

## Effect Of Intravenous Pulse Cyclophosphamtoe (CYC) Therapy On Systemic Sclerosis (Ssc) Inflammatory Lung Disease

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Deterioration of lung function is the most frequent cause of death in SSc. Alveolitis is considered the initiating event of SSc lung fibrosis. Effective therapy of SSc alveolitis is, therefore, of paramount importance. Here we assessed the use of intravenous CYC on pulmonary function testing (PFT) and high resolution computerized tomography (HRCT) abnormalities in 15 patients with SSc and alveolitis.

### Methods

Fifteen patients with SSc and alveolitis diagnosed by clinical, bronchoalveolar lavage, or HRCT findings were treated with intravenous pulse CYC (750 mg/m<sup>2</sup> of body surface area) monthly for 6 months. If there was no improvement on PFT or HRCT findings, patients were continued with 6 monthly pulses. If the condition was stable or improved, patients received 3 additional bimonthly pulses. After the first year, patients continued with CYC pulses every 3 months for one year. PFT and HRCT of the chest were obtained during and following therapy to determine the efficacy of CYC treatment. Seven patients received low-dose oral corticosteroids for reasons other than interstitial lung disease.

### Results

A regression analysis was performed after normalizing the DLCO (diffusion capacity of the lung for carbon monoxide), FVC (forced vital capacity), and TLC (total lung capacity) values to the values obtained immediately prior to initiation of treatment. This analysis revealed a 61% improvement in the DLCO at 36 months, following initiation of CYC therapy. The FVC and the TLC improved by 28% and 27%, respectively, during the same period. These results were in contrast to a 35% decline in DLCO during the 36 months prior to therapy. FVC and TLC were essentially unchanged in the pre-treatment period. HRCT showed improvement in the ground glass appearance in 80% of the cases, while 20% were unchanged. Fibrosis improved in 67% and remained unchanged in 33%. Honeycombing, if present, disappeared in every case.

### Conclusion

Intravenous pulse CYC treatment alone or accompanied by low dose corticosteroid therapy was an effective and well tolerated treatment of SSc inflammatory lung disease. The greatest improvement was observed in DLCO, although there was improvement in FVC and TLC values, as well.