REFERENCES

- [1] van Roon AM, Smit AJ, van Roon AM, Bootsma H, Mulder DJ. Digital ischaemia during cooling is independently related to nailfold capillaroscopic pattern in patients with Raynaud's phenomenon. *Rheumatology (Oxford)* 2016;55:1083-1090.
- [2] Chikura B, Moore T, Manning J, Vail A, Herrick AL. Thumb involvement in Raynaud's phenomenon as an indicator of underlying connective tissue disease. *J Rheumatol* 2010;37:783-786.

Disclosure of Interests: Anniëk van Roon: None declared, Arie Van Roon: None declared, Alja J. Stel: None declared, Hendrika Bootsma 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 Diagnostix Technologies, the company which developed the AGE reader., Douwe J Mulder Grant/research support from: My University has received research grants for my research from: Boehringer Ingelheim and Actelion, Speakers bureau: My University has received speakers fee from: Sanofi

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.

DOI: 10.1136/annrheumdis-2019-eular.2739

AB0693 DUROMETRY: HARD FACTS IN SYSTEMIC SCLEROSIS – A SYSTEMATIC LITERATURE REVIEW

^{1,2}Amber Vanhaecke^{*}, Maurizio Cutolo³, Sven Verschuere¹, Sabrina Padino³, Carmen Pizzorni³, ^{1,2}Vanessa Smith⁴, ¹Ghent University, Department of Internal Medicine, Ghent, Belgium; ²Ghent University Hospital, Department of Rheumatology, Ghent, Belgium; ³IRCCS San Martino Polyclinic Hospital, University Of Genoa, Research Laboratory And Academic Division Of Clinical Rheumatology, Department Of Internal Medicine, Genoa, Italy; ⁴VIB Inflammation Research Center (IRC), Unit for Molecular Immunology and Inflammation, Ghent, Belgium

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'.

REFERENCES

- [1] Kissin & Schiller. *Arthritis Rheum* 2006;55(4):603-9
- [2] Falanga. *J Am Acad Dermatol* 1993;29(1):47-51
- [3] Merkel. *Arthritis Care Res* 2008;59(5):699-705
- [4] Kissin & Merkel. *Arthritis Rheum* 2006;54(11):3655-60
- [5] Moon. *Rheumatol Int* 2012;32(8):2465-70
- [6] Wiese. *Arthritis Care Res* 2014;66(11):1731-9

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.2564

AB0694 IS DUROMETRY A RELIABLE TOOL TO INVESTIGATE SKIN HARDNESS IN SYSTEMIC SCLEROSIS? A PILOT STUDY AND SYSTEMATIC LITERATURE REVIEW

^{1,2}Amber Vanhaecke^{*}, Maurizio Cutolo³, Sven Verschuere¹, Greta Pacini³, Veronica Vilela⁴, ^{1,2}Vanessa Smith⁵, ¹Ghent University, Department of Internal Medicine, Ghent, Belgium; ²Ghent University Hospital, Department of Rheumatology, Ghent, Belgium; ³IRCCS San Martino Polyclinic Hospital, University Of Genoa, Research Laboratory And Academic Division Of Clinical Rheumatology, Department Of Internal Medicine, Genoa, Italy; ⁴Universidade do Estado do Rio de Janeiro (UERJ), Disciplina de Reumatologia, Rio de Janeiro, Brazil; ⁵VIB Inflammation Research Center (IRC), Unit for Molecular Immunology and Inflammation, Ghent, Belgium

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.

Methods: Relevant full-text articles using durometry in SSc patients were identified through a systematic literature search in 3 electronic databases and a hand search of reference lists, according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines. All retrieved articles were screened on title, abstract and full-text level.

Additionally, a pilot study was conducted in a cohort of non-selected consecutive SSc patients to assess the intra-rater reliability of durometry, by having an anchor rater performing 3 cycles of durometry measurements at 17 skin sites approximating those of the mRSS. Reliability was described by calculating the intraclass correlation coefficient (ICC).

Results: A total of 94 records were identified through the systematic search of which a total of 40 unique titles were screened. After a thorough screening of all titles and abstracts, 7 manuscripts were eligible for full-text review. Finally, of these, only 2 manuscripts fulfilled the predetermined inclusion criteria to be included, as they documented the reliability of durometry as an outcome. Hence, both manuscripts were included in the final analysis for quality appraisal and data extraction. Regarding intra-rater reliability, Kissin et al. reported ICC values ranging from 0.86-0.94 in a small cohort study including 5 diffuse SSc patients (DcSSc) [1]. Regarding inter-rater reliability, ICC values ranged from 0.82-0.96 according to a multicentre randomised clinical trial (n=43 DcSSc) by Merkel et al. and 0.61-0.85 according to Kissin et al. [1,2]. Finally, only 5 fulfilled the predetermined eligibility criteria to be included for quality appraisal and data extraction.

