| Oropharyngeal dysfunction | Present (n=29) | absent (n=5) | p-values ($x^2$-test) |
|---|---|---|---|
| dcSSc | 8 | 0 | 0,027 |
| lcSSc | 22 | 4 | 0,027 |
| disease duration (mean) | 12,7 y | 12 y | 0,462 |
| mRSS<4 | 9 | 2 | 0,322 |
| mRSS>4 | 20 | 3 | 0,322 |
| Raynauds syndrom | 29 | 5 | 0,673 |
| digital ulcers | 19 | 2 | 0,812 |
| lung fibrosis | 11 | 2 | 0,596 |
| eosophageal dilatation | 15 | 2 | 0,566 |
| eosophageal dysmotility | 19 | 0 | 0,015 |
| GI-involvement | 23 | 1 | 0,007 |

**Conclusion:** Logopedic assessment revealed a high incidence of oropharyngeal dysfunction in our cohort of SSc patients. Oropharyngeal dysfunction was not associated with disease duration, skin- or lung-involvement or dcSSc/lcSSc differentiation. A logopedic training program seems to be of benefit for this currently not pharmacologically treatable problem.

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THU0351    **MUSCLE INVOLVEMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS AND ANTI-PM/SCL+ ANTIBODIES IS ASSOCIATED WITH CARDIAC AND PULMONARY INVOLVEMENT. ANALYSIS OF THE MULTICENTRE EUSTAR COHORT.**

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**Background:** Anti-PM/Scl antibodies positivity has been associated with frequent skeletal muscle involvement in patients with Systemic Sclerosis (SSc) in different studies, including the EUSTAR cohort (1). Moreover, although myositis has been previously associated with heart involvement in SSc patients (2), this issue has never been explored among anti-PM/Scl+ patients.
**Objectives:** To evaluate the cardiac involvement in anti-PM/Scl patients with SSc in the large multicentre EUSTAR database, with focus on the subgroup of patients with muscle involvement.
**Methods:** Patients from the EUSTAR database were included when the item anti-PM/Scl was fulfilled in at least one visit.
**Results:** Anti-PM/Scl status was available in 7,353 SSc patients from EUSTAR database: 295 were anti-PM/Scl+. After exclusion of 151 patients with multiple autoantibody positivity, 144 anti-PM/Scl + patients were compared with 7,058 anti-PM/Scl- patients. Among them, 3,120 (44.2%) were positive for ACA, 2,361 (33.5%) for anti-Topo I and 274 (3.88%) for anti-RNAP3.
Regarding the specific cardiac outcomes, in the anti-PM/Scl+ as compared to the anti-PM/Scl- group, a decreased rate of elevated sPAP at ECHO was recorded (12.8% vs 25.0%, p:0.001), while no differences were observed in the frequency of conduction blocks (26.2% vs 23.7%, p:0.526), abnormal diastolic function (33.9% vs 36.4%, p:0.582), pericardial effusion (10.2% vs 10.9%, p:1.000) and LVEF ≤50% (4.76% vs 6.11%, p:0.818). In multivariate analysis, adjusted for age at disease onset, sex, and disease duration, the negative association of anti-PM/Scl with elevated sPAP was not confirmed (p:0.061).
When comparing anti-PM/Scl+ patients with (n=47) and without (n=87) CK elevation, the former group had a higher frequency of conduction blocks (43.2% vs 17.5%, p:0.005; OR 95% CI 3.47, 1.51-7.97) and left ventricular dysfunction, both diastolic (45.6% vs 27.2%, p:0.050; OR 95% CI 2.25, 1.05-4.81) and systolic (LVEF ≤50% 13.3% vs 0%, p:0.018; OR 95% CI 16.8, 0.87-324). Moreover, anti-PM/Scl+ patients with CK elevation had significantly increased rate of lung fibrosis on HRCT (p:0.045), intestinal symptoms (p:0.017), joint contractures (p:0.045) and tendon friction rubs (p:0.034).
**Conclusion:** In the largest series of anti-PM/Scl positive SSc patients so far reported, muscle involvement in anti-PM/Scl+ patients (defined as increased serum CK) seems to represent a marker of a more severe disease phenotype, including a higher frequency of cardio-pulmonary involvement.

