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

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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.