

Autoimmune Hepatitis

National Digestive Diseases Information Clearinghouse



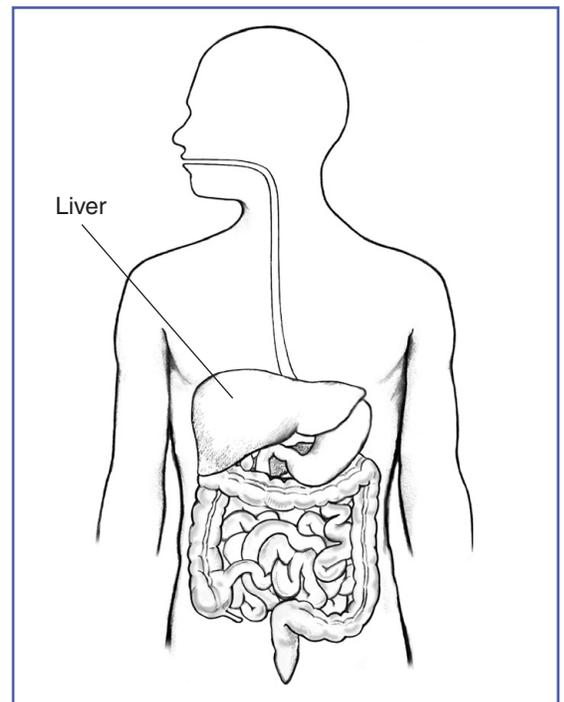
What is autoimmune hepatitis?

Autoimmune hepatitis is a chronic—or long lasting—disease in which the body’s immune system attacks the normal components, or cells, of the liver and causes inflammation and liver damage. The immune system normally protects people from infection by identifying and destroying bacteria, viruses, and other potentially harmful foreign substances.

Autoimmune hepatitis is a serious condition that may worsen over time if not treated. Autoimmune hepatitis can lead to cirrhosis and liver failure. Cirrhosis occurs when scar tissue replaces healthy liver tissue and blocks the normal flow of blood through the liver. Liver failure occurs when the liver stops working properly.

What are autoimmune diseases?

Autoimmune diseases are disorders in which the body’s immune system attacks the body’s own cells and organs with proteins called autoantibodies; this process is called autoimmunity.



Autoimmune hepatitis is a chronic disease of the liver.

The body’s immune system normally makes large numbers of proteins called antibodies to help the body fight off infections. In some cases, however, the body makes autoantibodies. Certain environmental triggers can lead to autoimmunity. Environmental triggers are things originating outside the body, such as bacteria, viruses, toxins, and medications.

What causes autoimmune hepatitis?

A combination of autoimmunity, environmental triggers, and a genetic predisposition can lead to autoimmune hepatitis.

Who is more likely to develop autoimmune hepatitis?

Autoimmune hepatitis is more common in females.¹ The disease can occur at any age and affects all ethnic groups.

What are the types of autoimmune hepatitis?

Autoimmune hepatitis is classified into several types. Type 1 autoimmune hepatitis is the most common form in North America. Type 1 can occur at any age; however, it most often starts in adolescence or young adulthood. About 70 percent of people with type 1 autoimmune hepatitis are female.¹

People with type 1 autoimmune hepatitis commonly have other autoimmune disorders, such as

- celiac disease, an autoimmune disease in which people cannot tolerate gluten because it damages the lining of their small intestine and prevents absorption of nutrients
- Crohn's disease, which causes inflammation and irritation of any part of the digestive tract

- Graves' disease, the most common cause of hyperthyroidism in the United States
- Hashimoto's disease, also called chronic lymphocytic thyroiditis or autoimmune thyroiditis, a form of chronic inflammation of the thyroid gland
- proliferative glomerulonephritis, or inflammation of the glomeruli, which are tiny clusters of looping blood vessels in the kidneys
- primary sclerosing cholangitis, which causes irritation, scarring, and narrowing of the bile ducts inside and outside the liver
- rheumatoid arthritis, which causes pain, swelling, stiffness, and loss of function in the joints
- Sjögren's syndrome, which causes dryness in the mouth and eyes
- systemic lupus erythematosus, which causes kidney inflammation called lupus nephritis
- type 1 diabetes, a condition characterized by high blood glucose, also called blood sugar, levels caused by a total lack of insulin
- ulcerative colitis, a chronic disease that causes inflammation and sores, called ulcers, in the inner lining of the large intestine

Type 2 autoimmune hepatitis is less common and occurs more often in children than adults.¹ People with type 2 can also have any of the above autoimmune disorders.

