

SECTION 16

IMMUNE DISORDERS

183 **Biology of the Immune System**1049
Nonspecific Immunity ▪ Specific Immunity ▪ Effects of Aging

184 **Immunodeficiency Disorders**.....1057
X-Linked Agammaglobulinemia ▪ Selective Antibody Deficiency ▪
Common Variable Immunodeficiency ▪ Transient
Hypogammaglobulinemia of Infancy ▪ Chronic Mucocutaneous
Candidiasis ▪ DiGeorge Anomaly ▪ Ataxia-Telangiectasia ▪
Severe Combined Immunodeficiency Disease ▪ Wiskott-Aldrich
Syndrome ▪ Hyperimmunoglobulinemia E Syndrome ▪ Chronic
Granulomatous Disease ▪ Immunodeficiency due to Spleen
Disorders

185 **Allergic Reactions**.....1063
Seasonal Allergies ▪ Year-Round Allergies ▪ Food Allergy ▪
Mastocytosis ▪ Physical Allergy ▪ Exercise-Induced Allergic
Reactions ▪ Hives and Angioedema ▪ Anaphylactic Reactions

186 **Autoimmune Disorders**1073

187 **Transplantation**.....1075
Principles of Organ Transplantation ▪ Kidney Transplantation ▪
Liver Transplantation ▪ Heart Transplantation ▪ Lung and Heart-
Lung Transplantation ▪ Pancreas Transplantation ▪ Stem Cell
Transplantation ▪ Transplantation of Other Organs

CHAPTER 183

Biology of the Immune System

The immune system is designed to defend the body against foreign or dangerous substances that invade it. Such substances include microorganisms (commonly called germs, such as bacteria, viruses, and fungi), parasites (such as worms), cancer cells, and even transplanted organs and tissues.▲ Substances that stimulate an immune response in the body are called antigens. Antigens may be

contained within or on bacteria, viruses, other microorganisms, or cancer cells. Antigens may also exist on their own—for example, as pollen or food molecules. A normal immune response consists of recognizing a foreign antigen, mobilizing forces to defend against it, and attacking it.

▲ see page 1075

Disorders of the immune system occur

- when the body generates an immune response against itself (an autoimmune disorder)▲
- when the body cannot generate appropriate immune responses against invading microorganisms (an immunodeficiency disorder)■
- when a normal immune response to foreign antigens damages normal tissues (an allergic reaction).★

The first line of defense against invaders is mechanical or physical barriers: the skin, the cornea of the eye, and the membranes lining the respiratory, digestive, urinary, and reproductive tracts. As long as these barriers remain unbroken, many invaders cannot penetrate them. If a barrier is broken—for example, if extensive burns damage much of the skin—the risk of infection is increased. In addition, the barriers are defended by secretions containing enzymes that can destroy bacteria. Examples are tears in the eyes and secretions in the digestive tract and vagina.

The next line of defense involves white blood cells that travel through the bloodstream and into tissues, searching for and attacking microorganisms and other invaders. This defense has two parts. The first part, called nonspecific (innate) immunity, involves several types of white blood cells that usually act on their own to destroy invaders. The second part, called specific (adaptive) immunity, involves white blood cells that work together to destroy invaders. Some of these cells do not directly destroy invaders but enable other white blood cells to recognize and destroy invaders.

Nonspecific immunity and specific immunity interact, influencing each other directly or through substances that attract or activate other cells of the immune system—part of the mobilization step in defense. These substances include cytokines (which are the messengers of the immune system), antibodies, and complement proteins (which form the complement system). These substances are not contained in cells but are dissolved in a body fluid, such as plasma, the liquid part of blood.

To be able to destroy invaders, the immune system must first recognize them. That is, the

immune system must be able to distinguish what is nonself (foreign) from what is self. The immune system can make this distinction because all cells have identification molecules on their surface. Microorganisms are recognized because they have unique, foreign identification molecules on their surface. In people, identification molecules are called human leukocyte antigens (HLA), or the major histocompatibility complex (MHC). HLA molecules are called antigens because they can provoke an immune response in another person (normally, they do not provoke an immune response in the person who has them). Each person has unique human leukocyte antigens. A cell with molecules on its surface that are not identical to those on the body's own cells is identified as being foreign. The immune system then attacks that cell. Such a cell may be a microorganism, a cell from transplanted tissue, or one of the body's cells that has been infected by an invading microorganism.

Some white blood cells—B lymphocytes—recognize invaders directly. But others—T lymphocytes—need help from other cells of the immune system—called antigen-presenting cells. These cells ingest an invader and break it into fragments. Antigen fragments from the invader are then “presented” in a way that T lymphocytes can recognize.

The immune system includes several organs in addition to cells dispersed throughout the body. These organs are classified as primary or secondary lymphoid organs. The primary lymphoid organs—the thymus gland and bone marrow—are the sites where white blood cells are produced. In the thymus gland, T lymphocytes—a type of white blood cell—are produced and trained to recognize foreign antigens and ignore the body's own antigens. (T lymphocytes are critical for specific immunity.) The bone marrow produces several types of white blood cells, including neutrophils, monocytes, and B lymphocytes. When needed to defend the body, the white blood cells are mobilized, mainly from the bone marrow. They then move into the bloodstream and travel to wherever they are needed.

The secondary lymphoid organs include the spleen, lymph nodes, tonsils, liver, appendix, and Peyer's patches in the small intestine. These organs trap microorganisms and other foreign substances and provide a place for mature cells of the immune system to collect, interact with each other and with the foreign

▲ see page 1073 ■ see page 1057
★ see page 1063

T lymphocyte surveillance bloodstream for foreign antigens. However, an antigen is “presented” to a white blood cell. Antigen-presenting cells include dendritic cells and macrophages.

1. By its size an antigen is recognized.
2. A cell dendritic cell



substances in response.

The lymphatic system is the body's network of lymphatic vessels. Lymph nodes are small, bean-shaped structures that filter out dead or dying cells and fight infection. Lymph nodes can evaluate cancer cells and cause inflammation.