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 6: woman diagnosed at 47 years old 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.

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AB0691

INCREASED CARDIOVASCULAR RISK IN MIXED CONNECTIVE TISSUE DISEASE: EVALUATION OF MACROVASCULAR INVOLVEMENT AND ITS PREDICTORS BY AORTIC PULSE WAVE VELOCITY

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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) (1) 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 ($p_{adj}=0.024$ 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.

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Figure 1 (a, b, c, d). Correlations of cfPWV (m/s) with age (years) and MAP (mm Hg) in the control and the MCTD group (all; * $p < 0.05$).

cfPWV carotid femoral pulse wave velocity, MAP mean arterial pressure, MCTD mixed connective tissue disease

AB0692

ASSESSING RECOVERY TIME AFTER COLD CHALLENGE AND THUMB INVOLVEMENT CAN HELP TO RULE OUT SYSTEMIC SCLEROSIS IN PATIENTS PRESENTING WITH RAYNAUD'S PHENOMENON

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Background: Distinguishing primary Raynaud's phenomenon (PRP) from Raynaud's phenomenon secondary to systemic sclerosis (SSc) is crucial in the early detection of SSc. Recently we reported that patients with more severe vasculopathy suffer from a prolonged ischemia time during Raynaud's attack. [1] Additionally, it appears that the thumb is more frequently involved in SSc, where it seems to be spared in PRP. [2] These two characteristics are easily recognized by patients and physicians, and can help rising awareness for SSc.

Objectives: The aim was to study if the recovery of a Raynaud's attack and involvement of the thumb are differentiators for SSc in patients with Raynaud's phenomenon (RP).

Methods: A stepwise cooling and recovery procedure was performed, provoking an RP attack, in patients with PRP and SSc. One hand was submerged up till the radiocarpal joint in water. The water temperature was lowered in steps of 3 degrees Celsius every four minutes, from 33 until at least 9 degrees Celsius. Afterwards ten minutes of recovery in room air of 23 degrees Celsius was observed. During the procedure perfusion of the fingertips was assessed by photo-electric plethysmography.

Results: In total 18 patients with SSc and 68 patients with PRP underwent the procedure. Seventeen (94%) SSc patients had no restoration of perfusion after ten minutes in one or more fingers, compared to 28 (41%) PRP patients (figure 1), with a negative predictive value of 98%. During cooling, 17 (94%) SSc patients developed abnormal perfusion in the thumb compared to 48 (71%) PRP patients ($p=0.036$), with a negative predictive value of 95%. There was no difference in involvement of the other fingers during cooling (all $p>0.05$). Positive predictive values were low.

Conclusion: In patients with RP, when there is restoration of perfusion in all fingers after ten minutes or when the thumb is spared, the presence of an underlying SSc is very unlikely. Although this objective measurement needs to be verified with patient's reports, these results suggest that these simple signs can help physicians to assess if the patient needs to be referred for additional tests.

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Abstract AB0692 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.

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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 truth: content validity was confirmed as durometry correlated well with histological findings (i.e. myofibroblast score and hyalinized collagen, $r=0.69$ and 0.78 respectively) [4], 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 intraclass 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.

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

Pillar	Subdomain	Reference
Truth	Face validity	[1]
	Content validity	[4]
	Construct validity	[1,3-6]
Discrimination	Sensitivity to change in the context of a RCT	[3]
	Detecting change in situations of change	
	- Sensitivity to change over time	[1]
	- Discrimination between disease subsets	[1,5]
	- Discrimination between SSc and HC	[1,2]
Feasibility	Stability in situations of no change	[1]
	Intra-rater reliability	[1,3]
	Inter-rater reliability	[1,3]

Green: a criterion was judged valid, if appropriate evidence from studies including SSc patients indicated that durometry met the requirements to be valid. **Amber:** a criterion was judged partially valid, if appropriate evidence from studies including SSc patients indicated that durometry only partially met the requirements to be valid. **White:** a criterion was judged with a white rating if appropriate evidence from studies including SSc patients was lacking. Exceptions are face validity and feasibility, in general evaluated by the judgement of experts as an appropriate measure rather than by specific studies.

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'.

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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.