

Rheumatologie GKJR - Autoimmunität / Kollagenosen

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**How is pulmonary function assessed in juvenile systemic scleroderma patient? Do we have a good clinical standard? Results from the juvenile scleroderma inception cohort-
www.juvenile-scleroderma.com**

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Juvenile systemic scleroderma (jSSc) is a rare, but potentially life-threatening disease. Around 45% of the patients develop interstitial lung disease and this can lead to relevant mortality. Pulmonary function tests (PFT). Including forced vital capacity (FVC) and lung carbon monoxide diffusion (DLCO), are key for the screening of interstitial lung disease. PFT require an optimal patient cooperation and coordination. For example, the patient has to hold the breath for 10 seconds. Additionally, the graph of the breath holding maneuver has to be carefully observed, if there is a premature expiration, numbers have to be judged with suspicion. We were interested how in the real world these problems are addressed in children.

Objective

to assess how PFT is assessed in the participating pediatric rheumatologic centers in the juvenile scleroderma inception cohort

Methods

We conducted a survey among the pediatric rheumatologists and associated pediatric pulmonologists, who are participating in the prospective juvenile scleroderma inception cohort (jSScC). We asked them some simple questions about the standard of care regarding assessment of the PFT.

Results

65% (26/40) of the surveyed participants of jSScC responded. From the participating centers 96% (25/26) assess the pulmonary function test in a specific pediatric setting. 65% (17/26) regularly assess DLCO. In 77% (20/26) of the centers the respiratory technician that conducts the PFT judges the level of cooperation of the child and the pediatric pulmonologist, who evaluates the results, also makes a judgement on the reliability of the test. We asked a specific question regarding the duration of the breath holding time setting in the lung function system. 32% (8/25) set the breath holding time under 7 seconds and 8% (2/26) at 7 seconds and 20% (5/25) between 7 and 10 seconds and 28% (7/25) exactly 10 seconds.

Discussion:

We could demonstrate that in 96% of the responding centers PFT are carried out in specialized pediatric pulmonology departments, which are aware of the methodological problems and change their breath holding time to an appropriate time to gain as many reliable tests as possible. There is still a wide range of the applied breath holding time and even in centers carrying for jSSc patients, 23% do not assess DLCO.

