presence of anti-cN-1A. Anti-cN-1A positivity had a sensitivity of 43.6% and a specificity of 91.8% for sIBM. The positive and negative predictive values were 36.4% and 93.8%, respectively.

There was no significant difference in gender, age at study entry, age at symptom onset, duration of symptoms or max creatine kinase (CK) levels during disease course between the anti-cN-1A positive and negative sIBM patients. Dysphagia was present in 19 (79%) of the anti-cN-1A positive and in 17 (55%) of the anti-cN-1A negative sIBM patients ($P = 0.06$).

Conclusion: Antibodies against cN-1A are the first and so far the only serological marker for sIBM. Our data showed that cN-1A autoantibodies are specific for sIBM and further corroborate the potential diagnostic role of cN-1A autoantibodies in this distinct subgroup of myositis.

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	All Patients n=83	ESC/ERS screen positive, n=8 (9.6%)	ESC/ERS screen negative, n=75 (90.4%)	DETECT screen positive, n=8 (9.6%)	DETECT screen negative, n=75 (90.4%)	ASIG screen positive, n=13 (15.7%)	ASIG screen negative, n=70 (84.3%)	Any of the algorithms, n=19 (22.9%)	Any of the algorithms, n=20 (24.1%)
Age, median [IQR]	51 (46.75-59.75)	66.5 (59-69.75)	48 (38.75-52)	66.5 (58.25-69.75)	48 (38.5-51.5)	61 (52.75-68.5)	48 (38-51)	57 (50.5-64)	48 (38-51)
Male, n (%)	100 (100)	100 (100)	100 (100)	100 (100)	100 (100)	100 (100)	100 (100)	100 (100)	100 (100)
Female, n (%)	13.5 (15-20.75)	52 (39-57)	65 (51-79.5)	24 (5-38)	12 (5-25)	14 (5-37)	13 (5-35)	14 (5-25)	12 (5-17.5)
Duration of SSc symptoms, year [IQR]	10 (4-20)	2 (0-7.5)	8 (0-13)	1 (0-13)	9 (2-17)	4 (1-13)	6 (2-17)	4 (1-13)	6 (2-17)
SSc pattern, n (%)									
Diffuse	24 (24)	6 (7.5)	18 (18)	6 (7.5)	18 (18)	6 (7.5)	18 (18)	6 (7.5)	18 (18)
Limited	24 (24)	6 (7.5)	18 (18)	6 (7.5)	18 (18)	6 (7.5)	18 (18)	6 (7.5)	18 (18)
SSc related serology, n (%)									
Anti-Scl-70 (+)	18 (14)	3 (3.75)	15 (15)	3 (3.75)	15 (15)	3 (3.75)	15 (15)	3 (3.75)	15 (15)
Anticentromere (+)	3 (3.75)	0 (0)	3 (3.75)	0 (0)	3 (3.75)	0 (0)	3 (3.75)	0 (0)	3 (3.75)
Presence of ILD, n (%)	37 (44.7)	4 (5)	33 (33)	4 (5)	33 (33)	7 (8.6)	30 (30)	7 (8.6)	30 (30)
Presence of digital ulcers (ever), n (%)	23 (28)	4 (5)	19 (19)	4 (5)	19 (19)	6 (7.5)	17 (17)	6 (7.5)	17 (17)
Presence of GIS involvement, n (%)	14 (14)	6 (7.5)	8 (8)	6 (7.5)	8 (8)	7 (8.6)	7 (7)	7 (8.6)	7 (7)
Presence of telangiectasia, n (%)	14 (14)	5 (6.25)	9 (9)	5 (6.25)	9 (9)	7 (8.6)	7 (7)	7 (8.6)	7 (7)
sPAP on TTE, median (mmHg) [IQR]	28.5 (23.5-33)	36.5 (29.25-46.25)	27 (21.75-31)	33 (25.5-44)	27 (21.75-31)	30 (25.5-40.5)	27 (19.5-31)	30 (25.5-40.5)	27 (19.5-31)
FVC, median (%) [predicted] [IQR]	86 (69.5-102.5)	92 (88.25-115.25)	83 (70-99)	92 (83.5-114)	79 (69.25-100)	92 (87-108.75)	76 (65.5-99)	90 (72-107.5)	74 (69.25-100)
DLCO, median (%) [predicted] [IQR]	53.5 (41.25-71)	47 (38.75-66.5)	62 (49-78.5)	55 (46-71)	52 (43-71)	50 (35.5-58.5)	63 (46-79)	51 (37.25-62.5)	62 (44-77)
FVC / DLCO ratio, median [IQR]	1.4 (1.1-1.74)	2.31 (1.58-2.18)	1.3 (1.1-1.4)	1.4 (1.28-2.18)	1.4 (1.1-1.58)	1.78 (1.35-2.12)	1.35 (1.1-1.43)	1.71 (1.29-1.44)	1.36 (1.04-1.44)
NT-proBNP, median (pg/mL) [IQR]	137 (63-256)	284 (162-778)	120 (52-165)	451 (120-887)	120 (52-158)	284 (124-25-695)	120 (38-158)	284 (124-25-695)	120 (38-158)
Presence of dyspnea, n (%)	7 (8.6)	7 (8.6)	7 (7)	7 (7)	7 (7)	7 (8.6)	7 (7)	7 (8.6)	7 (7)
WHO-FC, n (%)									
Class 1	1 (1.2)	0	0	0	0	0	0	0	0
Class 2	5 (7.2)	7 (8.6)	4 (5)	9 (10)	4 (5)	6 (7.5)	7 (7)	6 (7.5)	7 (7)
Class 3	1 (1.2)	0	0	0	0	0	0	0	0
Class 4	0	0	0	0	0	0	0	0	0

