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Title: Long-term follow-up in Churg-Strauss syndrome following IFN- α -induced remission

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Body: Churg-Strauss syndrome (CSS) is a small vessel systemic vasculitis associated with asthma, eosinophilia, and involvement of other organs. Interferon (IFN) represents a immunomodulatory cytokine that induces remission in CSS. Herein, we evaluated the long-term effect of IFN- α -induced remission after discontinuation of IFN- α treatment. We conducted a single-center, open-label pilot study using pegylated IFN- α (135 μ g/week s.c.) for induction of remission in p-ANCA-negative CSS patients with predominant pulmonary involvement (defined as severe corticosteroid-dependent asthma, chronic rhinosinusitis, peripheral eosinophilia). Written informed consent was obtained from all individuals. A total of 8 patients were treated with IFN- α over more than 2 yrs leading to full remission without immunosuppressive therapy. In three patients (2 females, 1 male; ages 50, 51 and 60, respectively) treatment was discontinued due to side effects (neuropathia, autoimmune hepatitis, anaemia) after 3, 4, and 10 years. At this time-point, IFN- α treatment had induced full normalisation of initially elevated (38%, 25% 23%) eosinophil counts and reduction of total IgE serum-levels (797 to 55, 377 to 233; 1170 to 133 kU/L; min/max values) in all patients. Following discontinuation of IFN- α -treatment, side effects disappeared. In addition, IgE-levels and eosinophil counts did not change and patients remained in remission without immunosuppression during follow-up for up to 4 years. **CONCLUSIONS:** Although reversible side effects occur, IFN- α is successful in inducing long-lasting remission even after discontinuation of therapy in patients with CSS.