

Immunodeficiency Diseases- Part III (Final Part)

In the last two articles, we read how the immune system works and how it could malfunction. In this article, we will find out how the malfunctioning immune system could be corrected or supported.

Immunology is one of the fastest growing fields in medicine. Everyday new exciting discoveries are made and we are beginning to understand both the complexity and the simplicity of the immune function. We are making inroads into discovering new diseases and their treatment options. About 40 years ago, only a handful of immunodeficiency diseases were known. Now, well over 100 immunodeficiency diseases are recognized. These diseases are transmitted by genes from parents to off-springs. Recent advances in genetics and molecular biology have made it possible to diagnose and even treat these conditions even before they manifest.

Treatment of immunodeficiency states

Obviously the treatment of any disease depends on its cause. Secondary immunodeficiency states caused by infections, cancer, malnutrition, protein-losing diarrhea, and liver and kidney diseases, prolonged use of steroids, chemotherapy, or radiation treatment get better as the underlying conditions get treated and improve.

Primary immunodeficiency diseases which are inherited and do not have an external cause are treated differently. Immunoglobulin G (IgG) deficiency and specific antibody deficiency (inability to make specific IgG antibodies against disease causing bacteria) are treated by IgG replacement therapy every 2-4 weeks. The replacement is given as an infusion through a vein over 4-5 hours under medical supervision in a clinic or hospital setting. More recently, such infusions are given under the skin (subcutaneously) over an hour at weekly intervals at home by patients who are trained to do this. The intravenous infusions are associated with side-effects such as headaches, fever, chills, and rarely anaphylaxis. Premedication with Tylenol and Benadryl and slowing the rates of infusions will minimize the problem. Subcutaneous infusions given at home by patients are rarely associated with such adverse outcomes and are equally effective.

Unfortunately, for antibody deficiencies caused by lack of Immunoglobulins A (IgA) and M (IgM) proteins, no such replacement therapy is available. In fact, IgG replacement therapy containing small amounts of IgA could cause serious allergic reactions in patients who are deficient in IgA. Such patients are managed conservatively with symptomatic treatment which is discussed below. Similarly persons with defects in white blood cell function and complement deficiency are also treated symptomatically. White blood cell (WBC) transfusions for patients with low WBC count and life-threatening infections are possible but are not widely available.

For lymphocyte deficiency, bone marrow transplantation (from siblings) is possible in centers which specialize in them. Bone marrow transplantation is sometimes curative; often patients need IgG infusion therapy even after the bone marrow transplantation.

Lymphocytes stimulate other lymphocytes and signal rest of the immune system by making cytokine proteins. Cytokine injections given subcutaneously three times a week are available for patients with rare lymphocyte and white blood cell problems. Apart from causing flu-like symptoms, such treatments are well tolerated by patients.

Supportive treatment for immunodeficiency

The following general treatment guidelines apply to most immunodeficiency conditions. Immunodeficient patients should avoid sick contacts and large gatherings and wash their hands frequently. Based on studies done in Canada many years ago, white blood cell function improves very well in people who take multivitamins at recommended doses. Administration of influenza vaccine, pneumonia vaccine and regular childhood vaccines may or may not work in immunodeficient patients. Administration of live vaccines is harmful to immunodeficient patients and should be avoided. Immunodeficient patients following exposure to chicken pox or shingles should get treated with antiviral and specific immunoglobulin preparations for chickenpox. Antibiotic prophylaxis taken at weekly intervals may help prevent frequent ear or sinus infections in some immunodeficient patients. Physicians should have low threshold for treating immunodeficient patients presenting with infections with antibiotics. Early, aggressive and appropriate treatment of infections in such patients will prevent high morbidity and mortality seen in such patients following infections. Immunodeficient patients who require blood transfusions should be given specially treated blood products.

Screening for leukemia, lymphoma, lupus and other associated conditions

Patients with immunodeficiency are at increased risk for developing leukemia and lymphoma and autoimmune diseases such as lupus. Their lung function also deteriorates rapidly. Regular screening for aforementioned conditions by doing blood tests and lung function tests will lead to earlier diagnosis and better outcome.

Family support

Like any other chronic disease, better awareness through education of family members will go a long way in helping patients with immunodeficiency. Many excellent support organizations are available on the Internet. One such organization, Immune Deficiency Foundation can be reached at www.primaryimmune.org on the worldwide web.

About the author:

Natarajan Asokan, M.D., F.A.A.P. is a board certified allergist and immunologist and a board certified pediatrician with over 25 years of experience as a physician and 7 years of experience as a practicing allergist & immunologist. He treats adults and children with various allergy & immunology problems. He can be reached at 1739, Beverly Ave, Suite 118, Kingman, AZ 86409, Tel: 928-681-5800, Fax: 928-681-5801, or www.trinityallergy.com