Abstract Number: 0796

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

Ivan Foeldvari,[1] Bernd Hinrichs,[2] Kathryn Torok,[3] Maria José Santos,[4] Ozgur Kasapcopur,[5] Amra Adrovic,[6] Valda Stanevicha,[7] Flavio Sztajnbok,[8] Maria Teresa Terreri,[9] Ekaterina Alexeeva,[10] Jordi Antón,[11] Maria Katsicas,[12] Vanessa Smith,[13] Tadej Avcin,[14] Rolando Cimaz,[15] Mikhail Kostik,[16] Thomas Lehman,[17] Walter Sifuentes-Giraldo,[18] Simone Appenzeller,[19] Mahesh Janarthanan,[20] Monika Moll,[21] Dana Nemcova,[22] Dieneke Schonenberg,[23] Christina Battagliotti,[24] Lillemor Berntson,[25] Blanca Bica,[26] Juergen Brunner,[27] Patricia Costa Reis,[28] Despina Eleftheriou,[29] Liora Harel,[30] Gerd Horneff,[31] Tilmann Kallinich,[32] Dragana Lazarevic,[33] Kirsten Minden,[34] Susan Nielsen,[35] Farzana Nuruzzaman,[36] Anjali Patwardhan,[37] Yosef Uziel,[38] and Nicola Helmus[1], [1]Hamburg Centre for Pediatric and Adolescence Rheumatology, Hamburg, Germany, [2]Asklepios Klinik Nord - Heidberg, Hamburg, Germany, [3]UPMC Children's Hospital of Pittsburgh, Pittsburgh, [4]Rheumatology department, Hospital Garcia de Orta, Almada, Portugal, [5]Department of Pediatric Rheumatology, Istanbul University-Cerrahpasa, Istanbul, Turkey, Istanbul, Turkey, [6]Cerrahpaşa Tıp Fakültesi, Istanbul, Turkey, [7]Riga Stradins University, Riga, Latvia, [8]Universidade do Estado do Rio de Janeiro, Rio de Janeiro, Brazil, [9]Pediatric Rheumatology Unit, Universidade Federal de São Paulo, São Paulo, Brazil, [10]National Medical Research Center of Children`s Health, Moscow, Russia, [11]Hospital Sant Joan de Déu, Barcelona, Spain, [12]Hospital de Pediatria, Buenos Aires, Argentina, [13]Dept. of Rheumatology, Ghent University Hospital, Ghent, Belgium; Dept. of Internal Medicine, Ghent University, Ghent, Belgium; Unit for Molecular Immunology and Inflammation, VIB Inflammation Research Center (IRC), Ghent, Belgium, Gent, Belgium, [14]Ljubljana University Medical Centre, Ljubljana, Slovenia, [15]University Hospital Meyer, Florence, Italy, [16]Saint-Petersburg State Pediatric Medical University, Saint-Petersburg, Russia, [17]Hospital for Special Surgery, New York, [18]Hospital Universitario Ramón y Cajal, Madrid, Spain, [19]UNICAMP Universidade Estadual de Campinas, Sao Paulo, Brazil, [20]Sri Ramachandra University, Chennai, India, [21]University of Tuebingen, Tuebingen, Germany, [22]General University Hospital, Prague, Czech Republic, [23]Amsterdam UMC, Amsterdam, Netherlands, [24]Children's Hospital Dr. Orlando Alassia, Santa Fee, Argentina, [25]Uppsala University Hospital, Uppsala, Sweden, [26]Hospital Universitário Clementino Fraga Filho (HUCFF/UFRJ), Rio de Janeiro, Brazil, [27]Tirol Kliniken, Innsbruck, Austria, [28]Hospital de Santa Maria, Lisbon, Portugal, [29]Great Ormond Street Hospital, London, United Kingdom, [30]Tel Aviv University, Tel Aviv, Israel, [31]Asklepios Clinic Sankt Augustin, Sankt Augustin, Germany, [32]Charite, Berlin, Germany, [33]KC Niš, Niš, Serbia, [34]German Rheumatism Research Center and Charité University Medicine, Berlin, Germany, [35]Department of Pediatrics, Rigshospitalet, Copenhagen University Hospital, Copenhagen, Denmark., Copenhavn, Denmark, [36]Stony Brook Children's Hospital, Stony Brook, NY, [37]University of Missouri, Columbia, [38]Meir Medical Center, Kfar Saba, Israel

SESSION INFORMATION

Session Date: Sunday, November 10, 2019

Session Title: Pediatric Rheumatology – ePoster I: Basic Science, Biomarkers, & Sclerodermic Fever

Session Type: Poster Session (Sunday)

Session Time: 9:00AM–11:00AM

Background/Purpose: Juvenile systemic sclerosis(jSSc) is an orphan disease with a prevalence in around 3 in a million children [1]. Pulmonary involvement in jSSc occurs in approximately 40 % in the inception cohort [2]. 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 ILD in children.

Methods: Utilizing a prospective international juvenile systemic scleroderma cohort (JSScC) [2], to determine if pulmonary screening with FVC and DLCO is sufficient enough to assess the presence of interstitial lung disease in comparison to CT evaluation.

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

Table 1. Diagnostic test properties of FVC as a test for ILD detection.

	Disease + (ILD Yes on HRCT)	Disease – (ILD No on HRCT)	Total
Test + (FVC <80%)	a) True positive = 13	c) False positive = 7	a+c =20 (+Test)
Test – (FVC>80%)	b) False negative = 20	d) True negative = 27	b+d =47 (-Test)
Total	a +b = 33 (+Disease)	c+d = 34 (- Disease)	

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. Table 1 presents these diagnostic test evaluations for the FVC. 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 sample cohort, supporting the use of HRCT for detection of ILD in children with SSc. In addition, the cut off utilized, of less than 80% of predicted FVC or DLCO could be too low for pediatric patients to exclude beginning ILD. This pilot data needs confirmation in a larger patient population.

Supported by the “Joachim Herz Stiftung”:

References:

1. Beukelman, T., F. Xie, and I. Foeldvari, *Assessing the prevalence of juvenile systemic sclerosis in childhood using administrative claims data from the United States.* Journal of Scleroderma and Related Disorders, 2018. **3**(2): p. 189-190.
2. Foeldvari, I., et al., *CHARACTERISTICS OF THE FIRST 80 PATIENTS AT TIMEPOINT OF FIRST ASSESSMENT INCLUDED IN THE JUVENILE SYSTEMIC SCLEROSIS INCEPTION COHORT.* WWW.JUVENILESCLERODERMA.COM. Journal of Scleroderma and Related Disorders, 2019. **4**: p. 49-61.

Disclosure: I. Foeldvari, Beyer, 5, BMS, 5, Glaxo, 5, Inventa, 5, Novartis, 5; **B. Hinrichs**, None; **K. Torok**, None; **M. Santos**, AbbVie, 8, Biogen, 8, Novartis, 8, Pfizer, 8, Roche, 8; **O. Kasapcopur**, AbbVie, 2; **A. Adrovic**, None; **V. Stanevicha**, None; **F. Sztajnbok**, None; **M. Terreri**, None; **E. Alexeeva**, None; **J. Antón**, AbbVie, 2, Bristol-Myers Squibb, 2, Gebro, 2, GlaxoSmithKline, 2, Novartis, 2, Novimmune, 2, Pfizer, 2, Roche, 2, Sobi, 2; **M. Katsicas**, None; **V. Smith**, None; **T. Avcin**, None; **R. Cimaz**, None; **M. Kostik**, None; **T. Lehman**, None; **W. Sifuentes-Giraldo**, None; **S. Appenzeller**, None; **M. Janarthanan**, None; **M. Moll**, None; **D. Nemcova**, None; **D. Schonenberg**, None; **C. Battagliotti**, None; **L. Berntson**, None; **B. Bica**, None; **J. Brunner**, None; **P. Costa Reis**, None; **D. Eleftheriou**, None; **L. Harel**, None; **G. Horneff**, Chugai, 5, 8, GlaxoSmithKline, 5, 8, Novartis, 5, 8, Sanofi, 5, 8; **T. Kallinich**, None; **D. Lazarevic**, None; **K. Minden**, AbbVie, 8, Abbvie, 8, German Arthritis Foundation, 2, GlaxoSmithKline, 5, GSK, 5, Sanofi, 8; **S. Nielsen**, None; **F. Nuruzzaman**, None; **A. Patwardhan**, None; **Y. Uziel**, None; **N. Helmus**, None.