

Topic Page: [Autoimmune diseases](#)

Definition: **autoimmune diseases** from *The Penguin Dictionary of Science*

Disorders of the ►immune system that result in the inability to differentiate between ‘self’ and ‘non-self’ antigens, leading to the production of an auto-antibody (an ►antibody against oneself). Such diseases include Hashimoto's disease of the thyroid, systemic lupus erythematosus, rheumatoid arthritis and Crohn's disease.

Summary Article: **Autoimmune Diseases (General)**

From *Encyclopedia of Global Health*

Autoimmune disease (AD) is a condition where the human body's immune system reacts against its own tissues. This process occurs in rheumatoid arthritis (RA), type 1 diabetes, systemic lupus erythematosus (SLE), Crohn's disease, and many other debilitating ailments. Worldwide, autoimmune diseases account for much suffering and morbidity. Many estimates indicate that between 20 to 50 percent of the world's population suffer from some variant of AD. There are too many autoimmune diseases to list here, so a few will be used for illustration.

There is a worldwide distribution of autoimmune conditions and many of the causes and treatments are still unknown. No region, country, race, or gender is invulnerable to AD and the burden of disease falls on all social and economic classes. Although some relationships have been drawn between viral infections, gender, geographic locations, and even diet or occupational exposures, much remains to be seen and understood in the fight against ADs and what must be done to treat them. AD is especially a burden in the developing world as diagnosis and proper treatment often elude those who need them most.

The human body's immune system is able to distinguish self tissues from foreign invading organisms. This is called the self-versus-nonsel self defense, or immune response. The immune system tissues and cells throughout the body's systems carry markers that enable cells of the immune system to identify these self cells. Cells encountered without markers initiate an immune response and are attacked and eliminated from the human body. This is the process that occurs when an invading organism enters the body. The initiation of this response is called an antigen. Antibodies, such as B cells that will invade and collaborate to kill the attacking bacteria, fungus, or virus, are then formed to continue the response.

AD occurs when the body is unable to properly distinguish self from nonself and initiates a response or an attack that eliminates its own healthy tissues. This attack can be on one organ or on a whole system of organs; for example, with SLE, antibodies are created against the patient's own DNA.

Lymph tissue found in lymph nodes throughout the body, such as the thymus, spleen, and bone marrow, are main centers for immune cells to grow and where signs or symptoms of immune system crisis can usually first be observed. Cells associated with the lymphatic and immune systems travel throughout the body in blood and lymphatic vessels.

Cells that make up the immune system must communicate with one another in order to be able to distinguish which cells are invading and which are not. T cells, B cells, and phagocytes are some major constituents in the immune system. These cells suppress activation of the immune system and maintain immune system homeostasis. T cells are one type of white blood cell that attacks virus-infected cells, foreign cells, and cancer cells. T cells also produce a number of substances that regulate

the immune response. Some T cells can seek and destroy invading foreign cells, while some are suppressor T cells. Suppressor T cells slow down and stop the immune response of B cells and other T cells. These suppressor cells effectively “shut off” the antibody production when an infection is under control. When T cells fail or function improperly, AD can result and cells of the immune system attack healthy cells in the body.

B cells are the other major class of lymphocytes, or white blood cells, involved in the immune response. B cells are white blood cells of the immune system that are derived from the bone marrow and spleen. B cells develop into plasma cells, which produce antibodies. When a B cell interacts with an antigen, it differentiates into antibody-secreting plasma cells and memory cells. It is the primary lymphocyte responsible for immunity, and is effective against pathogens such as bacteria, viruses, or fungi.

B cells secrete antibodies and are involved with memory of antigens. They have a long cell life, so that in the event the same antigen invades, the immune system can identify and initiate another response more quickly than the first time it was invaded. Phagocytes are large cells that “eat” or engulf other foreign invading pathogens. When the immune system becomes compromised, or is altered and can no longer function, immune cells can become blind and begin to attack self cells. These are the basic mechanisms of the immune response and what can go wrong in the presence of AD.

