

# European Respiratory Society Annual Congress 2012

**Abstract Number:** 1449  
**Publication Number:** P3653

**Abstract Group:** 1.5. Diffuse Parenchymal Lung Disease

**Keyword 1:** Idiopathic pulmonary fibrosis **Keyword 2:** Interstitial lung disease **Keyword 3:** Lung function testing

**Title:** Effect of pirfenidone on chronic interstitial pneumonia

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**Body:** Background: Several reports reveal the favorable effect of pirfenidone on early stage of idiopathic pulmonary fibrosis (IPF), but impacts on non-specific interstitial pneumonia (NSIP) and interstitial pneumonia (IP) associated with collagen vascular diseases (IP-CVD) are not clear. Objectives: To examine the effect of pirfenidone on chronic IP including IPF, NSIP and IP-CVD. Methods: Thirty-two patients were enrolled in the study evaluating the safety and efficacy of pirfenidone in IPF, NSIP and IP-CVD. Clinical diagnosis are IPF(14), NSIP(14), IP associated with scleroderma(3), and rheumatoid arthritis(1). Based on PaO<sub>2</sub> at rest and SpO<sub>2</sub> after 6 minutes walk test (6MWT), disease severity of those were classified into four groups [Table 1]. We retrospectively analyzed subjective symptom in British Medical Research Council scale, pulmonary function, KL-6, SP-D and CT findings before and during pirfenidone administration.

Table 1. Disease severity of patients

	PaO <sub>2</sub> at rest (mmHg)	SpO <sub>2</sub> after 6MWT	n
I	>80		9
II	70-79	When<90%, to III	8
III	60-69	When<90%, to IV	4
IV	<60	unnecessary to perform 6MWT	11

Results: Correlations was observed between subjective symptom and VC, but was not between subjective

symptom and KL-6, SP-D, CT findings. After administration of pirfenidone, VC improved above 5% in 22%, 50%, 25%, 45% of patients (severity grade I, II, III, IV, respectively). Subjective symptoms highly improved in patients with scleroderma (improvement ratio: 67% in scleroderma vs. 14%, 14%, 0% in IPF, NSIP, RA, respectively). Conclusions: Pirfenidone might have the favorable efficacy independent of disease severity, without severe adverse events, in chronic IP, especially in IP associated with scleroderma.