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Title: A retrospective cohort study of interstitial lung diseases in Denmark

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Body: Introduction Interstitial lung diseases are a heterogeneous group of diseases with varying degrees of inflammation and fibrosis. Epidemiological data based on the current diagnostic criteria are sparse.

Objectives The aim of this study was to characterize the distribution of ILD subtypes in a cohort of Danish patients referred to a tertiary referral hospital. Methods We included 431 patients diagnosed with interstitial lung disease (sarcoidosis excluded) and first visit at our department between April 1. 2003 and April 1. 2009. All diagnoses were re-evaluated according to current diagnostic criteria including the 2011 ATS/ERS IPF-guidelines. Details on diagnostics, lung function, comorbidity and treatment were recorded at baseline and throughout the follow-up period. Results A total of 186 patients were diagnosed with idiopathic interstitial lung diseases. In this group IPF (n=121) was the most common diagnosis (108 definite and 13 probable IPF) followed by NSIP (n=30), and DIP (n=20). Other large groups were unclassifiable fibrosis (n=62), end stage fibrosis (n=43), extrinsic allergic alveolitis (n=32) and ILD in collagen vascular disease (n=54). Of patients with CVD-ILD 14 showed a UIP pattern, 22 a NSIP pattern and 18 an unclassifiable fibrosis pattern. The remaining 54 patients in the cohort had 15 different diagnoses. Mean age at diagnosis was 61 years (SD=14), and mean symptom duration before referral was 2.5 years. Thoracoscopic biopsy was performed in 39%. The estimated incidence rate is 4.1 per 100,000/year. Conclusion The preliminary results of the study of this well-characterised cohort show that IPF is the most common diagnosis, and that a definite IPF-diagnosis can be made in the majority of these cases.