Background: It is estimated that the gastrointestinal (GI) tract is involved in about 90% of patients with Systemic Sclerosis (SSc).¹ The UCLA Scleroderma Clinical Trials Consortium Gastrointestinal Tract 2.0 (UCLA SCTC GIT 2.0) questionnaire has been validated in several countries as a useful tool in the assessment of GI symptoms in this pathology.²

Objectives: To assess whether the application of the questionnaire has an added value in clinical practice by comparing the frequency of GI symptoms found during the clinical evaluation in the consultation vs. application of the questionnaire.

Methods: A cross-sectional study was carried out in a cohort of patients with SSc. During the consultation, patients were asked about the presence of GI symptoms and the UCLA SCTC GIT 2.0 questionnaire was handed in for completion. Subgroups that analyse reflux, bloating, dirt, diarrhea and constipation were evaluated and their responses were transformed into dichotomous variables (present/absent). Clinical data was obtained and analysed.

Results: 27 patients were included, most of them female [n = 23 (85.2%)], with an average age of 53.3 ± 13.5 years. All patients met the classification criteria of Leroy/Medsgers of 2001 or ACR/EULAR of 2013: 14 (51.9%) had limited ES, 6 (22.2%) had Overlap Syndrome, 4 (14.8%) had diffuse ES and 3 (11.1%) had early SS. The prevalence of symptoms in all evaluated subgroups was higher in the questionnaire than in the clinical evaluation, with a statistically significant difference in the reflux subgroup [n = 16 (59.3%) vs. n = 13 (48.1%), p = 0.018]. In the remaining subgroups, this difference, although not statistically significant, was also found [abdominal distension n = 20 (74.2%) vs. n = 1 (3.7%), p = 1.0; dirt n = 2 (7.4%) vs. n = 0 (0%); diarrhea n = 22 (91.7%) vs. n = 3 (11.1%), p = 1.0; constipation n = 11 (40.7%) vs. n = 4 (14.8%), p = 0.273]. In the clinical evaluation, 12 patients said they were asymptomatic from the GI point of view, but all of them reported some GI symptoms when filling out the questionnaire. The subgroups in which the disagreement between the answers during the clinical evaluation and the filling out of the questionnaire were more frequent were distension [n = 19 (70.4%) and diarrhea [n = 19 (79.2%)].

Conclusion: The results of the present study reinforce the usefulness of the UCLA SCTC GIT 2.0 questionnaire, usually more applied in clinical trials, in clinical practice, concluding that it allows to find more GI symptoms in patients with SSc than the clinical evaluation in a consultation, this difference being statistically significant in the subgroup that evaluates reflux. The considerable difference found in the subgroups that assess symptoms of the low GI tract, such as diarrhea and constipation, although non statistical significant, may be due to the fact that the patient does not feel comfortable talking about it, a struggle that can be overcome with application of the questionnaire. On the other hand, this tool can also be a means of quantifying the severity of symptoms, monitoring their progress and making the consultation time more profitable. More studies with larger samples are needed to continue studying its role in clinical practice.

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AB0418

SHEAR WAVE ELASTOGRAPHY FOR THE ASSESSMENT OF LIVER FIBROSIS IN PATIENTS WITH SYSTEMIC SCLEROSIS AND SYSTEMIC SCLEROSIS/PRIMARY BILIARY CHOLANGITIS OVERLAP COMPARED TO PRIMARY BILIARY CHOLANGITIS

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Background: Systemic sclerosis (SSc) is an autoimmune disease characterized by vasculopathy and skin as well as organ fibrosis. The lungs, skin, and gastrointestinal care frequently affected. Primary biliary cholangitis (PBC) is an autoimmune disease of the liver associated with potential progression to liver fibrosis. More recently, these two disorders have been described as an overlapping entity, especially in patients with limited, anti-centromere (CENP-B) positive SSc (1). Here, we report the first results of a pilot study of shear wave elastography (SWE) in patients with SSc and SSc/PBC compared to PBC patients.

Objectives: To assess liver stiffness in patients with SSc, SSc/PBC overlap, and PBC with shear wave elastography.

