

L-carnitin: a potential treatment for hyper-IgE syndrome

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Dear Editor,

Hyper-IgE syndrome (previously Job's syndrome) is a complex disorder characterized by extreme elevation of serum IgE level, chronic dermatitis, repeated lung and skin infection, atopic dermatitis¹ and *Staphylococcal pneumoniae*.² These patients are also at increased risk of osteoporosis and spontaneous bone fracture.¹ Ulceration is also frequently present and lymphadenopathy may be complicated by lymph nodes abscess.²

Non-immunological features of the condition include coarse facies with a wide nasal bridge, large head, joint laxity and a high incidence of scoliosis.² After the newborn period, skin findings include retroauricular fissure, external otitis, infected dermatitis of axilla and groin, and folliculitis of the upper back and shoulder.¹

Peripheral blood lymphocyte subsets are generally normal and no consistent abnormality of T-cells has yet been identified.³ In a significant proportion of patients there is a failure of antibody response to polysaccharide antigens, which contributes to the susceptibility to infection.² Patients are not neutropenic and their neutrophils ingest and kill bacteria normally. However, neutrophil chemotaxis is reduced and it has been shown that mononuclear cells from hyper-IgE patients produce a factor that inhibits neutrophil chemotaxis.³

The mainstay of treatment is long term antiStaphylococcal antibiotic prophylaxis.^{1, 4} Intravenous immunoglobulin, which is an expensive treatment modality, is also employed successfully for the treatment of this illness.⁵ Severe cases may need bone marrow transplantation.¹

Cimetidine is often given to those who respond poorly to antibiotics alone, for its stimulatory effects on neutrophil chemotaxis.^{1, 6} Recently, Namazi and Handjani speculated

the potential efficacy of lithium, which enhances PMN motility, for treatment of this order.¹

L-carnitin is a natural biomolecule found in all animal tissues, which acts as an essential cofactor in fatty acid metabolism.⁷ It has been shown that L-carnitin restored lymphocytic proliferative response⁸ and the lytic activity of macrophages in aged rats.⁹ L-carnitin increases human lymphocyte proliferative response following mitogenic stimulation and increases polymorphonuclear chemotaxis.¹⁰ Furthermore, it improved decreased neutrophil chemotactic activity in aging rats.⁷

In conclusion, therefore, given the safety of L-carnitine we suggest to our colleagues to consider L-carnitin treatment while facing a patient suffering from this conundrum.

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