Abstract AB0663 Table 1: Outcome measure scores (SF-36 and HAQ-DI) of patients with SSc

SF-36 Scores Whole group (n=78) DcSSc (n=60) LcSSc (n=18)

PCS 35.65 ± 23.65 34.56 ± 22.90 39.31 ± 26.36

MCS 41.36 ± 25.94 39.82 ± 26.27 46.51 ± 24.84

HAQ-DI scores

HAQ-DI 1.40 ± 0.95 1.43 ± 0.99 1.31 ± 0.78

Abstract AB0663 Figure 1: Correlation HAQ-DI with physical component summary ($r=0.641$, $p<0.001$)

Figure 2: Correlation HAQ-DI with mental component summary ($r=-0.360$, $p=0.001$)

Abstract AB0663 Figure 3: Distribution of SSc patients by functional disability (n=78)

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AB0664

REYNOLDS SYNDROME: ABOUT 7 CASES

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Background: Reynolds syndrome is a rare condition defined by the association of primary biliary cholangitis (PBC) with CREST syndrome. The frequency of scleroderma during PBC varies between 3 and 17%. The diagnosis is most often done during systematic search for autoimmune diseases associated with CBP.

Objectives: To determine the epidemiological, clinical, paraclinical and evolutionary features of Reynolds syndrome

Methods: We report 7 cases of Reynolds syndrome collected in the our department between 1996 and 2014

Results: There were 7 middle-aged women with an average age of 49.2 years (32-61 years old). The diagnosis of CREST syndrome with anti centromeric positive antibodies was retained in 6 cases. Extensive trunk sclerosis associated with diffuse sclerodacty and anti centromeric antibodies was noted in 1 case. An isolated acrosyndrome was noted in 1 case. The hepatic assessment found non icteric cholestasis in 6 patients. Anti-mitochondrial antibodies type M2 were positive in 6 cases. Liver biopsy showed histologic lesions consistent with primary biliary cholangitis PBC in 5 cases. Sjörger's syndrome was confirmed histologically in 1 case. Autoimmune diseases (rheumatoid arthritis and lymphocytic thyroiditis) were associated with Reynolds Syndrome in 2 cases. The treatment consisted of ursodeoxycholic acid alone (N = 4), or combined with D-penicillamine (N = 1), azathioprine-associated prednisone (N = 1) or a calcium channel blocker alone (N = 1). The evolution was favorable in 2 cases. In the other 3 patients, worsening of cholestasis and complications such as oedemato-ascitic decompensation and gastrointestinal haemorrhage were noted during the course of the study. One patient died of cataclysmic gastrointestinal bleeding by rupture of oesophageal varices.

Conclusion: Reynolds syndrome is a rare entity that should be discussed in the presence of liver function disturbances in patients with CREST syndrome. Conversely, the CREST syndrome should be referred to and sought systematically in case of PBC.

Disclosure of Interests: None declared

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AB0665

OUTCOME OF INTERSTITIAL LUNG DISEASE IN PATIENTS WITH SCLERODERMA, A 5 YEAR PROSPECTIVE COHORT STUDY

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Background: Systemic sclerosis (SSc) is a multi-system disorder characterized by a disturbance in fibroblast function, microvascular disease, and immune system activation, resulting in fibrosis of the skin and internal organs.

Objectives: To study changes in lung function in patients with scleroderma with ILD on treatment followed up prospectively over a period of 5 years

Methods: In this study, 73 patients with scleroderma with ILD were prospectively followed up from July 2013 to July 2018. Patients' clinical features, investigations, treatment and complications were recorded at regular interval

Results: Of 73 patients, 68 were females and 5 were males. 4 had only one visit and 11 had incomplete data. Baseline characteristics are shown in table 1. Mean PAH at baseline was 35.95±14.26 mmHg and at end of study was 39.44±14.44 mmHg. Immunosuppressant's used were MMF in 52, cyclophosphamide in 15, azathioprine in 13, and methotrexate in 17 patients. 61 patients received oral prednisolone with mean dose of 6.46±7.10 mg/day during maintenance and mean dose 15±8.95 mg/day during exacerbation of ILD. There was no statistically significant difference in mean FVC of 59 patients at baseline (59.76±16.91) and at end of the study (57.03±16.77) and in subgroup analysis of patients who received MMF ($p > 0.05$). Mean 6MWD at baseline was 370±58.83 meters and at end of study was 377±88.96 meters. Comorbidities seen were hypertension in 15, DM in 6, osteoporosis in 23 (of which one had vertebral fracture), AVN in 3 patients. 5 patients died because of ILD. 13 patients needed home O2.

