should be aware of such specific features as similar survival rates in cutaneous and ssSSc pts, and late recognition of PAH in pts with ssSSc, as well as its similarity with IPAH.

Disclosure of Interest: None declared

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AB0792 AUTOIMMUNE AND INFLAMMATORY DISTURBANCES IN PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION ASSOCIATED WITH SYSTEMIC SCLEROSIS

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Background: The prognosis of patients with pulmonary arterial hypertension associated with systemic sclerosis (SSc-PAH) is significantly worse, than other forms of PAH, and mechanisms of this phenomenon are unknown. Therefore, the isolation of autoimmune disorders is of great importance for early diagnosis and differential diagnosis, as well as the search for new therapeutic targets.

Objectives: To identify the autoimmune disorders in patients with SSc-PAH.

Methods: The study includes 52 pts with idiopathic pulmonary arterial hypertension (IPAH), 51 pts with SSc-PAH, 65 pts with SSc without PAH. Serum concentrations of the C-reactive protein (CRP), anticentromere antibodies (ACA) and antibodies to topoisomerase-I (anti-Scl-70) were routinely measured. The control group consists of 146 volunteers. Statistical analysis includes univariable logistic regression, ROC analysis and Kaplan-Mayer method.

Results: The average age of patients with IPAH was 37.9±10.5 years, SSc-PAH – 52.3±12.7 years, SSc without PAH – 51.2±13.2. Patients did not differ in functional class (FC), which was the main criterion of comparability. Mean values of FC in groups with SSc-PAH and IPAH also did not differ (2.7±0.8 and 2.6±0.7, respectively). ACA was associated with a 15.2-fold increased odds of developing PAH in SSc (OR 15.2, 95% CI 5.4–43.0), on the contrary, presence of anti-Scl-70 associated with low risk of PAH (OR 0.5, 95% CI 0.01 to 0.21). The level of CRP in the serum was significantly higher in patients with PAH than in the control group: 4.1 (1.9, 10.0) and 0.61 (0.25, 1.9), $p < 0.000001$, and also in comparison with patients without PAH (1.9 (0.8, 6.4), $p = 0.02$). In pts with PAH, the level of CRP correlated with FC and right atrium pressure and 6 min walk test distance. The level of CRP was significantly higher in patients with FC III-IV compared with FC I-II and in non-surviving patients. The Kaplan-Mayer analysis showed that pts with CRP level more than 4.75 mg/L at the time of diagnosis of PAH had a significantly lower survival rate (median 48 months) than pts with normal values (median 91 months) ($p < 0.005$), with 67% sensitivity and 61% specificity.

Conclusions: SSc-PAH is a unique phenotype combining the manifestations of SSc and PAH, the pathogenetic mechanisms of which modify the course of these states. It is based on a feature of autoimmunity with the predominance of ACA and a low occurrence of anti-Scl-70. An increase in the concentration of CRP, as well as the effect of its baseline level on survival, attests to the significant role of autoimmunity and inflammation in the pathogenesis of this fatal SSc complication.

Disclosure of Interest: None declared

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AB0793 DESCRIPTIVE ANALYSIS OF A COHORT OF PATIENTS DIAGNOSED WITH INFLAMMATORY MYOSITIS IN A TERTIARY HOSPITAL

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Background: Inflammatory myopathies are a group of rare systemic diseases characterised by muscle weakness and inflammation. Clinical manifestations, course and prognosis of these pathologies are very heterogeneous.

Objectives: The aim is to describe the main characteristics of patients diagnosed with inflammatory myositis fulfilling Bohan and Peter criteria.

Methods: Descriptive analysis of a cohort of 34 patients from the same hospital with follow-up between January 2010 and December 2017. We recorded demographic characteristics, clinical manifestations, treatment, comorbidity and mortality.

Results: 34 patients (73% female) were recruited with an average age at diagnosis of 56.3 years^{27–83} among adults and 10 years^{4–15} among children. Most of them were Caucasian (94%). 18% were smokers and 15% previous smokers. The most frequent type was dermatomyositis (DM) (40%) followed by antisynthetase syndrome (ASS) (15%), necrotizing myopathy (12%), inclusion body myopathy (12%), overlap myositis (9%) and polymyositis (9%). 2 patients (out of 4) with necrotizing myopathy were treated with statins.

Clinical manifestation included muscle weakness (84%) and skin manifestations (48%) mainly among DM patients. 8 patients (24%) showed interstitial lung disease (4 non-specific interstitial pneumonia, 3 usual interstitial pneumonia and 1 cryptogenic organising pneumonia), especially among patients with overlap syndrome (n=3), DM (n=2) and ASS (n=2). Pulmonary hypertension occurred in 7 patients (21%), 30% among patients with overlap myositis associated to systemic sclerosis. The rest of extramuscular manifestations are expressed in the table 1. Muscle biopsy was performed in 57% of patients (77% compatible with myopathy). MRI was carried out in 45% (100% with active myositis). EMG was performed in 94% of patients with myopathic findings in 67% of them. 20 patients (60%) presented positive antinuclear antibodies, being the most frequent antiPML-SCL (18%), antiJo1 (18%), antiRo (12%) and anti-MDA5 (9%).

All patients were treated with corticosteroids. Only 2 responded to corticosteroids in monotherapy. More than 90% needed additional immunosuppressive treatment and 65% received 2 or more immunosuppressants. The most commonly used drugs were methotrexate (72%), rituximab (28%), azathioprine (25%), immunoglobulins (21%) and cyclophosphamide (21%). Only in 12% treatment could be stopped because of sustained remission.

3 cases of cancer (9%) were reported: myelodysplastic syndrome, lung neoplasm (in the case of paraneoplastic myositis) and lymphoma.

During the follow-up period 4 deaths were registered (12%) due to infections and cancer.

38% of patients required a multidisciplinary approach.

	n	%	Dermatomyositis	Polymyositis	Inclusion Body Myopathy	Necrotizing Myopathy	Antisynthetase syndrome	Overlap syndrome
Raynaud	7	21,2%	2	0	0	1	2	2
Cardiac Manifestations	7	21,2%	1	2	1	2	1	0
Arrhythmia	4	12,2%	1	1	1	0	1	0
Cardiomyopathy	1	3%	0	0	0	1	0	0
Heart attack	1	3%	0	1	0	0	0	0
Aortic stenosis	1	3%	0	0	0	1	0	0
Gastrointestinal Manifestations	6	18,1%	1	1	2	1	1	0
Dysphagia	3	9,1%	1	1	0	0	1	0
Chronic gastritis	2	6%	0	0	1	1	0	0
Constipation	1	3%	0	0	1	0	0	0
Severe Infections	5	15,1%	3	1	0	0	1	0
Hematologic Manifestations	5	15,1%	0	0	2	1	1	1
Anaemia	4	12,1%	0	0	2	1	1	0
Anaemia + thrombocytopenia	1	3%	0	0	0	0	0	1
Arthritis	4	12,1%	1	0	0	0	1	2

Conclusions: Inflammatory myopathies have frequent multiorgan involvement and represent a heterogeneous group of systemic diseases as shown in our registry and in the literature. Most patients need chronically combined immunosuppressive treatment and few achieve sustained remission. In consequence the collaboration of several specialties is necessary for the diagnosis and management of these pathologies.

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AB0794 MYOSITIS DAMAGE INDEX IN A MYOSITIS POPULATION COHORT

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Background: Idiopathic inflammatory myopathies (IIM) are a heterogeneous and uncommon group of diseases characterised by muscular involvement and many systemic manifestations. They usually have a chronic course, and despite treatment they often develop functional impairment. MDI-MYODAM a score to assess damage has been recently developed. Few publications in this regard have reported a prevalence of 92%. There are no studies in Mexican-mestizo population describing the extent of damage and its main determinants.

Objectives: To investigate the prevalence of damage measured with the MDI-MYODAM tool in patients with IIM and its relationship with characteristics of the disease and treatment

Methods: A cross-sectional study was conducted in an IIM cohort from a national reference hospital. MDI questionnaire was applied to all patients. Demographic, disease characteristics, IIM subgroup (dermatomyositis, polymyositis and overlap), comorbidities (Charlson index), medical treatment were collected. Descriptive statistics were applied. Bivariate Pearson correlation test was conducted to

analyse associations between MDI score and clinical characteristics. P value <0.05 was considered as a statistically significant.

Results: Fifty subjects participated, 75% were women, mean age at diagnosis was 40.7 years (SD $\pm$) and mean duration of the disease was 11.8 $\pm$ 8.3 years. Thirty-one percent of patients had polymyositis, 31% dermatomyositis, 12% juvenile dermatomyositis, 10% overlap myositis (mainly systemic lupus erythematosus, scleroderma and rheumatoid arthritis). The mean Charlson index was 2.7 $\pm$ 2.4. Eighty-one percent had a score greater than 0 for at least one category; majority within the endocrinological systems (55%), skeletal (51%), gastrointestinal (35%). The mean total damage score (MDI) was 3.6 $\pm$ 2.9. The mean score for the total damage according to the MYODAM score was 6.9 $\pm$ 5.1. A moderate correlation between the MDI score and the use of glucocorticoids (r 0.3, CI 95% 0.1–0.6, $p<0.01$). No associations between damage and activity of the disease or IIM subgroup were observed.

Abstract AB0794 – Figure 1

Conclusions: Most patients with IIM develop some kind of damage, with greater impact on the endocrinological, skeletal and gastrointestinal systems. There increases with the time of evolution and the use of steroids in a non-linear way. Therefore, these results should be taken into account in the decision-making process in long-term treatment.

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AB0795 LONG-TERM EFFICACY AND SAFETY OF RITUXIMAB IN SYSTEMIC SCLEROSIS REFRACTORY TO CONVENTIONAL IMMUNOSUPPRESSANTS

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Background: Recent studies^{1 2} have shown that B-cells are involved in the pathogenesis of systemic sclerosis (SSc) suggesting that B-cell depletion might have a role in the treatment of the disease³.

Objectives: To describe the long-term efficacy and safety profile of Rituximab (RTX) in SSc patients who failed previous conventional immunosuppressive drugs.

Methods: We retrospectively analysed medical records of all SSc patients treated with Rituximab in our Rheumatology Unit from July 2011 to December 2017. Disease manifestations, particularly skin, pulmonary and articular involvement, were evaluated at baseline (T0), at 8 $\pm$ 2 months (T1) and at the last follow-up visit (T2).

Results: Thirteen patients, 11 females and 2 males, were treated with 32 RTX cycles (median per patient 2.5, range 1–4). Eleven patients had diffuse and 2 limited cutaneous form; all had positive ANA, 9 anti-TopoI, 2 anti-RNA polIII, and 1 anti-SSA.

The mean age at first RTX infusion was 49.3 $\pm$ 13.6 years and the mean disease duration was 6.5 $\pm$ 3.3 years. The main indications to RTX were: interstitial lung disease (11/13), worsening of skin involvement (6/13) and arthritis (5/13), refractory to at least one previous immunosuppressive drug (median 2, range 1–3). Twelve patients were treated with mycophenolate mofetil, 5 with cyclophosphamide, 4 with methotrexate and 2 with azathioprine.

All patients were treated with RTX (1000 mg twice, two weeks apart). Twelve patients received more than one cycle; the interval between cycles was 12.7 $\pm$ 5.4 months. The mean follow-up period was 41.4 $\pm$ 21.9 months.

Five patients had arthritis and we observed complete remission of arthritis in all of them. 8 months after the first RTX administration arthritis relapsed in 4 patients who were successfully retreated.

We did not observe a significant decrease in modified Rodnan Skin Score over time (T0: 15.6 $\pm$ 6.9, T1: 14.6 $\pm$ 7.8, and T2: 13.1 $\pm$ 8.3; $p=ns$).

Lung involvement, detected in 11/13 patients, remained radiologically stable in 8 patients and worsened in 3. No significant changes in the pulmonary function tests was observed from T0 to T2.

Five patients had a follow-up longer than 33 months after the last RTX cycle: 3 patients had a stable disease, 1 developed severe PAH and 1 lung neoplasia.

CD19 cell depletion ($<2\%$) was observed after each RTX cycle, without significant changes in immunoglobulin levels. No infusion reactions were observed. We registered only one severe side effect: hand cellulitis in a patient with digital ulcer, requiring hospitalisation.

Conclusions: Our experience suggests that RTX is well tolerated and could be effective in the management of severe and refractory SSc manifestations.

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AB0796 SHORT TERM TREATMENT RESPONSE TO INTRAVENOUS PULSE CYCLOPHOSPHAMIDE TREATMENT IN CONNECTIVE TISSUE RELATED INTERSTITIAL LUNG DISEASE

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Background: Interstitial lung disease (ILD) is a major cause for morbidity and mortality in patients suffering from connective tissue diseases. Early detection and prompt recognition of symptoms with appropriate treatment is necessary for effective control of the disease and for better prognosis and long-term survival. Various treatments have been approved based on observational studies. We have very few studies as regards ILD treatment from the Indian subcontinent. In a resource constrained country like India, we mainly rely on cheap drugs like cyclophosphamide. We have done a study to assess the short-term treatment response of connective tissue related interstitial lung disease to cyclophosphamide in a tertiary centre in India.

Objectives: 1.To assess short treatment outcome for Connective tissue disease related interstitial lung disease treated with intravenous pulses of cyclophosphamide (CYP) based on improvement in-

Lung function 1) Forced vital capacity (FVC)% predicted, forced expiratory volume in first second (FEV1)% predicted 2) Dyspnoea Borg scale 3) Cough visual analogue score

2. To determine the factors affecting treatment outcome like age, sex, duration of the connective tissue disease, type of connective tissue disease, HRCT type and presence of PAH.

Methods: A cohort study was conducted in the Department of General Medicine, Rheumatology and Pulmonology in Amala Institute of Medical Sciences, Thrissur, Kerala from November 2015 to June 2017, that evaluated 74 patients who were having connective tissue disease related ILD. Detailed history and a thorough clinical examination was done. Treatment with 6 intravenous pulses of cyclophosphamide was done and response was assessed after 6 months. Response to treatment and associations were analysed using SPSS software (version 15). Method used for statistical analysis was Chi –Square test and paired t test.

Results: