The analysis showed ANA 1/640 centromere pattern, anticontromere antibodies, rheumatoid factor (RF) (652 U/mL) and C3 of 86.4 mg/dL. Previously to the BMT he had negative ANA, but we do not know the rest of the previous autoimmunity. On the videocapillaroscopy we observe an active scleroderma pattern (IMAGE 2–3).

He was diagnosed with systemic sclerosis based on Raynaud's phenomenon, digital ulcers, anticontromere antibodies and abnormal nailfold capillaries.

He had not familiar background of connective tissue diseases, but his mother presented Raynaud's phenomenon since she was thirty. So we studied her.

She presented facial telangiectasia, puffy fingers and fingertip pitting scars and the same autoantibodies: ANA 1/320 centromere pattern, anticontromere antibodies, RF (159 U/mL) and consumption of C3 (74.3 mg/dL) and C4 (7.6). We do not have previous autoimmune studies of her. On the videocapillaroscopy we observed a late scleroderma pattern (IMAGE 4–5).

She was also diagnosed with systemic sclerosis, based on Raynaud's phenomenon, puffy fingers, fingertip pitting scars and facial telangiectasia, anticontromere antibodies and abnormal nailfold capillaries.

Conclusions: Experimental animal studies and human clinical reports have described the transfer of immune-mediated diseases from affected donors to unaffected recipients, because of that the importance of screening this diseases in the donor before a BMT².

To our knowledge, this will be the first described case of transmission of systemic sclerosis by this mechanism. However, other explanations should also be taken into consideration. This include recipient's own persistent intrathymic lymphocyte population that may produce autoantibodies and autoimmunity in the context of cGVHD. However, this last hypothesis is not supported since there is a familiar background and presence of anticontromere antibodies.

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AB0722 CAPILLAROSCOPY AND PULMONARY ARTERIAL HYPERTENSION IN SYSTEMIC SCLEROSIS: A SYSTEMATIC REVIEW

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Background: At this very moment, no systematic review evaluating the role of nailfold videocapillaroscopy (NVC), with standardised definitions, in pulmonary arterial hypertension (PAH) has been published.

Objectives: To systematically identify and review all available literature evaluating the role of capillaroscopy in PAH in SSc, according to the definitions of the EULAR study group on microcirculation in Rheumatic diseases.

Methods: A systematic literature search was performed in Pubmed, EMBASE and Web of Science. All retrieved articles were screened on title, abstract and full-text level. Reference lists and google scholar were additionally searched. Original research papers that documented an association between NVC and PAH (confirmed on right heart catheterization [RHC]) in SSc were included. Subsequently, NVC parameters were subdivided in quantitative (density, dimension, morphology and haemorrhages), semi-quantitative (NVC score) and qualitative assessment (presence of scleroderma pattern, severity of scleroderma pattern and worsening of scleroderma pattern) according to the definitions of the EULAR study group on microcirculation in Rheumatic diseases.

Results: The systematic search identified 215 unique search results, of which 171 references were withheld after title screening. Abstract screening resulted in 51 references, only 19 were eligible for full-text review. Finally, 9 references were included in the final analysis after full-text screening (n=7) and bibliographic and google scholar search (n=2) (see table 1).

Regarding cross-sectional studies, density has been evaluated in 5 studies, with no unequivocal results for mean density; avascular score has been unequivocally associated with PAH. Dimension has been evaluated in 4 studies, with no unequivocal results. Morphology has been evaluated in 1 study and has been unequivocally associated with PAH. Haemorrhages and scleroderma pattern has been evaluated in 1 and 2 studies respectively, with no association. Severity of

scleroderma pattern has been evaluated in 3 studies and has been unequivocally associated with PAH.

Regarding longitudinal studies, density (i.e. capillary loss) has been evaluated in 2 studies and has been unequivocally associated with incident PAH. Dimension and haemorrhages have been evaluated in 1 study both, with no association. Morphology has been evaluated in 2 studies, with no unequivocal results. Worsening of scleroderma pattern has been evaluated in 2 studies and has been unequivocally associated with incident PAH.

