can be associated with significant morbidity and mortality related to systemic involvement or treatment-related complications.

Objectives: This study aimed to describe the clinical features and survival of patients with inflammatory myositis in our centre.

Methods: We performed a single-centre, retrospective study on patients with inflammatory myositis (polymyositis [PM], dermatomyositis [DM], or Anti-Jo1 syndrome) seen from 2000 to 2014, noting their demographic data, clinical features and outcome until December 2016. The primary outcome assessed was all-cause mortality. Cumulative mortality rates were estimated using the Kaplan-Meier test; the Log Rank (Mantle-Cox) test was used to compare subgroup differences in survival.

Results: Seventy-four patients (19 PM, 28 DM, 27 Anti-Jo1 syndrome) were available for the study. Median age at diagnosis was 47 years (min 17, max 75) and median follow-up time was 93 months (min 4, max 311). Sixty-one patients (82, 4%) were female and 13 (17, 6%) were male. Malignancy was found in 4 patients (2 invasive ductal adenocarcinoma of breast, 1 over cancer and 1 non-small cell lung cancer) and they were all female DM patients. Nineteen patients (25,7%) died at the end of the follow-up.

The 19 patients with PM consisted of 15 female and 4 male, with a median age at diagnosis of 45 years (min 22, max 74) and median follow-up time of 88 months (min 5, max 204). The 28 patients with DM consisted of 24 female and 4 male, with a median age at diagnosis of 52 years (min 17, max 75) and median follow-up time of 80 months (min 4, max 288). The 27 patients with Anti-Jo1 syndrome consisted of 22 female and 5 male, with a median age at diagnosis of 50 years (min 22, max 63) and median follow-up time of 117 months (min 5, max 311).

Overall survival rates of the whole group were 91%, 83%, and 76% for 1, 5, and 10 years, respectively. The survival rates at 1, 5 and 10 years from the diagnosis were respectively 88%, 82%, %82 for PM, %88, 80%, %80 for DM and 96%, 88%, 74,5% for Anti-Jo1 syndrome. But there was no significant difference between the survival rates of the diagnose groups ($p=0,734$). Also there wasn't a significant difference between the survival rates of sex and age groups ($p=0,503$), ($p=0,112$). But the survival rates were significantly lower in patients with the time from diagnose less than 8 years ($p=0,000$).

Figure 1. Survival probability of all patients with inflammatory myositis (Graph A) and according to the diagnose* (Graph B). *1. Polymyositis; 2. Dermatomyositis; 3. Anti-Jo1 syndrome

Conclusions: Our study involved 74 patients followed up for a median of 7, 7 years and is one of the largest cohorts of patients with inflammatory myositis in Turkey. Survival was quite similar with the literature. Usually mortality has been expected in the first years after the diagnosis.

Disclosure of Interest: None declared

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SAT0339 LUNG TRANSPLANTATION IN PATIENTS WITH INTERSTITIAL LUNG DISEASE ASSOCIATED WITH ANTISYNTHEASE AND ANTI-MDA5 SYNDROMES. EXPERIENCE FROM A REFERENCE SPANISH HOSPITAL

E. Trallero-Araguás¹, C. Berastegui², M. López-Corbeto¹, I. Bello³, A. Román², A. Selva-O'Callaghan⁴. ¹Rheumatology Unit; ²Pneumology Department; ³Thoracic Surgery Department; ⁴Internal Medicine Department, Hospital Vall d'Hebron, Barcelona, Spain

Background: Interstitial lung disease (ILD) is the most characteristic feature and prognosis determinant of patients with antisynthetase (AS) and anti-MDA5 syndromes. Despite immunosuppressive treatment, ILD sometimes progresses to an end-stage lung disease, for which lung transplantation (LT) is the only therapeutic option. There is scarce data about post-LT outcome in this group of patients.

Objectives: To describe clinical characteristics and post-LT outcome of patients with ILD associated to AS and anti-MDA5 syndromes included in the LT program of the Vall d'Hebron Hospital of Barcelona.

Methods: We performed a review of patient records listed in the LT program.

Results: From 1990 to 2016 ten patients (5 women) with ILD related to AS or anti-MDA5 syndromes were included in the LT program (3% of patients with ILD accepted for LT). Nine patients (2 anti-MDA5, 4 anti-Jo1, 2 anti-PL12 and 1 anti-PL7) received LT while 1 patient (anti-Jo1) was still in list at the end of this study. Median age (range) of disease diagnosis was 39 years (25–55). Six patients had

