Children's Liver
Disease Foundation
fighting childhood
liver disease

Autoimmune Liver Disease

A Guide

An overview of autoimmune liver disease, including autoimmune hepatitis, autoimmune sclerosing cholangitis and AIH after a liver transplant (de novo AIH)

This information has been written for:

- parents and carers of children and young people with autoimmune liver disease

Others who may also find this information useful:

- young people with autoimmune liver disease
- healthcare professionals who would like to find out more about autoimmune liver disease

It provides information on:

- the different types of autoimmune liver disease
- diagnosis
- symptoms
- treatment
- relapse
- monitoring over time

You may also find it helpful to read the following CLDF leaflets:

- Introduction to Liver Disease
- Portal Hypertension and Ascites
- Liver Transplantation

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QR Code to view
our resources**

Key facts about autoimmune liver disease

1. Autoimmune diseases are conditions in which the immune system mistakenly attacks and damages the body's healthy cells.
2. In autoimmune liver disease, the immune system attacks healthy liver cells, bile duct cells or both.
3. There are 3 autoimmune liver diseases that can happen in children.
4. Autoimmune hepatitis is split into type 1 (AIH-1) and type 2 (AIH-2).
5. Blood tests, scans and biopsies help with the diagnosis.
6. Treatment involves using medicines to make the immune system less active. This is called immunosuppression.
7. The length of treatment will vary from child to child. Treatment is likely to last for several years, and often longer (sometimes for life).
8. Relapses can happen suddenly, at any point. But they are most likely to happen during the first 2 years of treatment and during puberty.
9. Long-term monitoring is important due to the risk of relapse. Your child will need regular blood tests.
10. In a small number of children, damage to the liver becomes worse over time. This can lead to severe scarring of the liver (cirrhosis).

What are autoimmune diseases?

Autoimmune diseases are conditions in which the immune system mistakenly attacks and damages the body's healthy cells.

The immune system usually defends us against bacteria and viruses that make us ill. It does this by making proteins called antibodies.

In autoimmune conditions, the immune system starts to make antibodies that stick to healthy cells. They are called autoantibodies. Each different antibody will only stick to one very specific marker. So autoantibodies will stick to one type of cell in the body. When this happens, the immune system will treat those cells as dangerous and attack them. This causes damage to cells and organs.

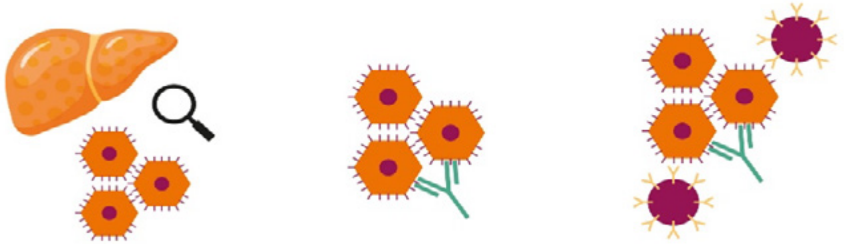
The body creates autoantibodies by mistake. It is not clear why the immune system acts in this way. There may be some factors that work together to trigger the reaction, including:

- a problem with the immune system
- genetics – it can run in families due to inherited genes
- causes outside the body, such as getting a virus or taking certain medicines

But some autoantibodies have no direct cause or trigger.

In autoimmune liver disease, the immune system mistakenly attacks and damages healthy cells. This may be liver cells (hepatocytes), bile duct cells (cholangiocytes) or both. This leads to inflammation in the liver.

Autoimmune liver diseases are NOT caused by alcohol or other toxins.



Liver cells

Autoantibodies wrongly mark the liver cells as foreign

Immune cells attack and destroy the liver

Immune system attack on liver cells

There are three autoimmune liver diseases that can happen in children:

- autoimmune hepatitis (AIH)
- autoimmune sclerosing cholangitis (ASC)
- autoimmune hepatitis after liver transplantation for a non-autoimmune liver disease. This is also known as de novo autoimmune hepatitis.

We know that autoimmune liver disease is rare in children. But we don't know the exact numbers affected. But we do know that:

- autoimmune hepatitis happens in less than 1 in every 100,000 children each year
- autoimmune hepatitis is more common in girls than in boys

What is autoimmune hepatitis (AIH)?

Autoimmune hepatitis is a rare, long-term (chronic) disease of the liver. It happens when the body's immune system causes damage to its own healthy liver cells. This leads to inflammation in the liver. AIH is more aggressive in children than in adults.

Types

Autoimmune hepatitis is split into two types. Each type has different autoantibodies present in the blood.

Type 1 (AIH-1)

This is the most common type of autoimmune liver disease in children. Two out of every three cases of AIH in children are type 1. This type usually starts in teenagers.

Type 2 (AIH-2)

This type of autoimmune liver disease is less common. It is more likely to affect young children, including infants. It is often more severe and gets worse (progresses) at a faster rate.

Hepatitis is the medical term for inflammation of the liver. There are many causes of hepatitis. Some are more well-known than others.

Autoimmune hepatitis is different to hepatitis caused by viruses such as hepatitis A, B and C. You can't catch autoimmune hepatitis.

Treatment for AIH involves using medicines. They help make the immune system less active. If left untreated, AIH can cause serious scarring of the liver (cirrhosis). This could lead to liver failure in childhood or in adulthood.

With treatment, the outlook for most children is very good.

What is autoimmune sclerosing cholangitis (ASC)?

Autoimmune sclerosing cholangitis is similar to AIH. You may hear it called juvenile sclerosing cholangitis or AIH/PSC overlap syndrome.

Children with ASC have inflammation of the liver (hepatitis). They have the same autoantibodies that are present in AIH-1. But they also have inflammation and infection of the bile ducts. This is known as cholangitis.

Autoimmune sclerosing cholangitis (ASC) =

autoimmune inflammation of the liver (autoimmune hepatitis)

\+

inflammation and infection of the bile ducts (cholangitis)

ASC affects boys and girls in equal numbers.

ASC is often linked with inflammatory bowel disease (IBD). The main types of IBD are Crohn’s disease and ulcerative colitis. They cause inflammation in the bowel (intestine). The main symptoms are severe tummy pain, diarrhoea and bloody poo (stools).

Most cases of ASC are type 1. ASC is rarely linked with AIH-2.

What is autoimmune hepatitis after liver transplant (de novo AIH?)

This is a rare complication of liver transplant. It happens in around 2% to 6% of children who have a transplant for conditions other than autoimmune liver disease. In AIH after liver transplant, the immune system attacks the new liver. The condition looks identical to autoimmune hepatitis. It is treated in the same way as this condition.

The cause of AIH after liver transplant is unclear. There may be a link to the medicines used after transplant. They may trigger an autoimmune reaction in a small number of children.

It is important to treat AIH after transplant quickly to make sure that the new liver stays healthy.

What are the symptoms of autoimmune liver disease?

The symptoms of autoimmune liver disease will vary from child to child. Some children will appear well and active. They may show no symptoms. Others will be very ill. They may show signs of liver failure.

The most common symptoms of autoimmune liver disease are:

- tiredness (fatigue)
- generally feeling unwell
- loss of appetite

Other symptoms may include:

- feeling sick (nausea) or being sick (vomiting)
- tummy (abdominal) pain
- yellowing of the whites of the eyes and skin (jaundice)
- dark wee (urine) and pale poo (stools)
- joint and muscle pain
- weight loss
- itching (pruritus)
- high temperature (fever)
- nose bleeds, bleeding gums and bruising easily
- delayed starting of periods or periods that stop once started (amenorrhoea)
- loose, watery poo (diarrhoea) - (bowel symptoms are more common in ASC)

Symptoms that can appear as the condition progresses

- swollen tummy (ascites)
- swelling, especially in the lower legs, ankles and feet (oedema)
- irritability, confusion and mood changes

Children with AIH after a transplant are usually well with no symptoms. The condition is often picked up on blood tests.

How is autoimmune liver disease diagnosed?

There is no single test for autoimmune liver disease. The diagnosis is based on a combination of factors. This means a diagnosis can take time. Many children are diagnosed after having tests for another health problem.

The symptoms of autoimmune liver disease can be similar to those seen in other liver diseases. The medical team need to rule these out before making a diagnosis.

