Jonathan Hoggatt Shareholder of: Magenta, Grant/research support from: Magenta, Consultant of: Magenta, David Scadden Shareholder of: Magenta, Consultant of: Magenta, Haley Howell Shareholder of: Magenta, Employee of: Magenta, Veit Schmelmer Shareholder of: Magenta, Employee of: Magenta, Jason Neale Shareholder of: Magenta, Employee of: Magenta, Tony Boitano Shareholder of: Magenta, Employee of: Magenta, Michael Cooke Shareholder of: Magenta, Employee of: Magenta, Dwight Morrow Shareholder of: Magenta, Employee of: Magenta, Glen Raffel Shareholder of: Magenta, Employee of: Magenta, Will Savage Shareholder of: Magenta, Employee of: Magenta, Kevin Goncalves Shareholder of: Magenta, Employee of: Magenta, Pat Falahee Shareholder of: Magenta, Employee of: Magenta, John Davis Shareholder of: Magenta, Employee of: Magenta

DOI: 10.1136/annrheumdis-2020-eular.3750

FRI0236

FIRST PILOT STUDY OF T1 MAPPING MRI WITH ECV MEASUREMENT OF PERIPHERAL MUSCLE IN SYSTEMIC SCLEROSIS PATIENTS SHOWS DIFFUSE FIBROSIS

R. B. Dumitru¹, A. Goodall^{2,3}, D. Broadbent², A. Kidambi⁴, S. Plein⁴, F. Del Galdo¹, A. L. Tan¹, J. Biglands², M. H. Buch^{1,5}. ¹Leeds Institute of Rheumatic and Musculoskeletal Medicine, Leeds, United Kingdom; ²Medical Physics and Engineering, Leeds Teaching Hospitals NHS Trust, Leeds, United Kingdom; ³Medical Imaging and Medical Physics, Sheffield Teaching Hospital Trust, Sheffield, United Kingdom; ⁴Leeds Institute of Cardiovascular and Metabolic Medicine, Leeds, United Kingdom; ⁵Centre for Musculoskeletal Research, School of Biological Sciences, Manchester, United Kingdom

Background: Peripheral myositis in systemic sclerosis (SSc) is associated with poor prognosis and myocarditis¹ but non-inflammatory myopathy, which also represents a significant cause of disability in SSc remains poorly understood. Cardiovascular magnetic resonance (CMR) T1 mapping studies in asymptomatic SSc patients show increased extracellular volume (ECV), suggestive of diffuse fibrosis in both the myocardium and thoracic muscle².

Objectives: To evaluate the feasibility of T1 mapping MRI and determine ECV in peripheral muscle of SSc patients with and without myopathy and explore the association between cardiac and peripheral muscle T1 mapping in SSc.

Methods: This was a hypothesis-generating pilot and feasibility study. SSc patients, fulfilling the 2013 ACR/EULAR criteria, with no cardiovascular disease or myositis but either minimal muscle symptoms (non-inflammatory myopathy) or no muscle involvement and healthy volunteers (HV) underwent peripheral muscle T1 mapping-MRI for native T1 and extracellular volume (ECV) quantification of the dominant thigh. Patients also had T1 mapping CMR, and creatine-kinase (CK) measured. Non-inflammatory myopathy was defined as current/history of minimally raised CK (<600 IU/l) +/- presence of clinical signs-symptoms (including proximal myasthenia and/or myalgia) +/- a Manual Muscle Testing (MMT) score <5 in the thighs.

Results: 12 SSc patients and 10 HV were recruited. SSc patients had a median (IQR) age of 52 (41,65) years, 9/12 had limited cutaneous SSc, 4/12 interstitial lung disease, 7/12 non-inflammatory myopathy. Higher skeletal muscle ECV was recorded in SSc patients compared to HV [mean (SD) 23(11)%, vs 11(4)% p=0.04]. Skeletal muscle native T1 values were comparable between the 2 groups although modestly higher in SSc patients [mean (SD) 1396ms (56) vs 1387ms (42)] (Figure 1A).

Peripheral muscle ECV associated with CK ($R^2 = 0.307$, $\rho = 0.554$, $p = 0.061$) and was higher in SSc patients with evidence of myopathy compared to those with no myopathy [28 (10)% vs 15 (5)%, $p = 0.023$] (Figure 1B). An ECV of 22% was determined to best identify myopathy with a sensitivity of 71% and a specificity of 80%.

SSc patients had raised myocardial ECV and native T1 with means (SD) of 31 (3) % and 1287 (54) ms respectively (normal reference range ECV $\leq 29\%$, native T1 ≤ 1240 ms). No clear association between myocardial and peripheral muscle ECV ($\rho = -0.485$) or between myocardial ECV and peripheral muscle native T1 ($\rho = -0.470$) of SSc patients was observed.

Conclusion: This pilot study to determine ECV-MRI of the peripheral muscle showed higher ECV in SSc patients compared to HV, suggesting the presence of diffuse fibrosis in the peripheral muscle of SSc patients. These data support further investigation to understand the pathophysiological involvement and relationship of peripheral and cardiac muscle in SSc.

References:

- [1] Follansbee WP et al, Am Heart J. 1993
- [2] Barison A et al, Eur Heart J Cardiovasc Imaging. 2015

Figure 1A. Peripheral muscle native T1 and ECV in HV compared to SSc patients. 1B. Mean (SD) ECV and native T1 in those with and without myopathy

Disclosure of Interests: Raluca-Bianca Dumitru: None declared, Alex Goodall: None declared, David Broadbent: None declared, Ananth Kidambi: None declared, Sven Plein: None declared, Francesco Del Galdo: None declared, Ai Lyn Tan: None declared, John Biglands: None declared, Maya H Buch Grant/research support from: Pfizer, Roche, and UCB, Consultant of: Pfizer; AbbVie; Eli Lilly; Gilead Sciences, Inc.; Merck-Serono; Sandoz; and Sanofi

DOI: 10.1136/annrheumdis-2020-eular.5724

FRI0237

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

M. Erdogan¹, B. Kilickiran Avci², C. Ebrun², Y. Ersoy², Z. Ongen², G. Ongen³, V. Hamuryudan¹, G. Hatemi¹. ¹Istanbul University -Cerrahpasa Medical Faculty, Internal Medicine, Division of Rheumatology, Istanbul, Turkey; ²Istanbul University -Cerrahpasa Medical Faculty, Department of Cardiology, Istanbul, Turkey; ³Istanbul University -Cerrahpasa Medical Faculty, Department of Pulmonary Diseases, Istanbul, Turkey

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: Sixty-nine consecutive pts fulfilling ACR/EULAR 2013 SSc criteria have been screened for PH until now, 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 and 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 (LVD) were not included. RHC was performed in all patients with positive screening according to any one of the screening algorithms. Pts 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 69 SSc pts, 27 were excluded due to ILD (n=6), LVD (n=6), already diagnosed PH (n=4) no measurable TRV (n=5), lung cancer (n=2), pulmonary embolism (n=1) and nephrotic syndrome (n=1). Among the remaining 42 patients, 17 required RHC according to at least one of the screening algorithms (Table 1). Number of patients who had suspected pulmonary hypertension and required RHC according to ESC/ERS 2015, DETECT and ASIG were 7 (17%), 13 (31%), and 12 (29%) respectively (Figure 1). Among the 17 pts. who had RHC, PH was present in 3 pts according to the 25-mmHg cut-off (Group 2 in 2, Group 3 in 1) and in 9 pts according to the 20-mmHg cut-off (Group 1 in 5, Group 2 in 3, Group 3 in 1). The sensitivity and specificities were presented in Table 2. ASIG and DETECT had better sensitivity for 25-mmHg cut-off and was better with ASIG for 20 mmHg cut-off. The specificity was better with ESC/ERS for both cut-off values.

Conclusion: The ASIG algorithm has a better sensitivity and ESC/ERS algorithm has a better specificity 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.

Table 1. Clinical Features of Patients

	All Patients n=42	ESC/ESR screen positive, n=7 (17%)	ESC/ESR screen negative, n=35 (83%)	DETECT screen positive, n=13 (31%)	DETECT screen negative, n=29 (69%)	ASIG screen positive, n=12 (29%)	ASIG screen negative, n=19 (59%)
Age, median [IQR]	51 [46-75]	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]
Male/Female, n (%)	3/39	-	3/32	2/11	1/28	0/12	3/27
Duration of SSc symptoms, year [IQR]	13.5 [5-20.75]	52 [39-57]	65 [51-79.5]	24 [5-38]	12 [5-25-16.75]	14 [5-37]	13 [5-19]
Diffuse Limited	12 (28)	2 (29)	10 (29)	2 (15)	10 (34)	4 (67)	8 (27)
Anti-Scl70 (+)	20/33 (61)	3/6 (50)	17/27 (63)	5/11 (44)	15/22 (68)	5/9 (56)	15/24 (62.5)
Anticentromere (+)	8/39 (21)	3/7 (43)	5/32 (16)	6/12 (50)	2/27 (7)	4 (33)	4/27 (15)
Presence of IRI, n (%)	25 (60)	4 (57)	21 (60)	9 (69)	16 (55)	7 (58)	18 (60)
Presence of digital ulcer(ever), n (%)	26 (62)	3 (43)	23 (66)	8 (62)	18 (64)	8 (67)	18 (62)
Presence of GIS involvement, n (%)	20 (48)	5 (71)	15 (43)	6 (52)	14 (50)	6/11 (55)	14 (50)
Presence of telangiectasia, n (%)	23 (55)	5 (71)	18 (58)	9 (75)	14 (54)	8/10 (80)	15/28 (54)
sPAP on TTE, median (mmHg) [IQR]	30 [26-34]	41 [34-46]	28 [26-32]	32 [30-43]	28 [26-34]	38 [30-42]	28 [26-32]
FVC, median (% predicted) [IQR]	87.5 [71.5-100.25]	94 [62-117]	86 [72-99]	82 [71-96]	79 [69.25-100]	94 [69.25-115.25]	84.5 [71.5-97.5]
DLCO, median (% predicted) [IQR]	60.50 [50.75-72.5]	52 [39-57]	64 [51-78]	57 [43.5-73.5]	61 [53-76]	53 [43-59.25]	65 [51.75-78.5]
FVC / DLCO ratio, median [IQR]	1.39 [1.18-1.72]	2.11 [1.58-2.18]	1.35 [1.16-1.55]	1.64 [1.29-2.13]	1.36 [1.13-1.57]	1.84 [1.41-2.12]	1.35 [1.0-1.47]
NT-proBNP, median (pg/mL) [IQR]	141 [62.75-196]	310 [210-887]	120 [52-168]	4210 [133.5-546]	120 [45-163.5]	260 [149-593.5]	114.5 [48.5-165.75]
Presence of dyspnea, n (%)	16 (38)	7 (100)	9 (26)	6 (46)	10 (35)	7 (58)	9 (30)
Class 1	14 (33)	6 (86)	8 (100)	5 (83)	9/9 (100)	6 (86)	8 (100)
Class 2	1 (3)	1 (14)	-	1 (7)	-	1 (14)	-
Class 3	-	-	-	-	-	-	-
Class 4	-	-	-	-	-	-	-

Figure. Number of patients who required RHC according to the algorithms

Table 2. Performance of the algorithms for screening PHT in patients						
	25 mmHg cut-off			20 mmHg cut-off		
	ESC/ESR	ASIG	DETECT	ESC/ESR	ASIG	DETECT
Sensitivity, %	67	100	100	56	89	56
Specificity, %	64	36	29	75	50	0
PPV, %	29	25	23	71	67	39
NPV, %	90	100	100	60	80	0
PPV: Positive predictive value NPV: Negative predictive value						

Disclosure of Interests: Mustafa Erdogan: None declared, Burcak Kilickiran Avci: None declared, Cansu Ebrun: None declared, Yagmur Ersoy: None declared, Zeki Ongun: None declared, Gul Ongun: None declared, Vedat Hamuryudan Speakers bureau: Pfizer, AbbVie, Amgen, MSD, Novartis, UCB, Gulen Hatemi Grant/research support from: BMS, Celgene Corporation, Silk Road Therapeutics – grant/research support, Consultant of: Bayer, Eli Lilly – consultant, Speakers bureau: AbbVie, Mustafa Nevzat, Novartis, UCB – speaker

DOI: 10.1136/annrheumdis-2020-eular.3826

FRI0238

AUTOLOGOUS FAT GRAFTING IN THE TREATMENT OF FACIAL SCLERODERMA: A SINGLE - CENTRE EXPERIENCE

P. Faggioli¹, L. Castelnovo¹, A. Tamburello¹, M. Falaschi¹, E. Saporiti¹, A. M. Lurati², A. Laria², A. Mazzone¹. ¹Ospedale di Legnano, Legnano, Italy; ²Ospedale Giuseppe Fornaroli, Magenta, Italy

Background: Systemic sclerosis (SSc) is a systemic autoimmune disease characterized by progressive cutaneous and internal organ fibrosis. Orofacial manifestations are disabling and treatment options are limited. Fat Tissue Grafting (FTG) can be used for treating facial manifestations of the fibrosis.

Objectives: In this study, we aimed to assess the safety and efficacy of FTG of our cohort of patients with SSc.

Methods: We enrolled 20 SSc (18W, 2M) patients, from 2016 to 2019, suffering from facial sclerosis and restricted mouth opening capacity. FTG was carried out in accord with modified Coleman's procedure (1): fat tissue was taken from periumbilical or trochanteric areas and was injected in 8 different points around the mouth. No side effects or adverse reactions have been documented. Evaluations included mouth opening capacity by measuring interincisal distance, oral functionality (MHISS scale) and patient global satisfaction (by Global Health scale).

Results: A 11 mm (8 - 18mm range) median increase of interincisal distance was reported at month 6 and in 80% of patients at month 12, too (p<0.03). A significant improvement in MHISS scale was also observed (p<0.03). The patient satisfaction questionnaire showed 95% positive results and 80% of the patient replied affirmatively to the question about the repetition of FTG but only 2 patients required new FTG after 12 months.