Capillaroscopic parameter	Significant association		Non-significant association	CONCLUSION
	Mean density	Incident PAH (HR 0.001225 (p=0.001225) mRAP (p=0.58, p=0.039)225	PAH (NS)4µ	
Quantitative assessment	Capillary loss	Incident PAH (HR 18.53 (95%CI 1.28-78.35) p=0.033)508 Incident PAH* (p=0.04)80K	N/A	Regarding cross-sectional studies: *Avascular score unequivocally associated with PAH
	Avascular score	PAH (p=0.0017)2 mRAP (p=0.63, p=0.031)712	N/A	Regarding longitudinal studies: *Capillary loss unequivocally associated with incident PAH
	Dimension	Width PAH (p=0.051)15 Giant PAH (p=0.051)15	PAH (NS)225, 131E(4)µ Incident PAH (HR 0.31 (95%CI 0.15-0.67, p=0.003)508	Regarding cross-sectional studies: *No unequivocal results
Qualitative assessment	Morphology	Normal N/A Abnormal PAH (p=0.013)15 Incident PAH* (p=0.04)80K	N/A Incident PAH (HR 1.14 (95%CI 0.225-86, p=0.872)508	Regarding longitudinal studies: *Morphology unequivocally associated with PAH
	Haemorrhages	N/A	PAH (NS)315 Incident PAH* (HR 0.44 (95%CI 0.05-3.35, p=0.44)508	Regarding cross-sectional studies: *No association
	NVC score	PAH (p=0.031)712 mRAP (p=0.58, p=0.005)712	N/A	Regarding longitudinal studies: *NVC score unequivocally associated with PAH in cross-sectional studies
Qualitative assessment	Scleroderma pattern vs non-scleroderma pattern	N/A	PAH (NS)815 PAH (HR 0.5 (95%CI 0.1-4.5, p=0.46)508	No association in cross-sectional studies
	Severity of scleroderma pattern (early/active/late)	PAH (p=0.051)15 PAH (p=0.05)815 PAH (p=0.031)712	N/A	Severity of scleroderma pattern unequivocally associated with PAH in cross-sectional studies
	Worsening of scleroderma pattern	Incident PAH* Cvs A (p=0.031)508 Incident PAH* Cvs B (p=0.05)508 Incident PAH* (HR 5.12 (95%CI 1.33-21.27)508	N/A	Worsening of scleroderma pattern unequivocally associated with incident PAH in longitudinal studies

Abstract AB0722 – Figure 1

Conclusions: This systematic literature review, on behalf of the EULAR study group on microcirculation in Rheumatic diseases, is the first to investigate unequivocal associations between (incident) PAH and capillaroscopic alterations in a standardised way. Unequivocal associations were found in cross-sectional studies between avascular score, morphology, NVC score and severity of scleroderma pattern and PAH and in longitudinal studies between capillary loss and worsening of scleroderma pattern and incident PAH.

Disclosure of Interest: None declared

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AB0723 SEROPREVALENCE OF EPSTEIN-BARR VIRUS AND CYTOMEGALOVIRUS IN SYSTEMIC SCLEROSIS PATIENTS: PRELIMINARY RESULTS

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Background: Epstein-Barr Virus (EBV) and cytomegalovirus (CMV) are among the most diffused virus in humans, with prevalence of 90% and 80% respectively in adult immunocompetent population^{1,2}. Hypothesis for a role of EBV and CMV in systemic sclerosis pathogenesis was reported³.

Objectives: To evaluate the prevalence of EBV and CMV in a population of patients affected by SSc. As an exploratory aim, correlations with available clinical variables were evaluated.

