

Results: SSc patients started to suffer of CP in younger ages compared to AR/CH cohort but patients suffering of CH have higher mean scores in all questionnaires compared to AR/SSc. CH patients, have higher mean score in SFMPQ-sensory and affectory. Hundred percent (100%) of AR patients suffered of chronic pain. They, generally, had higher scores than SSc patients with a prevalence of the affectory component. The 83.9% (67/75 pts) of SSc patients experienced chronic pain [SF-MPQ PRI: 6.25±8.34DS; SF-MPQ PPI: 1.69±1.34DS; BPI-fattore1: 13.37 ±11.26DS; VAS: 40.7±29.6DS; NRS: 4.08±2.98DS] and in a great majority (84%) that pain interfered with their working activities and social lifes. Fortyeight percent of SSc patients had digital ulcers and 41.3% had musculoskeletal involvement. Pain used to correlate with both of them [p=0.004; p=0.041]. In patients with DUs, affectory component of pain overuled on the sensory one

Conclusion: SSc patients frequently experience chronic pain and particularly those who have a history of active DUs and musculoskeletal involvement. By the way, it seems that the BPI questionnaire could be more suitable than VAS or NRS in assessing DUs' pain. Furthermore it provides useful information on the impact of CP on social life and work impairment. Otherwise, SF_MPQ allows clinicians to better discriminate affective or sensory components of pain.

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Disclosure of Interests: Luca Magnani: None declared, Federica Lumetti: None declared, Emanuele Cocchiara: None declared, Amelia Spinella: None declared, Carlo Baraldi: None declared, Luigialberto Pini: None declared, Lanfranco Pellesi: None declared, Gianluigi Baiocchi: None declared, Carlo Salvarani Grant/research support from: Roche, Consultant for: Eli Lilly and Company, Roche, Abbvie, Dilia Giuggioli: None declared
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SAT0286

EVALUATION OF SWALLOWING ALTERATIONS IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

Emiliano Marasco¹, Giulia Bertino², Ludovico De Stefano¹, Lorenzo Volpiano¹, Pietro Canzi², Marco Benazzo², Carlomaurizio Montecucco¹, Lorenzo Cavagna¹.
¹Rheumatology Department, IRCCS Policlinico S. Matteo Foundation, Pavia, Italy;
²Otorinolaringoiatria, IRCCS Policlinico S. Matteo Foundation, Pavia, Italy

Background: Dysphagia represents a frequent and disabling symptom in patients with Idiopathic Inflammatory Myopathies (IIMs) and it has been recently included in the new ACR/EULAR classification criteria for IIMs. Despite the clinical relevance, dysphagia assessment in IIMs is not currently standardized and evaluation tools are highly variable between different centers. Functional Endoscopic Evaluation of Swallowing (FEES) is an exam that allows, by using nasal endoscopy, the direct evaluation of anatomic structures and swallowing abilities in different swallowing phase. In fact, FEES could help the identification of different problems leading to dysphagia occurrence and to related dysphagia problems. Thanks to these characteristics, FEES for ENT specialists is the gold standard technique for evaluating swallowing functions. However, no studies so far have investigated the role of FEES in the assessment of IIMs and we are completely lacking a semeiotic description of FEES findings in these patients.

Objectives: To provide the first semeiotic description of swallowing alterations evidenced by FEES in a cohort of IIMs patients.

Methods: We retrospectively reviewed the FEES findings of IIMs patients performed at our hospital.

Results: We enrolled 19 patients with a diagnosis of IIMs (10 patients were positive for a myositis specific antibody), of these 16 (84%) reported symptomatic dysphagia. We divided patients into 3 groups based on levels of peripheral muscle strength. Six patients (32%) had no clinical sign of active muscle disease (MRC scale 5, median CK 51 mU/ml, IQR 35-235), 5 patients (26%) had a mild reduction in muscle strength (MRC scale 4, median CK 76, IQR 54-220) and 8 patients (42%) showed a moderate-severe reduction in muscle strength (MRC scale ≤3, media CK 1440, IQR 628-6180). The 67% of patients without muscle disease activity showed an impairment in the oral phase of swallowing for solids and the 33% for fluids; 33% had a reduction in the activation of the pharyngeal phase of swallowing for both fluids and solids; only 17% of patients showed any sign of penetration, aspiration or pharyngeal residue for both solids and fluids. In the group of patients with moderate

muscle activity, 80% of patients showed impairment in the oral phase of swallowing for solids and 40% for fluids; 60% had a reduction in the activation of the pharyngeal phase of swallowing for solids while 40% for fluids; 40% of patients showed signs of penetration, aspiration or pharyngeal residue for both solids and fluids. Finally, in the group of patients with severe muscle disease activity, 88% of patients showed an impairment in the oral phase of swallowing for solids and 50% for fluids; 63% had a reduction in the activation of the pharyngeal phase of swallowing for solids while 50% for fluids; 75% of patients showed signs of penetration, aspiration or pharyngeal residue for both solids and fluids. The 15% of all patients (3 cases, 1 from each group of muscle activity) showed a dysfunction in the upper esophageal sphincter. Of note, 3 patients (15%; 1 with moderate and 2 with severe muscle disease activity) required nutrition through nasogastric tube.

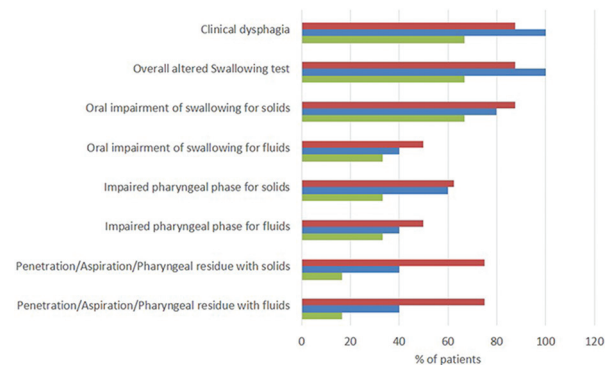


Figure 1: swallowing alteration for solids and fluids in patients divided by muscle disease activity: no signs of muscle disease activity (green), moderate muscle disease activity (blue) and severe muscle disease activity (red).

Conclusion: We showed that FEES study identified swallowing dysfunctions in both the oral and pharyngeal phases of swallowing. Swallowing dysfunctions were more prevalent in patients with greater muscle involvement; however, alterations were not rare also in patients with no clinical signs of muscle activity and, in particular, in the few patients without reported symptoms of dysphagia. FEES appears as a useful tool for the evaluation of dysphagia in IIMs.

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SAT0287

SERUM CYTOKINE PROFILE IDENTIFIES PATHOMECHANISM AND EFFICIENT BIOMARKERS OF DISEASE ACTIVITY AND PROGNOSIS IN INTERSTITIAL PNEUMONIA COMBINED WITH POLYMYOSITIS/ DERMATOMYOSITIS

Shogo Matsuda¹, Takuya Kotani¹, Takaaki Ishida¹, Youhei Fujiki¹, Takayasu Suzuka¹, Koji Nagai¹, Kenichiro Hata¹, Takeshi Shoda², Kentaro Isoda², Shigeki Makino¹, Tohru Takeuchi¹, Shigeki Arawaka¹.
¹Osaka Medical College, Internal Medicine (IV), Takatsuki, Japan; ²Yodogawa Christian Hospital, Rheumatology, Osaka, Japan

Background: Polymyositis (PM) and dermatomyositis (DM) are idiopathic inflammatory myopathies that mainly involve the muscles, skin, lungs, and heart. PM/DM are frequently complicated by interstitial lung disease (ILD) that causes increased mortality. Anti-aminoacyl tRNA synthetase (ARS) antibody and anti-melanoma differentiation-associated gene 5 (MDA5) antibody are associated with complications of ILD. Anti-ARS antibody-positive PM/DM-ILD responds well to immunosuppressive therapy and has a good short-term prognosis but a high rate of relapse over the long term. In contrast, anti-MDA5 antibody-positive PM/DM-ILD responds poorly to immunosuppressive therapy, and its prognosis is poor. Recently, a number of cytokines have been implicated in the pathomechanism and

outcome of PM/DM-ILD. High levels of serum IL-6, IL-8, IL-10, IL-18, CCL2, CXCL10, TNF- α , and IFN- α were detected in PM/DM-ILD cases, suggesting the pathological involvement of activated macrophages, type 1 T helper (Th1) cells, and neutrophils (ref 1, 2). However, investigation of the pathomechanism using serum cytokines remains insufficient in PM/DM-ILD. We hypothesised that multiple inflammatory cytokine pathways related to the above inflammatory cells would be involved in the pathomechanism of PM/DM-ILD.

