Anti-U1RNP was associated with muscle-skeletal manifestations (OR: 10.7 95% CI 0.92–20.44) and with overlap syndromes (OR: 15.2 95% CI 4.7–29.1). Pts with anti-Th/To and anti-RNA-polymerase III had lcSSc subtype. Vascular manifestations, oesophageal involvement and calcinosis cutis were the main manifestations, respectively. Table 1 shows detailed clinical manifestations and antibody profile.

Conclusions: In our cohort, ACA and anti-Scl70 were the commonest antibodies and were associated with lcSSc and dsSSc phenotype, respectively. ACA positivity conferred a higher risk of vascular disease and had a protective effect for ILD, while anti-Scl70 was associated with ILD.

Pts with anti-U1 RNP and anti-PM/Scl had mainly muscle-skeletal manifestations. This study confirms an association between immunological profile and clinical manifestations, reinforcing the importance of antibody profile and raising awareness for possible disease complications. Larger national studies would be desirable, specially for a better understanding of major organ involvement associated with least common antibodies.

Disclosure of Interest: None declared

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AB0728 NAILFOLD CAPILLAROSCOPY IN SYSTEMIC SCLEROSIS – SIX YEARS IN REVIEW

A.C. Duarte, A. Cordeiro, M.J. Santos. *Rheumatology, Hospital Garcia de Orta, Almada, Portugal*

Background: Microvascular dysfunction is a dynamic process that is crucial in systemic sclerosis (SSc) pathogenesis. Nailfold capillaroscopy (NCP) is a rapid, non-invasive exam that illustrates the early capillary changes in SSc and monitor their evolution. It is extremely useful in clinical practice and has been recognised in 2013 ACR/EULAR classification criteria for SSc.

Objectives: Evaluate the prevalence and evolution of NCP scleroderma pattern in SSc patients and analyse possible associations with disease-phenotype.

Methods: NCP of SSc patients followed in our centre were reviewed; clinical and demographic features were collected. A descriptive analysis was performed and nonparametric tests compared patients with and without SSc pattern.

Results: In total, 70 out of 117 SSc patients had at least 1 NCP available during the last 6 years. Most of these patients (62.9%) had limited cutaneous SSc, 21.4% diffuse cutaneous SSc, 11.4% very early diagnosis SSc and 4.3% overlap syndromes; mean disease duration was 10.7±9.6 years.

At the moment of the first NCP, 46 patients (39.4%) had a scleroderma pattern, 12 (10.3%) had non-specific(NS) NCP abnormalities and 12 had a normal NCP. During the 6 years follow-up, NCP changed in 5 patients as illustrated in figure 1. However, none had concomitant development/worsening of other clinical manifestations.

At the end of the follow-up, 49 (70%) patients had a NCP scleroderma pattern. Early pattern was present in 13 (26.5%) patients, active pattern in 21 (42.9%), active/late pattern in 3 (6.1%) and late pattern in 12 (24.5%).

When comparing patients with and without scleroderma specific patterns (table 1), the presence of scleroderma pattern was associated with the presence of current/previous digital ulcers (OR 1.49 95% CI 1.17–1.92). However, this difference was not confirmed between the different scleroderma patterns.

Regarding, major organ involvement, although there were no statistical differences between both groups, patients with scleroderma pattern had a higher prevalence of oesophageal involvement.

Abstract AB0728 – Table 1. Comparison between patients with and without scleroderma pattern

NCP pattern	Non-scleroderma		Scleroderma				p
	Normal (n=10)	Non-specific abnormalities (n=11)	Early (n=13)	Active (n=21)	Active/Late (n=3)	Late (n=12)	
Female	100%	72.7%	100%	85.7%	66.7%	91.7%	0.623
Age	55.3 ±16.2	66.7±5.7	58.2 ±20.7	53.8 ±15.8	44.9 ±18.2	64.9 ±9.3	0.219
Disease duration (years)	9.4±6.6	8.3±4.4	7.6±6	10.2 ±7.1	4.2 ±0.6	19.6 ±16.6	0.947
Diffuse cutaneous disease	10%	27.3%	7.7%	14.3%	33.3%	33.3%	0.689
Digital ulcers	10%	0%	30.8%	28.6%	33.3%	33.3%	0.021
Pulmonary arterial hypertension	0%	0%	0%	0%	0%	0%	NA
Interstitial lung disease	10%	18.2%	7.7%	9.5%	33.3%	25%	0.948
Oesophageal involvement	40%	45.4%	46.2%	71.4%	66.7%	50%	0.148
Anticentromere + Antitopoisomerase I +	50%	45.4%	69.2%	57.1%	0%	58.3%	0.557
	20%	27.3%	15.4%	19%	33.3%	33.3%	0.971

Abstract AB0728 – Figure 1. Progression of nailfold capillaroscopy alterations during follow-up

Conclusions: This study demonstrates how NCP can illustrate the dynamic vascular damage in SSc. In our data, a NCP scleroderma pattern was significantly associated with a higher number of digital ulcers and these patients had a higher percentage of oesophageal involvement.

In daily clinical practice, NCP is useful not only for corroborating SSc diagnosis, but also for monitoring endothelial injury and potential macrovascular/systemic damage. Although our sample is too small to demonstrate possible associations between specific NCP alterations and internal organ involvement, some studies have already identify NCP patterns as predictive factors for organ damage.

Disclosure of Interest: None declared

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AB0729 QUALITY OF LIFE ASSESSMENT IN SYSTEMIC SCLEROSIS PATIENTS TREATED WITH AUTOLOGOUS STEM CELL TRANSPLANTATION: A LONGITUDINAL STUDY

A.L.C. Guimaraes¹, K. Costa-Pereira², J.B. Elias², D.A. Moraes², E.A. Oliveira-Cardoso¹, J.T. Garcia¹, M.C. Oliveira³, V. Leopoldo³, A.F. Zombrili³, T.M. Costa², M. Vasconcelos², M.C. Oliveira². ¹Ribeirao Preto School of Philosophy, Sciences and Literature; ²Division of Clinical Immunology, Ribeirao Preto Medical School; ³Ribeirao Preto School of Nursing, University of Sao Paulo, Ribeirao Preto, Brazil

Background: Autologous Hematopoietic Stem Cell Transplantation (AHSCT) has been explored as a therapeutic option for patients with systemic sclerosis (SSc) that do not respond to conventional treatment.

Objectives: To investigate changes in quality of life of severe and rapidly progressive SSc patients treated with AHSCT.

Methods: This is a longitudinal and comparative study. Patients were evaluated before (n=27), and at 6 (n=27) and 12 months (n=21) after AHSCT. The Generic Questionnaire for Evaluation of Quality of Life Medical Outcomes Study 36 Item Short-Form Health Survey (SF-36) was applied individually, face-to-face, under patient written consent. This questionnaire evaluates eight domains: physical functioning (PF), role-physical (RP), bodily pain (BP), general health (GH), social functioning (SF), vitality (VT), role-emotional (RE) and mental health (MH). Results were transformed into a 0–100 scale, where zero corresponds to a worse health condition and 100 to the best possible score, and submitted to statistical analysis. Significance was defined as p<0.05.

Results: Most participants were females (n=24), with mean age of 33 years (standard deviation, SD=10.33) and mean time from diagnosis of 34.4 months (SD=34.89). Before AHSCT, the mostly impaired aspects were: PF (mean=8.33, SD=18.34), and RP (mean=38.52, SD=21.56), while MH (mean=61.63, SD=15.46) and SF (mean=56.87, SD=27.17) were mostly preserved. At 6 and 12 months post-AHSCT, there was significant improvement of the SF-36 scores in the following domains: PF (6 months, p<0.01, 12 months, p<0.01); RP (6 months, p<0.01, 12 months, p<0.01); BP (6 months, p<0.01, 12 months, p<0.01); GH (6 months, p<0.01, 12 months, p=0.02); VT (6 months, p<0.01, 12 months, p<0.01); MH (6 months, p<0.01, 12 months, p<0.01). The SF domain showed significant increase only at 12 months (p=0.02). The only domain in which there was no significant change was RE.

Conclusions: Increases in the physical components of quality of life are more evident in the initial periods that follow AHSCT, while improvements in mental state, which are also associated with social aspects, are detected on longer follow-up. These data reinforce the relevance of AHSCT upon patient quality of life, signalling the importance of psychotherapeutic evaluations and follow-up.

