

European Respiratory Society Annual Congress 2012

Abstract Number: 1445

Publication Number: P3652

Abstract Group: 1.5. Diffuse Parenchymal Lung Disease

Keyword 1: Interstitial lung disease (connective tissue disease) **Keyword 2:** No keyword **Keyword 3:** No keyword

Title: The efficacy of pirfenidone in scleroderma related interstitial lung disease

Ms. Yukiko 13584 Miura s7081@nms.ac.jp MD ¹, Mr. Yoshiya 13585 Tsunoda chabo448@yahoo.co.jp MD ¹, Mr. Toru 13586 Tanaka toru10302000@nms.ac.jp MD ¹, Mr. Hiroyuki 13587 Takoi taco155485@hotmail.com MD ¹, Mr. Yohei 13588 Yatagai yoheiyatagai@gmail.com MD ¹, Mr. Shigen 13591 Rin owenlin1031@yahoo.co.jp MD ¹, Mr. Akimasa 13593 Sekine akimasa.sekine@gmail.com MD ¹, Mr. Kenji 13594 Hayashihara dora@nms.ac.jp MD ¹, Mr. Takefumi 13595 Saito takefumisaito@yahoo.co.jp MD ¹, Mr. Akihiko 13596 Gemma agemma@nms.ac.jp MD ² and Mr. Arata 13597 Azuma azuma_arata@yahoo.co.jp MD ². ¹ Department of Respiratory Medicine, National Hospital Organization Ibarakihigashi Hospital, Ibaraki, Japan and ² Division of Pulmonary Medicine, Infection and Oncology, Nippon Medical School, Internal Medicine, Tokyo, Japan .

Body: Introduction: The major cause of death in Systemic Sclerosis (SSc) is interstitial lung disease, and cyclophosphamide is an only agent which significantly demonstrated a beneficial effect on lung function in patients with scleroderma-related interstitial lung disease (SSc-ILD), however the effect was quite modest, and it is necessary to identify a reasonable alternative. Objectives: TGF-beta1 plays a critical role in the pathophysiology of pulmonary fibrogenesis. Pirfenidone exerts its antifibrotic effect through regulation of lung TGF-beta1 levels. This raises the possibility that agents targeting TGF-beta1 may be beneficial for SSc-ILD. Methods: We administered pirfenidone to 3 patients with SSc-ILD and evaluated pulmonary function. Results: Case 1 is a 62 year-old female. Vital capacity (VC) improved by pirfenidone. The change rate was +27.3% (+0.51L) for 5 months. Case2 is a 75 year-old female. VC improved remarkably, at the change rate of +44.4% (+0.32L) for 25 months. Case 3 is a 66 year-old female. VC improved at the rate of +8.3% (+0.17L) for 26 months. Conclusion: All of 3 patients with SSc-ILD demonstrated the favorable efficacy of VC by pirfenidone without severe adverse events. The previous studies documented that deteriorating lung function was associated with increased mortality in SSc-ILD. Therefore, it is necessary to identify and treat early stages of patients with SSc-ILD for the prevention of pulmonary function impairment. Pirfenidone exerts its antifibrotic effect through regulation of TGF-beta1, which is one of the important inducers of fibrogenesis in SSc. We suggest pirfenidone may be a possible option for SSc-ILD.