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THU0352    **DIAGNOSTIC PERFORMANCES OF HAND ULTRASOUND PARAMETERS AND THEIR IMPACT ON THE 2013 ACR/EULAR CLASSIFICATION CRITERIA FOR SYSTEMIC SCLEROSIS.**

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**Background:** Recent studies have highlighted that ultrasound (US) examination could offer a better assessment of hand manifestations of systemic sclerosis (SSc). Indeed, US allows a simultaneous evaluation of vascular, fibrotic and inflammatory hand features of the disease. Power Doppler US can especially explore macrovascular involvement characterized by an obliteration of digital arteries or ulnar arteries. Ulnar artery occlusion (UAO) is especially frequent in SSc patients and could be a relevant marker of the severity of SSc-associated vasculopathy. Among other hand manifestations of SSc, US evaluation can notably explore tenosynovial involvement such as fibrotic tenosynovitis (TS), which is considered to be SSc-specific.
**Objectives:** This study aims to assess the diagnostic performances of these hand US parameters for the diagnosis of SSc.
**Methods:** 244 patients with suspected SSc were consecutively included. They all had US evaluation assessing the presence of fibrotic TS and UAO. The final diagnosis of SSc was based on the evaluation of an expert, independently from US results and from any pre-established classification criteria.
**Results:** 166 patients were finally diagnosed as SSc. 62 SSc and 8 non-SSc patients had UAO (uni or bilateral) (p=0.001). 23 SSc and 1 non-SSc patient had US fibrotic TS (p=0.007). A US SSc-pattern (presence of UAO and/or fibrotic TS) was reported in 73 SSc patients and 9 non-SSc patients (p<0.001). UAO had an area under ROC curve (AUC) for the diagnosis of SSc of 0.618 (95%CI 0.539-0.697); with Se= 0.373 (0.304-0.449) and spe=0.862

(0.751-0.928). The presence of a US fibrotic TS had an AUC of 0.561 (0.480-0.643); with Se= 0.139 (0.094-0.199) and spe=0.983 (0.909-0.997). The US-SSc pattern had a AUC of 0.641 (0.563-0.695), with Se=0.440 (0.367-0.516) and spe=0.845 (0.731-0.916). When used as a diagnostic tool, the original 2013 classification criteria had an AUC of 0.982 (0.969-0.996) with Se= 0.946 (0.900-0.971) and spe=0.931 (0.836-0.973). Including UAO and fibrotic TS in this classification had few impact (AUC of 0.979 (0.962-0.996) with Se= 0.940 (0.893-0.967) and and spe=0.931 (0.836-0.973)) but allows the substitution of some items (such as capillaroscopy) by US parameters with similar performances for diagnosis.

**Conclusion:** The use of hand US parameters may help to refine the diagnostic strategy of SSc and their inclusion in addition with the items of the ACR/EULAR classification could be discussed.

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| THU0353 | ENDOCAN AND CIRCULATING PROGENITOR CELLS IN SYSTEMIC SCLEROSIS: ASSOCIATION WITH PULMONARY HYPERTENSION |

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**Background:** Systemic sclerosis (SSc) is characterized by early vascular involvement and by varying degrees of fibrosis in skin, lungs, and other tissues. Vascular manifestations include Raynaud's phenomenon, digital ulcers, and pulmonary hypertension (PAH). The prevalence of PAH is 7.85–13% in SSc and it is the second most common cause of mortality in SSc. Circulating CD34+ cell number is associated to cardiovascular health status in several chronic conditions, including inflammatory disease. CD34+ cell number was found inconstantly reduced in SSc. Endocan is a proteoglycan expressed by endothelial cells likely interacting with white blood cells, recently suggested as a marker of vascular stress.

**Objectives:** to evaluate CD34+ cell number and endocan as a markers od cardiovascular involvement in SSc.

**Methods:** Standard transthoracic echocardiography, Rodnan skin score were performed. Vitamin D levels, CD34+ cell number, markers of inflammation, red blood cells distribution width (RDW) and Endocan plasma levels were also determined.

**Results:** 37 female SSc patients (11 diffuse/26 limited) and 35 matched healthy controls (HC) were enrolled. CD34+ cell count was lower as compared to controls (2.3±0.73 vs 2.9±0.6; p<0.001), CRP and ESR were significantly higher in SSc patients than controls. Vitamin D3 was lower in SSc patients with respect to controls (18±4.5 vs 27.26±10.98; p<0.001), Rodnan skin score was 28.89±10.24. Endocan was two times greater than controls and serum endocan levels were higher in patients with diffuse Ssc as compared with limited ones. We found no correlation between Endocan and: Rodnan skin score, ESR, fibrinogen; we found a trend of correlation between Endocan and Vitamin D levels (r= -0.315), RDW (r= 0.310), CRP (r= 0.310), but statistical significance was not reached likely due to the small sample size. We found an association of PAPs with Endocan levels (rho= 0.442, p<0.01) and CD34+ count (-0.349, p<0.05. A significant correlation was also found between Endocan and CD34+ cell number (rho= -0.605, p<0.01).

