

ort a time as possible. When
h for more than 3 to 4 weeks,
ny, sometimes serious side ef-

f of the people with chronic
s disappear without treatment
For some adults, the antide-
n, which is also a potent anti-
s relieve chronic hives.
ioedema results in difficulty
eathing or in collapse, prompt
ment is necessary. Affected
ways carry a self-injecting sy-
ine and antihistamine tablets
ediately if a reaction occurs.
llegic reaction, such people
: hospital emergency depart-
y can be checked and treated

Lactic Reactions

tions (anaphylaxis) are sud-
potentially severe and life-
ic reactions.

actions are most commonly
(such as penicillin), insect
ods, and allergy injections
therapy). But they can be
rgen. Like other allergic re-
ylactic reaction does not
the first exposure to an al-
ur after a subsequent expo-
ny people do not recall a
y allergen that causes an
on in a person is likely to
n with subsequent expo-
res are taken to prevent it.

tions begin within 1 to 15
to the allergen. Rarely, re-
1 hour. The heart beats
may feel uneasy and affect-
d pressure may fall, caus-
ymptoms include tingling
sensations, itchy and
ng in the ears, coughing,
l swelling (angioedema).
e difficult and wheezing
the windpipe (upper air-
omes swollen.
action may progress so
to collapse, cessation of
ad loss of consciousness
The reaction may be fa-

tal unless emergency treatment is given im-
mediately.

Prevention and Treatment

People who are allergic to unavoidable aller-
gens (such as insect stings) may benefit from
long-term allergen immunotherapy.▲

If an anaphylactic reaction occurs, an
epinephrine injection should be given imme-

diately. People who have these reactions
should always carry a self-injecting syringe of
epinephrine and antihistamine tablets for
prompt treatment. Usually, this treatment
stops the reaction. Nonetheless, after a severe
allergic reaction, such people should go to the
hospital emergency department, where they
can be closely monitored and treatment can be
adjusted as needed.

Merck Manual,
2nd Home Edⁿ, 2003.

CHAPTER 186

Autoimmune Disorders

*An autoimmune disorder is a malfunction of
the body's immune system, causing the body
to attack its own tissues.*

Normally, the immune system can distin-
guish what is self from what is not self (or for-
eign)■ and reacts against foreign substances
called antigens. Antigens may be contained
within or on bacteria, viruses, other microor-
ganisms, or cancer cells. Or antigens may ex-
ist on their own—for example, as pollen or
food molecules. Sometimes the immune sys-
tem malfunctions, interpreting the body's
own tissues as foreign and producing abnor-
mal antibodies (called autoantibodies) or im-
mune cells that target and attack particular
cells or tissues of the body. This response is
called an autoimmune reaction. It results in
inflammation and tissue damage. Different
cells or tissues are targeted in different au-
toimmune disorders.

Causes

Autoimmune reactions can be triggered in
several ways:

- A substance in the body that is normally
confined to a specific area (and thus is hidden
from the immune system) is released into the
bloodstream. For example, a blow to the eye
can cause the fluid in the eyeball to be re-
leased into the bloodstream. The fluid stimu-
lates the immune system to attack.

- A normal body substance is altered, for ex-
ample, by a virus, a drug, sunlight, or radi-
ation. The altered substance may appear for-
eign to the immune system. For example, a
virus can infect and thus alter cells in the

body. The virus-infected cells stimulate the
immune system to attack.

- A foreign substance that resembles a natu-
ral body substance may enter the body. The
immune system may inadvertently target the
similar body substance as well as the foreign
substance.

- The cells that control antibody produc-
tion—for example, B lymphocytes (a type of
white blood cell)—may malfunction and pro-
duce abnormal antibodies that attack some of
the body's cells.

Heredity may be involved in some autoim-
mune disorders. Susceptibility, rather than the
disorder itself, may be inherited. In suscepti-
ble people, a trigger, such as a viral infection
or tissue damage, may cause the disorder to
develop. Hormonal factors may also be in-
volved, because many autoimmune disorders
are more common among women.

Symptoms and Diagnosis

Autoimmune disorders commonly cause a
fever. However, symptoms vary depending on
the disorder and the part of the body affected.
Some autoimmune disorders affect certain
types of tissue throughout the body—for ex-
ample, blood vessels, cartilage, or skin. Other
autoimmune disorders affect a particular or-
gan. Virtually any organ, including the kid-
neys, lungs, heart, and brain, can be affected.
The resulting inflammation and tissue dam-
age can cause pain, deformed joints, weakness,

▲ see page 1064

■ see page 1050

SOME AUTOIMMUNE DISORDERS

DISORDER	MAIN TISSUES AFFECTED	CONSEQUENCES
Autoimmune hemolytic anemia	Red blood cells	Anemia with fatigue, weakness, and light-headedness develops, and the spleen enlarges. The anemia can be severe and even fatal.
Bullous pemphigoid	Skin	Large blisters, surrounded by red, swollen areas, form on the skin. Itching is common. With treatment, the prognosis is good.
Graves' disease	Thyroid gland	The thyroid gland is inflamed, stimulated, and enlarged, resulting in high levels of thyroid hormones (hyperthyroidism). With treatment, the prognosis is good.
Hashimoto's thyroiditis	Thyroid gland	The thyroid gland is inflamed and damaged, resulting in low levels of thyroid hormones (hypothyroidism). Lifelong treatment with thyroid hormone is necessary.
Type 1 diabetes	Beta cells of pancreas (which produce insulin)	The beta cells are destroyed, so the body lacks insulin. Treatment with insulin is needed lifelong even if the reaction terminates because the cells in the pancreas have been destroyed.
Lupus (systemic lupus erythematosus)	Joints, kidneys, skin, lungs, heart, and brain	The affected tissues are inflamed and often damaged, but the joints, although inflamed, do not become deformed. The prognosis varies widely, but most people can lead an active life despite occasional flare-ups of the disorder.
Myasthenia gravis	The connection between nerves and muscles (neuromuscular junction)	Muscles, particularly those of the eyes, weaken and tire easily, but the weakness varies in intensity. The pattern of progression varies widely, but drugs can usually control the symptoms. Rarely, the disorder is fatal.
Pemphigus	Skin	Large blisters form on the skin. The disorder can be life threatening.
Pernicious anemia	Cells in the stomach's lining and red and white blood cells	Because the stomach's lining is damaged, it is less able to absorb vitamin B ₁₂ (which is necessary for the production of mature blood cells). Anemia results, and nerves are damaged. Without treatment, the spinal cord may be damaged. The risk of stomach cancer is increased. Otherwise, with treatment, the prognosis is good.

