

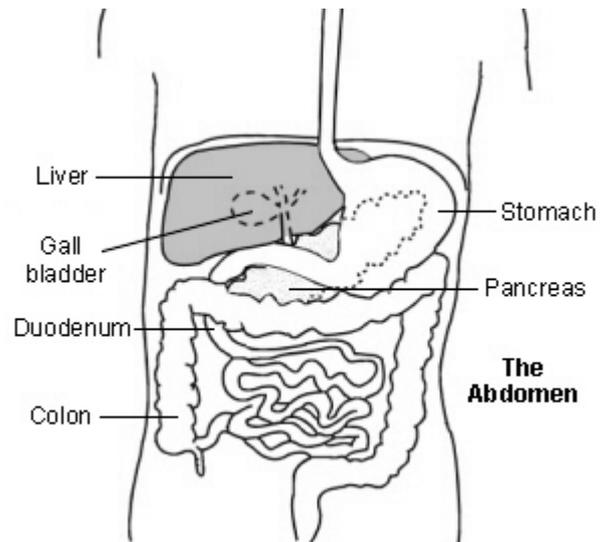
Autoimmune Hepatitis

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), which is eventually fatal. With treatment, the outlook is good. Treatment is with steroids and/or other drugs 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 and helps to digest fats.



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 results 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 foreign 'germs'. In people with autoimmune diseases the immune system attacks part or parts of the body as if it was '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. 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. About 1 in 100,000 people in the UK develop autoimmune hepatitis each year. About 7 in 10 cases are in women. It most commonly develops in women aged 15-40. However, it sometimes occurs in young children and older adults. Men are affected more commonly than women in older age groups.

People with autoimmune hepatitis have a high chance of also having one or more other autoimmune diseases such as pernicious anaemia, type 1 diabetes, overactive thyroid, Sjögren's syndrome, vitiligo, etc.

What are the symptoms of autoimmune hepatitis?

In many cases the symptoms develop gradually over weeks or months.

- Common early symptoms include: fatigue (tiredness), generally feeling unwell, mild pain in the upper abdomen over the liver, joint and muscle pains.
- Jaundice develops when the liver inflammation becomes worse. Jaundice is when you 'go yellow'. You tend to first notice it when the whites of the eyes become yellow. It is due to a build up of the chemical bilirubin which is made in the liver. This spills into the blood in some liver conditions. (With jaundice due to hepatitis your urine goes dark, your faeces (stools) may go pale, and you tend to itch.)
- Without treatment, in time the persistent inflammation causes liver damage called 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*'.)

In some cases, the symptoms develop quickly over a few days with an 'acute hepatitis'. (Acute means sudden onset or 'short duration'). This can cause a fairly sudden onset of fever, abdominal pain, jaundice, feeling sick, vomiting, and generally 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, if left untreated, most people with autoimmune hepatitis would die within 10 years of the onset of the disease due to liver failure caused by cirrhosis.

With treatment, the inflammation can usually be controlled, the outlook is good, and most people survive long-term.

How is autoimmune hepatitis diagnosed?

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 '*Liver Function Tests*'. These tests can confirm that you have hepatitis, but not usually the cause of the hepatitis.
- An ultrasound scan of the liver may help.
- Other blood tests can measure various auto-antibodies which occur in various autoimmune diseases. For example, antinuclear antibodies (ANA) or smooth muscle antibodies (SMA). 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 may 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 '*Liver Biopsy*'. The type of cells involved in the inflammation usually help to confirm the diagnosis of autoimmune hepatitis, and rule out other causes of hepatitis.

What is the treatment for autoimmune hepatitis?

Treatment aims to reduce inflammation and suppress the immune system with 'immunosuppressant' drugs.

- 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. (A separate leaflet discusses steroid tablets in more detail.)
- Azothiaprime is an immunosuppressant drug that works in a different way to steroids. It may be used in addition to the steroid. It may not be started until the inflammation has been brought under control with the steroid. A steroid plus azothiaprime tends to work better than either alone. Also, the dose of steroid needed may be less if you also take azothiaprime. This means that any side-effects from steroids may be less severe.

A steroid or azothiaprime are sometimes used alone, depending on various circumstances such as the severity of the condition, how well the condition responds to treatment, side effects, etc.

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.

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.

Once the condition has completely settled, after a year or two, your doctor may advise a trial without treatment. However, in many cases the condition flares up again if treatment is stopped. If it does flare-up again, treatment can be re-started. In many cases, long-term 'maintenance' treatment is then needed.

If steroids and/or azothiaprime do not work well, or cause severe side effects, other immunosuppressant drugs may be tried such as cyclosporin or tacrolimus.

For the few people who do not respond to drug treatment, or 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.

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 small amounts. If you already have liver inflammation, alcohol may increase the risk and speed of developing cirrhosis.

Further help and information

British Liver Trust

2 Southampton Road Ringwood BH24 1HY

Tel: 0870 770 8028 Web: www.britishlivertrust.org.uk

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Comprehensive patient resources are available at www.patient.co.uk