Tests may include:

Blood tests:

- liver blood tests (also known as liver function tests or LFTs)
- blood tests to check for the autoantibodies
- blood tests to rule out other liver diseases

To do these tests, a blood sample is taken and tested in a laboratory. The results will help the doctor understand your child's general health. They are also a good way of seeing if the liver is injured and how well it is working.

Abdominal ultrasound scan

An abdominal ultrasound scan uses sound waves to create a picture of the inside of the body. It may be used to check and monitor:

- the size and texture of the liver
- the size of the spleen
- the blood flow into and out of the liver

Magnetic resonance imaging (MRI) / magnetic resonance cholangiopancreatography (MRCP)

These scans use strong magnets and radio waves to make pictures of the internal organs. In autoimmune liver disease they help doctors see the bile duct tree. They can help find inflammation, narrowing or even blockages.

Liver stiffness measurements, e.g. FibroScan

Special scans are used to monitor the amount of stiffness in the liver. Healthy liver tissue is soft, so stiffness shows that damage has occurred.

Liver biopsy

Your child will need a liver biopsy to confirm the diagnosis and check the severity of the liver disease.

During this test a very thin needle is inserted through the tummy (abdominal) wall and into the liver. The needle takes a small sample of liver tissue. This is sent to a laboratory to be studied under a microscope.

How is autoimmune liver disease treated?

Treatment for autoimmune liver disease involves using medicines to suppress the immune system to make it less active. This is called immunosuppression. These medicines stop the immune system from attacking the liver. The aim of treatment is to stop liver inflammation. This will also prevent or reduce liver scarring (fibrosis).

Doctors use a combination of medicines to suppress the immune system. The amount of medicine needed (the dose) will be different for each child. Doctors aim to use the lowest dose they can. This helps to reduce side effects.

Steroids are usually the first treatment. They are drugs that reduce swelling (inflammation). They also suppress the immune system.

Blood tests will be done at least weekly when treatment begins. This is to check if the medicines are effective.

Prednisolone

Prednisolone is the main steroid used to treat autoimmune liver disease. Sometimes it is simply called “pred”. The response to this treatment is usually fast. It is very effective for most children.

High doses are usually needed when treatment starts. The dose will be reduced gradually (usually over 6-8 weeks). But this will depend on blood test results and symptoms.

Azathioprine

Azathioprine is another drug used to treat autoimmune liver disease. Like prednisolone, it reduces the response of the immune system. But it works in a different way to steroids.

Azathioprine is used in addition to prednisolone. The amount of steroid needed is often lower when azathioprine is added. This means that the side effects from the steroid may be less severe. They may even stop completely.

Azathioprine is usually started a few weeks after steroid treatment. It is started at a low dose and gradually increased if needed. Feeling sick (nausea) is a common side effect at the start of treatment. This usually eases after a few weeks.

Prednisolone and azathioprine are effective in most children. But it can take a year or more to get the disease fully under control. Most children lead a good quality of life while taking medication.

These drugs may not work in a small number of children. Other treatments may be needed if your child:

- experiences too many side effects from treatment
- has lots of relapses when trying to reduce treatment

The medical team will discuss other treatment options with you if they are needed.

Additional treatments used in autoimmune sclerosing cholangitis

In ASC, the autoimmune hepatitis part of the condition responds well to immunosuppression treatment. The bile duct inflammation part does not. This gets worse in about half of children.

Children with ASC may need other treatments to manage:

- inflammation and infection of the bile ducts (cholangitis)
- inflammatory bowel disease

Inflammatory bowel disease will be managed by a paediatric gastroenterologist.

Extra medicines used in ASC may include:

Ursodeoxycholic acid

Ursodeoxycholic acid is also known as urso or UDCA. It is a bile salt that occurs naturally in small amounts in bile. Taking it as a medicine can help improve bile flow out of the liver.

Mesalazine (also known as 5-ASAs or aminosalicylates)

In ASC, the wall of the bowel can become inflamed and irritated. This drug reduces inflammation and allows damaged tissue to heal.

Antibiotics

Antibiotics may be used to prevent or treat inflammation and infection of the bile ducts (cholangitis)

Anti-TNF medicines

These drugs may be used if the inflammatory bowel disease is very severe or difficult to treat.

Other treatments:

Endoscopic retrograde cholangio-pancreatography (ERCP)

ERCP is a medical procedure. It is used to diagnose and treat conditions of the liver, bile ducts, pancreas and gallbladder.
It may be used if there is narrowing in the bile duct tree. The medical team use ERCP to stretch the narrowed bile ducts. The aim of the treatment is to improve bile flow.

Treatment side effects

Steroid treatment may cause side effects in some children.
Some side effects happen straight away.

Others happen after weeks or months. The medical team will monitor your child and will do regular blood tests.

Possible side effects from using steroids include:

- being more likely to pick up an infection
- increased appetite and weight gain
- acne
- bone weakness (osteoporosis)
- anxiety and mood changes
- roundness of the face
- increased body hair (hirsutism)
- slower growth

High doses of steroids may be needed when starting treatment. The medical team will reduce the dose as quickly as possible. They will aim for a dose that controls the disease with as few side effects as possible. High doses may also be used during a period of relapse.

High doses can cause more severe complications including:

- obesity
- raised blood sugar (hyperglycaemia)
- difficulty sleeping (insomnia)
- high blood pressure (hypertension)

Azathioprine treatment can also cause side effects. The most common side effects include:

- feeling sick (nausea)
- being sick (vomiting)
- loose stools (diarrhoea)
- loss of appetite
- tiredness (fatigue)

Tell your medical team about any side effects. They may be able to help by changing medicines or altering the dose.

Living with a liver disease can be hard. Your child will be coping with physical symptoms. They may also struggle with the side effects of treatment.

Some children and young people find it hard to keep taking medicines. But it is very important not to stop any medicines without talking to the medical team. Stopping medicines suddenly can cause a relapse.

Staying healthy on immunosuppressants

Immunosuppressants affect the way your child's immune system works. This means they may be more at risk of infections.

Here is some advice to help your child stay healthy when taking these medicines:

Avoid anyone who has or may have:

- measles
- shingles
- chickenpox
- flu
- COVID
- respiratory syncytial virus (RSV)

If your child becomes ill, make a doctor's appointment as soon as possible.

Protect skin from strong sun and use at least SPF30 sunscreen. This is because immunosuppressants can slightly increase the risk of developing skin cancer and some other cancers.

Your child may not be able to have 'live' vaccines during treatment. They may also need to avoid live vaccines for 3-6 months after finishing treatment. The medical team will make sure your child's vaccinations are up-to-date before starting immunosuppressants. Your child may also need extra vaccinations including:

- flu vaccine
- COVID vaccine
- pneumococcal vaccine

Other tips for staying healthy on immunosuppressants:

- taking prednisolone in the morning can help with problems sleeping
- reduce salt intake and drink more water
- get plenty of sleep

Weight gain can be a distressing side effect for children and young people. Help your child eat well without increasing portion sizes. Regular exercise will also help your child keep their weight stable.

How long does treatment continue for?

The length of treatment will vary from child to child. It will also depend on:

- the type of autoimmune liver disease
- the response to treatment
- whether the disease gets worse again (relapses)

Treatment is likely to last for several years, and often longer (sometimes for life).

Treatment will only be reduced or stopped if symptoms improve or go away for enough time. This is called remission. It will only be considered after at least two to three years of treatment. The medical team will also want to see completely normal blood tests during the last year. Your child will also need a repeat liver biopsy. This is used to check that the inflammation in the liver has disappeared.

If the decision to stop treatment is made, the dose is decreased very slowly. This is because liver tests can get worse again if:

- doses are reduced too quickly
- medicines are stopped suddenly

Symptoms may also come back. If this happens, your child will need to re-start treatment at a higher dose.

- Around 1 in 5 children with type 1 AIH can eventually stop treatment.
- Very few children with type 2 AIH can ever stop treatment. Relapse is common in type 2.

What is a relapse and how is it managed?

A relapse is when the disease becomes active again after a period of remission. Relapses can happen suddenly, at any point. But they are most likely to happen during the first two years of treatment. They are also more common during puberty.

Relapse happens in around 2 out of every 5 children.

A relapse is usually picked up on routine blood tests. Or it may be noticed due to symptoms starting again.

A relapse can also happen if medicines are not taken as prescribed. This is called non-adherence. It is crucial that children and young people understand the importance of taking their medicine. There are usually no symptoms at the start of a relapse. This may give the false impression that it is safe not to take the medicine.

