Patients who experienced their first non-RP feature of the disease before the onset of RP were included with a simultaneous onset.

Conclusions: Several clinical and serological differences were evident between the three racial groups. Asians had high prevalences of Scl-70, PH and of a reduced FVC. Black patients in contrast had fast disease onset and a high prevalence of diffuse skin involvement.

Disclosure of Interest: None declared


SAT0476


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Background: Systemic Sclerosis (SSc) is a complex and heterogenous chronic inflammatory disease characterised by widespread fibrosis of the skin and visceral organs, microvascular injury and evidence of immune system activation. Diagnosis can be challenging in the absence of specific laboratory markers or diagnostic criteria.

Objectives: To determine the incidence, prevalence and mortality of physician diagnosed SSc in a population based US cohort and evaluate the performance of the ACR/EULAR 2013 Classification criteria in comparison to the 1980 ACR criteria in classifying patients with SSc.

Methods: Medical records of patients with a diagnosis or suspicion of SSc in Olmsted County, Minnesota from January 1, 1980 to December 31, 2016 were reviewed to identify incident cases of SSc (defined by physician diagnosis). Prevalent cases of SSc in Olmsted County on January 1, 2015 were also identified. Incidence and prevalence rates were age and sex adjusted to the 2010 US white population. Survival rates were compared with the expected rates in the population of Minnesota. Fulfilment of the 1980 and 2013 classification criteria was ascertained.

Results: A total of 79 incident cases of SSc from 1980 through 2016 and 49 prevalent cases on Jan 1, 2015 were identified. Of these, 71 (90%) were females, 68 (87%) were caucasians, age at diagnosis 55.8±15.9 years (mean ±SD). The overall age- and sex-adjusted annual incidence for 1980–2016 was 2.7 (95% CI: 2.1–3.3) per 1.00 000 population. The age-adjusted incidence was 4.6 (95% CI: 3.5–5.7) per 1.00 000 for females and 0.6 (95% CI: 0.2–1.1) per 1.00 000 for males, with no change in incidence over time. The age- and sex-adjusted prevalence on January 1, 2015 was 47.4 (95% CI: 34.1–60.7) per 1.00 000 population.

64 of 79 (81%) patients fulfilled the 2013 classification criteria, while only 48% fulfilled the 1980 criteria. All but 1 patient that fulfilled the 1980 criteria, also fulfilled the 2013 criteria. All 79 patients had Raynaud’s, 38 had cardiopulmonary involvement (pulmonary artery hypertension and/or interstitial lung disease), 33 had digital ulcers/pitting scars, 66 had telangiectasias and 69 had sclerodactyly. 66 patients had limited cutaneous SSc, 11 had diffuse cutaneous SSc and only 2 had SSc sine scleroderma. 39 patients had a positive autoantibody for SSc: anti-centromere antibodies (12), anti-U1-RNP antibodies (9), anti-topoisomerase 1 antibodies (12), anti-RNA-polymerase III (3) and anti-PL7 antibodies (5). The autoantibody profiles were rare and were not significantly higher in patients with the 2013 classification criteria compared to the 1980 classification criteria.

Conclusions: Incidence and prevalence rates were age and sex adjusted to the 2010 US white population. Survival rates were compared with the expected rates in the population of Minnesota. Fulfilment of the 1980 and 2013 classification criteria was ascertained.

Disclosure of Interest: None declared


INTENSIFIED B-CELL DEPLETION THERAPY IN DISEASE-SPECIFIC AUTOANTIBODIES ASSOCIATE WITH PROGRESSIVE SYSTEMIC SCLEROSIS PATIENTS: 24 MONTHS FOLLOW-UP

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Background: Systemic sclerosis (SSc) is a connective tissue autoimmune disease with systemic involvement and a serious medical condition with a high rate of mortality, especially due to interstitial lung disease (ILD). The exact pathophysiology is still unclear, but B cells seem to play a crucial role in the initiation and the progression of the disorder. Therefore, the use of Rituximab (RTX) might have a rationale in the treatment of SSc.

Objectives: We aimed to investigate the outcomes of SSc patients treated with RTX after a follow-up of 24 months.

Methods: We retrospectively collected data from SSc patients resistant or intolerant to previous therapies, treated with intensified B-depletion therapy, between 2013 and 2016. Therapeutic protocol comprehends: RTX 375 mg/sm on days 1, 8, 15, 22, and two more doses after one and two months, associated with two intravenous administrations of 10 mg/kg of cyclophosphamide and three methylprednisolone pulses (15 mg/kg) followed by oral prednisone (0.8 mg/kg/day, rapidly tapered to 5 mg/day by the end of the 3rd month after RTX).

Results: We included 20 SSc patients (18 females and 2 males; mean age 66.7±11.0 years). Patients presented with severe multorgan involvement: ILD (19/20, 95%), pulmonary hypertension (12/20, 60%), and skin thickening (17/20, 85%). After a follow-up of 24 months, we observed a decrease in the levels of NT-proBNP (mean baseline: 385±4517, mean at 24 months: 283±648, p<0.05), and in the Modified Rodnan Skin Score (mRSS) (mean mRSS baseline: 14.4±10.5, mean after 24 months of follow-up: 12.9±10.0, p<0.05). Four out 19 (21%) patients experienced a significant improvement of ILD, as assessed by high-resolution computed tomography, while in 12/19 (63%) patients the intensified B-cell depletion therapy was associated with a stabilisation of the imaging features with no sign of progression. Three out of 19 (16%) patients showed a deterioration of the ILD.

Patients showed no significant decrease in forced vital capacity (FVC) (mean baseline FVC: 0.83±0.19, mean after 24 months of follow-up: 0.22±0.23), no significant decrease in forced expiratory volume in one second (FEV1) (mean baseline FEV1: 85.5±15.6, mean at 24 months: 87±21.2), no significant decrease in diffusing capacity (DLCO) (mean baseline DLCO values: 58.8±8.6, mean at 24 months: 60.3±14), no significant change in the ejection fraction (EF) (mean baseline EF values: 62.8±6.4, mean EF at 24 months: 58.6±7.1). In pulmonary artery pressure (PAP) (mean baseline PAP: 30.2±10.5, mean at 24 months: 31.1±11.05).

Conclusions: Despite recent advances in the treatment of SSc, ILD heavily affects prognosis and life expectancy of these patients. Our data suggest that the intensified B-depletion therapy protocol might represent a promising tool for the management of SSc in terms of controlling the progression of the disease, especially when considering pulmonary and skin manifestations. Further prospective studies are needed in order to confirm our results.

References: