

Intermittent Epoprostenol Infusions in Systemic Lupus Erythematosus Associated Pulmonary Hypertension—A Series of Three Cases

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Abstract

Introduction: Pulmonary hypertension (PHT) associated with systemic lupus erythematosus (SLE) has a grave prognosis. Continuous prostacyclin infusion significantly reduces pulmonary arterial pressure (PAP) and may improve survival in patients with primary and secondary PHT. We report our experience with intermittent epoprostenol (EPO) infusion in SLE patients with PHT. **Clinical Characteristics:** We reviewed patients with SLE associated PHT who were treated with intermittent EPO infusions in 1998. All 3 patients had severe PHT, with maximum systolic PAP (PASP) of 58, 96 and 67 mm Hg, respectively, when measured using Doppler echocardiography, and were in New York Heart Association functional class III. **Treatment:** All patients were given 6 infusions of EPO monthly via peripheral venous access without significant side effects. **Outcome:** The PASP of all patients remained stable during the therapy. The therapeutic response as measured by patient symptoms and PASP was variable. No patient had significant reduction of PASP during the 6-month treatment period. Two patients developed rebound elevation of PASP after cessation of treatment, while the PASP of the other patient remained stable for 10 months. **Conclusion:** Although intermittent EPO infusion did not substantially reduce PASP, it prevented further rise in PAP during the treatment period, suggesting that this regimen may benefit a subset of SLE patients with PHT.

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