Methods: A total of 49 patients (44 females and 5 males, age 65.6±9 years, 34 with limited cutaneous involvement and 14 with diffused cutaneous involvement) diagnosed according to the 2013 ACR/EULAR criteria⁴ were randomly enrolled at their follow-up visit for a further more detailed analysis. After consent, all participants underwent serological testing for VCA-IgM, VCA-IgG and EBNA-IgG and CMV IgM and IgG, to determine prior or actual EBV and/or CMV infection. All SSc patients undergone also to clinical examination and instrumental evaluations according to SSc clinical standard follow up.

Results: Results for EBV seroprevalence were: VCA-IgM negative=43 (87%), positive=4 (8,2%), borderline=2 (4,1%) in all SSc patients. VCA-IgG resulted positive in 48 patients (98%) and EBNA-IgG resulted positive in 40 (81,6%) patients, borderline in 4 (8,2%) and negative in 5 (10,2%) patients. Forty-six (93,9%) patients resulted positive for CMV-IgG and 22 (44,9%) showed detectable CMV IgM. Consequently, EBV and CMV seroprevalence was significantly higher (p=0.01 and p=0.032 respectively) than that calculated in general population (90% and 80% respectively). Interestingly, the presence of higher CMV-IgM correlated linearly with higher pulmonary arterial pressure values (p=0.036) and was different in patients with or without sicca syndrome (p=0.012) and arthralgias

($p=0.04$). Finally, higher CMV-IgG correlated with diffused cutaneous SSc form ($p=0.023$) and nephropathy ($p=0.036$).

Conclusions: The seroprevalence of EBV and CMV positivity was found significantly higher in the sample of SSc patients. EBV seropositivity (VCA-IgG) was present nearly in the totality of patients (98%). Of relevance, the high presence of CMV-IgG (93%) and CMV-IgM (44.9%) in SSc patients: the last correlated linearly with higher pulmonary arterial pressure values. EBV and CMV infections, among the supposed triggers in several autoimmune diseases, might also play a role in SSc patients, at least with progressive disease.

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AB0724 CARDIO-PULMONARY DISEASE MANAGEMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS: CARDIO-RHEUMATOLOGY CLINIC AND PATIENT CARE STANDARDISATION PROPOSAL

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Background: Systemic sclerosis (SSc) is a chronic connective tissue disease characterised by endothelial dysfunction, dysregulation of fibroblasts with excessive fibrosis of the skin and internal organs and autoimmune abnormalities. Cardio-pulmonary manifestations are common in SSc and their detection in the early stage of the disease as well as their careful follow-up are mandatory in order to counteract their impact on the overall disease outcome. Despite the need of establishing a proper methodology, literature provides few reports about this issue.

Objectives: To evaluate the activity of our Cardio-Rheumatology Clinic in order to optimise diagnostic management of cardio-pulmonary disease in SSc patients.

Methods: We retrospectively analysed data from 350 consecutive SSc patients referred to our University-based Rheumatology Centre and SSc Unit (F/M 308/42; lc/dcSSc 45/305; mean age 50.8±14.7 years; mean disease duration 10.9±7.0 years). All patients underwent general and cardio-pulmonary assessment, in particular they were evaluated in the Cardio-Rheumatology Clinic. The following parameters were considered: physical examination; past and current drugs; blood tests, in particular Erythrocyte sedimentation rate-ESR, C-reactive protein-CRP, CPK enzymes, troponin, NT-pro-BNP, d-dimer, serum autoantibodies, 25-OH-vitamin D; capillaroscopy; pulmonary function tests; high resolution scan of the lungs (HRCT); standard electrocardiogram (ECG) and 24 hour Holter ECG monitoring; Doppler echocardiography; cardiac stress test; coronary angiography and right heart catheterization (RHC); cardiac MRI and CT; vascular ultrasound (intima-media-thickness, carotid-femoral and brachial-ankle pulse-wave-velocity). The clinicians decided to perform these examinations according to clinical picture and current methodologies.

