

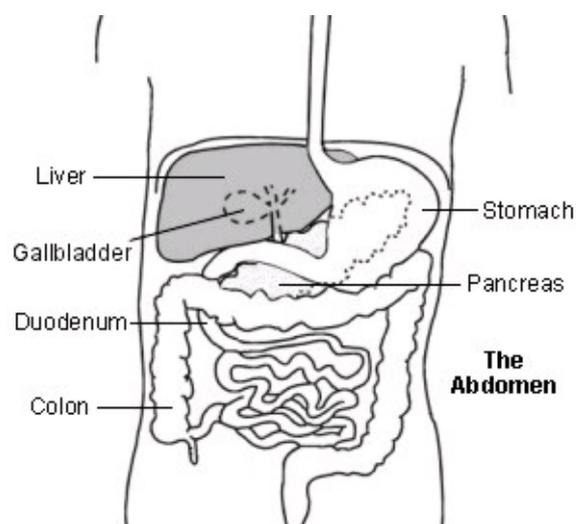
Autoimmune Hepatitis

Autoimmune hepatitis is an uncommon cause of chronic hepatitis (persistent liver inflammation). The cause is not known. If left untreated, the inflammation causes cirrhosis (scarring of the liver). However, with treatment, the outlook for people with this condition is very good. Treatment is usually with steroids and other medicines which suppress inflammation.

What does the liver do?

The liver is in the upper right part of the abdomen. It has many functions which include:

- Storing glycogen (fuel for the body) which is made from sugars. When required, glycogen is broken down into glucose which is released into the bloodstream.
- Helping to process fats and proteins from digested food.
- Making proteins that are essential for blood to clot (clotting factors).
- Processing many medicines which you may take.
- Helping to remove or process alcohol, poisons and toxins from the body.
- Making bile which passes from the liver to the gut down the bile duct. Bile breaks down the fats in food so that they can be absorbed from the bowel.



What is autoimmune hepatitis?

Hepatitis means inflammation of the liver. There are many causes of hepatitis. For example, alcohol excess and infections with various viruses are the common causes of hepatitis.

Autoimmune hepatitis is an uncommon cause of chronic hepatitis. Chronic means that the inflammation is persistent or long-term. The chronic inflammation gradually damages the liver cells, which can result in serious problems.

What causes autoimmune hepatitis?

The cause is not clear. It is thought to be an autoimmune disease. Our immune system normally defends us against infection from bacteria, viruses and other germs. In people with autoimmune diseases the immune system attacks part or parts of the body as if it were foreign. This causes inflammation and may damage the affected part or parts of the body.

The immune system includes white blood cells and antibodies (special proteins) which attack and destroy foreign germs and materials. There are various different autoimmune diseases. For example, rheumatoid arthritis, myasthenia gravis, some forms of thyroid disease, and autoimmune hepatitis. The symptoms of each disease depend on which part or parts of the body are attacked by the immune system. In some autoimmune diseases it is antibodies which do the damage. In autoimmune hepatitis, it is white blood cells called lymphocytes which are mainly involved. The lymphocytes attack the liver cells (hepatocytes) which causes inflammation and damage.

It is not known why autoimmune hepatitis or other autoimmune diseases occur. Some factor may trigger the immune system to attack the body's own tissues. There are various theories as to possible triggers. For example, a virus, a chemical or some other factor may trigger the disease. There is also an inherited factor which makes some people more prone to autoimmune diseases. (That is, you may need to be genetically susceptible for a trigger to set off the disease.)

Who gets autoimmune hepatitis?

Autoimmune hepatitis is uncommon. It affects around 1 in 10,000 people in the UK. About 7 in 10 cases are in women. It most commonly develops in women aged around 45 years but it can affect people at all ages (including children and the elderly).

What are the symptoms of autoimmune hepatitis?

In many cases the symptoms develop gradually over weeks or months. At the beginning of the disease, many people have no symptoms at all. The most common early symptoms include feeling more tired than usual, feeling generally unwell and also joint and muscle pains which are usually worse in the mornings. It is common to feel nauseated (feel sick).

Jaundice can develop when the liver inflammation becomes worse. Jaundice is when you go yellow. You tend to notice it first when the whites of the eyes become yellow. This is due to a build-up of the chemical bilirubin which is made in the liver and spills into the blood in some liver conditions. (With jaundice due to hepatitis your urine goes dark, your faeces (stools) may go pale and and you tend to itch.)

Without treatment, in time the persistent inflammation causes liver damage and can lead to cirrhosis. Cirrhosis is like a scarring of the liver, which can cause serious problems and liver failure when it is severe. See separate leaflet called '*Cirrhosis*' for more detail.

In some cases, the symptoms develop quickly over a few days with an acute hepatitis. (Acute means sudden in onset or short duration.) This can cause a fairly sudden onset of fever, abdominal pain, jaundice, feeling sick, vomiting and feeling unwell. In some of these cases, the acute hepatitis settles down to the more common type of chronic hepatitis. In some cases, the acute hepatitis becomes severe and rapidly leads to liver failure.

If left untreated, the time it takes from the onset of the condition until severe cirrhosis develops varies from person to person. However, with treatment, the inflammation can usually be controlled and the outlook is good.

How is autoimmune hepatitis diagnosed?