¹Manns MP, Czaja AJ, Gorham JD, et al. Diagnosis and management of autoimmune hepatitis. *Hepatology*. 2010;51(6):1–31.

What are the symptoms of autoimmune hepatitis?

The most common symptoms of autoimmune hepatitis are

- fatigue
- joint pain
- nausea
- loss of appetite
- pain or discomfort over the liver
- skin rashes
- dark yellow urine
- light-colored stools
- jaundice, or yellowing of the skin and whites of the eyes

Symptoms of autoimmune hepatitis range from mild to severe. Some people may feel as if they have a mild case of the flu. Others may have no symptoms when a health care provider diagnoses the disease; however, they can develop symptoms later.

How is autoimmune hepatitis diagnosed?

A health care provider will make a diagnosis of autoimmune hepatitis based on symptoms, a physical exam, blood tests, and a liver biopsy.

A health care provider performs a physical exam and reviews the person's health history, including the use of alcohol and medications that can harm the liver. A person usually

needs blood tests for an exact diagnosis because a person with autoimmune hepatitis can have the same symptoms as those of other liver diseases or metabolic disorders.

Blood tests. A blood test involves drawing blood at a health care provider's office or a commercial facility and sending the sample to a lab for analysis. A person will need blood tests for autoantibodies to help distinguish autoimmune hepatitis from other liver diseases that have similar symptoms, such as viral hepatitis, primary biliary cirrhosis, steatohepatitis, or Wilson disease.

Liver biopsy. A liver biopsy is a procedure that involves taking a piece of liver tissue for examination with a microscope for signs of damage or disease. The health care provider may ask the patient to temporarily stop taking certain medications before the liver biopsy. He or she may also ask the patient to fast for 8 hours before the procedure.

During the procedure, the patient lies on a table, right hand resting above the head. A health care provider will apply a local anesthetic to the area where he or she will insert the biopsy needle. If needed, he or she will give sedatives and pain medication. Then, he or she will use a needle to take a small piece of liver tissue, and may use ultrasound, computerized tomography scans, or other imaging techniques to guide the needle. After the biopsy, the patient must lie on the right side for up to 2 hours and is monitored an additional 2 to 4 hours before being sent home.

A health care provider performs a liver biopsy at a hospital or an outpatient center. The liver sample is sent to a pathology lab where the pathologist—a doctor who specializes in diagnosing disease—looks at the tissue with a microscope and sends a report to the patient’s health care provider.

A health care provider can use liver biopsy to diagnose autoimmune hepatitis and determine if cirrhosis is present. People often have cirrhosis at the time they are diagnosed with autoimmune hepatitis. A health care provider can also use liver biopsy to look for changes in the severity of liver damage prior to ending treatment for autoimmune hepatitis.

How is autoimmune hepatitis treated?

Treatment for autoimmune hepatitis includes medication to suppress, or slow down, an overactive immune system. Treatment may also include a liver transplant.

Treatment works best when autoimmune hepatitis is diagnosed early. People with autoimmune hepatitis generally respond to standard treatment and the disease can be controlled in most cases. Long-term response to treatment can stop the disease from getting worse and may even reverse some damage to the liver.

Medications

People with autoimmune hepatitis who have no symptoms or a mild form of the disease may or may not need to take medication. A health care provider will determine if a person needs treatment. In some people with mild autoimmune hepatitis, the disease may go into remission. Remission is a period when a person is symptom-free and blood tests and liver biopsy show improvement in liver function.

Corticosteroids. Corticosteroids are medications that decrease swelling and reduce the activity of the immune system. Health care providers treat both types of autoimmune hepatitis with a daily dose of a corticosteroid called prednisone. Treatment may begin with a high dose that is gradually lowered as the disease is controlled. The treatment goal is to find the lowest possible dose that helps control the disease.