Methods: We analyzed a sample of 10 SSc to 11 PBC patients. In all patients, a baseline US examination of the liver and liver stiffness measurement by 2D SWE were performed using a GE logiq E10 ultrasound machine. Normal values for

SWE in healthy people have recently been published (2). Liver stiffness measurement was performed according to the recommendations of the manufacturer and the recommendations of the current "EFSUMB Guideline and Recommendation on the Clinical Use of Liver Elastography" (3). In addition, age, body mass index (BMI), and antibody profiles were assessed.

Results: Of 8 SSc patients without PBC, 6 were anti-CENPB pos., 1 had Scl70, and 1 PmScl antibody. Median age was 59.5 (47-71). Median BMI was 23.1 (19.6-25). 1 patient had SSc/PBC overlap, 1 had hepatic steatosis. 1 was positive for CENP-B/AMA-M2 antibodies, the other patient was Scl70 positive. Median age of these 2 patients was 55.5 (55-56), BMI was 23.86 (20.9-26.8). 11 patients with PBC were positive for AMA-M2 antibodies, median age was 58 (41-78) years, BMI was 27.8 (15.8-50.3). The differences were not statistically different. Liver stiffness is expressed in kPa. Measurements had an interquartile range/median ratio <30%, indicating sufficient quality of the measurement. Liver stiffness was higher in patients with SSc/PBC overlap/hepatic steatosis (9.4 ± 0.18 , $**p=0.0133$) and PBC alone (7.359 ± 2.51 , $**p=0.0163$) compared to SSc alone (4.526 ± 0.89 kPa). The results indicate that PBC is the main driver of liver stiffness in patients with SSc, and SSc alone may not necessarily lead to an increased liver stiffness (figure 1).

Figure 1.

Conclusion: Our results indicate that SWE is a useful tool in for the non-invasive assessment of liver stiffness in SSc and SSc/PBC overlap. We will further increase the sample size, especially of patients with SSc/PBC overlap. Of note, other liver diseases, such as hepatic steatosis, have to be kept in mind when SWE is performed as they may contribute to liver stiffness.

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AB0419

ACCESS TO TREATMENT FOR RAYNAUD'S PHENOMENON AND DIGITAL ULCERS FOR PATIENTS WITH SYSTEMIC SCLEROSIS DOESN'T FOLLOW EULAR/EUSTAR GUIDELINES

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Background: Patients with Raynaud's Phenomenon (RP) from systemic sclerosis (SSc) may experience severe complications. Digital ulcers (DUs), occur in approximately half the patients with SSc, and cause hand dysfunction, severe pain, and decreased quality of life. DUs lead to increased healthcare utilization and systemic economic burden through hospitalizations, ED visits, and ambulatory services (1). However, access to medications such as PDE5 inhibitors and prostacyclins that are within the EULAR/EUSTAR SSc guidelines (2) in a country with global health care but patchy pharmacare such as Canada has not been studied.

Objectives: The purpose of this study was to elucidate the access to treatment of medications for RP and DU in patients with SSc in Canadian provinces through identifying the provincial and private insurance coverage of PDE5 inhibitors (PDE5i) and prostanoids, the timelines and procedures of requesting these medications, and the process of administering IV prostanoids if required for patient care.

Methods: We designed an online survey and collected data through the Survey Monkey platform. The survey was administered to rheumatologists affiliated with the Canadian Scleroderma Research Group (CSRG) from December 2020 to January 2021. Responders were asked to report if the province or private insurance automatically provided PDE5i for patients with RP and DU or if a dedicated process was required to attain these medications. Additionally, responders were asked to describe the process of administering Iloprost, Epoprostenol and Alprostadil and the barriers inherent to their administration. Of note, there is no DIN number for Iloprost in Canada so every time it is used there must be an application to Health Canada.

Results: The survey was completed by 100% of CSRG researchers (17/17), representing 8 provinces in Canada. None of the provincial governments provided coverage for PDE5i without special requests that were adjudicated on a case by case basis with approximately half the provinces paying for PDE5i upon special request if a patient was eligible for provincial drug insurance (ex elderly, youths, low income families). Two provinces, Quebec and Saskatchewan, provided PDE5i "all the time". Whereas NS, MB, ON, BC, and AB provided them "sometimes"; NFLD provided them "never". Provincial governments and private insurance fulfilled requests "within 1 month" 62% of the time and the other requests took longer to be answered. Private insurance approved coverage with special request in AB, MB, QC, ON, and NS. Respondents described administration of IV prostanoids as "inconsistent", requiring "a lot of work", and that patients in most jurisdictions be admitted as in-patients for provinces to cover these medications.

Conclusion: Most jurisdiction within Canada do not provide coverage for PDE5i and the process to obtain access for patients is delayed, non-uniform, and often not approved. Intravenous prostanoid infusions are difficult to obtain and have system barriers. Advocacy and cost effectiveness data should be used to advocate for access to medications that are recommended within SSc recommendations.

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AB0420

CIRCULATING MONOCYTES HAVE DISTINCT PHYSICAL PROPERTIES THAT CORRELATE WITH DISEASE ACTIVITY AND SEVERITY AND PREDICT PROGRESSION IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is associated with high morbidity and is one of the autoimmune rheumatic diseases with the highest mortality. However, tools to evaluate disease activity, response to treatment or to predict disease progression are scarce. Dysregulated immune responses are major pathogenic players at the onset and in the progression of SSc. Recent evidence demonstrates that mechanical properties of circulating leukocytes reflect their states and functions, and during activation ensure their adaptation to the changing physical requirements (e.g. softening to extravasate and migrate in the tissues) (1). Real-time fluorescence and deformability cytometry (RT-FDC) is a novel technique that allows the identification of cells from a heterogenous population by marker expression, with their subsequent mechanical phenotyping in a high-throughput manner (2, 3).

Objectives: Here we characterized the physical properties of circulating immune cells in SSc patients, aiming to identify disease-related changes in their phenotypes, clinical correlates of these changes and their potential to predict disease progression.

Methods: 51 patients fulfilling the 2013 ACR/EULAR classification criteria for SSc and 17 age- and sex-matched healthy controls were included in the study. Blood was collected from the donors between 05.2019 and 10.2020. Peripheral blood mononuclear cells (PBMCs) were isolated and stained with antibodies against major circulating lymphoid (CD8⁺, CD4⁺ T cells, B cells, NK cells, NKT-like cells) and myeloid subpopulations (classical, intermediate and inflammatory monocytes, conventional dendritic cells and plasmacytoid dendritic cells). Each subpopulation was identified in RT-FDC by standard gating based on its marker expression and its area, deformation and apparent Young's modulus (a measure of cell stiffness) were determined. The analysis was conducted using a custom Python script. For the patients included, demographic and clinical data were collected at every visit. Correlations with clinical parameters were analyzed in R.

Results: All three subpopulations of monocytes identified by expression of HLA-DR, CD14 and/or CD16 had higher deformation and cross-sectional area in SSc patients as compared to healthy controls. From the SSc patients, monocytes had higher deformation and area in those with diffuse cutaneous SSc, extensive lung fibrosis and active disease as compared to those with limited cutaneous SSc, limited lung fibrosis and stable disease, respectively. Moreover, monocyte deformation and area significantly correlated with the EUSTAR activity index, with mRSS, with the extent of lung involvement on HR-CT (positive correlation), with DLCO and FVC (negative correlation). Follow-up data collected one year after the measurements showed that a higher monocyte deformation and cross-sectional area at baseline predicts future progression of lung disease, defined according to the INBUILD study, as well as future progression of skin fibrosis.

Conclusion: We demonstrated that circulating subsets of monocytes in SSc patients show an increase in deformation and cross-sectional area, that these changes correlate with current disease activity and can identify patients with high risk of future progression of skin or lung fibrosis. These changes might reflect an activated state of circulating monocytes in SSc that facilitate their tissue migration. Mechanical phenotyping of monocytes by RT-FDC might thus serve as a useful tool for clinical evaluation of SSc patients.

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