Abstract AB0724 – Figure 1

Results: In the last 12 months we assessed 300 patients with 1st-level screening (cardio-rheumatologic evaluation, standard ECG, Doppler echocardiography, pulmonary function tests, thoracic imaging). Among 2nd-level, 30 procedures of 24 hour Holter ECG and 15 RHC tests were performed. Cardiac MRI, coronary CT angiography and vascular ultrasound were assessed, when requested, as 3rd-level examinations (30 procedures). After 1 year we observed a mean time of 10±5 days between request and clinical cardio-rheumatologic evaluation, 20±12 days to perform 1st-level screening, 25±15 days to execute the 2nd-level examinations. Figure-1 shows Cardio-Rheumatology algorithm for the management of SSc cardio-pulmonary disease.

Conclusions: The activity of our Cardio-Rheumatology Clinic optimises the cardio-pulmonary SSc assessment, determining an early detection of these harmful complications with reduced waiting times which are critical issues. Screening algorithms are useful to stratify the risk and to establish the most appropriate diagnostic-therapeutic protocols, improving outcome of scleroderma patients. The development of a cardio-pulmonary risk score and the standardisation of a patient care approach, according to international quality indexes, could represent further tools to optimise SSc management.

Disclosure of Interest: None declared

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AB0725 SEVERE DYSPHAGIA “POOR PROGNOSTIC MARKER” IN IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: In Idiopathic Inflammatory Myopathies (MII), 18%–20% of patients have dysphagia.

Objectives: To evaluate the frequency of dysphagia in patients with MII, association with other manifestations of the disease, treatment and evolution.

To evaluate clinical characteristics and evolution of severe dysphagia.

To compare clinical characteristics and evolution of mild-moderate versus severe dysphagia.

Methods: Retrospective, observational study. Patients with a diagnosis of MII were included according to modified classification criteria of Bohan and Peter. Demographic, clinical and complementary studies were recorded. Serious dysphagia was considered: contraindication of oral feeding. Descriptive statistics were performed. Chi2 test, Student's test or Mann Whitney as appropriate.

Results: We included 91 of 106 patients evaluated from 1992 to 2017: 76% female, mean age at diagnosis 48±14 years. 53% presented dysphagia: mild/moderate 62.5% (30/48 pts), severe 37.5% (18/48). Idiopathic dermatomyositis was the most frequent MII in these patients (71%). In patients with dysphagia, proximal muscle weakness was 90%, weakness of neck muscles 45%, weakness of respiratory muscles 27%.

A significant association was found between dysphagia and weakness of respiratory muscles, weak neck muscles, glucocorticoid pulses, gammaglobulin, grave infections and death. (Data not shown in the summary).

In patients with **severe dysphagia**, we observed a significant association with the requirement for mechanical ventilation, hospitalisation in an intensive care unit, serious infections and death (table 1).

When comparing mild-moderate dysphagia vs severe dysphagia, a statistically significant association was found with neck muscles weakness, respiratory muscle weakness, glucocorticoid pulses, gamma globulin use, requirement for mechanical ventilation, hospitalisation in an intensive care unit, severe infections and mortality (table 2).

Abstract AB0725 – Table 1

	Severe dysphagia (SI) 18/91	Severe dysphagia (NO) 72/91	p	OR	IC 95%
Weakness of respiratory muscles	8/18	6/63	0.00054	7,6	2,1–26,6
Weak neck muscles	8/16	6/63	0.00129	8	2,5–26
Glucocorticoid pulses	12/18	8/69	<0,0001	15	4,4–51,9
Gammaglobulin	10/18	7/72	<0,0008	11,60	0,8–10
Intensive therapy unit	7/18	8/72	0,0046	5	1,5–16,8
Mechanical respiratory assistance	6/18	5/72	0,002	6,70	1,7–25
Grave Infections	10/17	11/66	0,0003	7	2,2–22,8
Death	12/18	9/72	<0,0001	14	4,2–46,6