EPIDEMIOLOGY

In the United States, prevalence of autoimmune disorders has been estimated as ranging from at least 10 million to as many as 50 million people. This difference reflects the current lack of consensus about which diseases meet the definition of AD. Classification of diseases as AD or not is still under review in much of the world.

In the early 1980s so much of HIV/AIDS was misunderstood that some scientists classified it as an autoimmune disease. Because more of the mechanisms of HIV/AIDS became known after intense study, scientists realized it is not an AD at all. Other such mis-classifications may remain in practice until more is understood about the many ADs that occur throughout the world. Standardization of this classification will bring more clarity to disease epidemiology.

Furthermore, correct diagnosis is difficult even in developed nations and may happen years after medical care is first sought for an ailment. It is thought that there are many undiagnosed cases in developing nations, and this likely contributes to an inaccurate estimation of prevalence.

One consensus in the medical community is that AD disproportionately affects women. Some estimates are that 75 percent of all cases first appear in women between 15 and 44 years of age. Because of this, the burden of disease in the developing world is significant. Women bear more responsibility for child rearing, daily-life activities, and are accountable for many other liabilities in the family unit. Furthermore, AD can occur during childbearing years, leading to further problems with their children and community. Some ADs affect African-American, American-Indian, and Latina women more often than white women in the United States. Sex-specific hormones may further amplify this hyperimmune response in women leading to the increased prevalence. Much is misunderstood and much more research needs to be done to find more causes of AD and reasons for the disproportionate number of female sufferers. ADs, thus, contribute disproportionately to morbidity and mortality among young to middle-aged women.

SLE, thyroiditis, scleroderma, RA, Crohn’s disease and type 1 diabetes affect both women and men.

SLE is a chronic inflammatory connective tissue disease. It is marked by skin rashes; joint pain and swelling; inflammation of the kidneys; inflammation of the fibrous tissue surrounding the heart, or pericardium; and causes the skin to lose its elasticity and can lead to total hair loss. Thyroiditis is an inflammation of the thyroid gland that causes it to become underactive and results in fatigue, weakness, weight gain, cold intolerance, and muscle aches.

Sclerosis is a hardening within the nervous system, especially of the brain and spinal cord, resulting in degeneration of the nervous system and leading to eventual death. RA is a disease that attacks joints. It causes hot, painful swelling and deformity mainly to the joints of the fingers, hands, and knees.

Crohn's disease is an ongoing disorder that causes inflammation of the digestive tract. Crohn's disease is an inflammatory bowel disease, the general name for diseases that cause swelling in the intestines. Crohn's disease affects men and women equally and seems to run in some families, leading it to most likely be a genetically inherited AD.

Type 1 diabetes, also known as insulin-dependent diabetes, is diagnosed mainly in childhood or early adolescence. This AD requires daily insulin injections for the body to metabolize sugar. The most common form is caused by the destruction of beta cells in the pancreas by an AD process. Beta cells are those found in the pancreas and are responsible for making insulin, which controls the amount of sugar in the blood by moving it into the cells in the body for energy. When the immune system attacks these cells, the pancreas is unable to produce insulin and the body is unable to process sugar from foods to obtain energy.

Treatment for type 1 diabetes in the developed world is readily available and prognosis when the treatment regimen is kept can be quite positive. Daily insulin injections, strict blood glucose monitoring, diet plan, daily exercise, and good patient and doctor follow-up are needed for good outcomes with diabetes type 1. These treatments are not readily available in the developing world.

Specific incidence and prevalence rates vary among the ADs. For example, in the United States an estimated 5 percent of the population are affected by tissue-specific or systemic ADs. Most ADs can occur at any age of development and ethnic and geographic differences in incidence of specific ADs have been documented. However, specific groups may be at higher risk for some diseases and at lower risk for others, as illustrated by geographic region and women in general.

For example, genetic predisposition and hormones play a part in ADs. The incidence of multiple sclerosis, Crohn's disease, and type 1 diabetes are highest in northern Europe but nearly disappear the closer to the equator the population lives. There are some common traits among many ADs, but there are also important demographic differences between them. Disease-specific research and studies that focus on other potentially related diseases are still underway.