**Conclusion:** In our study population, we found a significant correlation between CD34+ cell number and Endocan plasma levels and PAPs; Endocan and CD34+ progenitor cells might be suggested as potential marker of pulmonary arterial hypertension in SSc patients.

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| THU0354 | CHARACTERIZATION OF ANTI-MYOSITIS ANTIBODY RELATED MYOPATHIES. DESCRIPTIVE STUDY IN A MULTICENTRIC COHORT. |

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**Background:** Idiopathic inflammatory myopathies (IIM) are a group of rare diseases consisting on immune-mediated muscle damage. About 40 to 60% show specific-myositis antibodies; additionally, 20-40% can show myositis-related (non-specific) antibodies. The profile of antibodies can help to divide patients into subgroups with more homogeneous clinical characteristics and prognosis.

**Objectives:** This study characterizes patients with IIM with specific or related anti-myositis antibodies, in five hospitals in the Alicante health area.

**Methods:** This is an observational study, carried out in five hospitals with a reference population of 1.083.463 people. Patients with positive anti-myositis antibodies between October 2015 and May 2018 were selected from the database of the Clinical Laboratory of the University Hospital of Alicante. We considered the following antiboides: anti-myositis specific antibodies (anti-TIF1γ, anti-MDA5, anti-Mi-2, anti-PmScl75, anti-PMScl100, anti-NXP2, anti-SRP), anti-synthetase antibodies (anti-PL7, anti-PL12, anti-Jo1, anti-OJ), myositis-related antibodies (anti-Ro52, anti-Ku). Clinical records were examined, identifying those patients with a diagnosis of IIM acording to their clinician, rheumatologist. Epidemiological and clinical data were obtained.

**Results:** 291 patients with positive anti-myositis antibodies were identified. Among them, 40 patients had a diagnosis of IIM. Median age was 59.5 (IQR 41.5, 70) years and 68% were women. Within the subgroups, the most frequent diagnosis were dermatomyositis (n=22; 55%) and polimyositis (n=9; 22%). The most common antibody detected was anti-TIF-γ among specific antibodies, and anti-Jo-1 among the anti-synthetase antibodies. The most common extramuscular feature was skin involvement. The presence of interstitial lung disease was reported in about one third of patients, being UIP the most commong pattern. Regarding treatment, the use of steroids was generalized; methotrexate was the most used inmunosupresant agent. Eight patients had a cancer related myopathy.

| | DM (N=22) | PM (n=9) | DMJ (N=3) | IMNM (n=1) | AS (n=5) |
|---|---|---|---|---|---|
| **Age, median (IQR)** | 65,5(46, 76) | 57(34, 66) | 15,7 (14, 18) | 83 (-) | 61(56, 64) |
| **Female n(%)** | 15(68,2) | 6(66,6) | 2(66,6) | 1(100) | 3(60) |
| **Muscular feat. n(%)** | 16(77,7) | 9(100) | 1(33,3) | 1(100) | 5(100) |
| **ILD n(%)** | 1(4,5) | 3(33,3) | 0 | 0 | 5(100) |
| **Disfagia n(%)** | 3(13,6) | 1(11,1) | 0 | 0 | 0 |
| **Skin feat n(%)** | 17(77,3) | 0 | 2(66,6) | 0 | 1(20) |
| **arthritis n(%)** | 1(4,5) | 0 | 2(66,6) | 0 | 4(80) |
| **Raynaud n(%)** | 4(19) | 1(11,1) | 0 | 0 | 1(20) |
| **Calcinosis n(%)** | 1(4,5) | 1(11,1) | (66,6) | 0 | 0 |
| **Puffy hands n (%)** | 2(9) | 0 | 0 | 0 | 2(40) |
| **Sicca synd n(%)** | 3(13,6) | 1(11,1) | 0 | 0 | 0 |
| **Cáncer n(%)** | 7(31,8) | 1(11,1) | 0 | 0 | 0 |
| **Ck máx(UI/l), median (IQR)** | 432(171, 706) | 809(350, 1653) | - | - | 665(396, 2300) |

**Conclusion:** This register allows us to characterise patients with inflammatory myositis in our area. It is important to make multicentric and prospective registers in infrequent diseases such as IIM in order to have more detailed and representative information about clinical and socio-demographic characteristic as well as prognostic data from these patients.

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