Objectives: We measured serum cytokine levels before and during treatment of patients with PM/DM-ILD and examined the associated pathomechanism.

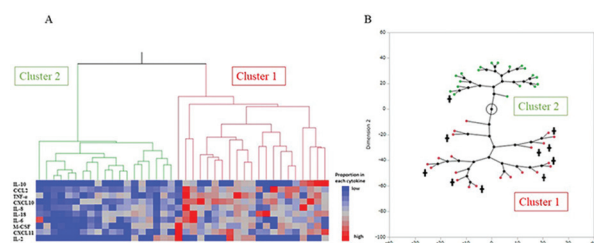
Methods: Serum cytokines were collected from 40 PM/DM-ILD patients. Principal components analysis (PCA) and cluster analysis were used to classify patients into subgroups. We compared cytokine profile of the survivors and dead patients as well as anti-MDA5 antibody-associated ILD and anti-ARS antibody-associated ILD. We also examined the association of various cytokines with disease activity indicators and prognosis of ILD.

Results: PCA revealed that the diversity of cytokines was driven by three groups: (1) neutrophilic and M1-macrophage-driven cytokines, (2) Th1 cell-driven and M2-macrophage-induced cytokines, and (3) M2-macrophage-driven cytokine. Based on cluster analysis, patients were classified into two subgroups according to the cytokine levels of all groups (Figure A). Ninety percent of patients who died of ILD were included in clusters with high cytokine levels (Figure B). Serum cytokine levels of all groups were significantly higher in the anti-MDA5 antibody-positive patients than in the anti-ARS antibody-positive patients. Factors of poor prognosis in PM/DM-ILD correlated significantly with serum cytokine levels of groups 1 and 2. Among the 3 groups, serum cytokine levels of group 1 were significantly higher initially and at 2 and 4 weeks in the death group.

Conclusion: These findings suggest that the activation of monocytes, macrophages, and Th1 cells, and neutrophils plays a role in the pathomechanism. Group 1 cytokines could be efficient biomarkers for predicting prognosis of PM/DM-ILD.

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SAT0288

A SYSTEMATIC REVIEW OF SYSTEMIC SCLEROSIS CLINICAL TRIALS SINCE 2005. LACK OF EARLY DISEASE TARGETING, CONFUSION RELATED TO DISEASE DURATION DEFINITION AND LOW LEVEL OF INCORPORATION OF THE NEW CLASSIFICATION CRITERIA

Konstantinos Melissaropoulos¹, Dimitrios Daoussis², Ioannis Antonopoulos¹, Alexandra Filippopoulou¹, Stamatis-Nick Lioussis². ¹Patras University Hospital, Rheumatology, Patras, Greece; ²Patras University Hospital, University of Patras Medical School, Rheumatology, Patras, Greece

Background: It is still unknown whether treatment in very early systemic sclerosis (SSc) can affect long term outcomes.

Objectives: To systematically review clinical trials in SSc during the last 14 years aiming at answering the following questions: 1) how many clinical trials in SSc have targeted early disease and whether treatment of patients with early disease leads to better clinical outcomes, 2) how is disease duration defined in SSc clinical trials and whether data on duration since Raynaud's onset are provided and 3) whether the new 2013 ACR/EULAR SSc criteria have been incorporated in clinical trials throughout the last 5 years.

Methods: A search in published studies indexed in MEDLINE was performed from Jan-2005 to Dec-2018. The *Clinical Trial* filter was activated and the terms "systemic sclerosis treatment" used. Studies were included

if they evaluated adult patients with systemic sclerosis, concerned systemic treatments, presented data for clinical endpoints assessing fibrosis and were prospective in design.