Table: Mean age, duration of disease and age and follow up.

Parameter	Mean	SD
Mean age at enrollment (yrs)	43.24	11.70
Mean duration of disease (yrs)	10.02	6.86
Mean duration of ILD (yrs)	6.02	2.94
Mean duration of prospective follow up (yrs)	3.80	2.28

Figure 1. Change in FVC of 59 patients of SSc with ILD after 5 years of follow up

Figure 2. Change in FVC of 46 patients of SSc with ILD taking MMF after 5 years of follow up

Conclusion: This prospective study was carried out with aim to determine progression of ILD in patients with systemic sclerosis with ILD over period of 5 years. In our study, there was no statistically significant change of FVC over period of time. Worsening of lung function with deterioration in FVC >10% was seen in only 13 patients (22%) ($P < 0.01$). Our findings are similar to study done by Le Gouellec et al in which 75 patients with scleroderma and ILD were followed over period of 72 months of which 26% patients showed significant worsening of FVC ($\geq 10\%$)¹. In our study there statistically significant decline in DLCO and increase in PAH over period of 5 years ($p > 0.001$). In study carried out by Le Gouellec et al studying Scleroderma/ILD, there was significant decline in DLCO over period of 72 months of follow up¹. In study done by Stephen C. Mathai et al, there was progressive increase in PAH when patients with scleroderma and ILD with PAH were followed up for period of 4 years².

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AB0666

ALL-CAUSE AND CAUSE-SPECIFIC MORTALITY TREND IN PATIENTS WITH SYSTEMIC SCLEROSIS: A POPULATION-BASED STUDY

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Background: Systemic sclerosis (SSc) is a disease with one of the highest mortality rates among connective tissue diseases with 5-year survival ranging from 68-88%[1]. Recent research has shown an improvement in survival for some rheumatic diseases at the population level [2, 3]. Contemporary data on mortality trends in SSc is scarce, especially at the general population level.

Objectives: 1) To investigate all-cause and cause-specific mortality in patients with newly diagnosed systemic sclerosis (SSc) compared to the general population.

2) To assess mortality trends between 2 calendar time periods, 1997–2004 and 2005–2012

Methods: Using an administrative health database, we compared all patients with new diagnosis of SSc with non-SSc individuals randomly selected from the general population matched for sex, age, and time of entry into the study. The study cohorts were divided into 2 groups based on the year of diagnosis ("early cohort [1997–2004] and "late cohort" [2005–2012]). The outcome was death (all-cause and cause specific including cancer, cardiovascular disease [CVD], infections, renal disease, pulmonary disease and other causes) during the follow-up period. Hazard ratios (HR) were estimated using Cox proportional hazards models, first adjusted for age, sex, and entry cohort time and then adjusted for selected baseline covariates (health care resource utilization, medications, pre-existing comorbidities, and socioeconomic status), based on a purposeful selection algorithm.

Results: We included 874 patients with SSc (83.6% female and mean age of 54.19 in the early cohort, and 81.7% female and mean age of 55.32 in the late cohort) and 8,740 non-SSc individuals in this study, contributing 4,083.8 and 47,515.8 person-years of follow-up, respectively. We observed 168 deaths in the SSc cohort, and 504 deaths occurred in the non-SSc cohort during the follow-up. Overall, the age-, sex-, and entry time-adjusted all-cause mortality HR in the SSc cohort was 4.47 (95% CI 3.72–5.37), and the fully adjusted remained significant [HR=2.87 (95% CI 2.35–3.50)]. Moreover, there was excess mortality due to CVD-related causes in the age-, sex-, and entry time-adjusted model (HR=2.65 [95% CI 1.68 – 4.18]), but it did not persist in the fully adjusted model (HR= 1.52 [95% CI 0.93–2.48]). There was no significant difference in other cause-specific mortalities, including cancer mortality (HR=0.91 [95% CI 0.56–1.49]). As a consequence of the residual disclosure policy of Population Data BC to ensure patient confidentiality, we were not allowed to report on the deaths due to infection, renal or pulmonary disease, as <5 deaths were associated with each outcome. There was no improvement in all-cause mortality between the early and late cohort (fully adjusted HR= 3.73 and 95% CI 2.49–5.59; HR 2.51 and 95% CI 1.88–3.34, respectively).

Conclusion: This population-based study showed that people with SSc have a 3-fold increase risk of death when compared with the general population. The risk of death has not improved over time and this calls for further research to close this gap.

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