When a relapse happens, the dose of medicine will need to be increased. The aim is to try and bring the disease back under control.

How is autoimmune liver disease monitored?

Children with AIH or ASC will need regular blood tests. When first diagnosed, testing will be done at least weekly. This helps the doctors check the liver and see how quickly the dose of steroids can be reduced. As your child's condition stabilizes, blood tests will be needed less often (at least every 3 months).

Long-term monitoring is important due to the risk of relapse.

What happens if the disease gets worse over time?

A small number of children will not respond to treatment. If treatments are not effective, damage to the liver can become worse over time.

In ASC, bile duct disease gets worse over time in about half of children.

Complications of long-term (chronic) liver disease include:

- weight loss or poor weight gain
- fluid in the tummy (ascites)
- high blood pressure in the blood vessels around the liver (portal hypertension)
- yellowing of the whites of the eyes and skin
- itchy skin (pruritus)
- bile duct infection (cholangitis)

Liver transplant

Liver transplant is only considered if children don’t respond to medicines and the liver is damaged beyond repair. The rate of liver transplant is higher in children with ASC.

A liver transplant is an operation to remove a liver that does not work. It is replaced with a healthy liver from another person (donor). A successful liver transplant will help a child survive when the liver is failing. It will also significantly improve a child’s quality of life.

A liver transplant is a major operation. It will only be done after the benefits and risks have been carefully weighed up. Following a successful liver transplant children need lifelong medicines and follow up.

Autoimmune liver disease that comes back after transplant

Autoimmune liver disease can come back in the new liver following transplant. Doctors called this recurrence. It is different to AIH following transplant for a different type of liver disease (de novo AIH).

If autoimmune hepatitis comes back, treatment for AIH will be re-introduced. Children usually respond well to treatment. But some research suggests that children with AIH type 1 have a better outcome than those with AIH type 2. It is important to treat AIH after transplant quickly. This helps the new liver stay healthy.

Autoimmune sclerosing cholangitis can also come back after transplant. This is often accompanied by inflammatory bowel disease (IBD). This may get worse over time (progress).

There are no standard treatments for ASC after transplant. Up to two thirds of children will eventually need another liver transplant.

Useful resources

Glossary of liver terms

Many medical terms may be used by those caring for children and young people with liver disease. Follow this QR code for a list of terms.

Other CLDF resources

CLDF produces a wide range of information for children and young people up to the age of 25 with liver disease, their families and the healthcare professionals who look after them.

Information can be downloaded or ordered from:
Website: **www.childliverdisease.org**
Email: **info@childliverdisease.org**
Tel: **0121 212 3839**

Follow this QR code to see our resources.

How we produce information

Our information resources are written, edited and reviewed with the help of medical experts and families living with liver disease.

Thanks

We would like to thank staff at each of the specialist paediatric liver centres: Birmingham Children's Hospital, King's College Hospital and Leeds Children's Hospital. We would also like to thank the families who helped us produce this information.

PIF TICK accreditation

We have gained PIF TICK accreditation for our information production process. Follow this QR code for more details.

Disclaimer

This resource provides general information but does not replace medical advice. It is important to contact your medical team if you have any worries or concerns.

Feedback and information sources

Your feedback is important to us and will help us improve our information. To provide feedback, or for more information on how it was developed, email: info@childliverdisease.org.

This leaflet was reviewed in June 2025.

It is due to be reviewed by June 2028.

Notes

Children's Liver Disease Foundation

Children's Liver Disease Foundation (CLDF) is the UK's leading organisation dedicated to fighting all childhood liver diseases.

CLDF provides information and support services to young people up to the age of 25 with liver conditions and their families and is a voice for everyone affected.

CLDF is here for you, whether you want to talk, share with others, or just belong to a group which cares, knows what it's like and is fighting to make a difference.

Are you a young person up to the age of 25?

Contact the Young People's Team:
Phone: 0121 212 6024 **Email:** youngpeople@childliverdisease.org

Are you the parent or carer of a child or young person with a liver condition?

Contact the Families Team:
Phone: 0121 212 6023 **Email:** families@childliverdisease.org

Would you like to help us support the fight against childhood liver disease? Please help us to continue to support children, young people and families now and in the future.

Contact the Fundraising Team:

Email: fundraising@childliverdisease.org

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