Disclosure of Interest: None declared

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AB0730

COMPARISON OF A SINGLE-CENTRE IDIOPATHIC INFLAMMATORY MYOPATHY COHORT FROM ARGENTINA WITH THE EUROMYOSITIS INTERNATIONAL REGISTRY

A.S. Brailard Poccari, R. Gomez, M. Pino, M. Garcia Carrasco, D. Dubinsky.
Hospital de Clinicas "Jose de San Martin", Ciudad Autonoma de Buenos Aires, Argentina

Background: The idiopathic inflammatory myopathies (IIM) are rare systemic autoimmune diseases that affect the muscle and other organs. Traditionally, IIM encompasses polymyositis (PM) and dermatomyositis (DM), but progressively inclusion body myositis (IBM), Immune-mediated necrotising myopathy (IMNM), the antisynthetase syndrome (ASS) and connective tissue diseases-over myositis (CTD-OM) have been recognised within the IIM spectrum

Objectives: To compare the clinical characteristics and treatment in a IIM cohort from an Argentinian university hospital with the international IIM cohort EUROMYOSITIS

Methods: Descriptive, retrospective study. IIM patients defined by expert opinion followed in our centre between October 2007 and October 2017 were included. ASS was defined by the presence of arthritis, raynaud's phenomenon, mechanic hands, elevated CK, muscle weakness, interstitial lung disease and/or presence of antisynthetase antibodies and, as in EUROMYOSITIS, patients with IIM with positive antisynthetase antibodies were reclassified as ASS. CTD-OM was defined as patients with IIM fulfilling classification criteria for other CTD.

Demographic data, accumulated clinical features, time interval between disease onset and diagnosis, IIM subtype, treatment and presence of neoplasm were evaluated.

Ethnicity was defined using the same classification as in EUROMYOSITIS

Results: 58 patients were included: DM 24, PM 4, ASS 10, CTD-OM 20. 89.6% Hispanic, mean age 48.4±15.2 years, median time interval between disease onset and diagnosis 5 months (IQR 2–11 months), been higher in AAS (8.5 months, IQR 1.5–18.2 months). 6.89% (4/58) patients presented associated neoplasm, 3 with DM and 1 with CTD-OM. Table 1 shows the demographic and clinical features of our IIM cohort and EUROMYOSITIS. Table 2 shows treatments received in our cohort and EUROMYOSITIS.

Abstract AB0730 – Table 1. Demographic and clinical features

	DM		PM		ASS		CTD-OM	
	HCJSM (n=24)	EUROMYO (n=949)	HCJSM (n=4)	EUROMYO (n=813)	HCJSM (n=10)	EUROMYO (n=512)	HCJSM (n=20)	EUROMYO (n=358)
Gender % M/F	20/4	30/70	75/25	29/71	40/60	31/69	25/75	21/79
Mean Age at Onset, years (SD)	46.73 (15.29)	49 (15)	43 (13.95)	40 (15)	40.5 (13.89)	48 (15)	40.9 (15.98)	48 (15)
Mean Age at Diagnosis, years (SD)	46.74 (15.36)	51 (15)	44.75 (15.39)	52 (15)	41.9 (15.34)	49 (15)	41.35 (15.83)	48 (15)
Median interval between onset and diagnosis, months (IQR)	4 (1.7-5)	5 (2-11)	5.5 (3.25-65.75)	8 (3-19)	6.5 (1.5-19.25)	7 (3-13)	5 (2.25-9.75)	11 (4-24)
Muscle weakness (%)	79.16	52	100	98	60	50	95	94
Rayn (%)	91.6	100	0	0	60	44	40	32
Raynaud's phenomenon (%)	33.3	25	50	29	50	51	65	60
Periungual erythema (%)	20.88	52	0	6	10	32	15	33
Actinias (%)	33.3	20	50	20	60	50	40	40
Mechanic's hands (%)	16.6	22	0	8	20	38	5	16
Calcinosis (%)	29.16	7	50	1	50	3	30	10
Ulceration (%)	8.33	13	0	2	10	3	5	10
Interstitial Lung Disease (%)	16.6	21	25	17	90	71	50	32
Cardiac involvement (%)	16.6	9	0	9	20	11	10	12

Conclusions: DM was the most frequent IIM subtype in both cohorts. In our group, CTD-OM was second and ASS was third.

Muscle weakness was found less frequently in our DM and AAS than reported in EUROMYOSITIS. However, calcinosis was more frequent. This could be explained by our mostly Hispanic population and/or by frequent Systemic Sclerosis overlap in our patients. It's important to remark that the ethnic variety defined as Hispanic in EUROMYOSITIS has a complex composition in Latin America, due to interbreeding.

