Conférence laurentienne de rhumatologie
Laurentian Conference of Rheumatology

Abstract #: 2416

Category: Travail de recherche / Research work

Primary author: Sabrina Hoa **Institution**: Centre hospitalier de l'Université de Montréal, Montréal, Quebec

Additional Authors: Maggie Larche, McMaster University, Hamilton, Ontario; Nouha Lahmek, Université de Montréal, Montreal, Québec; Janet Pope, University of Western Ontario, London, Ontario; Mohamed Osman, University of Alberta, Edmonton, Alberta; May Choi, University of Calgary, Calgary, Alberta; Carter Thorne, Southlake Regional Health Centre, Newmarket, Ontario; Joshua Levy, Université de Montréal, Montreal, Quebec; Marie Hudson, McGill University, Montreal, Quebec

Title: **Characterization of incident interstitial lung disease in late systemic sclerosis**

Objective(s): Interstitial lung disease (ILD) is a common and potentially severe complication of systemic sclerosis (SSc). Screening by HRCT is recommended in all SSc patients, particularly in the presence of risk factors, including early disease. Little is known on the incidence, characteristics, risk factors, evolution and treatment outcomes of ILD presenting in late SSc. The objective of this study was therefore to characterize late-onset ILD in comparison to earlier-onset ILD.

Method(s): Subjects enrolled in the Canadian Scleroderma Research Group (CSRG) cohort from 2004 to 2020 without prevalent ILD were included. Demographic, clinical and serological characteristics were compared between patients with incident vs no incident ILD on HRCT stratified by disease duration above (late) and below (early) 7 years from first non-Raynaud manifestation. Incidence rates (and 95% CI) for ILD diagnosis and progression were calculated based on Poisson distribution. Survival analyses with unadjusted Kaplan-Meier and adjusted Cox proportional hazard models were used to assess the association between disease duration and time to ILD progression. Exploratory analyses were done to evaluate the effect of immunosuppressive drugs on lung disease progression in late-onset SSc-ILD.

Result(s): Out of 1117 CSRG patients without prevalent ILD at baseline, 197 (18%) developed incident ILD over a median [IQR] duration of 2.4 [1.2, 4.3] years. The incidence rate in late-onset SSc-ILD was 3.7 (95% CI: 3.1-4.3) per 100 person-visits, which was lower than in early-onset SSc-ILD at 5.4 (95% CI: 4.2-6.9) per 100 person-years (RR 0.68, 95% CI: 0.51-0.92, p=0.01). Risk factors for incident ILD included presence of anti-topoisomerase I autoantibodies, absence of anti-centromere autoantibodies and higher C-reactive protein levels. Patients with late-onset ILD were also more frequently non-Caucasian and more often had diffuse cutaneous involvement, arthritis, myositis and anti-RNA polymerase III autoantibodies. Lung disease characteristics and severity were similar in late-onset and early-onset ILD. The incidence of lung disease progression was 14.1 per 100 person-visits and not different between late- and early-onset ILD (log rank p=0.8, adjusted HR 1.11, 95% CI: 0.58-2.10). Exploratory analyses showed no significant difference in effect of immunosuppressive drugs on ILD progression, although the effect was numerically more protective in early-onset SSc-ILD.

Conclusion(s): ILD can present in late SSc, particularly in patients with anti-topoisomerase I autoantibodies, diffuse skin involvement and high C-reactive protein levels. The rate of lung disease progression was similar in late- and early-onset SSc. Screening for ILD should continue in late SSc. Frequency and modality of screening remain to be defined.