## AB0620 ORAL HEALTH IN PATIENTS WITH SYSTEMIC SCLEROSIS: AN EUSTAR CENTER EXPERIENCE

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**Background:** Although the orofacial manifestations are commonly reported among patients with systemic sclerosis (SSc), only few studies have adequately addressed the issue of oral health-related quality of life (QoL) in such pathobiological settings.
**Objectives:** The aim was to characterize the oral health status (OHS) of adults with SSc and to compare with the general population.
**Methods:** A cross-sectional prospective observational study in 37 consecutive SSc in a EUSTAR cohort (EUSTAR 162 center) and 37 gender and age-matched controls without SSc.
A standardized oral exam meaning OHS (periodontal, dental, mucosal and microbial), oral health-related behaviors and oral HRQoL (Oral Health Impact Profile, OHIP) were evaluated in all recruited individuals, while oral manifestations such as size of oral aperture, oral dryness, manual dexterity for oral hygiene and HRQoL only in SSc.
Multivariable regression analysis was done to evaluate association between SSc, oral abnormalities, oral health status and QoL.
**Results:** Overall, SSc had significantly reduced oral HRQoL compared with controls ($p<0.05$).
We demonstrated lower resting salivary flow rates ($p<0.05$) and pH ($p<0.05$), reduced maximal mouth opening ($p<0.05$), smaller interincisal distance ($p<0.05$) in SSc group as compared to their controls. Moreover, despite comparable oral health-related behaviors (e.g. same level of daily tooth-brushing), the majority of SSc experienced caries ($p<0.05$) and presented with more periodontal disease ($p<0.05$) comprising periodontal pockets (pocket depth PD>3 mm) ($p<0.05$), clinical attachment level (CAL) $\geq$5.5 mm ($p<0.05$).
Finally, SSc was documented as a significant independent predictor of OHIP, missing teeth, periodontal disease, interincisal distances as shown by multivariate regression ($p<0.05$).
**Conclusions:** SSc patients are at risk to develop impaired oral health and oral HRQoL compared with the general population.
**Disclosure of Interest:** None declared
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## AB0621 DEVELOPMENT OF SYSTEMIC SCLEROSIS IN TRANSGENDERED FEMALES: A CASE SERIES

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**Background:** Scleroderma (SSc) is an autoimmune connective tissue disease with a female preponderance (female to male ratio of 9.7:1) [1]. Sex hormones are thought to play a role in the susceptibility to autoimmune diseases [2].
**Objectives:** We report 3 cases of SSc in male-to-female transsexuals diagnosed following their male-to-female transition
**Methods:** Medical records of 3 patients diagnosed with SSc after male-to-female transition were reviewed. Disease features, hormonal therapies and surgical interventions related to gender reassignment were collected.
**Results:** At our tertiary University Hospital clinical service, 3 male-to-female transsexual patients were diagnosed with SSc following their surgery between May 1997 and October 2016. All 3 patients had started their transition before the onset of the disease and had not been diagnosed with any autoimmune disease before either starting the hormonal therapy required for the transition or before undergoing plastic surgery interventions. The first case was diagnosed with anti-RNA pol III +ve diffuse cutaneous SSc at the age of 35 yearsa year after the first surgical intervention for infected silicone buttock implants and approximately 5 years of hormonal therapy with combination of mestranol and norethisterone (Norinyl-1®). She experienced scleroderma renal crisis 2 years after the diagnosis and severe vascular involvement with frequent and severe digital ulcers (DU), Raynaud's phenomenon (RP), gastroesophageal reflux disease (GERD), pulmonary arterial hypertension and telangiectasiae. She required chronic dialysis and was treated with mycophenolate mofetil (MMF), rapamycin, bosentan and proton-pump inhibitors (PPI). She eventually died 8 years after the diagnosis. The second case was diagnosed at the of 49 years, approximately 6 months following her gender reassignmentand 5 years after having started hormonal therapy initially with conjugated estrogen isolated from pregnant mares (Premarin®) and later with ethinylestradiol and gestodene (Femodene®). She developed ANA –ve, antiCCP antibody and rheumatoid factor positive limited cutaneous SSc/rheumatoid arthritis overlap syndrome and experienced severe DUs, RP, GERD, inflammatory arthritis and pulmonary fibrosis (PF). She was treated with MMF, PPI, calcium-channel blocker (CCB) and low-dose steroid therapy with good response. The third case was diagnosed at the age of 43 years, 2 years after having started triptorelin (Decapeptyl®) and oestradiol valerate. She had not undergone any surgical intervention prior to her diagnosis. She developed anti-PM/Scl +ve limited cutaneous SSc/myositis overlap syndrome and experienced RP, GERD and PF. She has only been treated with PPI and CCB prior to her reconstructive surgery.
**Conclusions:** Although still debated, the role of sex hormones in triggering autoimmunity has been suggested in animal models and human studies [2]. The development of SSc in our case series supports the notion that altered profile of sex hormones may modulate autoimmunity in genetically susceptible individuals with distinct clinical and laboratory characteristics.
**References:**
[1] Barnes J et al. Epidemiology of systemic sclerosis: incidence, prevalence, survival, risk factors, malignancy, and environmental triggers. Curr Opin Rheumatol 2012.
[2] Pennell LM et al. Sex affects immunity. J autoimmun 2012.
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## AB0622 RENAL RESISTIVE INDEX (RRI): PROPOSAL FOR AGE-ADJUSTED CUT-OFF VALUES IN SYSTEMIC SCLEROSIS PATIENTS

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**Background:** Renal resistive index (RRI) by Doppler ultrasound, reflects changes in both renal vascular and tubular-interstitial compartments and systemic vascular compliance related to physiological (age) and pathological conditions among which hypertension, diabetes mellitus, hyperuricaemia, dyslipidaemia and chronic kidney disease play a major role [1]. Because of the age-related changes in RRI reported in literature [2,3] the use of a 0.70 cut-off to detect renal damage, as proposed [4], was questioned: renal injury in younger decades (<60yrs) may occur also for RRI value <0.70 and be underestimated. In systemic sclerosis (SSc), RRI was previously correlated with disease duration, glomerular filtration rate and nailfold-videocapillaroscopy pattern [5–7], although tested on small samples and not investigating the possible confounding role of age-related RRI values.
**Objectives:** to describe RRI in a larger scleroderma population and to test both the fixed 0.70 RRI cut-off and age-adjusted cut-offs in reflecting renal and other disease-related organ damage.
**Methods:** SSc patients attending classified according to ACR/EULAR 2013 criteria were enrolled. Data on renal arteries Doppler ultrasound (RRI), autoantibodies status and biochemical tests for renal function/damage, subset and extent of skin fibrosis, instrumental assessment for internal organ involvement were collected and analysed as appropriate with SPSS vers 20.0. Considering that age-adjusted mean values were higher in the SSc population compared to literature values for the general population, we created SSc-specific age-adjusted pathologic cut-offs dividing our SSc population in quartiles and considering RRI values above the 75th percentile as pathologic (Table 1).
**Results:** 190 SSc patients (age 56.3±15.0 years, disease duration 6±8,20 men) were eligible for the study. In the SSc population significant positive correlations between RRI and age, as well as significant associations between RRI and above mentioned general population comorbidities [1], were confirmed. When considering absolute value of RRI, the 0.70 pathologic cut-off and age-adjusted cut-offs validated in the general population [1], only renal function, systolic PAP, DLCO and late nailfold scleroderma pattern were associated with RRI (Figure 1). Pathologic RRI identified according to age cut-offs could not detect early renal damage, but was significantly associated with various fibrotic [interstitial lung disease (p=0.015), tendon friction rubs (p=0.032), skin fibrosis vs no skin involvement ($p<0.001$),

| Age | Proposed SSc age-adjusted pathologic cutoff |
|---|---|
| 1st quartile, ≤48 years | ≥0,68 |
| 2nd quartile, 49–58 years | ≥0,69 |
| 3rd quartile, 59–67 years | ≥0,75 |
| 4th quartile, ≥68 years | ≥0,77 |

