progression and skin involvement, we started MP with prednisone (mean dose 8mg. a day), observing no significant changes after treating for 8 months on skin or lung disease, only improving the hands edema.

Case: A woman diagnosed with diffuse SSC, having telangiectasias, scleroderma, positive ATA and NSIP. At 100 months from diagnosis, after 6 cyclophosphamide cycles followed by azathioprine (both ineffective) due to lung function (FVC 76%, FEV 83%, DLCO 26%) and skin (mRSS 30) worsening, MP and prednisone (mean dose 15mg. a day) were started. Currently, we are still waiting to assess clinical response.

Conclusion: Despite it is not included in EULAR current recommendations for SSC complications, our patients have remarkably improved with MP, especially skin involvement, with a good safety profile. These data reaffirm those obtained in previous studies and encourages us to continue considering its use.

Disclosure of Interests: None declared

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Background: Macrovascular involvement and cardiovascular (CV) risk have not been sufficiently studied in patients with mixed connective tissue disease (MCTD). In particular, the gold standard assessment method of aortic stiffness carotid-femoral pulse wave velocity (cfPWV) has never been evaluated in patients with this disease.

Objectives: Aims of the present study were to examine cfPWV in MCTD and to evaluate its associations with MCTD associated parameters and traditional CV risk factors.

Methods: cfPWV measurements were performed in 43 MCTD patients and 107 healthy controls. The difference between cfPWV in the two groups was statistically examined and subsequently controlled for the effect of possible confounding factors. Association of cfPWV with MCTD associated organ involvement, routine laboratory parameters and immunoserological markers was also evaluated. Finally, relationship of cfPWV with medications and traditional CV risk factors was examined.

Results: Adjusted statistical analyses for confounding factors showed significantly higher cfPWV values in MCTD patients in comparison to controls (p.adj<0.001). cfPWV correlated in both the patients and the control group significantly with age (rho=0.69, p<0.001 and rho=0.67, p<0.001 respectively), diastolic arterial pressure (rho=0.024 and p.adj=0.032 respectively) and mean arterial pressure (rho=0.44, p=0.004 and rho=0.49, p=0.001 respectively). Moreover, cfPWV correlated in the control group with systolic arterial pressure (p.adj<0.001) (Fig. 1). Higher cfPWV values could be documented in the subset of MCTD patients without lung involvement (p=0.049).

Conclusion: To our knowledge, this is the first study to show that patients with MCTD have significantly higher aortic stiffness and thus CV risk in comparison to controls. Except the disease itself, age and blood pressure were the main predictors of cfPWV.

REFERENCES

Disclosure of Interests: Konstantinos Triantafyllias: None declared, Michele De Blasi: None declared, Freya Lütgendorf: None declared, Lorenzo Cavagna: None declared, Marco Stortz: None declared, Julia Weinmann-Menke: None declared, Stavros Konstantinides: None declared, Peter Galie: None declared, Andreas Schwarting: Grant/research support from: GSK, Pfizer, AbbVie, Novartis, Roche, Speakers bureau: GSK, Novartis
REFERENCES

Disclosure of Interests: Anniek van Roon: None declared, Arle Van Roon: None declared, Alja J. Stel: None declared, Hendrika Bootma Grant/research support from: Unrestricted grants from Bristol-Myers Squibb and Roche, Consultant for: Roche, Bristol-Myers Squibb, Novartis, Medimmune, Union Chimique Belge, Speakers bureau: Bristol-Myers Squibb, Novartis, Andries Smit Shareholder of: Has been co-founder, and is still shareholder of Diagnostics Technologies, the company which developed the AGE reader., Douwe J Mulder Grant/research support from: My University has received the AGE reader., Douwe J Mulder: Shareholder of: Has been co-founder, and is still shareholder of Diagnostics Technologies, the company which developed the AGE reader.

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Figure 1. Percentage of patients with and without restoration of perfusion in all fingers during 10 minutes of recovery, measured in one hand by photoplethysmography.


AB0693

DUROMETRY: HARD FACTS IN SYSTEMIC SCLEROSIS – A SYSTEMATIC LITERATURE REVIEW

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Background: Fibrosis represents one of the main characteristics of systemic sclerosis (SSc), with skin and internal organ involvement being its main clinical expressions. The modified Rodnan Skin Score (mRSS), using clinical palpation to estimate skin thickness, is considered the current ‘gold standard’ to measure skin involvement in a semi-quantitative way. The mRSS has been judged as fully valid through the Outcome Measures in Rheumatology Clinical Trials (OMERACT) Filter, however a high risk of observer bias exists. Therefore, a new challenge for the SSc community is to define a reliable tool, able to more precisely investigate skin involvement in SSc patients, which would be of great value in clinical trials. Durometry, able to objectively investigate skin hardness, might address this need.

