA 8, JDM 1). Table 2 details the treatment being received at time of study. Seven JIA patients were receiving methotrexate, as well as low doses of prednisone in four; two lupus patients were treated with cyclophosphamide pulses plus prednisone during pulmonary evaluations.

In four cases (28.6%, SLE 2, JIA 2) we found abnormal test (Table 3). One patient with SLE showed forced vital capacity (FVC) and forced expiratory volume in 1 s (FEV<sub>1</sub>) lower than reference values compatible with restrictive defects; in this case, the other lung function test and radiology test were abnormal too. She had reduction in DLCO and evidence of hypoventilation in blood arterial gases at rest and exercise, and the value of maximal inspiratory pressure (Pi max) was lower than the reference values. In the other SLE

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