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| **A Retrospective Analysis and Descriptor of Patients with Progressive Pulmonary Fibrosis in an Australian Population** |
| Daniel Bird1,2, Robert Sheehy1, Greg Keir1 |
| *1Department of Respiratory Medicine, Princess Alexandra Hospital, QLD, Australia* *2 School of Medicine, Griffith University, QLD, Australia* |
| **Introduction/Aim:** Progressive pulmonary fibrosis (PPF) is a relatively newly described phenotype of interstitial lung disease (ILD). It is a heterogeneous group of conditions with different causes and disease behaviours. It results in a rapid and irreversible scarring of the lungs that may cause respiratory failure and eventual death. Until recently, immune suppression along with best supportive cares have been the only treatments available with variable results. An antifibrotic medication, Nintedanib, recently expanded its listing to cover other types of ILD with a PPF phenotype. There is a paucity of Australian specific data, and this study aims to improve our understanding of the prevalence, predictors and characteristics of these patients. **Methods:** This was a single institution retrospective descriptive analysis. The Princess Alexandra Hospital lung function respiratory database (Respiro) was used to screen for patients with a diagnosis of ILD and who had 2 or more lung function tests. The PBS criteria derived from the IN-BUILD trial was used for patient inclusion. ​The investigators then manually checked the electronic medical records, imaging and serial lung function to exclude patients and collect demographic data.**Results:** 251 patients were screened with 227 excluded, leaving 24 patients for inclusion and analysis. The most common types of ILD to develop a PPF phenotype were connective tissue disease-ILD (CTD-ILD) (38%), idiopathic interstitial pneumonia (21%) and fibrotic hypersensitivity pneumonitis (21%). Patients were predominantly caucasian (75%) and female (71%) in keeping with the high rate of CTD-ILD. The average age was 70.5yo and 67% had background immunosuppression. The annual rate of FVC decline observed was 265.9mL. This is comparable to the decline seen in trial patients from the IN-BUILD and SENSCIS trials. The annual reduction in DLCO predicted was 7.8%.**Conclusion:** We have demonstrated that this Australia Cohort is comparable to those published in the literature and highlighted the rapid deterioration in lung function these patients experience which is comparable to patients with IPF and SSc-ILD. |