Conclusion: Our results showed that FTG improved mouth opening capacity and that aesthetic and functional results were satisfying to about 90% of the patients; long-term effects of this type of treatment are currently unknown. However, our and literature data at 12 months follow-up seems to confirm the benefits in long term, despite the filling effect is over.

This study – that's one of the largest case series described right now (2) - supports the possible therapeutic role of autologous FTG in improving facial scleroderma both in aesthetic and in functional aspects.

References:

- [1] Coleman SR. Structural fat grafting: more than a permanent filler. *Plast Reconstr Surg.* 2006; 118: 108S-20S
- [2] Fat Grafting for the Treatment of Scleroderma. Strong AL, Rubin JP, Kozlow JH, Cederna PS. *Plast Reconstr Surg.* 2019 Dec;144(6):1498-1507. doi: 10.1097/PRS.0000000000002691.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.1527

FRI0239

ANTI-NXP2 ANTIBODIES: CLINICAL AND SEROLOGICAL ASSOCIATIONS IN A MULTICENTRIC ITALIAN STUDY

M. Fredi¹, I. Cavazzana¹, A. Ceribelli², M. G. Lazzaroni¹, S. Barsotti³, M. Benucci⁴, L. Cavagna⁵, L. De Stefano⁵, A. Doria⁶, G. Emmi⁷, M. Fornaro⁸, F. Furini⁹, R. Gerli¹⁰, M. G. Giudizi⁷, M. Govoni⁹, A. Ghirardello⁶, L. Iaccarino⁶, F. Iannone⁸, M. Infantino⁴, A. Mathieu¹¹, E. Marasco⁵, P. Migliorini¹², B. Palterer⁷, P. Parronchi⁷, M. Piga¹¹, F. Pratesi¹², A. Radice¹³, C. Selmi², V. Ricciari¹⁴, M. Tampoia¹⁵, G. Zanframundo⁵, A. Tincani¹, F. Franceschini¹. ¹Rheumatology Unit Spedali Civili Università di Brescia, Brescia, Italy; ²Rheumatology Unit Humanitas, Rozzano, Italy; ³Rheumatology Unit AOU Pisana, Pisa, Italy; ⁴Rheumatology Unit Ospedale S. Giovanni di Dio, Firenze, Italy; ⁵Rheumatology Unit Policlinico San Matteo, Pavia, Italy; ⁶Rheumatology Unit Università Studi di Padova, Padova, Italy; ⁷AO Universitaria Careggi, Firenze, Italy; ⁸Rheumatology Unit Università di Bari, Bari, Italy; ⁹Rheumatology Unit Università di Ferrara, Ferrara, Italy; ¹⁰Rheumatology Unit Università di Perugia, Perugia, Italy; ¹¹Rheumatology Unit, Università di Cagliari, Cagliari, Italy; ¹²Immunologia, AO Universitaria Pisana, Pisa, Italy; ¹³Microbiologia e Virologia Ospedale San Carlo Borromeo, Milano, Italy; ¹⁴Policlinico Umberto I, Roma, Italy; ¹⁵Laboratorio Patologia Clinica Policlinico Bari, Bari, Italy

Background: anti-NXP2 antibodies is considered a serological marker of dermatomyositis (DM), with calcinosis, severe myositis and, in some series, cancer. Historically, these associations have been detected with immunoprecipitation (IP), but in the last few years commercial lineblot (LB) assay have been released.

Objectives: to analyze the clinical features associated to anti-NXP2 antibodies, including the onset of concomitant cancers, both with LB and homemade IP

Methods: clinical and serological data from medical charts of 213 patients with a diagnosis of inflammatory myositis without anti-NXP2 (NXP2-), followed-up by two third-level Centers, and 61 anti-NXP2+ patients from 10 Rheumatological centers were analyzed. Anti-myositis specific (MSA) and anti-myositis associated antibodies (MAA) were detected in single centers by LB (Euroimmun Autoimmune Inflammatory Myopathies 16 antigens). Anti-NXP2 was confirmed by protein and RNA IP, as previously described (1)

Results: clinical diagnosis of anti-NXP2+ positive with LB were 42 DM, 11 PM, inclusion body myositis (IBM) 4, necrotizing myositis and overlap (OM) 1 each. Anti-NXP2+ showed a lower age at onset (p<0.0001) more frequent diagnosis of DM (68.8%vs30%,OR5.2) and IBM (6.5%vs0.49%,OR14.8), typical skin manifestations, myositis (93%vs79% OR3.3), concomitant presence of another MSA (12.7%vs2%, OR6.41) and lower rate of features associated with OM or anti-synthetase syndrome. Serum from 49 NXP2+ was available and IP analysis