BACKGROUND: Intestinal lung disease (ILD) can be the first manifestation of connective tissue disease (CTD) and rheumatoid arthritis (RA). Pulmonologists are usually the first referral in these patients.

OBJECTIVES: To determine: 1) the prevalence of ILD as initial manifestation of CTD or RA and 2) clinical characteristics of such patients.

METHODS: From a database of consecutive patients with CTD or RA referred to our academic hospital from 2009 and 2017, we selected all the patients with ILD as initial manifestation of the disease. Periodic multidisciplinary evaluations were performed during a median follow-up of 48 (35-50) months.

RESULTS: 1) Fifty-four of the 427 patients with CTD or RA (12.6%) had ILD as initial manifestation (mean age: 63.9±12.9 yrs, F/M ratio: 20/34). Autoimmune myositis was diagnosed in 16/54 patients (29.6%), systemic sclerosis in 11 patients (20.4%), RA in 9 patients (16.7%), Sjogren syndrome in 9 patients (16.7%) and SLE in 3 patients (5.5%). Six patients remained classified as IPAF (11%). Among the 427 Rheumatology patients we followed-up in the same period, autoimmune myositis had the highest prevalence of ILD as initial manifestation (33.6%), followed by Sjogren syndrome (20.5%), systemic sclerosis (20.0%), RA (3.4%) and SLE (3.3%). 2) Patients with initial ILD were all firstly evaluated by the Pulmonologist and the main reasons for Rheumatology referral were positivity for one or more autoantibodies (57.4%), mainly ANA≥1:320, and joint pain (29.6%). Thirty-six patients (66.7%) received steroid and immunosuppressive therapy. In the 3 months before the first Rheumatology visit to relieve respiratory symptoms, Twenty of these patients (58.8%) had rheumatic manifestations during steroid tapering. ILD CT patterns were NSIP (25 patients, 52.1%), UIP (18 patients, 33.4%), and organizing pneumonia (5 patients, 9.3%). In four patients (4.5%), autoantibodies became positive during the follow-up. The final diagnosis of CTD or RA-ILD was made after a median period of 16.5 (6-39) months from the clinical onset.

CONCLUSION: In our study population, the prevalence of ILD as initial manifestation of CTD or RA was 12.6%. Autoimmune myositis, systemic sclerosis and Sjogren syndrome were the most frequent diagnoses. As our data confirmed, ILD is a major cause of death in patients with systemic autoimmune diseases and can progress despite immunosuppression [1]. Furthermore, clinical features may become evident even months after the disease onset. A multidisciplinary evaluation is therefore essential, not only at the time of diagnosis but also during the follow-up [2].

References: