



### **PF-ILD: Prevalence, Genetics, and Mortality**

Dr. Faverio from Italy and including data from two ILD hospitals, included 1,107 patients with non-IPF ILD, and they found 245 presenting fibrotic ILD and 75% with a progressive phenotype. And they analyzed the data related to the clinical features, including the drug that the patient received. And most drugs received were oral corticosteroids and immunosuppressive treatment.

The authors found that around 35% of the patients presented some adverse event associated with these two types of medications. And during the follow-up of two years, 25% died. So these are data similar to those data published 10 years ago for IPF. So again, the clinical features of these progressive fibrotic ILD patients are quite similar to those patients with IPF. And probably the therapeutic management will be similar to IPF and not to the entities that originally they are coming from.

These two studies coming from France and presented by the group of Leone, a low border related to the close national study. The first one, was focused on the data from 617 Fibrotic known IPF ILD patients from the French rare lung disease reference center in Leon, a single center study. And they found that the 165 patients, which represents 25% of all included Fibrotic ILDs showed progressive Palatine. The most predominant entities were autoimmune ILDs or unclassifiable ILDs is followed by chronic HP and other idiopathic interstitial pneumonia. The overall survival of BFI and li the three years was around 80% and the mortality was suited to the relative FVC decline at baseline higher than 10%. Therefore we can use this value of the decline of a FVC decline around 10% for identifying those cases with lung fibrosis different from IPA, from other types of fibrotic ILDs that are progressing, and we can act on them with the best treatment.

The other value related or associated with the progression of the disease, where the type of entity and those unclassifiable ILD cases idiopathic interstitial pneumonia and chronic or fibrotic HP cases had higher probability of progressing in the follow-up.

This was a very interesting eposter presented by Dr. Moreno evaluating the relatives of the sporadic IPF cases, especially looking for family aggregation through the medical history, the pulmonary function tests, a six minutes working test and the high resolution CT scan that there was performance in all of them and a total of 164 relatives from 94 IPF cases were evaluated and a total of 30 cases where they identify it as familial pulmonary fibrosis. What initially were identified as sporadic ones, up to 50% were new ILD diagnosis in siblings and 5% in decedents. Therefore, the complete study of relatives of the same generation as IPF patients could be useful for the ILDs screening or for identifying early ILD. Until now, only symptomatic relatives with IPF patients with familial aggregation were offered screening evaluation, and not in all the centers. After this study in line with the recent published data from Dr. Hunninghake, this advised the question "should we perform screening in the first degree relatives from all sporadic and familiar forms of IPF cases?" This is something that we have to, investigate more in depth, but definitely with this data on the table, we need to think at least about the potential cases in the first degree relatives, especially in siblings.