Conclusion: The period prevalence of AEs recorded during MMF treatment appeared as high as 71.7% in our real life setting. Even though we did not record any serious AEs, the retrospective nature suggests a close monitoring of this therapy in SSC.


OP0064 EVIDENCE-BASED CONSENSUS RECOMMENDATIONS FOR THE IDENTIFICATION AND MANAGEMENT OF INTERSTITIAL LUNG DISEASE IN SYSTEMIC SCLEROSIS

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Background: Interstitial lung disease in systemic sclerosis (SSc-ILD) occurs frequently and carries a high burden of morbidity and mortality. To date, there are no existing guidelines for screening, diagnosis and management of SSc-ILD that would aid early recognition and treatment and improve the care of these patients. Objectives: To develop expert consensus recommendations for the identification and management of SSc-ILD. Methods: Based on the results of a comprehensive systematic literature analysis conducted in line with NICE/CRD and IQWiG guidelines and PRISMA methodology, evidence-based statements on SSc-ILD risk, screening, diagnosis, treatment and follow-up were developed. A modified Delphi process was then used to establish consensus statements for the identification and management of SSc-ILD. Briefly, an expert panel of 27 European-based pulmonologists, rheumatologists and intensivists with experience in treating SSc-ILD was established. Between July and November 2018, the panel took part in 3 rounds of online surveys, a face-to-face discussion and a WebEx meeting to establish consensus-based recommendations for the management of SSc-ILD. Statements were categorised by topic: risk factors (including biomarkers); screening; diagnosis; assessment of severity; treatment utilisation; treatment options; diagnosis and treatment escalation; other management options. Panellists indicated their level of agreement with proposed statements on a scale of 1 (strong disagreement) to 7 (strong agreement), and consensus was considered achieved when ≥80% either disagreed (score of 1–3) or agreed (score of 5–7) with a statement. Based on panel feedback, statements that did not reach consensus were modified and re-voted in later rounds. Results: At the close of the Delphi process, the panel agreed on the following:

1. Risk factors: The presence of anti-topoisomerase I antibodies, male gender and diffuse cutaneous SSC all increase risk for ILD.

2. Screening: All SSc patients should undergo screening for ILD, using HRCT and lung function testing. Frequency of screening using HRTC should be guided by risk of developing ILD, in combination with clinical symptoms and lung function.

3. Diagnosis and assessment of severity: Use of HRTC to diagnose SSc-ILD and assess severity, with supporting findings from lung function testing and thoracic imaging should be recommended.

4. Treatment initiation and options: All patients with severe or progressive SSc-ILD should be considered for pharmacotherapy, with mycophenolate mofetil and cyclophosphamide recommended as treatments. Patients not receiving treatment should be followed closely for signs of disease progression.

5. Disease progression: Indicators of progression include sustained decline in lung function, worsening of clinical symptoms, and change in extent and/or pattern of fibrosis on HRTC.

6. Treatment escalation: Patients with inadequate treatment responses should be considered for treatment escalation. Suitability for lung transplant should be evaluated early, especially for patients diagnosed with advanced disease. Autologous haematopoietic stem cell transplant may be considered in carefully selected patients.

Conclusion: These evidence-based expert consensus recommendations, developed using a modified Delphi process, provide important guidance for the identification and management of SSc-ILD.

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OP0065 THE VERY EARLY DIAGNOSIS OF SYSTEMIC SCLEROSIS (VEDOSS) PROJECT: PREDICTORS TO DEVELOP DEFINITE DISEASE FROM AN INTERNATIONAL Multicentre STUDY

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Background: Early identification of patients is of key importance for the management and treatment of inflammatory rheumatic diseases. Objectives: The aim of the VEDOSS project (1) is to determine through an at-risk population the predictive factors for the progression toward a definite systemic sclerosis (SSc).

Methods: VEDOSS investigators prospectively recruited patients with Raynaud phenomenon (RP), with or without anti-nuclear antibodies (ANA) or for this longitudinal, observational study. Fulfilling the 2013 classification criteria at baseline was an exclusion criterion. Patients with primary Raynaud syndrome were recruited as controls. Patients had an annual assessment according to EUSTAR standards to determine organ involvement and severity. The endpoint was defined as fulfillment of the 2013 classification criteria. The time to fulfilling 2013 classification criteria was evaluated with Kaplan-Meier analysis, and predictors of evolution were determined by univariate and multivariate Cox regression.

Results: 735 patients with RP were recruited into the study. The sample is distributed as follows: i) 237 patients (143 with follow up) RP/ANA negative (ANA-/RP'); ii) 498 patients (401 with follow up) RP/ANA positive (ANA+/RP'); 27 had puffy fingers (PF), 199 had anti-centromere antibodies (Ab) positive, 45 had anti-topoisomerase I Ab positive and 182 had nailfold videocapillaroscopy (NVC) abnormalities at baseline. Out of 401 ANA+/RP' patients, 7.4% within 1 year, 29.3% within 3 and 44.1% within 5 years satisfied the 2013 classification criteria. Out of the 143 ANA-/RP' patients, none (0%) within 1 year, 4.6% within 3 years, and 4.6% within 5 years satisfied SSC criteria. After adjustment for age, the following baseline parameters were identified as independent predictors for progression into definite SSc by multivariate analysis: puffy fingers (OR=3.4 [2.05;6]), anti-centromere ab (OR=2.6 [1.64;1]) and anti-topoisomerase 1 ab (OR=3.1 [1.65;8]), and NVC abnormalities (OR=1.9 [1.32;9]). The presence of RP had a positive predictive value (PPV) of 79% and combination of PF + specific auto-antibodies showed 94% PPV to satisfy ACR/EULAR 2013 criteria within 5 years (p<0.01).

Conclusion: The data show that patients with very early SSc develop definite classification criteria fulfilling SSc within 5 years of follow up. The VEDOSS study

Scientific Abstracts

EFFECTIVENESS OF SPECIALIZED HAND/FACE THERAPY IN PATIENTS WITH SYSTEMIC SCLEROSIS – PRELIMINARY RESULTS OF A ONE-YEAR CONTROLLED STUDY

Maja Šprůtová1, Hana Smucrová2, Sabina Orséka1, Hana Štorkánová3, Barbora Heřmová2, Petr Česká1, Adéla Rathouská2, Olga Růžičková3, Karel Pavelka2, Ladislav Šenolt1, Jiří Vencovský1, Radim Bečvár1, Michal Tomkíč5

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Background: Systemic sclerosis (SSc) is characterized by stiffness and contraction of tissues, which leads to a limitation in the execution of day-to-day activities. The aim of our study was to investigate the impact of specialized physical-occupational therapy (POT) focused on the hands/face and QoL of SSc patients.

Methods: 25 patients were enrolled in the study. Objectives: The aim of our study was to investigate the impact of specialized POT focused on the hands/face and QoL of SSc patients. Parameters were stratified in patients with CTD- or SSc-PAH has not been established.


RESULTS: Patients were assessed by a physician and a physiotherapist blinded to intervention at months 0, 3, 6, and 12. Patients also filled out patient reported outcomes questionnaire and provided blood for routine laboratory analysis and bio-banking. Data analysis was done between groups and within the group.

Results: Compared to the observed statistically significant deterioration in the CG, we found a statistically significant improvement in the IG in objectively assessed function and strength of hand, distance between incisors and functional ability (FIS) – cognitive function. During the follow-up period, there was a significant deterioration or stagnation of the achieved positive results in the IG.

Conclusion: Our program led to a significant improvement in the observed parameters that was clinically significant in a substantial proportion of patients, and prevention of the expected worsening of hand/face handicap and QoL.

Acknowledgement: Supported by AZV-16-33574A, SVV for FTVS UK 2019-260466, MCR 023728.

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Op0067

UTILITY OF RISK STRATIFICATION IN PREDICTING OUTCOMES OF INITIAL MONOTHERAPY VERSUS COMBINATION THERAPY IN PULMONARY ARTERIAL HYPERTENSION ASSOCIATED WITH CONNECTIVE TISSUE DISEASE: A POST-HOC ANALYSIS OF THE AMBITION STUDY

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Background: The AMBITION study (NCT01178073) sponsored by GSK and Gilead demonstrated a reduced risk of clinical failure event in patients receiving initial combination therapy of ambrisentan and tadalafil (COMB) compared with initial monotherapy of each agent (MONO) in treatment-naive patients with pulmonary arterial hypertension (PAH)1. A similar treatment outcome was observed in connective tissue disease (CTD)-PAH or systemic sclerosis (SSc)-PAH subgroups in both the primary analysis set and modified intention to treat (mITT) population2-3. The 2015 ESC/ERS guidelines5 and 2018 WPSh consensus recommended the use of risk assessment (classified as low, intermediate, and high risk) based on the variables to predict prognosis, but the clinical utility of risk stratification in patients with CTD- or SSc-PAH has not been established.

Methods: Patients were assessed by a physician and a physiotherapist blinded to intervention at months 0, 3, 6, and 12. Patients also filled out patient reported outcomes questionnaire and provided blood for routine laboratory analysis and bio-banking. Data analysis was done between groups and within the group.

Results: Compared to the observed statistically significant deterioration in the CG, we found a statistically significant improvement in the IG in objectively assessed function and strength of hand, distance between incisors and functional ability (FIS) – cognitive function. During the follow-up period, there was a significant deterioration or stagnation of the achieved positive results in the IG.

Conclusion: Our program led to a significant improvement in the observed parameters that was clinically significant in a substantial proportion of patients, and prevention of the expected worsening of hand/face handicap and QoL.

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Parameter (unit) | Intervention group | Control group | Intergroup analysis (Friedman–Dunn) | Intragroup analysis (S/WA)
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<tr>
<td></td>
<td>Mean ± SEM</td>
<td>Mean ± SEM</td>
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<tr>
<td>dFTP, dominant hand (cm)</td>
<td>m0: 5.7 ± 0.5</td>
<td>m0: 6.8 ± 0.5</td>
<td>m0-3: p&lt;0.01</td>
<td>m0-3: NS m3:6: NS m0-6: p&lt;0.0001</td>
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<tr>
<td>Hand grip strength, dominant hand (kg)</td>
<td>m0: 17.2 ± 1.8</td>
<td>m0: 16.5 ± 1.2</td>
<td>m3-6: NS m0-6: p&lt;0.001</td>
<td>m6-12: NS</td>
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<tr>
<td>HAMIS, dominant hand</td>
<td>m0: 9.8 ± 1.3</td>
<td>m0: 9.1 ± 1.1</td>
<td>m0-3: p&lt;0.01</td>
<td>m0-3: NS m3:6: NS m0-6: p&lt;0.0001</td>
</tr>
<tr>
<td>Inter-incisor distance (cm)</td>
<td>m0: 2.9 ± 0.2</td>
<td>m0: 3.3 ± 0.1</td>
<td>m0-3: p&lt;0.01</td>
<td>m0-3: NS m3:6: NS m0-6: p&lt;0.001</td>
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<tr>
<td>SHAQ (mm)</td>
<td>m0: 28.8 ± 3.9</td>
<td>m0: 21.5 ± 2.1</td>
<td>m0-3: NS m3:6: NS m0-6: p&lt;0.001</td>
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