jaundice, itching, difficulty breathing, accumulation of fluid (edema), delirium, and even death.

Blood tests may detect an autoimmune disorder. For example, the erythrocyte sedimentation rate (ESR) is often increased, because proteins, produced in response to inflammation, interfere with the ability of red blood cells (erythrocytes) to remain suspended in blood. Typically, the number of red blood cells is decreased, resulting in anemia. Blood tests

can also detect different antibodies, some of which typically occur in people who have an autoimmune disorder. Examples of these antibodies are antinuclear antibodies (which attack the nuclei of cells) and rheumatoid factor.

Treatment

Treatment involves control of the autoimmune reaction by suppressing the immune system. However, many of the drugs used to control the autoimmune reaction interfere

with the body's ability to fight off especially infections. Treatments may also be needed.

Drugs that suppress the immune system (immunosuppressants) include cyclophosphamide, chlorambucil, cyclosporine, or methotrexate, given by mouth and often intravenously. However, these drugs suppress the immune reaction but do not prevent the body from defending itself against infections, including microorganisms and cancer cells. Corticosteroids are used to control inflammation and of certain

Often, corticosteroids are given, usually by mouth, to relieve inflammation and suppress the immune system. Corticosteroids have many side effects, and corticosteroids are usually given when the disorder is severe.

Transplantation is the replacement of tissues, or organs from one part of the body with tissues or organs from another part of the body.

The most common type of transplantation is a blood transfusion. Millions of people receive blood transfusions, and tissues can also be transplanted.

Tissues or organs from a living donor can be a living donor. If a living donor has recently died, tissues or organs from a living donor are preferred. Transplantation is more like a living donor are preferred.

Stem cells (from bone marrow) and kidneys are the tissues that are most commonly transplanted from a living donor. Usually, the donor is a living donor because the donor can function well. Donors can also donate organs from a living donor. An organ from a living donor can be transplanted within minutes.

with the body's ability to fight disease, especially infections. Treatment to relieve symptoms may also be needed.

Drugs that suppress the immune system (immunosuppressants), such as azathioprine, chlorambucil, cyclophosphamide, cyclosporine, or methotrexate, are often given, usually by mouth and often for a long time.▲ However, these drugs suppress not only the autoimmune reaction but also the body's ability to defend itself against foreign substances, including microorganisms that cause infection and cancer cells. Consequently, the risk of infection and of certain cancers increases.

Often, corticosteroids, such as prednisone, are given, usually by mouth. These drugs relieve inflammation as well as suppress the immune system. Corticosteroids given for a long time have many side effects.■ When possible, corticosteroids are used for a short time—when the disorder begins or when symptoms

worsen. However, corticosteroids must sometimes be used indefinitely.

Etanercept and infliximab block the action of tumor necrosis factor (TNF), a substance that can cause inflammation in the body. These drugs are very effective in treating rheumatoid arthritis and inflammatory bowel disease, but they may be harmful if used to treat certain other autoimmune disorders, such as multiple sclerosis.

Plasmapheresis is used to treat a few autoimmune disorders. Blood is withdrawn and filtered to remove the abnormal antibodies. Then the filtered blood is returned to the person.

Some autoimmune disorders resolve as inexplicably as they began. However, most autoimmune disorders are chronic. Drugs are often required throughout life to control symptoms. The prognosis varies depending on the disorder.

CHAPTER 187

Transplantation

Transplantation is the transfer of living cells, tissues, or organs from one person to another or from one part of the body to another.

The most common type of transplantation is a blood transfusion,★ which is used to treat millions of people each year. Some organs or tissues can also be transplanted.

Tissues or organs come from a donor. A donor can be a living person or a person who has recently died. Tissues and organs from a living donor are preferable because transplantation is more likely to be successful. However, some organs, such as the heart, obviously cannot be taken from a living donor.

Stem cells (from bone marrow or blood) and kidneys are the tissues most often donated by a living donor. Usually, a kidney can be safely donated because the body has two kidneys and can function well with only one. Living donors can also donate a part of the liver or a lung. An organ from a living donor is transplanted within minutes of being removed.

After a person dies, organs deteriorate quickly. Consequently, organs from a donor who has died usually come from a person who was expected to die and who had previously agreed to donate organs. Permission for donation may be given by the person's closest family member. Often, such donors are otherwise healthy people who have been in a major accident, rather than those who die of a disorder. Sometimes one donor can provide several people with transplants. For example, one donor could provide two people with corneas, two with kidneys, one with a liver, two with lungs, and another with a heart. Some organs last only a few hours outside the body. Other organs, if kept cold, last up to several days.

In the United States, a national organization (United Network for Organ Sharing) matches

▲ see table on page 1078 ■ see box on page 374

★ see page 982