Objectives: To appreciate the validation status of durometry in SSc patients, guided by a systematic literature review.

Methods: Relevant full-text articles using durometry in SSc patients were identified through a systematic literature search in PubMed, EMBASE and Web of Science, according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. Besides this systematic search, an additional hand search was performed through reference list screening. All retrieved records were screened by two raters based on title, abstract and full-text level to finally include manuscripts eligible for quality appraisal and data extraction. The finally included manuscripts were analysed according to the OMERACT Filter 2.0.

Results: The systematic search yielded 94 records, of which 50 were unique. Seven records were retained for full-text review and analysis, comprising one randomised clinical trial (RCT). The pillar feasibility was well documented in 3 studies [1-3]. Of note, the credibility (i.e. having face validity) of durometry was considered to be valid, as was stated by Kissin et al. [1]. Concerning the ‘pillar’ of construct validity, durometry was confirmed as a reliable tool, able to more precisely investigate skin involvement in SSc patients. In 5 studies (correlation coefficients ranging 0.59-0.81) [1, 3-6], construct validity was confirmed as a moderate to high significant correlation with the total mRSS was documented in 5 studies (correlation coefficients ranging 0.59-0.81) [1, 3-6]. Concerning the pillar discrimination, the sensitivity to change in the context of one RCT was confirmed [3], inter-rater reliability was confirmed as intra-class correlation coefficients (ICC) ranged from 0.61-0.91 in 2 studies, of which one RCT [1, 3], intra-rater reliability has not been confirmed as this was domain was only investigated in a cohort 5 SSc patients (ICC 0.86-0.94) and thus solid evidence was lacking [1]. The sensitivity to change in situations of change (i.e. change over time, discrimination between SSc patients or between SSc patients and controls) was not confirmed, since solid evidence was lacking in literature.

Conclusion: Current systematically identified evidence suggests partial validation of durometry in SSc patients according to the OMERACT Filter 2.0. Further dedicated studies are needed to completely validate this tool in SSc, more specifically concerning the pillar ‘discrimination’.

REFERENCES

Disclosure of Interests: None declared


AB0694

IS DUROMETRY A RELIABLE TOOL TO INVESTIGATE SKIN HARDNESS IN SYSTEMIC SCLEROSIS? A PILOT STUDY AND SYSTEMATIC LITERATURE REVIEW

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Background: Fibrosis of skin and internal organs is a hallmark feature of systemic sclerosis (SSc). The modified Rodnan Skin Score (mRSS) is routinely used to measure skin involvement, however this method has only an inter-rater reliability ranging from 0.38-0.92 and intra-rater reliability ranging from 0.74-0.76. Hence, finding a more reliable tool, capable of investigating skin involvement in SSc patients more reliably and precisely, would be of great significance. Durometry has been proposed as an objective technique to investigate skin hardness.

Objectives: To identify and critically appraise literature on the reliability of durometry in SSc patients by a systematic literature review and to perform a pilot study to investigate the intra-rater reliability of durometry in SSc patients.

Table 1: Validation of durometry according to the OMERACT Filter 2.0

<table>
<thead>
<tr>
<th>Pillar</th>
<th>Subdomain</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Truth</td>
<td>Face validity</td>
<td>[1]</td>
</tr>
<tr>
<td>Truth</td>
<td>Construct validity</td>
<td>[1, 3, 6]</td>
</tr>
<tr>
<td>Discrimination</td>
<td>Sensitivity to change in the context of a RCT</td>
<td>[1]</td>
</tr>
<tr>
<td>Discrimination</td>
<td>Sensitivity to change over time</td>
<td>[1]</td>
</tr>
<tr>
<td>Discrimination</td>
<td>Discrimination between disease subsets</td>
<td>[1, 3]</td>
</tr>
<tr>
<td>Discrimination</td>
<td>Discrimination between SSc and HC</td>
<td>[1, 3]</td>
</tr>
<tr>
<td>Feasibility</td>
<td>Stability in situations of no change</td>
<td>Inter-rater reliability</td>
</tr>
<tr>
<td>Feasibility</td>
<td>Inter-rater reliability</td>
<td>[1]</td>
</tr>
</tbody>
</table>

Conclusion: Current systematically identified evidence suggests partial validation of durometry in SSc patients according to the OMERACT Filter 2.0. Further dedicated studies are needed to completely validate this tool in SSc, more specifically concerning the pillar ‘discrimination’.