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Disease & Treatment

About Your Disease

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Inherited Immune System Disorders

Inherited immune system disorders are diseases in which children are born without an immune system or with an incomplete immune system. People with an immune system disorder are less able to fight infections. While these disorders are rare, there are about 100 different types, which range from mild to severe. About 50,000 people in the United States have some type of inherited immune system disorder.

Inherited immune system disorders are also called primary immune deficiency disorders. They are called "primary" because they are caused by a gene defect that affects the immune system. Genes carry an inherited code of instructions that tells the body how to make every cell and protein in the body. (Secondary immune deficiencies are caused by something outside the body, such as chemotherapy or infection with a virus.)

Immune system disorders and transplant

People with severe inherited immune system disorders are at high risk for lifethreatening infections. For many severe disorders, the only known cure is a bone marrow or cord blood transplant (also called a BMT). To learn more, read about two of the most severe inherited immune system disorders:

- ▶ Severe Combined Immunodeficiency (SCID) and Transplant
- ▶ Wiskott-Aldrich Syndrome (WAS) and Transplant

Transplant has also been used to treat some other inherited immune system disorders, including:

- ▶ Combined immunodeficiency associated with other syndromes: Severe DiGeorge Syndrome, X-linked immunodeficiency with normal or elevated IgM, X-linked lymphoproliferative disease, IPEX (Immune dysregulation, Polyendocrinopathy, Enteritis, and X-linked) syndrome
- ▶ Humoral immunodeficiency: Common variable immunodeficiency
- ▶ Other deficiencies: Chronic granulomatous disease, leukocyte adhesion

To find out whether transplant is a treatment option for you or your family member, talk to a doctor who is an expert in treating these disorders.

More information on inherited immune system disorders

You can get more information about inherited immune system disorders from disease-specific organizations, such as:

- ▶ National Primary Immunodeficiency http://www.info4pi.org/aboutPI/index.cfm? section=aboutPI&CFID=12977514&CFTOKEN=62075299
- ▶ Immune Deficiency Foundation http://www.primaryimmune.org/about_pi/about_pi.htm
- National Institute of Child Health & Human Development: Primary Immunodeficiency http://www.nichd.nih.gov/publications/pubs/primary_immuno.cfm

For other organizations that offer information and resources, see Organizations That Can Help: A Searchable Directory.

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