clinical myopathy [2 dermatomyositis (DM) and 4 polymyositis] whereas 1 patient was diagnosed with amyopathic DM. Four patients had associated pulmonary hypertension. Time between disease diagnosis and patient inclusion in LT list was higher in anti-Jo1 patients [median (range) 8.8 years (8.3–17.6)] than in the rest of the cohort [anti-MDA5 <1 year; anti-PL12 0.7 and 3.9 years; anti-PL7 1.3 years] ($p<0.05$). Four patients underwent bilateral LT and 5 unilateral LT. Three patients received an urgent LT (2 anti-MDA5 and 1 anti-Jo1). Six patients presented an histologic pattern of UIP and 3 of NSIP. Early complications (<45 days) included: primary graft dysfunction in 3 patients; phrenic paresis in 2 patients and infection in 6 patients. One patient suffered an acute rejection (AR). Six patients presented late complications (>45 days): one patient developed an acute cellular rejection (ACR) and an autoantibody mediated rejection 8 and 10 months from LT respectively; 4 patients suffered chronic lung allograft dysfunction (CLAD), 2 of whom are still alive; Infection was diagnosed in 6 patients. One patient developed a squamous skin carcinoma. No flare of myopathic or lung disease was observed after LT in any case. Four patients (44%) died: 2 of an acute respiratory failure in the immediate postoperative period (1 caused by suture dehiscence and 1 by refractory ACR); 1 of an invasive aspergillosis, 17 months after LT; and 1 of CLAD after 54 months. Median follow-up (range) of the rest of the cohort was 35 months (8–70). All patients who survived more than 45 days recovered an optimal functional capacity for daily activities with no request for long-term oxygen therapy.

Conclusions: LT should be considered a valid option to treat patients with end-stage or severe and rapidly progressive ILD associated to AS and anti-MDA5 syndromes. An early remission to LT referral centers for evaluation should be considered especially in non-Jo1 patients. No relapse of myositis or ILD was observed after LT. Mortality could not be attributed to the primary disease.

Disclosure of Interest: None declared

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SAT0340 SUBCLINICAL CARDIOVASCULAR DISEASE IN SCLERODERMA: A STUDY WITH CARDIOVASCULAR RISK CHARTS, CT CORONARY CALCIUM SCORE AND CAROTID ULTRASONOGRAPHY

I. Sanz Pérez¹, F. Martínez Valle¹, A. Guillén del Castillo¹, N. Pizzi², A. Roque Pérez³, H. Cuéllar Calábría³, A. Fernández Codina¹, E. Callejas Moragas¹, V. Fonollosa Pla¹, C.P. Simeón Aznar¹. ¹Internal Medicine; ²Nuclear cardiology; ³Radiology, Hospital Universitari Vall d'Hebron, Barcelona, Spain

Background: Recently published population-based cohort studies had shown a high prevalence of cardiovascular (CV) disease in Systemic Sclerosis (SSc) patients.

Objectives: The aim of this study is to compare three different methods to measure CV risk in patients with scleroderma.

Methods: We conducted a cross-sectional study in a single center that included 43 SSc patients without CV events. We used both CV risk assessment charts SCORE for populations with low CV risk and REGICOR algorithm adjusted to Spanish subjects. Coronary Compute Tomography (CT) with coronary arterial calcium score (CACscore) was performed considering several cut-off points as predictors of CV risk. Carotid doppler ultrasound was performed to measure the Carotid Intima Media Thickness (CINT) and for the detection of cholesterol plaques, according to Mannheim consensus criteria.

Results: Risk factors and SSc related features are described in table 1. None of the patients were catalogued as high risk according to SCORE chart (>5%). According to REGICOR chart, 17 patients (39,5%) were catalogued as intermediate risk and none as high risk. Twenty-two patients (51,2%) had carotid plaques (CP) and the CACscore of these patients was 283.4. In patients without CP CACscore was 53,2 ($p<0.05$).

Based on the presence of CP we performed ROC curve with CACscore. The AUC was 0.778. The best cut off point was 28 with a sensibility of 71% and a specificity of 82%. Kappa's coefficient was 0.54. Twenty patients (46,5%) had CACscore>28. Compared to patients with CACscore<28 statistical significance was found on CINTmax (0.93 vs 0.80mm; $p<0.01$), presence of CP (79% vs 25%, $p<0.01$) and number of CP (2.55 vs 0.43, $p<0.01$). There was no statistical significance on CINTm (0.71 vs 0.64mm p: 0.11).

Female	37 (82%)
Age	59 (33–83)
Disease duration	19 (3–57)
Limited cutaneous SSc	32 (74,4%)
Diffuse cutaneous SSc	10 (23,3%)
Sine Scleroderma SSc	1 (2,3%)
Arterial Hypertension	26 (60,5%)
Diabetes mellitus	5 (11,6%)
Statin treatment	22 (51,2%)
Smokers	5 (11,6%)
Digital ulcers	24 (55,8%)
Pulmonary Hypertension*	6 (14%)
Interstitial lung disease	20 (46,5%)
Altered conduction in Electrocardiogram	15 (34,9%)
Left Ventricular diastolic dysfunction**	32 (74,4%)
Left ventricular systolic dysfunction**	3 (7%)

*Mean pulmonary artery pressure >25 mm Hg at right heart catheterization. **Measured by echocardiography.