Table 2. Performance of the RHC screening algorithms

	Performance for 25-mmHg cut-off			Performance for 20-mmHg cut-off		
	ESC/ERS	ASIG	DETECT	ESC/ERS	ASIG	DETECT
Sensitivity, %	100	100	100	75	100	93
Specificity, %	94	9	40	67	17	0
PPV, %	37.5	23	33	75	61	44
NPV, %	100	100	100	67	0	0

FRI0321

PERFORMANCE OF DIFFERENT PULMONARY HYPERTENSION SCREENING ALGORITHMS IN PATIENTS WITH SYSTEMIC SCLEROSIS PATIENTS

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Background: Pulmonary hypertension (PH) is an important cause of morbidity and mortality in patients with systemic sclerosis (SSc). Different screening algorithms have been proposed for identifying patients who have a high probability of PH and require right heart catheterization (RHC), which is the gold standard for diagnosing PH.

Objectives: To compare the performance of PH screening algorithms in our patients with SSc.

Methods: Forty-eight consecutive patients, fulfilling ACR/EULAR 2013 SSC criteria, were screened for PH using the, 2015 ESC/ERS, DETECT and ASIG algorithms. Pulmonary function tests (PFT), diffusing capacity of the lung for carbon monoxide (DLCO), trans-thoracic echocardiography, serum NT-proBNP, uric acid assay and high-resolution computed tomography (HRCT) were performed as needed. Patients with known PH, severe interstitial lung disease and severe left ventricular dysfunction were not included. RHC was performed in all patients with positive screening according to any one of the screening algorithms. Patients with PH were classified according to the updated PH classification criteria. Sensitivity and specificity of the 3 screening algorithms were evaluated according to the established cut-off value of 25 mmHg for mean systolic pulmonary artery pressure and for the recently proposed cut-off value of 20 mmHg.

Results: Among the 48 SSc patients, 15 were excluded due to already diagnosed PH (n=4), left ventricular dysfunction (n=4), no measurable tricuspid regurgitation velocity (TRV) (n=5) and coexisting lung cancer (n=2). Among the remaining 34 patients, 16 required RHC according to at least one of the screening algorithms. Demographic and clinical features of remaining 34 patients were summarized in Table 1. Number of patients who had suspected pulmonary hypertension and required RHC according to ESC/ERS 2015, DETECT and ASIG were 8 (23.5%), 9 (26.5%), and 13 (38.2%) respectively (Figure 1). Among the 14 who had RHC, PH was present in 3 patients according to the 25-mmHg cut-off (Group 1 in 1, Group 2 in 1, Group 3 in 1) and in 8 patients according to the 20-mmHg cut-off (Group 1 in 5, Group 2 in 2, Group 3 in 1). The sensitivity and specificity of each algorithm is presented in Table 2. Sensitivity was similar at 100% for the 3 algorithms, but the ESC/ERS algorithm had better specificity, when PH was diagnosed with the 25-mmHg cut-off. For the 20-mmHg cut-off, both the sensitivity and the specificity were better with the ESC/ERS algorithm.

Conclusion: The ESC/ERS algorithm seems to have a better performance for detecting PH in patients with SSc. A limitation of this study was that RHC was not performed in patients who did not fulfill criteria according to any of the screening algorithms. The sensitivities may be lower than what we propose if there are patients with PH who are asymptomatic and not captured with any of the algorithms.

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FRI0322

AUTOANTIBODIES PROFILE INFLUENCE ON SYSTEMIC SCLEROSIS INTERSTITIAL LUNG DISEASE. A TERTIARY SPANISH HOSPITAL EXPERIENCE

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Background: Systemic Sclerosis (SSc) is a rare and heterogeneous connective tissue disease (CTD) characterized by skin fibrosis, vasculopathy/vascular damage and potential visceral impairment. Interstitial lung disease (ILD) constitutes the leading cause of mortality and requires close periodical assessment and follow-up. Diffuse cutaneous sclerosis and specific autoantibody profile (anti SCL70, anti Th/To, Anti U3 RNP and anti PmScI) are considered ILD development risk factors. In contrast, positivity to anticentromere has been considered a protective factor to develop a clinical significant ILD.

Objectives: To assess ILD frequency and severity (extension and functional impairment) in SSc patients, analyzing the association with the different autoantibodies.

Methods: Retrospective, descriptive study of patients meeting EULAR/ACR 2013 SSC criteria and had a HRTC performed at a tertiary Spanish hospital from 1975 to 2018. One hundred and eight patients were included. HRTCs were graded by two radiologists according to Goh et al semi-quantitative score. Three groups were established according to the presence of different autoantibodies: anticentromere (ACA), antitopoisomerase (ATA), and positivity to other ANAs (nucleolar pattern and other specificities).

Results: Clinical and laboratory characteristics are presented in table 1. Thirty-three patients had pulmonary involvement, 6 were ACA+, 18 ATA+ and 9 had other ANA specificities. The probability of not having pulmonary involvement among ACA+ and of having pulmonary involvement among ATA+ showed statistic significance ($p < 0.001$ and $p < 0.001$). Usual interstitial pneumonia (UIP) was the most frequently reported pattern (6 patients), followed by Non-Specific Interstitial Pneumonia (NSIP 11 patients); Six patients did not meet the radiological criteria for neither UIP or NSIP. No statistical significant difference was found among radiological pattern and autoantibody profile. Regarding extension: 15 patients