RIVER CITY
GASTROENTEROLOGY

Autoimmune Hepatitis (AIH)

Understanding your diagnosis and long-term management

WHAT IS AUTOIMMUNE HEPATITIS?

Autoimmune hepatitis (AIH) is a chronic liver disease in which the immune system mistakenly attacks the liver's own cells, causing ongoing inflammation. Left untreated, this inflammation can progress to cirrhosis (severe liver scarring) and liver failure. However, when diagnosed and treated appropriately, most people with AIH achieve remission and live a normal or near-normal lifespan.[1,2]

AIH is relatively uncommon, affecting approximately 1-2 people per 10,000. It can develop at any age but most commonly presents in young women and again in women over 40. It affects women approximately four times more often than men. AIH often occurs alongside other autoimmune conditions such as thyroid disease, type 1 diabetes, and rheumatoid arthritis.[1]

TYPES OF AUTOIMMUNE HEPATITIS

AIH is classified into two main types based on the autoantibodies present in the blood:[1]

- **Type 1 AIH (most common)**: associated with anti-nuclear antibodies (ANA) and anti-smooth muscle antibodies (ASMA). Can occur at any age.
- **Type 2 AIH**: associated with anti-liver-kidney microsomal antibodies (anti-LKM1) and anti-liver cytosol antibodies (anti-LC1). More common in children and young adults.

WHAT CAUSES AIH?

The exact cause is not fully understood. AIH is thought to result from a combination of genetic susceptibility and environmental triggers. Possible triggers include viral infections, certain medications (such as minocycline, nitrofurantoin, or herbal supplements), and other environmental factors that may initiate an abnormal immune response in genetically susceptible individuals.[1,2]

SYMPTOMS

Symptoms of AIH vary considerably. Some people are diagnosed incidentally on routine blood tests, while others present with acute or even fulminant (sudden, severe) liver disease. Typical symptoms include:[1]

- Fatigue (most common symptom)
- Jaundice (yellowing of the skin and whites of the eyes)
- Abdominal discomfort or pain in the upper right abdomen
- Nausea and loss of appetite
- Joint pains
- Skin rash (in some patients)
- Amenorrhoea (absence of menstrual periods) in women
- Dark urine and pale stools

A small number of people with AIH initially present with acute liver failure, which is a medical emergency requiring urgent hospitalisation and specialist care.

DIAGNOSIS

AIH is diagnosed through a combination of blood tests, liver biopsy, and exclusion of other liver diseases. There is no single definitive test; diagnosis is based on a scoring system incorporating multiple criteria.[1,2]

- **Blood tests (liver function)**: elevated transaminases (ALT, AST) indicating liver cell inflammation. Elevated bilirubin and reduced albumin in more advanced disease.
- **Autoantibody testing**: ANA, ASMA, anti-LKM1, anti-LC1, and anti-soluble liver antigen (anti-SLA) antibodies. Elevated immunoglobulin G (IgG) is a hallmark of AIH.
- **Viral hepatitis serology**: to exclude hepatitis B, C, and other viral liver diseases.
- **Liver biopsy**: a small tissue sample is taken from the liver and examined under a microscope. Characteristic findings in AIH include interface hepatitis (inflammation at the border of portal tracts and liver tissue), plasma cell infiltration, and rosette formation. Biopsy can assist with diagnosis and assessing the degree of fibrosis.
- **Imaging (ultrasound, FibroScan)**: to assess liver structure and the degree of fibrosis.

TREATMENT

The goal of treatment is to suppress the immune attack on the liver and achieve remission (normalisation of liver enzymes and control of inflammation). Treatment should be initiated promptly to prevent progression to cirrhosis.[1,4]

Induction of remission

- **Prednisone (prednisolone)**: a corticosteroid given at moderate to high doses initially (40-60 mg/day), then gradually reduced over weeks to months as liver enzymes normalise. Prednisone is highly effective, achieving biochemical remission in approximately 80% of patients within 2 years.[1]
- **Budesonide**: an alternative corticosteroid that acts locally in the liver with less systemic absorption, producing fewer side effects (such as weight gain, mood changes, and bone loss) compared with prednisone. It is preferred in patients without cirrhosis.[4]
- **Azathioprine**: an immunosuppressant medication added to steroid therapy. It allows the steroid dose to be reduced more quickly and maintained at a lower dose (steroid-sparing effect). Azathioprine alone is insufficient to induce remission but is highly effective for maintenance.

Maintenance therapy

After remission is achieved, most patients require long-term maintenance therapy to prevent relapse. Options include:[1,2]

- Low-dose azathioprine (often 1-1.5 mg/kg/day) as monotherapy, which is the preferred long-term approach for most patients
- Low-dose prednisolone in combination with azathioprine for patients who cannot tolerate azathioprine alone

Treatment is typically continued for at least 2-3 years. Withdrawal of treatment is associated with relapse in the majority of patients. Lifelong therapy is often required.

MONITORING

Regular monitoring is essential throughout the course of AIH:

- Liver function tests and IgG levels are checked regularly (initially every 1-3 months, then less frequently once stable)
- Monitoring for medication side effects: blood counts (azathioprine can suppress bone marrow), blood glucose and bone density (steroids can cause diabetes and osteoporosis)
- Bone density (DEXA) scan at baseline and periodically during corticosteroid therapy

- Liver ultrasound and AFP blood test every 6 months if cirrhosis is present (to screen for liver cancer)

LIFESTYLE RECOMMENDATIONS

- Avoid alcohol completely, as it causes additional liver damage
- Maintain up-to-date vaccinations, particularly hepatitis A and B (as additional liver infection could be serious) and COVID-19 and influenza vaccines while on immunosuppression
- Avoid herbal supplements and complementary medicines without discussing with your hepatologist, as many can damage the liver
- Discuss contraception with your doctor: azathioprine and some other medications are not safe during pregnancy; specialist advice is required before planning a pregnancy

RESOURCES AND SUPPORT

- **Australian Liver Association:** www.australianliver.com.au - Find a hepatologist and access information on autoimmune liver disease in Australia
- **Liver Foundation Australia:** www.liver.org.au - Patient information and support for people with autoimmune hepatitis
- **Autoimmune Association Australia:** www.autoimmune.org.au - Support and information for people with autoimmune conditions

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