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 ILO, 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 (93)	6 (86)	8 (100)	5 (83)	9/9 (100)	6 (86)	8 (100)
Class 2	1 (7)	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						

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FRI0238

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

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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
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FRI0239

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

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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