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Under Detection of Interstitial Lung Disease in Juvenile Systemic Sclerosis (jSSc) Utilizing Pulmonary Function Tests. Results from the Juvenile Scleroderma Inception Cohort

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SESSION INFORMATION

Session Date: Sunday, November 8, 2020

Session Title: Pediatric Rheumatology – Clinical Poster II: Systemic JIA, Autoinflammatory, & Scleroderma

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Background/Purpose: Juvenile systemic sclerosis (jSSc) has a prevalence in around 3 in a million children. Pulmonary involvement occurs in approximately 40 % in the international juvenile systemic scleroderma cohort (JSScC). Traditionally in jSSc, pulmonary function testing (PFT) with FVC and DLCO are used for screening and computed tomography (HRCT) was more reserved for those with abnormal PFTs. More recently, it has become apparent that PFTs might not be sensitive enough for detecting interstitial lung disease (ILD) in children.

Methods: JSScC database was queried for available patients with recorded PFT parameters and HRCT performed to determine sensitivity of PFTs detecting disease process.

Results: Of 129 patients in the jSScC, 67 patients had both CT imaging and an FVC reading from PFTs for direct comparison. DLCO readings were also captured but not in as many patients with tandem HRCT (n =55 DCLO and HRCT scan). Therefore, initial analyses focused on the sensitivity, specificity and accuracy of the FVC value from the PFTs to capture the diagnosis of interstitial lung disease as determined by HRCT.

Overall, 49% of the patients had ILD determined by HRCT, with 60% of patients having normal FVC (>80%) with positive HRCT findings, and 24% of patients having normal DLCO (> 80%) with positive HRCT findings. Fourteen percent (n = 3/21) of patients with both FVC and DLCO values within the normal range had a positive HRCT finding.

Conclusion: The sensitivity of the FVC in the JSScC cohort in detecting ILD was only 39%. Relying on PFTs alone for screening for ILD in juvenile systemic sclerosis would have missed the detection of ILD in almost 2/3 of the cohort, supporting the use of HRCT for detection of ILD. The cut off utilized, of less than 80% of predicted FVC or DLCO could be too low to exclude beginning ILD.

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Cardiovascular involvement as a clue for diagnosis of Juvenile Systemic Sclerosis *sine scleroderma*

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Background/Purpose: Juvenile Systemic Sclerosis (JSSc) is a rare condition in childhood and its variety with no skin involvement, systemic sclerosis *sine scleroderma* (JSSSS) is anecdotal as only two cases have been reported to date[1,2]. We describe a series of four patients from our Center and compare these six patients with a cohort of patients with standard JSSc.

Methods: Unselected consecutive patients with Juvenile Systemic Sclerosis (JSSc), diagnosed according with the PRES/EULAR/ACR criteria[3] were retrospectively evaluated. For every patient, we collected demographic, clinical and laboratory data, autoantibody profile and treatment. The following clinical-instrumental parameters were considered: skin involvement by the mRodnan Skin Score, Raynaud’s phenomenon (RP), chest x-ray, high-resolution computed tomography (HRCT), diffusing capacity for carbon monoxide (DLCO), forced vital capacity (FVC), musculoskeletal involvement, esophageal scintiscan or 24-hour pH-metry, and malabsorption test. Cardiac investigations included