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)

Disclosure of Interests: None declared

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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.