Conclusion: The discrimination function is a proposed screening algorithm for pts with SSc-ILD who could develop progression disease. Usage of discrimination function revealed that 84% pts with ILD-SSc have got poor prognosis and 9% of them have got fatal outcome. Usage the equation of prognosis is very useful to choice of treatment and observation pts SSc-ILD.

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AB0454

SPECIFICITY OF INTERSTITIAL LUNG DISEASE IN PATIENTS WITH SYSTEMIC SCLEROSIS POSITIVE FOR A-TOPO-1 AND A-RNP

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Background: The classification criteria of SSC (EULAR/ACR 2013) compared to the criteria of 1980 showed high sensitivity and allow to detect the full spectrum of systemic sclerosis (SSc). The use of the new criteria led to the fact that some of the patients who were diagnosed with MCTD began to meet the criteria of SSc. Since these patients have hyperproduction of antibodies to RNP, but no other SSc-specific autoantibodies, we believe that they represent a special clinical and immunological phenotype of SSc. Lung involvement in SSc impairs the quality of life and the prognosis, so the study of interstitial lung disease (ILD) in different phenotypes is relevant.

Objectives: To compare the main manifestations of ILD in two groups of patients who meet the classification criteria of SSc 2013 - positive for a-Topo-1 and for a-RNP.

Methods: The study included 100 patients. The first group - 50 patients positive for a-Topo-1 (45 women and 5 men, mean age 55±10 years, duration of the disease 10.2±8 years, subset of the disease: diffuse 52%, limited 48%) and the second group - 50 patients positive for a-RNP (43 women and 7 men, mean age 44.5±14 years, duration of the disease 11.7±8 years; form of the disease: diffuse 14%, limited 86). The diagnosis of ILD was established by HRCT.

Results: In patients of group I ILD was detected in 94% of cases, in group II - 68% (p<0.05). In both groups a decrease in functional lung tests was noted with a high frequency, but a severe decrease in the lung diffusion capacity was significantly more often observed in group I.

Table 1. Data of laboratory and instrumental parameters

Parameters	a-Topo-1- positive	a-RNP-positive	p
	n (%)	n (%)	
Decrease of DLCO < 80%	49 (98)	43 (86)	p<0,05
DLCO <55%	33 (66)	16 (32)	p<0,05
Decrease of FVC< 80 %	19 (38)	10 (20)	p<0,05
FVC<70%	13 (26)	7 (14)	NS
Ground glass opacity	25 (50)	26 (52)	NS
Honeycombing	17 (34)	8 (16)	p<0,05
SPAP >40mm.Hg.	6 (12)	10 (20)	NS
including PAH**	2 (4)	3 (4,6)	NS
ANA> 1:640	37 (74)	45 (90)	p<0,05

* ANA – antinuclear antibodies, PAH – pulmonary arterial hypertension, SPAP - systolic pulmonary artery pressure ** confirmed by the data of catheterization of the right heart

Typical changes according to HRCT were detected in both groups, but fibrotic changes dominates in group I. There were no significant differences in the frequency of occurrence of PAH. Patients of group II had high level of ANA more often.

Conclusion: In both groups, a high frequency and similar manifestations of ILD were noted. In patients with SSc positive for anti-U1RNP, despite the predominance of limited skin lesions (in 86% of patients), a high incidence of ILD was detected, while the frequency of PAH was comparable to SSc in general. Compared with a-Topo-1 positive patients, ILD was significantly less frequent in anti-U1RNP positive patients and was accompanied with a smaller reduction of DLCO and/or FVC and a lower frequency of severe lung fibrosis. These data confirm the similarity of lung involvement in compared phenotypes of SSc and indicate the importance for screening for ILD in patients with hyperproduction of a-RNP, especially at an early stage of the disease.

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AB0455

MODIFICATION OF PROGNOSIS INTERVALS WAS DERIVED BY THE DISCRIMINATION FUNCTION FOR SSc-ILD PATIENTS

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Background: Systemic sclerosis (SSc) is a multisystem autoimmune disease. Pulmonary alterations are the major cause of mortality in SSc. SSc-ILD has heterogeneous disease progression: many patients will have a chronic, indolent course while others may develop progressive, life-threatening disease.

Objectives: To assess prognosis intervals for pts with SSc-ILD.

Methods: It was a longitudinal study involving 140 pts with SSc-ILD. The mean age was 46.7±13.9 years, females 82%, SSc duration 6.2±5.8 years, diffuse subset - 54%. The mean duration of follow up was 73,2±27,8 months. Clinical, laboratory, and immunologic data were collected on all patients.

Based on analysis of discrimination function the equation of prognosis was developed. The equation is 3.14GGO (ground glass opacities) + 0,7 index EScSG - 1,326 FVC - 0,1 the maximum daily dose of glucocorticoids + 0,136 Gamma globulins + 1.066 CYP - 1.075 DLCO ≤ 5,8. Value of equation of prognosis to 5,8 corresponds to stabilization or good prognosis and value after 5,8 corresponds to poor prognosis. The equation of prognosis had 86% sensitivity and 56% specificity. The cut-off point 4,7 was represented 65% sensitivity and 67% specificity. The prognosis intervals were following: value of equation to 4,7 corresponds to good prognosis; value from 4,8 to 5,8 corresponds to stabilization and value after 5,8 corresponds to poor prognosis.

Results: Pts were distributed into groups depending on results of discrimination function: 14 pts (10%) - zone of good prognosis; 8 pts (6%) - zone of stabilization and 118 (84%) - zone of poor prognosis. We have decided to join pts from zone good prognosis and zone stabilization into united group (U-group) - 22 pts (16 %).

Table 1. Comparative characteristics of the groups

	Group 1 (n=14)	Group 2 (n=8)	U-Group (n=22)	Group 3 (n=118)
Age years	45,4±16,4	50,2±15,8	47,2±16	47,5±13
P>0,05				
Gender				
Male	3(21%)	1(12,5%)	4(18%)	21(18%)
Female	11(79%)	7(87,5%)	18(82%)	97(82%)
P>0,05				
Limited form	8(57%)	2(35%)	10(46%)	54(46%)
Diffuse form	6(43%)	6(75%)	12(54%)	64(54%)
P>0,05				
Duration of disease years	6,8±7,6	5±4,2	6,2±6,4	6,5±5,7
CRP mg/ml	10±10	8,5±10,6	9,4±10,4	12,6±23,9
p>0,05				
ESR mm/h	16,7±13,1	17,4±21,9	17±16,3	23,5±16,7
p>0,05				
ANA-hep-2	525±322*	420±190	487±281*	774±430*
p: Gr1 vs Gr3=0,038; Gr2 vs Gr3=0,02; Gr1,2 vs Gr3=0,032				
a-Scl-70 u/ml	59,9±82,7*	67±94,5	116±93**	62,5±85* **
p: Gr1 vs Gr3=0,03; Gr1,2 vs Gr3=0,013				
Ejection fraction(EF)(%)	67,3±5,3*	65,5±6,1	66,7±5,5**	63±6,8* **
p: Gr1 vs Gr3=0,02; Gr1,2 vs Gr3=0,017				
PAP mm hg	31,6±5,8	32,4±9,3	31,9±7	36,4±14
p>0,05				
6MWT m	490±110	477±104	485±104	430±100
(6 Minute walk test)				
p>0,05				

The age, SSc duration, gender proportion and SSc forms, were similar in the all groups (p>0,05). In the study mean dates of ANF-hep-2, a-Scl-70 were significantly lower and EF was significantly higher in groups 1 and U-group than in group 3. In groups 1,2 and U-group the mean levels of ESR, CRP, PAP were lower and value of 6MWT were higher than mean levels in group 3 accordingly. So separately group 2 didn't have significant differences with group 1 and 3.

Conclusion: In the study we didn't find any correlation between meanings of group 2 and groups 1,3 but the meanings of group 1 and U-group have had the

same correlation. It would be rational to highlight only one zone of prognosis before and after 5.8.

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AB0456

HYDROXYCHLOROQUINE MIGHT REDUCE MORTALITY IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a devastating disease that has a profound impact on life expectancy, reflected by a standardised mortality ratio of 3.5. There is still limited data regarding the predictive factors for mortality in patients with SSc. Determining those factors could guide in disease management and follow up.¹

Objectives: We aimed to identify the predictive factors for death in SSc.

Methods: Patients followed in a tertiary rheumatology clinic in the last 5 years were included in this retrospective study. All of the patients met the ACR / EULAR SSc 2013 criteria. Medical records of the patients were reviewed. Follow up time was defined as the time period from the first admission of the patient to our rheumatology clinic until the date of death or the date on which the study was performed. Candidate predictive factors for mortality were tested by Kaplan-Meier (with Log rank) and Cox-regression analyses.

Results: In total 146 patients (mean age 55.6±12.3 years, female 89.7%, diffuse cutaneous type SSc 45.2%) were included in the study (Table 1). The mean age at diagnosis of study group was 48±13.7 years. The median duration of follow up was 71 (6-228) months. Fourteen (10%) patients died during follow-up. The causes of death were: pulmonary (7), renal (2) and cardiac diseases (1), infection (3) and cancer (1).

Univariate analysis revealed that age at diagnosis (p=0.028), SSc subtype (p=0.035), the presence of interstitial lung disease (p=0.002), oesophageal involvement (on computed tomography) (p=0.030), pulmonary artery systolic pressure of ≥35 mmHg (measured by transthoracic echocardiography) (p=0.004), glucocorticoid (p=0.029), hydroxychloroquine (p=0.002) and cyclophosphamide (p=0.006) usage at any time were associated with mortality (Figure 1). Multivariate analyses model formed with age at diagnosis (B: 0.055, 95% CI, 1.005-1.112; p=0.033), SSc subtype (B: 0.963, 95% CI 0.541-12.684; p=0.231), glucocorticoid (B: 1.396, 95% CI, 0.487-33.507; p=0.196) and hydroxychloroquine usage (B: -1.50, 95% CI, 0.061-0.816; p=0.023) showed that age at diagnosis and hydroxychloroquine usage were independent predictive factors for mortality in patients with SSc.

Conclusion: The results of the study revealed for the first time that apart from the age at diagnosis hydroxychloroquine might reduce mortality in patients with SSc. Further studies are needed to prove of this information.

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Table 1. The demographic and clinical features in patients with systemic sclerosis.

Characteristic	Baseline
Age at diagnosis [*]	48±13.7
Female sex, n (%)	131 (89.7)
Duration of follow-up, months ^{**}	71 (6-228)
Disease subtype, n (%)	
Diffuse / Limited	66 (45.2) / 80 (54.8)
Autoantibodies, n (%)	
Anti-Scl70 antibody	50/143 (35.0)
Anti-Centromere antibody	62/143 (43.4)
Immunosuppressive medication, ever, n (%)	
Hydroxychloroquine	91/143 (63.6)
Mycophenolate mofetil	18/145 (12.4)
Azathioprine	47/145 (32.4)
Cyclophosphamide	24/145 (16.6)
Glucocorticoid	80/140 (57.1)
Others, n (%)	
ILD	68/130 (52.3)
Pericardial effusion, ever	26/133 (19.5)
Esophageal dilation (detected by CT)	51/128 (39.8)
sPAP ≥35mmHg, ever (measured by ECHO)	46/142 (32.4)

^{*}Parameter presented as mean±SD ^{**}Parameter presented as median (min-max)/CT, computed tomography; ECHO, echocardiogram; ILD, interstitial lung disease; sPAP, systolic pulmonary artery pressure

Fig. Cumulative survival of systemic sclerosis patients as determined by the Kaplan-Meier method.

SSc, systemic sclerosis; lSSc, limited cutaneous systemic sclerosis; dSSc, diffuse cutaneous systemic sclerosis; sPAP, systolic pulmonary artery pressure; ECHO, echocardiogram

Figure 1.

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AB0457

HIPPOCAMPAL SUBFIELDS VOLUMES REDUCTION IN PATIENTS WITH SYSTEMIC SCLEROSIS: A LONGITUDINAL MAGNETIC RESONANCE IMAGING (MRI) VOLUMETRIC STUDY

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Background: Systemic sclerosis (SSc) is a multisystem disease characterized by functional and structural abnormalities of small blood vessels, fibrosis of the skin and internal organs. In addition, volume reduction and shape abnormality of the hippocampus have been demonstrated in rheumatic and neurodegenerative diseases. However, the neuroanatomy of the hippocampus is complex and is not a uniform structure, consisting of subfields with distinct morphology: cornu ammonis (CA) subfields CA1–4, dentate gyrus (DG), fimbria, and adjacent subiculum and presubiculum [1].

Objectives: To investigate the hippocampal subfields volumes reduction in SSc patients using MRI.

Methods: In this study we included 37 SSc patients (33 women, mean age of 53.46, SD ± 12.29; range 30 - 78) and 37 healthy controls (HC) (31 women, mean age of 48.41, SD±12.20; range 29 - 80). Cognitive evaluation was performed using the Montreal Cognitive Assessment (MoCA), mood disorders were determined through Beck's Depression (BDI) and Beck's Anxiety Inventories (BAI). SSc patients were further assessed for clinical and laboratory SSc manifestations, disease activity (Valentini Activity Index), severity activity (Medsger Severity Index). MRI protocol consisted of: Sagittal T1-weighted images performed with a Philips 3.0T MRI scanner. MRI brain structure volumetric was done through volBrain [2]. After 48 months MRI acquisition were repeated in 26 SSc patients and 12 healthy volunteers. Statistics was performed according nature of the variable.

Results: We observed a significant reduction in hippocampal subfields volumes in SSc patients when compared to controls: Total hippocampi (SSc: mean volume = 4.78 cm³; SD = 0.38; HC: mean volume = 5.01 cm³; SD = 0.38; p = 0.033). Reduction in volume of the total hippocampi was associated with Raynaud's phenomenon (p = 0.006). A longitudinal study showed a reduction in volume of the hippocampus subfields volumes when compared to patient's baseline: Total hippocampi (mean initial volume = 4.78 cm³; mean follow-up volume = 4.50 cm³, p = 0.027); Total CA1 (mean initial volume = 1.59 cm³; mean follow-up volume = 1.58 cm³, p < 0.0001); Reduction in volume of the total hippocampi was associated with presence of current use of prednisone (p = 0.008). Reduction in left CA1 left associated with current use of prednisone (p = 0.014). Reduction in total subiculum was associated with presence of calcinosis (p = 0.023). No significant changes were observed in hippocampal subfields volumes in controls over the follow-up period.

Conclusion: This study provides evidence of hippocampus subfields volumes reductions in SSc patients when compared to controls and was associated with