Many people with autoimmune hepatitis have no symptoms, or only vague symptoms, for quite some time in the early stages of the disease. Therefore, the diagnosis is often made when you have tests for an unrelated condition. When symptoms suggest that you have hepatitis, your doctor will normally arrange tests to confirm hepatitis and to find the cause of the hepatitis. Tests usually include:

- Blood tests called liver function tests. These measure the activity of enzymes (chemicals) and other substances made in the liver. This gives a general guide as to whether the liver is inflamed, and how well it is working. See separate leaflet called '*Blood Test - Liver Function Tests*' for more detail. These tests can confirm that you have hepatitis but not usually the cause of the hepatitis.
- An ultrasound scan of the liver may be done.

- Other blood tests can measure various auto-antibodies which occur in various autoimmune diseases. For example, antinuclear antibodies (ANAs) or smooth muscle antibodies (SMAs). Some of these auto-antibodies are raised in autoimmune hepatitis and indicate that this may be the cause of the hepatitis.
- A biopsy (small sample) of the liver is likely to be taken to look at under the microscope. This can show inflammation and the extent of any cirrhosis in the liver. See separate leaflet called '*Biopsy - Liver*' for more detail. The type of cells involved in the inflammation usually help to confirm the diagnosis of autoimmune hepatitis, and to rule out other causes of hepatitis.

Various tests may also be done to rule out other liver disorders such as viral hepatitis.

What is the treatment for autoimmune hepatitis?

Treatment is almost always needed. Early treatment can improve symptoms, reduce the risk of complications, and also greatly improve your outlook. Treatment aims to reduce inflammation and suppress the immune system with immunosuppressant medicines:

- Steroid medication (usually prednisolone) is the usual first treatment. Steroids are good at reducing inflammation. A high dose is usually needed at first. The dose is then gradually reduced over a few weeks. The aim is to find the lowest dose needed to control the inflammation. The dose needed varies from person to person. See separate leaflet called '*Steroid Tablets*' for more detail.
- Azathioprine is an immunosuppressant medicine that works in a different way to steroids. It is usually used in addition to the steroid. A steroid plus azathioprine tends to work better than either alone. Also, the dose of steroid needed is usually less if you also take azathioprine. This means that any side-effects from steroids may be less severe.

Treatment works well in most cases. Usually, the inflammation settles and symptoms improve within a few months of starting treatment. However, it may take a year or more to get the disease totally under control. Azathioprine is usually taken for at least three years.

Side-effects from the treatment may be troublesome in some cases. The dose of treatment is kept as low as possible to keep the condition under control with the minimum of side-effects. If you cannot tolerate the azathioprine and/or steroids then alternative treatment may be given. This may include other medicines called ciclosporin or tacrolimus.

Side-effects from the steroids can occur. These can include osteoporosis (thinning of the bones), weight gain and glaucoma. Your doctor is likely to advise that you take vitamin D and calcium supplements while you are taking steroid medication. These work by strengthening the bones and lowering your risk of developing osteoporosis (thinning of the bones) in the future. It is likely that you will be advised to have a scan called a dual-energy X-ray absorptiometry (DEXA) scan before starting steroids and this is then repeated at regular intervals. This is done to assess the density of your bones. See separate leaflet called '*Preventing Steroid-induced Osteoporosis*'.

Once the condition has completely settled, your doctor may advise a trial without treatment. A liver biopsy is usually done at the end of the course of treatment, to ensure that the liver inflammation has fully resolved. However, only around one in five people can remain off treatment for long periods of time. This is because the hepatitis usually returns (relapses), which means you will have to then take medication again. In many cases, long-term maintenance treatment is needed. This means that the majority of people with autoimmune hepatitis will need to take low doses of immunosuppressant treatment for long periods of time, or even indefinitely.

For the few people who do not respond to treatment with the medicines mentioned above, or who are diagnosed in the late stage of the disease with severe cirrhosis or liver failure, a liver transplant may be an option. The long-term outlook after liver transplantation is good.

It is recommended that you are vaccinated against hepatitis A and B. You should also receive the annual influenza vaccination.

Diet and alcohol

Most people with autoimmune hepatitis will be advised to eat a normal healthy diet. Ideally, anybody with inflammation of the liver should not drink alcohol, or only in very small amounts. If you already have liver inflammation, alcohol may increase the risk and speed of developing cirrhosis.

What is the outlook?

With treatment, most people with autoimmune hepatitis have a normal life expectancy and feel well most of the time. The treatment used for autoimmune hepatitis has improved the outlook tremendously. It is very important that you do not stop your treatment too early without your doctor's knowledge, as your hepatitis may return.

There is a very small increased risk of developing liver cancer, especially if you also have cirrhosis due to your autoimmune hepatitis. Some doctors recommend a blood test and an ultrasound scan of your liver every so often to screen for this.

The treatment of autoimmune hepatitis is a developing area of medicine. New treatments continue to be developed and researched and the information above is very general. The specialist who knows your case can give more accurate information about the outlook for your particular situation.

Further help & information

British Liver Trust

2 Southampton Road, Ringwood, BH24 1HY

Tel: (Information Line) 0800 652 7330, (General) 01425 481320

Web: www.britishlivertrust.org.uk

Children's Liver Disease Foundation

36 Great Charles Street, Birmingham, B3 3JY

Tel: 0121 212 3839

Web: www.childliverdisease.org

Further reading & references

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