Methods: Thirty-two SSc patients underwent a cardiac MRI with dedicated lung scanning and chest HRCT on the same day. One-hundred-thirty-five regions of interest (ROIs) were identified, and STIR and T1 sequences were acquired [before (T0) and after 5 (T5), 10 (T10) and 15 (T15) minutes from gadolinium injection]. The ROIs were classified according to HRCT as normal, dependent areas (probably related to blood pooling in supine position) and pathological areas (ground glass ± reticulation on HRCT). Mean STIR and T1 times were also calculated for each patients, and correlated with FVC, DLco, B-lines on lung ultrasound and HRCT semi-quantitative scoring (Scleroderma Lung Study score). Patients were followed-up and lung worsening was defined on the basis of clinical judgement and at least >15% DLco decline.

Results: Mean STIR and mean T1 times were significantly different between normal, dependent and pathologic areas ($p < 0.001$ between groups). Patients' mean STIR showed a significant correlation with DLco ($R = -0.56$, $p < 0.01$), HRCT Scleroderma Lung Study score ($R = 0.52$, $p < 0.01$) and B-lines on lung ultrasound ($R = 0.63$, $p < 0.01$). The mean STIR of the 10 patients who developed a worsening pulmonary involvement had significantly different MRI signal intensity in comparison to the 25 patients without worsening pulmonary involvement (125 ± 46 vs 66 ± 37 msec, $p < 0.01$).

Conclusions: Our data highlight the usefulness of lung MRI in SSc patients to differentiate normal, dependent and pathologic areas, without need for contrast medium administration, and with good correspondence to other functional and imaging parameters. STIR values may have prognostic implications to predict lung worsening. Lung MRI, although still very preliminary, is a promising imaging tool that in the future may integrate HRCT in SSc-related ILD.

Disclosure of Interest: None declared

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FRI0440 RENAL INVOLVEMENT IN MIXED CONNECTIVE TISSUE DISEASE: A SINGLE CENTRE EXPERIENCE

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Background: Kidney injury in mixed connective tissue disease (MCTD) is an uncommon manifestation. Prevalence has been reported to be <4% in some cohorts. The frequency of renal involvement in Hispanic patients with MCTD is not known.

Objectives: We aimed to describe the prevalence, clinical characteristics and outcomes of renal involvement in Mexican patients with MCTD.

Methods: We conducted a retrospective single-centre study. We included patients with a diagnosis of MCTD according to the Alarcón-Segovia criteria who regularly attended to a referral centre in Mexico City (2003–2017) and we identified those with renal involvement defined as proteinuria >500 mg/d with or without active sediment, creatinine elevation 50% above baseline or development of glomerular filtration rate (GFR) <60 ml/min, with no other known cause. We collected demographics, clinical manifestations, follow-up time, treatment, outcomes and damage (SLICC/ACR-DI), renal function, serological and histological variables.

Results: One hundred and thirty one patients with MCTD were followed at our centre. We identified 14 patients with renal involvement with a prevalence of 10.7%. Among those patients, 13 were women (92.8%); mean age at onset of renal involvement was 44 ± 8 years. Most frequent manifestations were Raynaud's phenomenon in 13 (92.8%) patients, arthritis in 12 (85.7%), puffy hands in 12 (85.7%), sclerodactyly in 8 (77.1%), sicca syndrome in 8 (77.1%) and myositis in 7 (50%). Median time elapsed from MCTD diagnosis to renal involvement was 83 (2–365) months. In 3 patients, renal involvement was present at MCTD onset. Seven (50%) patients had other signs of MCTD activity at the time of renal involvement onset. Four (28.5%) patients presented with sub-nephrotic proteinuria, 3 (21.4%) with nephrotic range proteinuria and kidney injury, 2 (14.3%) with sub-nephrotic proteinuria and kidney injury, 2 (14.3%) with nephrotic range proteinuria, 1 only with nephrotic range proteinuria and 1 (7.1%) with end-stage renal disease. Microscopic hematuria was present in 9 (64.3%) patients and leukocyturia in 6 (42.8%). Renal biopsy was performed in 8 (57%) patients; pathological diagnoses were: crescentic and necrotizing glomerulonephritis (GN) (2 patients; one of these patients developed positive ANCA antibodies), GN ISN/RPS 2003 class III+V, 1 GN ISN/RPS 2003 class III+V with thrombotic microangiopathy, 1 GN ISN/RPS 2003 class IV+V and vasculopathy, 1 membranous GN, 1 minimal mesangial GN and chronic tubulointerstitial nephritis with vasculopathy. 1 Ten (71.4%) patients achieved either total or partial remission at a median follow up of 82 (1–367) months. Only one patient required dialysis. At last follow up the median SLICC/ACR-DI was 1.5 (0–4) points. Two patients died.

Conclusions: In our cohort of MCTD patients, prevalence of renal involvement was low, although higher than the one reported in other populations. Clinical presentation and pathological diagnoses were diverse. Renal biopsy was helpful, since glomerulonephritis, vasculopathy and overlap with ANCA associated

vasculitis were found in several patients; these options should be considered in the differential diagnoses of MCTD patients with renal involvement.

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FRI0441 IDIOPATHIC INFLAMMATORY MYOPATHIES: CLINICAL CHARACTERISTICS, SURVIVAL AND POOR PROGNOSTIC FACTORS OF 110 PATIENTS FROM TURKEY

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Objectives: In this study, we aimed to assess clinical features, poor prognostic factors and survival analysis of patients with Idiopathic inflammatory myopathy (IIM).

Methods: Records of 110 patients with IIM that had at least 6 months of follow-up, fulfilling Bohan and Peter's criteria were analysed for this study. Survival analysis was done by using Kaplan Meier method and multiple "Cox" regression analysis was applied to calculate the effect of multiple factors.

Results: Sixty-eight percent of 110 patients was female, the mean age of the patients was 46 years, and the average follow up time was 77.5 months. Diagnosis of these patients was dermatomyositis (DM) in 68%, polymyositis (PM) in 26%, autoimmune necrotizing autoimmune myopathy (NOM), inclusion body myositis (IBM) 6%. The percentage of periungual erythema, arthritis, dysphagia, respiratory muscle involvement and interstitial lung disease (ILD) 56, 22, 24, 32, 11 and 30. Malignancy was identified in 26% of patients. The percentages of malignancies diagnosed at the time of diagnosis, before the diagnosis and during the follow-up were 3.6, 11.8 and 8.2. The most frequent malignancy was breast cancer. Others are carcinoma of gastrointestinal tract, lung, and genitourinary tract. ANA was present in 36% and 12% of patients was positive for anti-Jo-1 antibody. The average daily dose of prednisolone was 7.5 mg, the average usage time was 35.5 months. Causes of death were aspiration pneumonia-sepsis (50%) and malignancy (25%). Significant associations with mortality was found between systemic symptoms, periungual erythema, respiratory muscle involvement, dysphagia, presence of malignancy. Mortality was higher in ANA negative patients ($p < 0.001$). Five and 10 year survival in these patients were 83% and 75%. Five year survival rate in patients with respiratory muscle involvement was 38% and 68% in those with dysphagia. The presence of systemic symptoms, and malignancies were identified as risk factors for mortality in multivariable analysis.

Conclusions: ILD and malignancies are frequent in our IIM cohort. Malignancies are mostly detected at diagnosis. The mortality rate was high and the most common cause was infection. 10 year survival rate was 79%. Malignancy, respiratory muscle involvement, dysphagia, negative ANA have a detrimental effect survival in IIM patients.

Disclosure of Interest: None declared

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FRI0442 LONG-TERM TREATMENT WITH RITUXIMAB IN INTERSTITIAL LUNG DISEASE RELATED TO SYSTEMIC SCLEROSIS: OUR CLINICAL EXPERIENCE

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Background: Systemic sclerosis (SSc) is an immune-mediated disorder characterised by abnormal fibrosis and diffuse microangiopathy with skin and internal organ involvement. Interstitial lung disease (ILD) represents one of the most challenging complication of SSc, difficult to manage and correlate with a poor prognosis. Chest Computed Tomography (CT) is the gold standard for detection and evaluation of SSc-ILD by means of semi-quantitative scoring of extent of lung involvement. Some preliminary data suggest that rituximab (RTX) may be usefully employed in the treatment of SSc patients.

Objectives: To investigate the role and effect of RTX on ILD in our SSc patients' series.

Methods: We retrospectively evaluated a series of 18 SSc patients (M/F 6/12, mean age 54.6 ± 17.6 SD years, mean disease duration 11.4 ± 6.5 SD years, L/D cutaneous subsets 6/12) who received one or more cycles of RTX (4 weekly infusions of 375 mg/m²) every 6 months for a total of 1–6 cycles. Lung involvement was studied by means of pulmonary function tests (PFTs) (18/18) and inspiratory