

## Autoimmune Hepatitis (AIH)

European Reference Network on Hepatological Diseases (ERN RARE-LIVER)

<https://rare-liver.eu>

Contact: [ern.rareliver@uke.de](mailto:ern.rareliver@uke.de)

### What is AIH?

AIH is a rare autoimmune liver disease. An autoimmune disease is a condition in which your immune system mistakenly attacks your body. The immune system of people with AIH mistakenly attacks the body's own liver cells causing damage and inflammation.

### Who is affected?

AIH can be diagnosed at any age and affects more females than males.

### Why do I have AIH?

The causes are still unknown. Research suggests that some people have a genetic predisposition for AIH and that the manifestation of the disease has a trigger, but it is not something that the patient has done. Despite this genetic predisposition, there is only a very small risk of a relative also having AIH.

### How is AIH diagnosed?

AIH can develop slowly in some individuals, with no symptoms or very mild, vague symptoms. In a few people, AIH develops rapidly and hospitalisation is required. In around a third of patients, severe liver damage (liver cirrhosis) may have already developed by the time the patient is diagnosed.

The following symptoms may occur:

- Fatigue, nausea, abdominal discomfort
- Joint pain (mostly involving multiple joints, no redness or swelling)
- Skin alterations (e.g. red colouring of the palm of the hand)
- Jaundice (yellowing of the skin or eyes) and dark urine in severe cases

There are not any specific tests that tell a doctor you have AIH. It is actually a matter of piecing together results (both positive or negative) from a range of tests (including blood tests and liver biopsy).

### Liver Blood Tests

If you have AIH, you are likely to have higher levels of the following:

- Aspartate aminotransferase (AST) / Glutamat-Oxalacetat-Transaminase (GOT) (shows liver injury)

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- Alanine aminotransferase (ALT) / Glutamat-Pyruvat-Transaminase (GPT) (shows liver injury)
- Immunoglobulin G (IgG) (shows liver inflammation)

In addition, you may test positive for autoantibodies, especially antinuclear antibodies (ANA), anti-smooth muscle antibodies (anti-SMA), anti-soluble liver antigen/liver pancreas antigen antibodies (anti-SLA/LP) and/or anti-liver-kidney microsome antibodies (anti-LKM).

### Liver biopsy

Liver biopsy is important to confirm the diagnosis, to determine the severity of liver damage and to rule out other liver diseases. It involves removing a tiny piece of the liver for examination.

Most hospitals perform liver biopsy using ultrasound to identify the exact location of the liver. Local anaesthetic is applied under your right ribs. A tiny piece of the liver is then removed with a fine needle for examination under a microscope.

Some hospitals perform liver biopsies that are mini-laparoscopically guided. For this procedure, you are sedated and the liver is examined with a small camera when the biopsy is taken. Another way to perform liver biopsy is by a transjugular intervention – for this technique a small catheter is put into your neck vein and pushed forward into one of the liver veins, and a small sample of the liver is taken.

After the biopsy you will be asked to lie still for four to six hours and might be kept in hospital overnight to make sure that there is no bleeding after the procedure. There is a small risk of bleeding after any liver biopsy and your doctor will discuss the benefits and risks of this procedure with you.

### What can my doctor do?

To control inflammation in the liver and lower the immune system's activity, your doctor will prescribe a combination of medications which include corticosteroids (e.g. prednisolone) and immunosuppressants (e.g. azathioprine).

The cortisone-like drug budesonide may also be used for treatment. Budesonide is metabolized very quickly in the liver so it has fewer side effects. However, it must not be given to individuals who have already developed liver cirrhosis.

Azathioprine allows the reduction of steroid doses and helps to suppress the inflammation of the liver in the long term.

Your doctor will adjust your medication doses over time to minimise side effects and maximise long term treatment success.

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AIH medications are effective if used correctly but can cause side effects, for example, nausea. It is important that you take all medications as prescribed and that you discuss any side effects with your doctor immediately so that different treatments or doses can be considered. **It is important not to stop or alter the dose of your medications without consulting your doctor.** This can cause disease reactivation with additional scar forming in the liver (liver cirrhosis).

Most people with AIH take these medications for life and do very well. However, in a small number of people, the liver damage worsens and a liver transplantation is necessary.

If you feel that you need some psychological support, please speak to your doctor about services that may be available to you.

### Follow-up care

Your blood tests should be monitored regularly (every 3-6 months or more frequently).

**Transient elastography** (e.g. FibroScan® if available) may be used to assess liver stiffness, an indication of severity of liver damage.

### Skin examination

Sun protection should be used. Regular dermatological screening for skin cancer can be considered.

**Bone density scan** may be used to check your bones, as some AIH medications can affect bone density.

### Warning signs

If your skin or whites of the eyes turn yellow (jaundice), you should consult your doctor immediately.

### Do I need a special diet?

There is no special diet for AIH. We recommend that you eat a healthy, well-balanced diet. Do not take any supplements without first checking with your doctor.

### Can I have a family?

YES. If you are female and if you wish to have children, speak to your doctor so that your pregnancy can be properly managed. If you are male, family planning is not known to be affected. Speak to your doctor if you have any questions.

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### Vaccinations

Vaccinations against viral hepatitis A and B and pneumococcal infections (bacteria) are recommended. You should discuss non-live vaccinations against herpes zoster (shingles) with your physician. We recommend vaccination against influenza as early as possible each autumn. Live vaccines must not be given to AIH patients who are undergoing immunosuppressive treatment.

### Is AIH an infectious disease?

No. AIH is entirely autoimmune. It is not infectious or contagious.

### How can I find a specialist?

AIH is a rare disease and as such not every doctor has experience in treating AIH. It needs to be managed by a specialist or a hospital with experience in treating AIH. Some hospitals are part of a national network for rare liver diseases and/or of the Europe-wide network for rare liver diseases, ERN RARE-LIVER. This means that patients seen at hospitals within the network can benefit from the expertise of specialists who work at other hospitals within the network. For more information about the ERN RARE-LIVER, visit <https://rare-liver.eu/>.

### Find information and support

Click on the section “patients” on the ERN RARE-LIVER website (<https://rare-liver.eu/>).

### Disclaimer

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