| | | RRI basal value | | | RRI general population cutoff 0,70 | | | proposed SSc age-adjusted pathologic cutoffs | | |
|---|---|---|---|---|---|---|---|---|---|---|
| Variables | value | | | | RRI≥0,70 (85pts) | RRI<0,70 (105 pts) | | yes (61pts) | no (129 pts) | |
| Creatinine Clearance | 86,63±31.34 | | p=-0,352 | p<0.001 | 77,85±28.33 | 94.16±3.87 | P<0.001 | | | N.S. |
| Proteinuria/24h | 84.46±65.66 | | | N.S. | | | N.S. | | | N.S. |
| classification (Very early SSc vs Early SSc+SSc) | 41 vs 159 (21,7 vs 78,3%) | 0,67±0,06 | 0,69±0,07 | p=0,051 | 13 (15%) | 28 (26%) | p=0,052 | 6 (9,8%) | 35 (27%) | p=0,007 |
| classification (SSc vs Very early SSc+ Early SSc) | 125 vs 65 (65,6% vs 34,4%) | 0,70±0,07 | 0,66±0,06 | p=0,002 | 61 (72%) | 64 (61%) | p=0,014 | 51 (83.6%) | 74 (57,4%) | p<0,001 |
| dcSSc+lcSSc | 134 (70,5%) | 0,70±0,07 | 0,67±0,06 | p=0,019 | | | N.S. | 55 (41) | 79 (59) | p<0,001 |
| mRSS | 2 (0-35) | | | N.S. | | | N.S. | 7,70±8,68 | 3,59±6,66 | p=0,001 |
| sPAP | 27,93±10,7 | | ρ=0,336 | p<0,001 | 31,08±13,00 | 25,55±6,05 | p=0,001 | | | N.S. |
| ILD | 72 (37,9%) | | | N.S. | | | N.S. | 31 (50,8%) | 39 (30,2) | p=0,015 |
| FVC | 104±22,1 | | | N.S. | | | N.S. | 99,09±22,14 | 106,12±21,76 | p=0,0141 |
| DLCO | 72,2±20,5 | | ρ=-0,207 | p=0,005 | 68,66±21,14 | 75,23±19,79 | p=0,032 | 67,07±22,57 | 74,72±18,44 | p=0,020 |
| DLCO/VA | 81,8±20,0 | | | N.S. | | | N.S. | | | N.S. |
| DU | 65 (34,2%) | | | N.S. | | | N.S. | 30 (49,2%) | 35 (28,2%) | p=0,006 |
| Late Scleroderma Pattern on NVC | 41 (21,5%) | 0,71±0,06 | 0,68±0,07 | p=0,004 | 24 (28%) | 17 (16%) | p=0,024 | 21 (34,4) | 20 (15,5) | p=0,002 |
| Tendon friction rubs | 9 (4,7%) | | | N.S. | | | N.S. | 6 (9,8) | 3 (2,3) | p=0,032 |
| Scl70 | 59 (31,1%) | | | N.S. | | | N.S. | 25 (40,9) | 34 (26,3) | p=0,044 |
| RNApolIII | 3 (1,6%) | | | N.S. | | | N.S. | 3 (4,9%) | 0 (0%) | p=0,033 |

higher mRSS (p=0.001)] and vasculopathic manifestations [late scleroderma pattern (p=0.002) and digital ulcers (p=0.006)] of the disease (Figure 1).
**Conclusions:** in clinical practice, different age-related or non-related RRI cut-offs must be used when looking for renal or extrarenal SSc-induced damages.
**References:**
[1] Boddi, Intern Emerg Med 2015;2)Boddi, Am J Hypertens 1996;3)Ponte, Hypertension 2014;4)Platt, Am J Roentgenol 1989;5)Rivolta,Arthritis Rheum 1996;6)Rosato, Seminar ARthritis Rheum 2012;7)Rosato, Arthritis Care Res 2014.
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## AB0623 PROGNOSTIC VALUE OF RENAL RESISTIVE INDEX (RRI) IN SYSTEMIC SCLEROSIS: PRELIMINARY DATA FROM A SINGLE CENTRE

