The immune system is vital for survival, because it protects us from infectious pathogens that abound in the environment. Predictably, immune deficiencies render individuals easy prey to infections. But the immune system is itself capable of causing tissue injury and disease. Examples of disorders caused by immune responses include *allergies* and reactions against an individual's own tissues and cells *(autoimmunity)*.

This chapter is devoted to diseases caused by too little immunity or too much immunologic reactivity. We also consider amyloidosis, a disease in which an abnormal protein, derived in some cases from fragments of immunoglobulins, is deposited in tissues. First, we review some of the important features of normal immune responses, to provide a foundation for understanding the abnormalities that give rise to immunologic diseases.

The Normal Immune Response

The classic definition of *immunity* is protection from infectious pathogens, and the normal immune response is best understood in this context. The mechanisms of defense against microbes fall into two broad categories (Fig. 6-1). *Innate immunity* (also called natural, or native, immunity) refers to the mechanisms that are ready to react to infections even before they occur, and that have evolved to specifically recognize and combat microbes. *Adaptive immunity* (also called acquired, or specific, immunity) consists of mechanisms that are stimulated by ("adapt to") microbes and are capable of recognizing microbial and nonmicrobial substances. Innate immunity is the first line of defense. It is mediated by cells and molecules that recognize products of microbes and dead cells and induce rapid protective host reactions. Adaptive immunity develops later, after exposure to microbes and other foreign substances, and is even more powerful than innate immunity in combating infections. By convention, the term *immune response* usually refers to adaptive immunity.

Innate Immunity

Innate immunity is always present, ready to provide defense against microbes and to eliminate damaged cells. The receptors and components of innate immunity have evolved to serve these purposes. Innate immunity functions in stages: recognition of microbes and damaged cells, activation of various mechanisms, and elimination of the unwanted substances.

Components of Innate Immunity

The major components of innate immunity are epithelial barriers that block entry of microbes, phagocytic cells (mainly neutrophils and macrophages), dendritic cells, natural killer (NK) cells, and several plasma proteins, including the proteins of the complement system.

- *Epithelia* of the skin and gastrointestinal and respiratory tracts provide mechanical barriers to the entry of microbes from the external environment. Epithelial cells also produce antimicrobial molecules such as defensins, and lymphocytes located in the epithelia combat microbes at these sites. If microbes do breach epithelial boundaries, other defense mechanisms are called in.

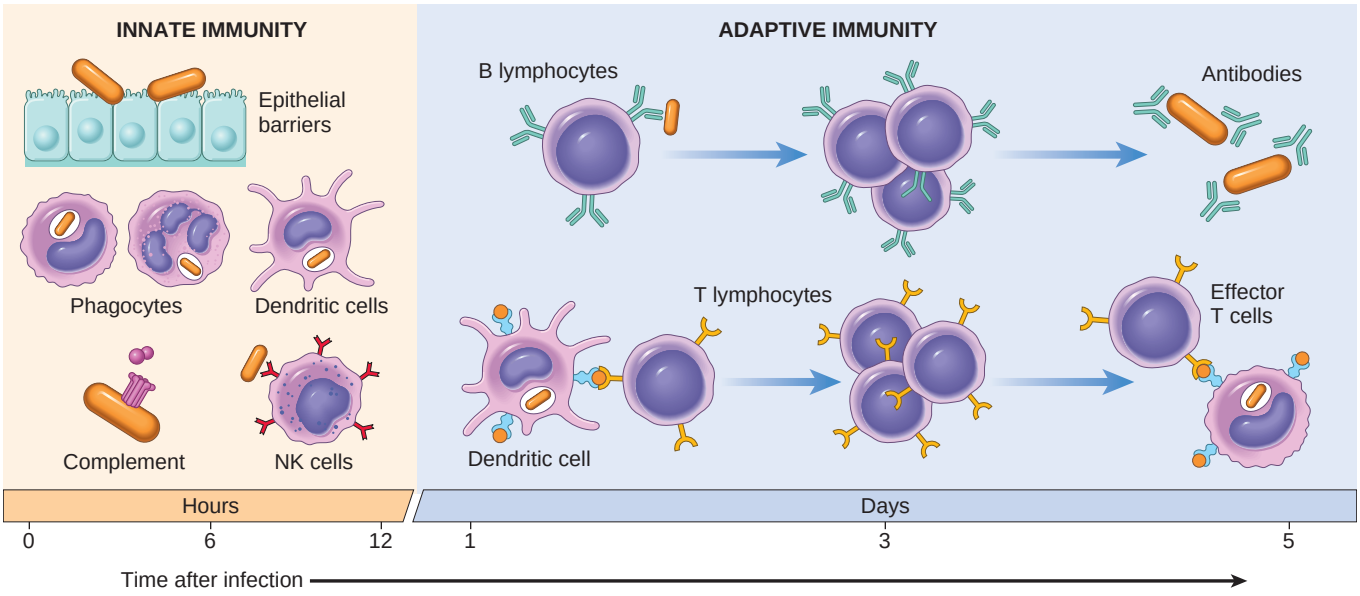

Figure 6-1 The principal mechanisms of innate immunity and adaptive immunity. NK cells, Natural killer cells.