

Immune Deficiency

Deficiencies of the immune system can occur in children and adults. Children with normal immune systems often have many infections that are an integral part of growing up. In children, allergic rhinitis and/or asthma are some of the most common diseases leading to repeated upper and lower respiratory tract infections. Adults with immune deficiencies often present with recurrent sinus and lung infections.

However, when these infections are recurrent or their severity is disproportionate to the virus or offending cause, an immunodeficiency may be suspected.

The warning signs that may indicate that a patient has an immunodeficiency include the following:

1. Eight or more new ear infections within 1 year.
2. Two or more serious sinus infections within 1 year.
3. Two or more months of antibiotics with little improvement.
4. Two or more pneumonias within 1 year.
5. Recurrent, deep skin or organ abscesses.
6. Failure of an infant to gain weight or grow normally.
7. Persistent thrush in mouth or elsewhere on skin, after age one.
8. Need for I.V. antibiotics to clear infections.
9. Two or more deep-seated infections such as meningitis, osteomyelitis, cellulitis or sepsis.
10. A family history of primary immune deficiency.

Infections that suggest an immunodeficiency

It's important to understand why some recurrent infections have characteristics that suggest they are more serious than just normal infections that are common in adults and children.

These characteristics can include the following:

Recurrence after 3-5 years of age, when most children have developed immunity to the most common childhood infections.

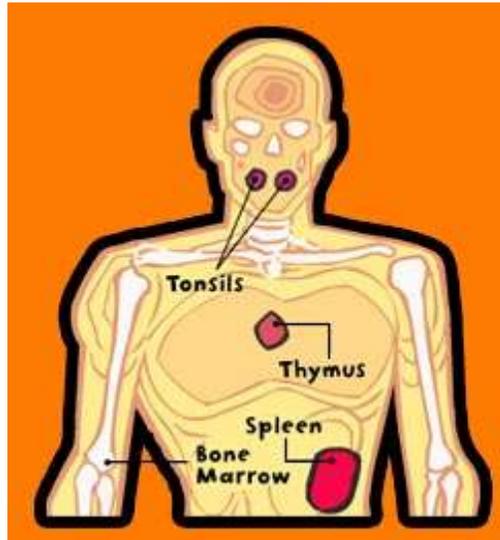
-Rapid recurrence of the infection despite completing antibiotic treatments.

-There is a lack of known predisposing factors. (Absence of sick contacts)

-The infection occurs at an unusual age of onset; unexpected site of the infection or it fails to respond to normal treatment.

Infections that are INFREQUENTLY associated with an immune defect

1. Recurrent strep infection.
2. Staph aureus and other bacterial infections in Atopic Dermatitis.
3. Arthritis and osteomyelitis except if caused by atypical mycobacteria.
4. Recurrent boils (especially due to MRSA)
5. Infectious Mononucleosis.
6. Line infections in patients with indwelling catheters.
7. Meningococcal meningitis in close contact outbreaks.
8. Pulmonary tuberculosis
9. Malaria
10. Most parasitic infections.



Primary immunodeficiency diseases occur in persons born with an immune system that lacks the ability to fight infections normally. These are caused by genetic or hereditary defects and can affect anyone, regardless of age or sex. An estimated 400 children a year are born in the U.S. with a serious primary immune deficiency. There are approximately 250,000 children and adults with primary immune deficiency.

The immune system is composed of a variety of different cell types and proteins. Each part of the system performs a specialized task aimed at recognizing and reacting to foreign material. These **foreign materials or antigens can include bacteria and viruses, and allergens.**

The major components of the immune system are:

B Lymphocytes (**B cells**)

T lymphocytes (**T cells**)

NK cells

Phagocytes (macrophages & neutophils)

Complement

When B cells are stimulated by antigens they form antibodies. When antibodies recognize a foreign antigen they set off a complex reaction that destroys the foreign antigen. The chemical name for antibody proteins is “Immunoglobulins” There are 4 major classes of antibodies. **IgG, IgA, IgM, and IgE.**

Each immunoglobulin has special characteristics and functions.

IgG is formed in large quantities and travels from the blood stream to the tissues easily. It’s the only class of Immunoglobulins, which crosses the placental and passes immunity from the mom to the newborn.

IgA antibodies are produced near mucous membranes, where they protect against infection in the respiratory tract and intestines.

IgM antibodies are the first antibodies formed in response to infection. They are important in protection during the early days of infections.

IgE antibodies are responsible for allergic reactions.

The long life of B Cells enables us to retain immunity to viruses and bacteria that infected us many years ago.

T cells Do not produce antibody molecules. They directly attack foreign antigens such as viruses, fungi and regulate the immune system. T cells develop from stem cells in the bone marrow. Early in fetal life these immature T cells move to the thymus where they become mature T cells. T cells cannot develop if the fetus does not have a thymus. T cells migrate to the spleen, lymph nodes, bone marrow and blood. T cells can react against virtually any antigen.

NK cells: Natural killer cells destroy cells infected with viruses. They are derived from the bone marrow and are extremely important in defending against viral infections. Many people believe they act to prevent cancer. They are very important in the defense against herpes viruses. This family of viruses include the traditional cold sore form of herpes, Epstein Barr virus (cause of most mono infections), and varicella virus, which causes chicken pox and shingles.

Phagocytes (neutrophils & macrophages) are specialized cells whose primary function is to ingest and kill microorganisms. It is the phagocytic cell that increases in number in the bloodstream during infection and is in large part responsible for an elevated WBC during infection.

Complement system is composed of 30 proteins most of which are produced in the liver. When the complement components bind to microorganisms, a complex is created which can puncture the microorganism and cause it to burst and die.

There are over 150 primary immunodeficiency diseases. Some are common, while others are quite rare. The primary immunodeficiencies results from defects in T cells, B cells, NK cells, phagocytic cells, or the complement system.

Deficiencies of antibody-mediated immunity are by far the most frequently reported.

Many patients are unable to produce protective levels of antibody when immunized against certain vaccines, such as the pneumococcal vaccine.

Treatment

One of the primary goals is to have patients be as symptom-free as possible so that they may perform activities of daily living such as attending school and work.

The goal of therapy is also to prevent damage to the ears, sinuses and the lungs that might result in hearing loss or chronic lung disease, chronic sinusitis, or injury to other organ systems.

Prophylactic antibiotics may be recommended during the viral season in patients that are especially susceptible to ear, sinus and lung infections.

Individuals who are unable to produce adequate amounts of immunoglobulins may benefit from replacement therapy with Immunoglobulins. It replaces what the body should be making but doesn't help the immune system make more. Approximately 1/2 of the infused antibodies are metabolized over 3-4 weeks so repeat doses are required on a regular basis. All immunoglobulin products licensed in the U.S. are made from plasma collected in America. It's carefully screened for transmissible diseases.

Gammaglobulin replacement is not new. It was first used for treatment of primary immune deficiency in 1952.