Abstract #: 18

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Association Between Immunosuppressive Therapy and Course of Mild Interstitial Lung Disease in Systemic Scleroderma.

Objectives: Interstitial lung disease (ILD) is a leading cause of mortality in systemic sclerosis (SSc). Little is known about the benefits of immunosuppressive drugs in mild ILD. Our aim was to determine whether use of cyclophosphamide (CYC) or mycophenolate mofetil (MMF) was associated with an improved ILD course in patients with normal or mildly impaired lung function.
Methods: A retrospective cohort of SSc subjects with ILD, disease duration below seven years and no prior exposure to CYC or MMF was constructed from the Canadian Scleroderma Research Group registry. Subjects were categorized as having mild ILD if baseline forced vital capacity (FVC % predicted) was > 85%. The primary exposure was any use of CYC or MMF at baseline. FVC at one year was compared between exposed and unexposed subjects, using multivariate linear regression.
Results: Out of 294 eligible SSc-ILD subjects, 116 met criteria for mild ILD. In this subgroup, mean (SD) disease duration was 3.7 (2.0) years. 13 (11.2%) subjects were exposed to CYC or MMF at baseline. The one-year FVC was higher in exposed subjects compared to unexposed subjects, by a difference of 8.49% (95% CI: 0.01 to 16.98%). None of the exposed subjects experienced clinically meaningful progression over two years, whereas 24.6% of unexposed subjects did.
Conclusions: In this real-world setting, CYC/MMF exposure at baseline was associated with higher FVC values and a lower risk of progression among subjects with mild ILD. These data suggest a window of opportunity to preserve lung function in SSc-ILD.