**Lung Function Often Impaired in Juvenile Dermatomyositis**

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**FROM THE ANNALS OF THE RHEUMATIC DISEASES**

Juvenile dermatomyositis is associated with reduced lung volumes, restrictive ventilatory defects, and evidence of pulmonary abnormalities on high-resolution CT, a study has shown.

The pulmonary complications—even in the absence of lung symptoms—correlated with cumulative organ damage and patient-reported health status, which demonstrates the clinical relevance of the findings and the systemic nature of the chronic vasculopathic disease, reported Dr. Helga Sanner of the division of rheumatology at the University of Oslo and her associates.

Because of the scarcity of data on pulmonary involvement in juvenile dermatomyositis, Dr. Sanner and colleagues designed a case-control study to compare lung function in a cohort of patients with juvenile dermatomyositis (JDM) vs. matched controls. The investigators also determined the prevalence of and correlation between pulmonary function impairments and abnormalities on high-resolution computed tomography (HRCT).

The investigators enrolled 59 patients whose JDM was diagnosed between January 1970 and June 2006 at the Children’s Hospital, Denver. Another possible diagnosis is pyomyositis, noted Dr. Dominguez, a pediatric infectious diseases specialist at the hospital and the University of Colorado at Denver.

**Major Finding:** Pulmonary involvement is common in patients with juvenile dermatomyositis and is associated with cumulative organ damage and patient-reported health status.

**Data Source:** Case-control study comprising a retrospective inception cohort of 59 Norwegian patients diagnosed with JDM, and healthy controls.

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The JDM patients also underwent HRCT, anti–nuclear antibody analysis, measurement of disease activity using the Disease Activity Score (DAS) for JDM, assessment of cumulative organ damage using the Myositis Damage Index (MDI), measurement of physical health using the Short Form-36 physical component summary (PCS), and measurement of physical function using the Health Assessment Questionnaire (HAQ) for patients aged 18 years and older and the Child HAQ for patients younger than 18 years.

With respect to clinical lung involvement, three of the JDM patients had been diagnosed with interstitial lung disease (ILD) prior to the clinical examination; 6 months after her JDM diagnosis, one of the JDM patients developed mediastinal emphysema without evidence of ILD at the age of 13 years, the authors wrote, noting that none of the controls had lung symptoms.

Compared with the controls, the JDM patients had significantly lower total lung capacity (TLC), diffusion lung capacity of carbon monoxide (DLCO), forced vital capacity (FVC), and the forced expiratory volume in 1 second (FEV1). Approximately 26% of the JDM patients, compared with 9% of the controls, had a low TLC; 49% of the JDM patients, compared with 8% of the controls, had a low DLCO (defined for both measures as less than the fifth percentile of the predicted value), the authors reported (Ann Rheum Dis. 2010 Aug 30 [doi:10.1136/ard.2010.131433]).

With respect to HRCT findings in the JDM patients, 37% had evidence of pulmonary abnormalities, including changes compatible with interstitial lung disease (14%), airway disease (15%), pleural thickening (5%), and calcification in the chest wall (14%), the authors reported.

The correlation analyses showed that 50% of patients with an abnormal HRCT abnormality had a low TLC, compared with 12% of patients with normal HRCT findings, and 57% of patients with HRCT-detected calcification in muscle and/or fascia had a low TLC, compared with 22% of patients without that finding. The TLC percentage of predicted correlated with HRCT-detected airway disease, whereas the DLCO percentage of predicted did not correlate with any HRCT variables.

The association between chest wall calcification and restriction is not surprising, since calcium deposits might lead to respiratory muscle impairment, however the association between a low TLC and airway disease—for example, bronchiectasis—is more difficult to explain,” the authors wrote, noting the possibility of a type I error.

HRCT abnormalities also correlated with cumulative organ damage and poorer patient-reported health status as measured by total MDI, HAQ/child-HAQ, and SF-36 PCS, and a borderline association was found between HRCT-detected ILD and dyspnea on exertion. “Taken together, we believe this supports the clinical relevance of our findings,” the authors wrote.

Even though approximately 75% of the patients had impaired diffusion, restriction, or HRCT abnormality at follow-up, most of the patients did not report lung symptoms. It’s possible that the lung symptoms may have been masked by restricted functions in other organ systems, the authors speculated. For example, “if patients are not able to complete physical exercise due to muscle weakness, they will not experience shortness of breath even when they have reduced lung function,” they stated.

Follow-up studies are needed to investigate whether more patients with detectable HRCT abnormalities and pulmonary function test impairments will develop clinically manifest pulmonary disease in the future, they stressed.

The findings are limited by the study’s retrospective assessment of early disease variables and by the lack of longitudinal data on the outcome measures, according to the authors. Because experimental ILD can develop in the chronic phases of the disease, they wrote, “some of our latest diagnosed patients may still develop ILD.” The study is also limited by the retrospective assessment of early disease variables.