GENETICS AND POSSIBLE CAUSES

The cause of autoimmune disorders remains largely unknown. Recent evidence supports a role of environmental agents such as chemicals, lifestyles, and family history. The important question of whether genetics plays a large role is still under review, but most research indicates that it does. For the case of thyroid disease, a recent population-based study of risks stresses the increasing importance of environmental contributions to the development of autoimmunity.

For the case of RA, however, it has not yet been concluded that genetics does or does not contribute to the disease. Infection as a possible trigger for ADs has long been proposed as well. Viral infections

have received particular attention in SLE studies, with findings of virus-like inclusions in kidney biopsy tissue. These connections with infections and AD are still under review and are not conclusive.

Evidence in patients that susceptibility genes exists for autoimmunity comes from family studies. These studies are especially important with twins. Studies in type 1 diabetes mellitus, RA, multiple sclerosis, and SLE have shown that approximately more than a fifth of the pairs of monozygotic (one fertilized ovum separates into two identical zygotes or fetuses) twins show disease concordance, compared with less than 5 percent of dizygotic (two separate eggs by two separate sperm; nonidentical pair twins) twins. These data suggest there may be a genetic connection with AD.

OCCUPATIONAL AND PSYCHOSOCIAL RISKS

Cigarette smoke and passive or secondhand smoke are some of the worst occupational and daily-living hazards to affect world populations in the past 100 years. Contribution to cancer and many lung disorders is becoming more understood with more research. Cigarette smoke is a known carcinogen, or cancer-causing agent, and negative health effector. Furthermore, cigarette smoke has been associated with an increased risk of developing RA and possibly thyroiditis.

Also, some case-control studies from Japan, the United Kingdom, and Sweden detected a statistically significant increase in the risk of developing SLE among current smokers and identified cigarette smoking as a risk factor leading to the development of lupus nephritis. Lupus nephritis is an inflammation of the kidney caused by SLE.

Another major occupational exposure possibly causing AD is exposure to crystalline silica. Crystalline silica, or silicon dioxide, is an eye, skin, and lung irritant. It is a known carcinogen and is most dangerous in the dry form. Crystalline silica is used in some highly popular brands of cleansers, glass and ceramic manufacturing, cat litter, paints, and some powdered flea-control products for pets. Crystalline silica has been identified as a strong risk factor for developing AD, especially SLE. Occupational silica exposure is most frequently associated with dusty, nonfarming-related jobs. In areas where chemical regulation and poor compliance persist, many workers may be heavily exposed to crystalline silica.

Some research suggests that emotional stress from major life events may contribute to the onset of AD, and specifically RA. The cause may be different hormone levels or another unknown cause. New case-control studies provide several new insights into the contribution of psychosocial stress to the onset of AD and RA. For example, one study showed that in women, arguments with their partners during a five-year period preceding the RA symptoms and the five years preceding the RA diagnosis were significantly associated with the onset of RA compared with controls. The same study found men who reported problems at work preceding the onset of symptoms and economic problems within five years of diagnosis of RA. These data may not be entirely conclusive, but they offer insight into further areas of research on the causes of AD.

This connection between social environmental factors and medical status is not new. Much research in the past has made the connection between the onset of ADs such as type 1 diabetes. Psychological mechanisms are directly linked by hormonal and nervous system signals, influencing the need for insulin. Stress has also been shown to modulate immune responses in a number of studies.

TREATMENT

At present, there is no known way to prevent ADs. However, with proper medication and careful monitoring, many people are able to live fairly normal lives. There are four general immunologic

approaches to AD treatment: altering thresholds of immune activation, modulating antigen-specific responses, reconstituting the immune system with autologous (one's own) or allogeneic (genetically different) stem cells, and sparing of target organs. Each treatment is different for each AD.

Treatment of autoimmune diseases can focus on either suppressing the induction of autoimmunity, restoring normal regulatory mechanisms, or inhibiting the effector mechanisms. To eliminate autoreactive cells, immunosuppressive or ablative therapies are most commonly used. In recent years, cytokine blockade has been demonstrated to be effective in preventing immune activation in some diseases.

