

Autoimmune Hepatitis

What is autoimmune hepatitis?

Autoimmune hepatitis is a disease in which the body's immune system attacks liver cells. This immune response causes inflammation of the liver, also called hepatitis. Researchers think a genetic factor may make some people more susceptible to autoimmune diseases. About 70 percent of those with autoimmune hepatitis are female.

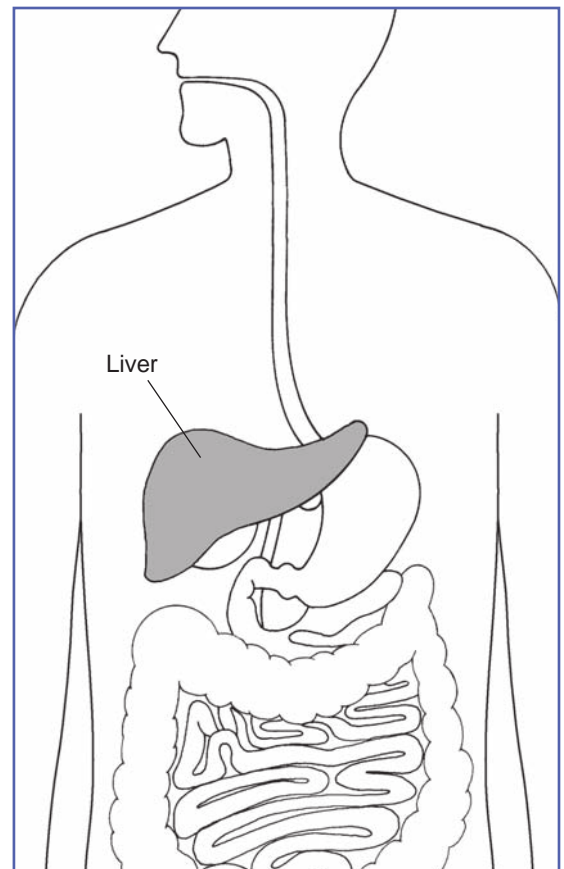
The disease is usually quite serious and, if not treated, gets worse over time. Autoimmune hepatitis is typically chronic, meaning it can last for years, and can lead to cirrhosis—scarring and hardening—of the liver. Eventually, liver failure can result.

Autoimmune hepatitis is classified as type 1 or type 2. Type 1 is the most common form in North America. It can occur at any age but most often starts in adolescence or young adulthood. About half of those with type 1 have other autoimmune disorders, such as

- type 1 diabetes
- proliferative glomerulonephritis, an inflammation of blood vessels in the kidneys
- thyroiditis, an inflammation of the thyroid gland
- Graves' disease, the leading cause of overactive thyroid
- Sjögren's syndrome, a syndrome that causes dry eyes and mouth

- autoimmune anemia
- ulcerative colitis, an inflammation of the colon and rectum leading to ulcers

Type 2 autoimmune hepatitis is less common, typically affecting girls aged 2 to 14, although adults can have it too.



Autoimmune hepatitis affects the liver.

What is autoimmune disease?

One job of the immune system is to protect the body from viruses, bacteria, and other living organisms. The immune system usually does not react against the body's own cells. However, sometimes it attacks the cells it is supposed to protect; this response is called autoimmunity. Researchers think certain bacteria, viruses, toxins, and drugs trigger an autoimmune response in people who are genetically susceptible to developing an autoimmune disorder.

What are the symptoms of autoimmune hepatitis?

Fatigue is probably the most common symptom of autoimmune hepatitis. Other symptoms include

- an enlarged liver
- jaundice
- itching
- skin rashes
- joint pain
- abdominal discomfort
- spider angiomas, or abnormal blood vessels, on the skin
- nausea
- vomiting
- loss of appetite
- dark urine
- pale or gray-colored stools

People in advanced stages of the disease are more likely to have symptoms related to chronic liver disease, such as fluid in the abdomen—also called ascites—and mental confusion. Women may stop having menstrual periods.

Symptoms of autoimmune hepatitis range from mild to severe. Because severe viral hepatitis or hepatitis caused by a drug—for example, certain antibiotics—have the same symptoms as autoimmune hepatitis, tests may be needed for an exact diagnosis. Doctors should also review and rule out all medicines a patient is taking before diagnosing autoimmune hepatitis.

How is autoimmune hepatitis diagnosed?

The doctor will make a diagnosis based on symptoms, blood tests, and a liver biopsy.

- **Blood tests.** A routine blood test for liver enzymes can help reveal a pattern typical of hepatitis, but further tests, especially for autoantibodies, are needed to diagnose autoimmune hepatitis. Antibodies are proteins made by the immune system to fight off bacteria and viruses. Autoantibodies attack the body's cells. In autoimmune hepatitis, the immune system makes one or more types of autoantibodies. The most common are antinuclear antibodies (ANA), smooth muscle antibodies (SMA), and antibodies to liver and kidney microsomes (anti-LKM). People with type 1 have ANA, SMA, or both, and people with type 2 have anti-LKM.

Blood tests also help distinguish autoimmune hepatitis from other diseases that resemble it, such as viral hepatitis B or C or a metabolic disease such as Wilson disease.

- **Liver biopsy.** A tiny sample of liver tissue, examined with a microscope, can help doctors accurately diagnose autoimmune hepatitis and tell how serious it is. This procedure is done in a hospital or outpatient surgical facility.

How is autoimmune hepatitis treated?

Treatment works best when autoimmune hepatitis is diagnosed early. With proper treatment, autoimmune hepatitis can usually be controlled. In fact, studies show that sustained response to treatment stops the disease from getting worse and may reverse some of the damage.

The primary treatment is medicine to suppress, or slow down, an overactive immune system.

Both types of autoimmune hepatitis are treated with daily doses of a corticosteroid called prednisone. Treatment may begin with a high dose of 30 to 60 mg per day and be lowered to 10 to 20 mg per day as the disease is controlled. The goal is to find the lowest possible dose that will control the disease.

Another medicine, azathioprine (Imuran) is also used to treat autoimmune hepatitis. Like prednisone, azathioprine suppresses the immune system, but in a different way. Treatment may begin with both azathioprine and prednisone, or azathioprine may be added later, once the disease is under control. The use of azathioprine allows for a lower dose of prednisone, which in turn reduces prednisone's side effects.

In about seven out of 10 people, the disease goes into remission within 3 years of starting treatment. Remission occurs when symptoms disappear and lab tests show improvement in liver function. Some people can eventually stop treatment, although many will see the disease return. People who stop treatment must carefully monitor their condition and promptly report any new symptoms to their doctor. Treatment with low doses of prednisone or azathioprine may be necessary on and off for years, if not for life.

Some people with mild forms of the disease may not need to take medication. Doctors assess each patient individually to determine whether those with mild autoimmune hepatitis should undergo treatment.

What are the side effects of prednisone and azathioprine?

Both prednisone and azathioprine have side effects. Because high doses of prednisone are often needed to control autoimmune hepatitis, managing side effects is very important. However, most side effects appear only after a long period of time.

Some possible side effects of prednisone are

- weight gain
- anxiety and confusion
- thinning of the bones, a condition called osteoporosis
- thinning of the hair and skin
- diabetes
- high blood pressure
- cataracts
- glaucoma

Azathioprine can lower white blood cell counts and sometimes causes nausea and poor appetite. Rare side effects are allergic reaction, liver damage, and pancreatitis, which is an inflammation of the pancreas gland with severe stomach pain.

Are other treatments for autoimmune hepatitis available?

People who do not respond to standard immune therapy or who have severe side effects may benefit from other immunosuppressive agents such as mycophenylate mofetil, cyclosporine, or tacrolimus. People who progress to end-stage liver disease—also called liver failure—or cirrhosis may need a liver transplant. Transplantation has a 1-year survival rate of 90 percent and a 5-year survival rate of 70 to 80 percent.

Points to Remember

- Autoimmune hepatitis is a long-term disease in which the body's immune system attacks liver cells.
- The disease is diagnosed using various blood tests and a liver biopsy.
- With proper treatment, autoimmune hepatitis can usually be controlled. The main treatment is medicine that suppresses the body's overactive immune system.

Hope through Research

Scientists are studying various aspects of autoimmune hepatitis to find out who gets it and why and to discover better ways to treat it. Basic research on the immune system will expand knowledge of autoimmune diseases in general. Epidemiologic research will help doctors understand what triggers autoimmune hepatitis in some people. Research on different steroids, alternatives to steroids, and other immunosuppressants will eventually lead to more effective treatments.