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**Background:** Renal Resistive Index (RRI- measured with Renal arteries Doppler ultrasound) is a useful technique to evaluate vascular and tubular-interstitial damage in both general and systemic sclerosis (SSc) population, where increased RRI values correlates with longer disease duration [1], lower glomerular filtration rate and more advanced nailfold-videocapillaroscopy pattern [2]. Moreover, higher RRI values were seen in SSc patients with new occurrence of digital ulcers [3].
**Objectives:** to test the prognostic value of RRI [absolute, ≥0.70 and SSc age-adjusted pathologic value (Table1)] and RRI delta change in predicting general and organ-specific worsening in scleroderma patients.
**Methods:** SSc patients classified according to ACR/EULAR 2013 criteria were enrolled. Demographics data and renal ultrasound data were collected. Data on clinical worsening had been collected as herewith specified: a) Skin worsening as an increase of mRSS≥5 units, b) Peripheral vascular worsening as the appearance of new digital ulcers or the worsening of nailfold videocapillaroscopy scleroderma pattern, c) Lung worsening as decline of FVC≥15% or FVC<80% with new detection of ILD on chest HRCT or worsening of HRCT-ILD extent, d) Cardiac worsening as new onset of left ventricular failure requiring treatments or new onset of PAH confirmed on RHC or detection of severe ventricular arrhythmias on 24h EKG, e) Renal worsening as a new scleroderma renal crisis or reduction of creatinine clearance ≤30 ml/min. General worsening was recorded in case of death due to SSc or for any of the above organ-specific worsening. Data were analysed as appropriate with SPSS vers. 20.0.
**Results:** 190 SSc patients (age 56.3±15.0 yrs, 170 women, disease duration 6±8yrs, 65 with a follow up RRI measurement after 2.8±0.9 years) were enrolled. After a mean clinical follow-up of 3.6±2.6 years, 89 (46.8%) pts showed general worsening. Skin, peripheral vascular, cardiac, lung and renal worsening were detected in 14 (7.4%), 40 (21%), 32 (16.8%), 38 (20%) and 11 (5.8%) patients respectively. We registered 10 (5.2%) deaths and 43 (22.6%) patients with multiple organ worsening. Both absolute value of RRI and ≥0.70 RRI cut-off showed no significant association with organ or global clinical worsening, At the opposite, RRI cut-offs adjusted for age were associated with cardiac worsening (p=0.065 =- Figure 1). When in 65 patients the pattern of delta RRI and clinical worsening were analysed (Table 1), wider RRI changes were associated with general worsening (p=0.029) and cardiac worsening (p=0.006). The significance of these associations increased when sub-analyse was repeated focused on patients with normal SSc age-adjusted RRI values at baseline evaluation (p=0.017 and p<0.001 respectively, Figure 1).

| Age | Proposed SSc age-adjusted pathologic cutoff |
|---|---|
| 1st quartile, ≤48 years | ≥0,68 |
| 2nd quartile, 49–58 years | ≥0,69 |
| 3rd quartile, 59–67 years | ≥0,75 |
| 4th quartile, ≥68 years | ≥0,77 |

| | RRI basal value (190 pts) | | RRI>0,70 (85 pts) | | SSc age-adjusted pathologic (61 pts) | | ΔRRI (available 65 pts) | | ΔRRI in baseline SSc age-adjusted normal RRI (45 patients) | |
|---|---|---|---|---|---|---|---|---|---|---|
| | n (%) | p value | n (%) | p value | n (%) | p value | (n) mean±SD | p value | (n) mean±SD | p value |
| General Worsening | 89 (46,8) | N.S. | 37 (43,5) | N.S. | 31 (50,8) | N.S. | (33)+0,03±0,05 vs (42) 0,00±0,04 | 0,029 | (24)+0,04±0,04 vs (21) +0,01±0,03 | 0,017 |
| Cardiac Worsening | 32 (16,8) | N.S. | 16 (18,8) | N.S. | 15 (24,5) | p<0,001 | (13)+0,05±0,05 vs (52) +0,01±0,04 | p=0,006 | (7)+0,08±0,03 vs (38) +0,02±0,04 | p<0,001 |

**Conclusions:** increase in RRI could be used as a sentinel sign for general and cardiac worsening in SSc patients, especially when age-related RRI variations are taken into account.
**References:**
[1] Rivolta R, Arthritis Rheum. 1996.
[2] Rosato E, Semin Arthritis Rheum. 2012.
[3] Rosato E, Arthritis Care Res (Hoboken). 2014.
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## AB0624 INTERSTICTIAL LUNG DISEASE IN SCLERODERMA: SEVERITY ASSOCIATED FACTORS. OBJETIVES