New therapies are currently in clinical trials to target lymphoid cells more specifically, either by blocking a signal needed for T or B cell activation, by eliminating the effector T cells or B cells, or by using autoantigen to induce tolerance. Therapies that prevent target organ damage or support target organ function remain an important therapeutic approach to autoimmune disease. These therapies are still in early development.

There is currently considerable interest in new drugs that have been associated with the development of an autoimmune response with drug-related SLE. Two recent epidemics of scleroderma-like illness, one in Spain related to a contaminant of rapeseed oil and one in the United States related to a contaminant of L-tryptophan, have raised interest in potential environmental triggers of autoimmunity.

Nonsurgical treatment for the rheumatic ADs includes healthy diet, prompt treatment of infections, immunosuppressants, corticosteroids, and antiinflammatory medications. Current treatment protocols for the more severe outcomes of ADs include organ transplants and surgery to repair or replace damaged joints. It is predicted that genetic research will enable earlier and more precise diagnoses of ADs and highly individualized drug treatment.

As more information is published on both risk factors for the onset of autoimmunity and for the development of progressive disease, scientists are learning more about what makes AD start and function. Many potential exposures have not been thoroughly or consistently studied for their impact on both disease onset and disease progression. More research needs to be done and treatment must reach all patients in developed and less developed countries. AD will continue to be a major problem until it is better understood and more treatments become available.

SEE ALSO:

Acquired Immunity; Active Immunity; Childhood Immunization; Diabetes Type 1 (Juvenile Diabetes); Immune System and Disorders; Immunosuppression; Lupus; Rheumatoid Arthritis; Scleroderma; Sjogren's Syndrome.

BIBLIOGRAPHY

- American Autoimmune Related Diseases Association, "Questions and Answers," www.aarda.org (cited January 2007).
- M. A. Dooley; L. Hogan, "Epidemiology and Risk Factors for Autoimmune Disease," *Current Opinion in Rheumatology* (v.18, 2003).
- R. P. Kimberly, "Research Advances in Systemic Lupus Erythematosus," *Journal of the American Medical Association* (v.285, 2001).
- W. J. Koopman, "Research Advances in Rheumatoid Arthritis," *Journal of the American Medical Association* (v.285, 2001).

- Robert Lahita; Nicholas Chiorazzi, *Textbook of the Autoimmune Diseases* (Lippincott Williams & Wilkins, 2000).
- National Institutes of Health, "Press Release" (July 7, 1999).
- Noel Richard Rose; Ian R. Mackay, eds., *The Autoimmune Diseases*, 4th ed. (Academic Press, 2006).

John Michael Quinn V
University of Illinois at Chicago

APA

Chicago

Harvard

MLA

V, J. M. (2008). Autoimmune diseases (general). In Y. Zhang, *Encyclopedia of global health*. Thousand Oaks, CA: Sage Publications. Retrieved from https://search.credoreference.com/content/topic/autoimmune_disease

 Copyright © 2008 by SAGE Publications, Inc.

 Copyright © 2008 by SAGE Publications, Inc.

APA

V, J. M. (2008). Autoimmune diseases (general). In Y. Zhang, *Encyclopedia of global health*. Thousand Oaks, CA: Sage Publications. Retrieved from https://search.credoreference.com/content/topic/autoimmune_disease

Chicago

V, John Michael Quinn. "Autoimmune Diseases (General)." In *Encyclopedia of Global Health*, by Yawei Zhang. Sage Publications, 2008. https://search.credoreference.com/content/topic/autoimmune_disease

Harvard

V, J.M. (2008). Autoimmune diseases (general). In Y. Zhang, *Encyclopedia of global health*. [Online]. Thousand Oaks: Sage Publications. Available from: https://search.credoreference.com/content/topic/autoimmune_disease [Accessed 18 November 2019].

MLA

V, John Michael Quinn. "Autoimmune Diseases (General)." *Encyclopedia of Global Health*, Yawei Zhang, Sage Publications, 1st edition, 2008. *Credo Reference*, https://search.credoreference.com/content/topic/autoimmune_disease. Accessed 18 Nov. 2019.