



**Service Priorities and Programmes**  
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**Decline in lung function in patients with Idiopathic pulmonary fibrosis**

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**Introduction**

Idiopathic pulmonary fibrosis (IPF) is a rare disease. It is characterized by chronic progressive fibrosis of the lung. Patients will suffer from irreversible loss of lung function. The annual rate of decline in forced vital capacity (FVC) varies from 190ml in the IFIGENIA study to 428ml in the ASCEND trial. New treatment modalities such as Pirfenidone and Nintedanib were shown to slow down the decline in lung function in studies but not to reverse the disease progression. It is therefore important to understand the natural history of the disease in our population in order to guide treatment and to monitor treatment response.

**Objectives**

To determine the rate of decline in lung function in patients with idiopathic pulmonary fibrosis

**Methodology**

An IPF registry was set up in the Respiratory Medical Department (RMD) of Kowloon Hospital (KH) since 2015. All the patients with physician-diagnosed IPF who was followed-up in RMD since 2010 were included in this registry. Patient characteristics, lung function test results and comorbidities were recorded. These data was retrieved for the current retrospective, cross sectional descriptive study. Data was presented in mean+/- standard error.

**Result**

Thirty-three patients with physician diagnosed IPF were included in this study. Mean duration of follow-up was 1554 days. Mean age was 72 years. Twenty (60.6%) of them were male. Sixteen (48.5%) of them had a history of smoking (Smoker: 21.2%; ex-smoker: 27.3%; non-smoker: 51.5%). Seven (21.2%) of them had passed away at the time of data analysis. Thirty-two (97.0%) of them had a computed tomography of the thorax (CT thorax) being performed. Typical usual interstitial pneumonitis (UIP) pattern was noted in 69.7% of patients (23 out of 33). Probable UIP pattern was noted in 24.2% (8 out of 33) while other pattern was noted in 3% (1 out of 33). Surgical lung biopsy was performed in 3 of the patients (9.1%). Four patients was oxygen

dependent (12.1%). Seven (21.2%) of them were anti-nuclear factor (ANF) positive. Anti-ENA was positive in 3 subjects (9.1%). Rheumatoid factor was positive in 4 patients (12.1%). ANCA was positive in 4 patients (12.1%). One of the patient received lung transplant and none of them received Pirfenidone or Nintedanib. Twenty-one patients (63.6%) had at least 2 sets of lung function test being performed were included in the analysis of lung function decline. The mean forced vital capacity (FVC) was 2.28 liters (81.3% predicted). The mean diffusing capacity of the lungs for carbon monoxide (DLCO) was 11.86ml/min/mmHg (53.0% predicted). The mean rate of decline in FVC was 235ml per year. The mean percentage decline in FVC was 11.45% per year. The mean decline in DLCO was 14.5% per year.