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**Background:** Systemic sclerosis (SSc) can virtually affect any organ system (such as lungs, kidneys, gastrointestinal tract, and heart). However, it is the pulmonary manifestations that account for the majority of deaths, especially interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH).
**Objectives:** Our aim was to assess the differences between severe and mild-to-moderate ILD in SSc.
**Methods:** A descriptive study was performed, using the available data from the Spanish Scleroderma Study Group (RESCLE). ILD was deemed as serious when forced vital capacity (FVC) was <50%. Patients were classified attending the modified classification criteria proposed by LeRoy and Megdsger.
**Results:** Fourteen referral centers for SSc participated in the registry. By April 2014, 1374 patients with SSc had been enrolled, 541 of whom (39.4%) had ILD, which was severe in 72 of them (13.2%). There were no significant differences as far as sex and age at onset is concerned. Patients with diffuse SSc presented with severe ILD more frequently than those with limited SSc (57% vs. 35%, p=0.002), as well as those who had tested positive for ATA (51% vs. 33%, p=0.005). Additionally, prevalence of FVC<50% was higher in patients with myopathy (32% vs. 15%, p=0.002). Mean FVC was 40.2±6.4 in the severe ILD group, whilst it was 80.3±18.9 in the mild-to-moderate one (p<0.001), and mean DLco was 36.7±15.2 and 62.9±34.5, respectively (p<0.001). Likewise, DLco<70% was also more frequent among patients with severe ILD (100% vs. 69%, p<0.001), as well as mean DLco/VA (56.2±24.2 vs. 74.2±42.0, p=0.002). PAPs was equally higher when FVC<50% (42.2±18.2 vs. 35.1±13.4, p=0.034), and so was the frequency of PAPs>40mmHg (66% vs. 29%, p<0.001) and PAH by right heart catheterism (19% vs. 11%, p=0.050). Finally, by means of a multiple logistic regression, both ATA positivity [OR 0.17 (0.05–0.58), p=0.005] and low DLco [0.93 (0.91–0.95), p=0.000] were found to be related with FVC<50%
**Conclusions:** Patients with ACA positivity and with a limited variant of SSc seem to be at lower risk of severe interstitial lung involvement. Furthermore, the presence of myopathy may contribute to explain the decrease of FVC in SSc patients.
**References:**
[1] Steen VD, Medsger TA. Changes in causes of death in systemic sclerosis, 1972–2002. Ann Rheum Dis 2007; 66: 940–4.
[2] LeRoy EC, Medsger TA, Jr. Criteria for the classification of early systemic sclerosis. J Rheumatol 2001; 28: 1573–6.
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## AB0625 AUTOANTIBODY PROFILE IN PATIENTS DIAGNOSED WITH IDIOPATHIC INFLAMMATORY MYOPATHY: MULTICENTER REGISTRY ON INFLAMMATORY MYOSITIS FROM THE RHEUMATOLOGY SOCIETY IN MADRID, SPAIN

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**Background:** Inflammatory myopathies (IIM) are a heterogeneous group of autoimmune rheumatic diseases characterized by muscle inflammation and progressive weakness. They include any kind of primary inflammatory muscle disease that is not otherwise better explained by metabolic, toxic, infectious, neurologic or inherited causes. The presence of autoantibodies (AA) in IIM is variable and they can recognize nuclear and cytoplasmic cellular components.
**Objectives:** To evaluate the AA profile in patients diagnosed with IIM.
**Methods:** We evaluated 479 patients that included 12 hospitals belonging to the IIM registry of the Rheumatology Society in Madrid (SORCOM-REMICAM) with diagnosis from January 1980 to December 2014. All patients were diagnosed of IIM according to Bohan and Peter criteria. The AAs evaluated were ANA (n=476), anti-Jo1 (n=457), anti-RNP (n=427), anti-MI2 (n=159) and ACA (n=293), according to the standard techniques in the respective laboratories. The presence of ANA was considered valid with at least two positive determinations. The AAs were compared according to the classification I) as dermatomyositis (primary