Results: Seventy-three studies with a total number of 3078 patients met the inclusion criteria and were subjected to data extraction. The total weighted mean disease duration (adjusted to the number of patients in each study) was 38.55 months which is more than 3 years, the usual threshold for defining early disease. Baseline total weighted mean MRSS was 21.54 indicating that most patients had full blown fibrotic disease. Only 24 studies (32.9%) recruited early (<36 months disease duration) cohorts, with only 4 studies among them concerning very early disease (<18 months disease duration). We next focused on studies that did not specifically target early disease (n=49) and explored whether disease duration associated with clinical outcomes. We identified such an analysis only in 13 studies. In 8/13 studies there was no difference regarding outcomes according to disease duration. Nevertheless, in 4/13 studies investigators reported better outcomes in patients with shorter disease duration with only one study showing the opposite. Significant heterogeneity was found regarding disease duration definition. Four separate definitions were identified: 1) "From first non-Raynaud's symptom" (49.3%), 2) "From disease diagnosis" (11%), 3) "From skin thickening onset" (5.5%), 4) "From first symptom" (11%). The remaining studies (23.3%) presented no clear definition in their manuscript for disease duration in their cohorts. The stratification of the studies according to year of publication showed a tendency for greater consistency regarding the definition used in recent years since most studies published from 2015 and onwards use the definition "From first non-Raynaud's symptom". Data regarding the duration since Raynaud's onset were available only in 9 studies (12.3%). A separate analysis of all articles published from 2014 until 2018 was performed to explore the incorporation of the new 2013 ACR/EULAR criteria. Only 6 studies (25%) incorporating the new criteria were identified.

Conclusion: 1) The majority of patients recruited in clinical trials throughout the last 14 years do not have early disease, 2) Only one third of the studies were specifically designed to target early disease, 3) The question of whether early implementation of therapy may lead to better clinical outcomes cannot be definitely answered based on existing data, 4) there is confusion related to disease duration definition across SSc clinical trials but an obvious trend towards improvement was evident throughout the last few years and 5) there is still a very low level of incorporation of the new classification criteria in SSc trials.

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SAT0289 THE ROLE OF CAPILLAROSCOPY IN THE EARLY DIAGNOSIS OF SYSTEMIC SCLEROSIS

Atusa movasat Hajhkan, Henry Moruno, Ana Isabel Turrión, Cristina Bohórquez, Ana Pérez Gómez, Fernando Albarrán, Eduardo Cuende, Ana Sánchez Atrio, Lucía Ruiz, Paula Pretel, Laura Barrio, Valentina Emperiale, Adrián Abbasi, Melchor Álvarez de Mon. *Príncipe de Asturias University Hospital, Alcalá de Henares, Madrid, Spain*

Background: Systemic Sclerosis is a multisystem autoimmune disease characterized by fibrosis of the skin and internal organs and vascular damage. The diagnosis is usually made in the late stages, when irreversible damage has already occurred. Recently, we have established new criteria to detect early cases of the disease. Performing capillaroscopy and its findings are crucial in the diagnosis of early systemic Sclerosis or "prescleroderma" and there may be changes seen in capillaries of patients without skin manifestations.

Objectives: To analyze the nailfold capillaroscopy findings in patients with anti-centromere or anti-topoisomerase antibodies and evaluate their usefulness for the diagnosis of systemic sclerosis.

Methods: Of a total of 255 capillaroscopy performed in our Rheumatology department, those patients with positive anti-centromere or anti-topoisomerase were selected. In all cases, capillaroscopy was performed in eight fingers, always by the same observer. The following findings were considered pathological or scleroderma pattern: Local or global capillary loss (> 20%), hemorrhages: two or more in at least two fingers and enlarged capillaries: two or more capillary with double or more caliber in at least two different fingers.

Statistical analysis was performed with SPSS 19.0 program.

Results: The study included 69 patients: 5 (7.2%) males and 64 (92.8%) women, 14 (20.3%) smokers, 47 (68.1%) non-smokers and 8 (11.6%) quitters. The characteristics of the patients included in the study were as follows: 20 patients (30%) were previously diagnosed of systemic sclerosis (19 limited and one diffuse), of which 4 patients had concomitant primary biliary cirrhosis Syndrome (Reynolds), 1 patient with