CLINICAL AND IMMUNOLOGICAL EFFECT OF LOW-DOSE IFN-α TREATMENT IN PATIENTS WITH CORTICOSTEROID-RESISTANT ASTHMA


Purpose of the Study. To evaluate the clinical and immunologic effects of interferon (IFN)-α in patients with corticosteroid-resistant asthma with and without Churg-Strauss syndrome.

Study Population. Ten patients with severe steroid-resistant asthma, 3 of whom had Churg-Strauss syndrome, were studied.

Methods. Subjects were given 3 × 10^6 IU/day of recombinant IFN-α for at least 5 months. The prior systemic corticosteroid doses were maintained until clinical improvement was seen, and then they were decreased gradually. Spirometry, immunophenotyping of peripheral blood mononuclear cells, cytokine measurements, and lymphocyte proliferation assays were performed.

Results. IFN-α rapidly improved patient clinical status as assessed by improved lung-function parameters and decreased prednisone requirements. Immunologic changes included decreased leukocyte numbers, decreased numbers of eosinophils in patients with prior eosinophilia, increased relative numbers of CD4+ T cells, increased differentiation of T-helper (Th)1 cells, and increased interleukin 10 and IFN-γ levels in peripheral blood mononuclear cells.

Conclusions. Treatment with IFN-α in patients with steroid-resistant asthma with and without Churg-Strauss syndrome was associated with clinical improvement. Possible mechanisms of action include induction of anti-inflammatory interleukin 10 and establishment of a correct Th1/Th2 balance.

Reviewers’ Comments. Although this study involved only a few patients and additional elucidation of the underlying mechanisms is needed, these patients with steroid-resistant asthma improved with IFN-α treatment. Although this study involved only adults, the use of IFN-α as a potential steroid-sparing medication for use in children may also prove beneficial, especially given justified patient, parental, and physician concerns about using long-term oral corticosteroids in children because of the potential for significant toxicity. The use of IFN-α, however, would have to outweigh its inherent potential adverse effects including influenza-like symptoms, nausea, and liver toxicity, to name a few. This preliminary study, however, does make a case for the need for additional, longer-term clinical trials.

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Immunodeficiency

PRIMARY IMMUNODEFICIENCY

IMMUNODEFICIENCY AND INFECTIONS IN ATAXIA-TELANGIECTASIA


Purpose of the Study. To describe immunodeficiency in ataxia-telangiectasia (A-T) and its clinical manifestations and course.

Study Population. Patients with A-T who underwent multidisciplinary assessment at Johns Hopkins Hospital (Baltimore, MD).

Methods. Charts from the first 100 consecutive patients with A-T who were assessed at Johns Hopkins Ataxia-Telangiectasia Clinical Center were reviewed. Specific criteria for the diagnosis of A-T had to be met. Immunologic data were obtained by reviewing laboratory assessments of patients’ immune systems. Infections were determined by patient and family interviews and chart review.

Results. A large percentage of patients had immunoglobulin deficiencies at the time of first immunologic assessment: 65% had IgG4 deficiency, 63% had IgA deficiency, 48% had IgG2 deficiency, and 23% had IgE deficiency. Deficiencies did not correlate or progress with age. Lymphopenia occurred in 71% of patients. CD19 B lymphocytes were reduced in 75% of patients. CD4 T cells were decreased in 69% of the patients, and CD8 T cells were decreased in 51% of the patients. Patients had no untoward effects from live viral vaccines. Recurrent upper respiratory infections occurred in one third of the patients regardless of age. Lower respiratory tract infections increased with age. Viral and opportunistic infections were not common.

Conclusions. Patients with A-T have a wide array of laboratory-based immunodeiciencies. However, there seems to be no correlation between laboratory values and clinical manifestation of immunodeficiency in this population.

Reviewers’ Comments. This study confirms previously characterized immunodeficiencies in A-T patients. However, the large number of patients involved in this study allowed for a more extensive review of immunodeficiencies as well as clinical correlation of laboratory values. At this time it seems that clinical immunodeficiency is not common in A-T. Rather, the high rate of respiratory infections may be attributable to other factors of A-T such as neurologic deficits leading to aspiration.

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AUTOSOMAL RECESSIVE HYPERIMMUNOGLOBULIN E SYNDROME: A DISTINCT DISEASE ENTITY


Purpose of the Study. To describe the clinical and immunologic features of a distinct subgroup of patients with hyper-IgE syndrome (HIES) having autosomal recessive inheritance (AR-HIES) as distinct from the form having autosomal dominant inheritance (AD-HIES).
Clinical and Immunological Effect of Low-Dose IFN-α Treatment in Patients With Corticosteroid-Resistant Asthma
David Fleischer and Robert A. Wood
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