No difference was found in terms of most frequent treatments between both cohorts. However, use of IVIg was more frequent in our patients.

To our knowledge, this is the first comparative report of an argentinian single-centre IIM cohort and an international multi-centre cohort

Disclosure of Interest: None declared

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Abstract AB0730 – Table 2. Treatment

	DM		PM		ASS		CTD-OM	
	HCJSM (n=24)	EUROMYO (n=949)	HCJSM (n=4)	EUROMYO (n=813)	HCJSM (n=10)	EUROMYO (n=512)	HCJSM (n=20)	EUROMYO (n=358)
Steroids	95	98	50	100	90	98	85	96
Hydroxychloroquine	50	37	25	11	40	16	40	29
Methotrexate	37.5	69	50	76	20	60	25	72
Leflunomide	0	0	0	3	20	3	5	4
Azathioprine	33.3	44	25	52	60	60	35	51
Mycophenolate	0	20	25	20	30	31	10	29
Cyclophosphamide	8.33	15	0	14	20	39	10	18
IVIg	29.16	11	0	13	20	7	5	7

AB0731

TREATMENT ALGORITHMS FOR SYSTEMIC SCLEROSIS ACCORDING TO EXPERTS

A. Fernández-Codina^{1,2}, K.M. Walker³, J.E. Pope¹, on behalf of Scleroderma Algorithm Group. ¹Medicine, Rheumatology Division, University of Western Ontario, London, Canada; ²Internal Medicine, Systemic Autoimmune Diseases Unit, Hospital Universitari Vall d'Hebron, Universitat Autònoma de Barcelona, Barcelona, Spain; ³Rheumatology Division, The Ottawa Hospital, University of Ottawa, Ottawa, Canada

Background: Treatment for many aspects of systemic sclerosis (SSc) lacks agreement.

Objectives: To generate SSc treatment algorithms endorsed by high percentage of SSc experts.

Methods: Experts from the Scleroderma Clinical Trials Consortium and the Canadian Scleroderma Research group (n=170) were asked whether they agreed with SSc algorithms (from 2012¹). A further 2 consensus rounds refined agreement; 62 (36%), 54 and 48 experts completed surveys.

Results: For scleroderma renal crisis (SRC), 82% of the experts agreed (1st line ACEi, 2nd and 3rd adding: CCB or ARB). Pulmonary arterial hypertension (PAH) had 81% agreement. For mild PAH, PDE5i, then endothelin receptor antagonists plus PDE5i, then prostanooids; while for severe PAH prostanooids were first-line. Raynaud's phenomenon (RP) had 78% of agreement [mild (1st CCB, 2nd adding PDE5i, 3rd ARB or switching to another CCB, 4th prostanooids), severe (1st CCB, 2nd adding PDE5i, 3rd ERA, 4th prostanooids)]. Digital ulcer (DU) treatment had 69% agreement (1st CCB, 2nd PDE5i). Interstitial lung disease (ILD) had 65% agreement including induction (Mycophenolate mofetil (MMF) then intravenous cyclophosphamide then rituximab) and maintenance (1st line MMF). Skin involvement had 71% agreement. For a modified Rodnan skin score (mRSS) of 24 1st MTX, 2nd MMF; and for mRSS 32 1st MMF, 2nd MTX, 3rd intravenous cyclophosphamide (CYP), 4th hematopoietic stem cell transplantation. For inflammatory arthritis 79% agreed with 1st MTX, 2nd low dose glucocorticoids, 3rd hydroxy-chloroquine, 4th rituximab or tocilizumab. Cardiac and gastrointestinal algorithms had ≥75% agreement. The evolution of the agreement rates is shown in table 1:

Abstract AB0731 – Table 1

Algorithms for SSc treatment	Agreement 2012 ¹ (%)	Agreement 2017 (%)
Scleroderma renal crisis	69	82
Pulmonary arterial hypertension	45	81
Raynaud's phenomenon	66	78
Digital ulcers	58	69
Interstitial lung disease	64	65
Gastrointestinal involvement	NA	77
Skin involvement	56, 40, 36 [†]	71
Inflammatory arthritis	45	79
Cardiac involvement	NA	75

Agreement is the% who agreed to the algorithm.

SSc: Systemic Sclerosis; NA: Not applicable; [†]For modified Rodnan skin scores 10, 24 and 32, respectively

Conclusions: Total agreement for SSc algorithms was considerable. These SSc algorithms may guide treatment.

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