

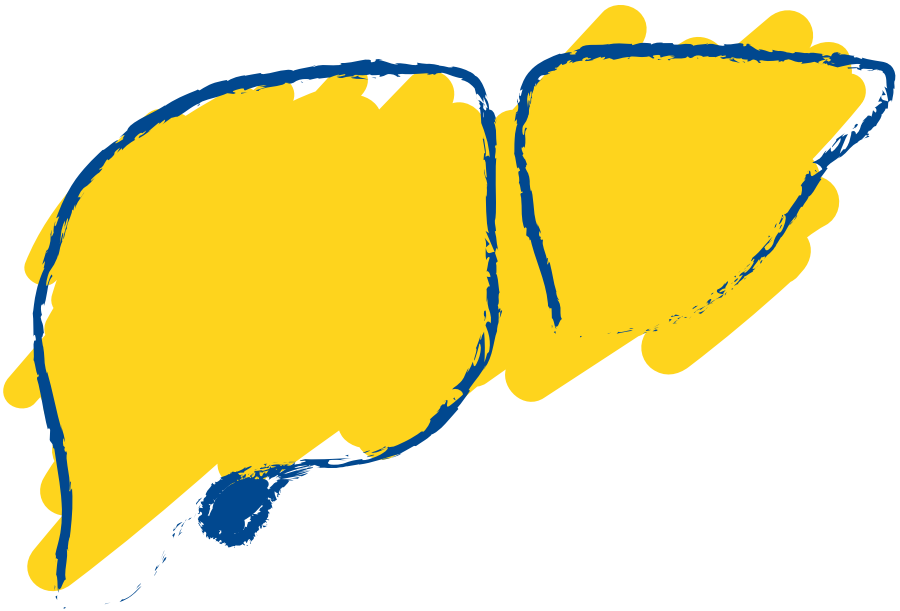


Children's Liver
Disease Foundation

fighting childhood
liver disease

Autoimmune Liver Disease

A Guide



An explanation of what autoimmune liver diseases are including autoimmune hepatitis and autoimmune sclerosis cholangitis

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This information has been written for:

- Parents/carers of a child diagnosed with autoimmune liver disease (this includes autoimmune hepatitis and autoimmune sclerosing cholangitis)

Others may also find this information useful:

- Young people diagnosed with autoimmune liver disease
- Healthcare professionals who would like to find out more about the condition

This leaflet aims to:

- Explain what autoimmune liver diseases are including autoimmune hepatitis and autoimmune sclerosing cholangitis
- Describe the signs and symptoms of autoimmune liver disease
- Discuss the diagnosis and treatment of these conditions

You may find it helpful to read the following CLDF leaflet:

- An Introduction to Liver Disease

What are autoimmune diseases?

Autoimmune diseases are caused by the body's immune system attacking itself.

An antibody is a protein, made by the immune system, which identifies and removes bacteria and viruses from the body. Sometimes, the antibody attacks the body's own tissues instead of a bacteria or a virus. These are called autoantibodies.

It is not known exactly what causes the autoantibodies to act in this way. It is thought that there may be a number of causes which include:

- A problem with the immune system
- An individual's genes
- Environmental factors

In children there are two liver conditions caused by autoimmune disorders:

Autoimmune hepatitis (AIH)

and

AIH/sclerosing cholangitis overlap syndrome known as autoimmune sclerosing cholangitis (ASC)

These are autoimmune liver diseases. They are similar conditions and are treated similarly.

What is autoimmune hepatitis (AIH)?

Hepatitis means inflammation of the liver. In AIH the inflammation is caused by the immune system.

What is autoimmune sclerosing cholangitis (ASC)?

ASC is similar to AIH but as well as hepatitis there is also damage to the bile ducts.

What are the symptoms of autoimmune liver disease?

It can be difficult for childhood autoimmune liver disease to be diagnosed because the symptoms are similar to many other liver disorders and they can vary greatly.

Some children and young people with autoimmune liver disease may appear well and active whilst others can be very ill and have signs of liver failure. Generally autoimmune liver disease in children and young people is more aggressive than it is in adults but treatment can be very successful.

The most common symptoms are:

- Tiredness and generally feeling unwell
- Loss of appetite

Other symptoms are:

- Nausea (feeling sick) or being sick
- Abdominal pain
- Jaundice with dark urine and pale stools (poo)
- Joint and muscle pain
- Weight loss
- Itching
- Fever
- Nose bleeds, bleeding gums and bruising easily
- Amenorrhoea (delayed starting of periods or they stop once started)
- Diarrhoea (bowel symptoms are more common in ASC)

Later symptoms which can appear as the condition progresses include:

- A swollen abdomen (ascites)
- Swelling, especially in the lower legs (oedema)
- Irritability and confusion

How is autoimmune liver disease diagnosed?

If a child or young person is displaying the symptoms of ALH there are a number of tests which are used to confirm a diagnosis:

- Blood tests
- Liver biopsy
- Ultrasound scan
- MRI/MRCP scan
- Upper and lower gastrointestinal endoscopy if there are bowel symptoms

More information regarding these routine investigations can be found in the CLDF leaflet “An Introduction to Liver Disease”.

Children with autoimmune liver disease usually have high immunoglobulin G levels (a type of antibody). They also usually have positive autoantibodies which help define the type of autoimmune liver disease they have.

What are the different types of AIH?

There are two types of AIH. Each type has different types of autoantibody present:

Type 1 = Anti-nuclear (ANA) and/or anti-smooth muscle (SMA) antibodies

Type 1 makes up two out of three of all cases of AIH. It normally starts around puberty.

Type 2 = Liver kidney microsomal (LKM) antibodies

Type 2 is mainly found in younger children.

Children who have AIH are usually positive for ANA and/or SMA autoantibodies. Very few children will have LKM autoantibodies.

How is autoimmune liver disease treated?

Prednisolone

Prednisolone is a steroid and is normally the first treatment which is used. Sometimes it is simply called “pred”.

High doses of steroids are required when treatment starts. The dose will gradually be reduced depending on blood results and symptoms. The overall aim of treatment is to ensure the disease is treated using as little medicine as possible.

Side effects of steroids include:

- Being more likely to pick up an infection
- Increased appetite and weight gain
- Acne
- A risk of bone weakness
- Behavioural changes
- Slower growth
- Diabetes in some individuals

Azathioprine

Azathioprine is another medicine which can be given to help the steroid to work. This is a type of immunosuppressive medicine which reduces the response of the immune system.

Other medicines are available if prednisolone and azathioprine don't successfully treat the disease. These options will be discussed with you by your medical team if they are necessary.

Other medications

Another medicine called ursodeoxycholic acid (known as urso) is also usually given to children with ASC. The aim of this is to improve bile flow through the bile ducts.

If colitis (an inflammation of the large intestine) is present, different medicines may be prescribed; this includes a drug called mesalazine. Your healthcare team will talk to you about these if they are needed.

It's important to note that most children and young people respond well to treatment.

For young people the side effects of treatment can be distressing, especially cosmetic changes such as weight gain. CLDF's young people's team is here to support young people over the age of 11 and can be contacted at ypo@childliverdisease.org or on **0121 212 6024**.

The psychology teams at the liver centres can also provide help and support. Get in touch to find out about the support available.

How long does treatment continue for?

This can depend on a variety of things but treatment usually has to be continued for a long time, normally years.

Remission is the aim of treatment. This means blood tests measuring liver function and inflammation are within a normal range.

Relapse

A relapse is where the disease becomes active again after remission. Relapses can occur suddenly, particularly during puberty. This may be picked up on blood tests or may be noticeable due to symptoms reappearing. Relapse can also happen when medicines have not been taken as prescribed (non-adherence).

It is really important that young people understand the importance of taking their medication properly, even if they feel OK.

During a relapse medicine dosages may need to be increased to bring the disease back under control.

Considering stopping treatment can only be done if there have been at least two years of completely normal blood tests and often only if a repeat liver biopsy shows that the inflammation in the liver has disappeared. If treatment is decreased, it is done very slowly.

About 20% of children/young people with type 1 autoimmune hepatitis who respond well to prednisolone and azathioprine can eventually stop treatment.

Very few children/young people with type 2 autoimmune hepatitis can ever stop treatment.

How is autoimmune liver disease monitored?

Those with AIH/ASC require regular blood tests. You can read more about these in CLDF's information called "An Introduction to Liver Disease".

Monitoring the condition is really important. Regular blood tests are taken to measure AST (aspartate aminotransferase) and ALT (alanine aminotransferase). These are enzymes which are normally present in the liver and their levels can be used to monitor the health of the liver.

When first diagnosed, testing may be done weekly to see how quickly the level of steroids can be reduced. As the condition stabilises, blood tests will be needed less often (every 3-6 months).

What happens if the disease progresses?

Complications of chronic liver disease include:

- Weight loss/poor weight gain
- Ascites (swelling of the tummy)
- Portal hypertension (high blood pressure in the blood vessels around the liver)
- Jaundice

CLDF has leaflets on ascites, portal hypertension and pruritus (itchiness). They can be ordered from CLDF or downloaded from the CLDF website.

If the function of the liver is affected by the disease then other medicines may be given. For example vitamin K may be given which helps the blood to clot.

Liver transplantation is considered in cases that don't respond to medicine or when the liver is damaged beyond repair and the complications are life threatening. Rate of liver transplantation is higher in cases of ASC and recurrence of autoimmune liver disease may occur following liver transplantation, especially in cases with ASC.

Most children can lead a good quality of life whilst taking medication.

CLDF has a leaflet on liver transplantation for further information.

CLDF produces a wide variety of information resources for children and young people up to the age of 25 with liver disease, their families and the healthcare professionals who look after them. This information can be downloaded or ordered from CLDF's website www.childliverdisease.org. For further enquiries regarding CLDF's information please contact the Health and Research Information Manager by email at hrim@childliverdisease.org or call 0121 212 6029.

Thanks

This booklet has been written, edited and reviewed with the help of staff at each of the specialist paediatric liver centres: Birmingham Children's Hospital, King's College Hospital and Leeds Children's Hospital. Thank you to all of the staff involved who have made the production of this leaflet possible.

Disclaimer

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

Feedback and Information Sources

Information within this leaflet has been produced with input from the three specialist paediatric liver centres in the UK. To provide feedback on this leaflet, for more information on the content of this leaflet, including references and how it was developed, contact Children's Liver Disease Foundation: info@childliverdisease.org

This leaflet has been reviewed in June 2017. It is due to be reviewed by June 2020.

What is Children's Liver Disease Foundation (CLDF)?

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, funds vital research into childhood liver disease and is a voice for everyone affected.

Are you a young person up to the age of 25 with a liver condition or a family member? CLDF's Families and Young People's teams are here for you, whether you want to talk about issues affecting you, meet and share with others, or just belong to a group which cares, knows what it's like and is fighting to make a difference. You are not alone.

If you are a parent/carer or family member then get in touch with CLDF's Families Team:

Phone: 0121 212 6023 **Email:** families@childliverdisease.org

If you are a young person and want to find out more about CLDF's services you can contact CLDF's Young People's Team:

Phone: 0121 212 6024 **Email:** youngpeople@childliverdisease.org

CLDF have a social media platform called **HIVE** for 11 – 25 year olds with a liver disease/transplant to make new friends, connect and share stories
www.cldfhive.co.uk

Would you like to help us support the fight against childhood liver disease?

All of CLDF's work is funded entirely through voluntary donations and fundraising. Please help us to continue to support young people, families and adults diagnosed in childhood now and in the future. To find out more about fundraising and how you can join the fight against childhood liver disease you can visit www.childliverdisease.org/get-involved. Alternatively you can email the Fundraising Team at fundraising@childliverdisease.org or call them on **0121 212 6022**

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