Side effects of prednisone may include

- weight gain
- weakness of the bones, called osteoporosis or osteomalacia
- thinning of the hair and skin
- acne
- diabetes
- high blood pressure
- cataracts, a clouding in the lens of the eyes
- glaucoma, elevated pressure in the eyes
- anxiety and confusion

A health care provider will closely monitor and manage any side effects that may occur, as high doses of prednisone are often prescribed to treat autoimmune hepatitis.

Immune system suppressors. Medications that suppress the immune system prevent the body from making autoantibodies and block the immune reaction that contributes to inflammation. In most cases, health care providers use azathioprine (Azasan, Imuran) in conjunction with prednisone to treat autoimmune hepatitis. When using azathioprine, a health care provider can use a lower dose of prednisone, which may reduce prednisone's side effects.

Side effects of azathioprine include

- low white blood cell count
- nausea
- vomiting
- skin rash
- liver damage
- pancreatitis, or inflammation of the pancreas

Azathioprine is an immune system suppressor, so people taking the medication should undergo routine blood tests to monitor their white blood cell counts. A low white blood cell count can lead to bone marrow failure. Bone marrow is the tissue

found inside bones that produces new blood cells, including platelets. A health care provider will also check the platelet count when blood tests are done.

A person may need to discontinue prednisone or azathioprine if they cause severe side effects. The risk of side effects is higher in people who also have cirrhosis.

A health care provider may gradually reduce the dose of medication in people who show improvement, although the symptoms can return. When a person discontinues treatment, a health care provider will perform routine blood tests and carefully monitor the person's condition for a return of symptoms. Treatment with low doses of prednisone or azathioprine may be necessary on and off for many years.

People who do not respond to standard immune therapy or who have severe side effects from the medications may benefit from other immunosuppressive agents such as mycophenolate mofetil (CellCept), cyclosporine, or tacrolimus (Hecoria, Prograf).

Medications that suppress the immune system may lead to various forms of cancer. People on low doses of azathioprine for long periods of time are at slight risk of developing cancer.

Liver Transplant

In some people, autoimmune hepatitis progresses to cirrhosis and end-stage liver failure, and a liver transplant may be necessary. Symptoms of cirrhosis and liver failure include the symptoms of autoimmune hepatitis and

- generalized itching
- a longer-than-usual amount of time for bleeding to stop
- easy bruising
- a swollen stomach or swollen ankles
- spiderlike blood vessels, called spider angiomas, that develop on the skin
- abdominal bloating due to an enlarged liver
- fluid in the abdomen—also called ascites
- forgetfulness or confusion

Liver transplant is surgery to remove a diseased or an injured liver and replace it with a healthy one from another person, called a donor. A team of surgeons performs a liver transplant in a hospital. When possible, the patient fasts for 8 hours before the surgery. The patient stays in the hospital about 1 to 2 weeks to be sure the transplanted liver is functioning properly. The health care provider will monitor the patient for bleeding, infections, and signs of liver rejection. The patient will

take prescription medications long term to prevent infections and rejection. Liver transplant surgery for autoimmune hepatitis is successful in most cases.

Read more in *Liver Transplantation* at www.digestive.niddk.nih.gov.

What is a possible complication of autoimmune hepatitis and cirrhosis?

People with autoimmune hepatitis and cirrhosis are at risk of developing liver cancer. A health care provider will monitor the person with a regular ultrasound examination of the liver. Ultrasound uses a device, called a transducer, that bounces safe, painless sound waves off organs to create an image of their structure. A specially trained technician performs the procedure in a health care provider's office, an outpatient center, or a hospital, and a radiologist—a doctor who specializes in medical imaging—interprets the images; anesthesia is not needed. The images can show the liver's size and the presence of cancerous tumors.

Eating, Diet, and Nutrition

Researchers have not found that eating, diet, and nutrition play a role in causing or preventing autoimmune hepatitis.

Points to Remember

- Autoimmune hepatitis is a chronic—or long lasting—disease in which the body’s immune system attacks the liver and causes inflammation and damage.
- Autoimmune hepatitis is a serious condition that may worsen over time if not treated. Autoimmune hepatitis can lead to cirrhosis and liver failure.
- Autoimmune hepatitis is more common in females. The disease can occur at any age and affects all ethnic groups.
- Autoimmune hepatitis is classified as type 1 or type 2.
- A health care provider will make a diagnosis of autoimmune hepatitis based on symptoms, a physical exam, blood tests, and a liver biopsy.
- A person usually needs blood tests for an exact diagnosis because a person with autoimmune hepatitis can have the same symptoms as those of other liver diseases or metabolic disorders.
- Treatment for autoimmune hepatitis includes medication to suppress, or slow down, an overactive immune system.
- Treatment works best when autoimmune hepatitis is diagnosed early.
- People with autoimmune hepatitis generally respond to standard treatment and the disease can be controlled in most cases.
- In some people, autoimmune hepatitis progresses to cirrhosis and end-stage liver failure, and a liver transplant may be necessary.

Hope through Research

The National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) and other components of the National Institutes of Health (NIH) conduct and support basic and clinical research into many digestive disorders, including autoimmune hepatitis.

Clinical trials are research studies involving people. Clinical trials look at safe and effective new ways to prevent, detect, or treat disease. Researchers also use clinical trials to look at other aspects of care, such as improving the quality of life for people with chronic illnesses. To learn more about clinical trials, why they matter, and how to participate, visit the NIH Clinical Research Trials and You website at www.nih.gov/health/clinicaltrials. For information about current studies, visit www.ClinicalTrials.gov.

For More Information

American Association for the Study of Liver Diseases

1001 North Fairfax Street, Suite 400
Alexandria, VA 22314
Phone: 703-299-9766
Fax: 703-299-9622
Email: aasld@aasld.org
Internet: www.aasld.org

American Liver Foundation

39 Broadway, Suite 2700
New York, NY 10006
Phone: 1-800-GO-LIVER
(1-800-465-4837) or 212-668-1000
Fax: 212-483-8179
Internet: www.liverfoundation.org

Acknowledgments

Publications produced by the Clearinghouse are carefully reviewed by both NIDDK scientists and outside experts. This publication was originally reviewed by Willis Maddrey, M.D., University of Texas Southwestern Medical Center at Dallas, and Edward Krawitt, M.D., University of Vermont College of Medicine.

You may also find additional information about this topic by visiting MedlinePlus at www.medlineplus.gov.

This publication may contain information about medications and, when taken as prescribed, the conditions they treat. When prepared, this publication included the most current information available. For updates or for questions about any medications, contact the U.S. Food and Drug Administration toll-free at 1-888-INFO-FDA (1-888-463-6332) or visit www.fda.gov. Consult your health care provider for more information.

The U.S. Government does not endorse or favor any specific commercial product or company. Trade, proprietary, or company names appearing in this document are used only because they are considered necessary in the context of the information provided. If a product is not mentioned, the omission does not mean or imply that the product is unsatisfactory.

National Digestive Diseases Information Clearinghouse

2 Information Way
Bethesda, MD 20892-3570
Phone: 1-800-891-5389
TTY: 1-866-569-1162
Fax: 703-738-4929
Email: nddic@info.niddk.nih.gov
Internet: www.digestive.niddk.nih.gov

The National Digestive Diseases Information Clearinghouse (NDDIC) is a service of the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). The NIDDK is part of the National Institutes of Health of the U.S. Department of Health and Human Services. Established in 1980, the Clearinghouse provides information about digestive diseases to people with digestive disorders and to their families, health care professionals, and the public. The NDDIC answers inquiries, develops and distributes publications, and works closely with professional and patient organizations and Government agencies to coordinate resources about digestive diseases.

This publication is not copyrighted. The Clearinghouse encourages users of this publication to duplicate and distribute as many copies as desired.

This publication is available at www.digestive.niddk.nih.gov.



National Institute of
Diabetes and Digestive
and Kidney Diseases

NIH Publication No. 14-4761
January 2014