Subsequently to this systematic review, an additional pilot study investigating the intra-rater reliability was performed. Seventy-four SSc patients (61 women; 13 LSSc, 53 LcSSc and 8 DcSSc) underwent durometry assessment in this pilot study. The ICC values for intra-rater reliability ranged between 0.93-0.99.

Conclusion: The systematic literature review revealed that reliability, in particular intra-rater reliability, of durometry in SSc is nearly unexplored territory. The findings of our very pilot study, demonstrating as first an excellent intra-rater reliability of durometry measurements in a large unselected cohort of SSc patients, could confirm the explorative findings previously reported in a small cohort study. Hence, our results stepped forward to the need for intra-rater validation of durometry.

REFERENCES

- [1] Kissin. *Arthritis Rheum* 2006;55(4):603-9
- [2] Merkel. *Arthritis Care Res* 2008;59(5):699-705

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.2547

AB0695

LONG-TERM OUTCOMES OF AFRICAN AMERICAN PATIENTS WITH SYSTEMIC SCLEROSIS-RELATED INTERSTITIAL LUNG DISEASE

¹Elizabeth Volkmann¹, Li Ning¹, Dinesh Khanna², Philip Clements¹, Daniel Furst¹, Shervin Assassi³, Michael Roth¹, Robert Elashoff¹, Donald Tashkin¹. ¹University of California, Los Angeles, Los Angeles, United States of America; ²University of Michigan, Ann Arbor, United States of America; ³University of Texas Health Science Center at Houston, Houston, United States of America

Background: Observational studies have demonstrated that African American patients with Systemic Sclerosis (SSc) have a more unfavorable prognosis compared with non-African Americans. However, no studies have evaluated racial disparities using data from a randomized controlled trial (RCT) where all patients have equal access to care and standard treatment and follow-up during the trial.

Objectives: To compare morbidity and mortality in African American and non-African American patients who participated in the Scleroderma Lung Study (SLS) I and II.

Methods: SLS I randomized 158 SSc patients with interstitial lung disease (ILD) from 13 US SSc centers to 1 year of oral CYC (cyclophosphamide) versus placebo. SLS II randomized 142 SSc-ILD participants from 14 US SSc centers to 1 year of oral CYC, followed by 1 year of placebo, versus 2 years of mycophenolate (MMF). Up to 12 (SLS I) and 5 (SLS II) years after randomization, we contacted enrolled patients or designated surrogates to assess the following: mortality, cause of mortality, and development of organ failure. We used cox proportional hazard modeling to determine the variables associated with survival.

Results: Baseline characteristics of the SLS I and II cohorts were similar. In SLS I, African American participants (N=26) were younger than non-African American participants (N=132) (43.1 vs. 49.5 years, P=0.015). In SLS II, African American participants (N=33) had slightly increased

baseline forced vital capacity (FVC) (69.2 vs 65.6, P=0.038), compared with non-African American participants (N=109). There were no significant baseline differences in the extent of cutaneous sclerosis (Modified Rodnan Skin Score [MRSS]), presence of diffuse cutaneous disease or serological profiles between racial groups. After adjusting for age, MRSS, FVC, there was no difference in long-term mortality outcomes (due to all causes or due to respiratory failure) in African American versus Non-African American SSc-ILD participants in SLS I or II (Figure 1). There was also no difference in time to the development of respiratory failure in African American versus Non-African American SSc-ILD participants in SLS I or II.

Conclusion: Data from two of the largest RCTs in SSc-ILD demonstrated that African American patients with SSc-ILD have similar morbidity and mortality outcomes compared with non-African American SSc-ILD patients, even after adjusting for age and baseline disease severity. These findings contrast with the racial disparities described in previous observational studies and warrant further investigation.

REFERENCES

- [1] Tashkin, et al. *NEJM* 2006;354:2655.
- [2] Tashkin, et al. *Lancet Resp Med* 2016;4:708.

Figure 1a. Time to death in African American participants of SLS I (red line) and non-African American participants of SLS I (blue line).

Figure 1b. Time to death in African American participants of SLS II (red line) and non-African American participants of SLS II (blue line).

Disclosure of Interests: Elizabeth Volkmann Shareholder of: Pfizer, Inc., Consultant for: Boehringer Ingelheim, Speakers bureau: Boehringer Ingelheim, Ning Li: None declared, Dinesh Khanna Shareholder of: Eicos Sciences, Inc, Grant/research support from: Bayer, BMS, Pfizer, Horizon, Consultant for: Actelion Acceleron, Arena, Bayer, BI, BMS, CSL Behring, Corbus, Cytrol, GSK, Genentech/Roche, Galapagos, Employee of: Elcos Sciences, Inc, Philip Clements: None declared, Daniel Furst Grant/research support from: F. Hoffmann-La Roche, Genentech, Shervin Assassi: None declared, Michael Roth Grant/research support from: Genentech/Roche, Robert Elashoff: None declared, Donald Tashkin Consultant for: EMD Serono

DOI: 10.1136/annrheumdis-2019-eular.1380