

WHAT CAN YOUR DOCTOR DO?

The aim of treatment is to 'dampen' the immune response and reverse inflammation. The mainstay of treatment remains cortico-steroids such as prednisolone. Whilst the body's own adrenal gland produces similar substances, greater amounts are required for treatment. Initially large doses are prescribed (ie between 20-30mg of prednisolone) which can then be tapered to inflammatory markers (ALT, level of immunoglobulin G). Other immunosuppressives used are those such as azathioprine (doses 0.5-1mg/kg) which allow lower doses of steroid. Other drugs such as cyclosporin and tacrolimus are sometimes used depending on the clinical situation; as yet there no large trials suggesting they should be routinely used. Budesonide, a steroid with less systemic effects, shows promise as an immunosuppressive in AIH.

Steroids such as prednisolone have variable side-effects which are marked at higher doses. It can increase the appetite and cause weight gain, fluid retention, high blood pressure, diabetes in susceptible individuals, mood swings and irritability can all occur. Over a longer period of treatment osteoporosis (thinning of the bones) can occur, as can glaucoma and thinning of the skin. However the aim of treatment is to utilise the smallest dose of prednisolone to control inflammation (as measured by the liver function tests especially the ALT). The maintenance dose is usually 3-6mg and at these levels long term complications are minimised. The majority of patients require ongoing treatment although a few can be weaned off all immunosuppressives. Follow up requires regular monitoring of liver function tests, assessment and prevention of treatment side effects and review of symptoms. Patients who are stable can be seen 3-6 monthly for review. Some patients (a small minority) have little liver reserve and such cases are put forward for transplantation.

HOW DO YOU LIVE WITH AIH?

Patients with AIH who have been assessed, diagnosed and treated nearly always return to a good level of functioning with excellent quality of life. Care should be taken not to drink alcohol excessively as this can damage the liver, likewise inappropriate use of medication such as analgesics (painkillers) is to be avoided. Pregnancy is safe on stable immunosuppressive doses of steroids and or azathioprine. Generally medications prescribed should be checked with your own family doctor. A healthy varied diet and moderate exercise is to be recommended.

LIVERNORTH can help you

As well as having many of the Freeman's health professionals as members, we have access to extensive information on all liver diseases and treatments.

Information leaflets are available explaining each specific liver disease; please telephone for details.

We support each other in our free newsletter, at our meetings and via our helpline. To join our mailing list and receive your own free copy of our bi-monthly newsletter which lists all the meeting times and local helpline numbers.

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Autoimmune Hepatitis

WHAT IS CHRONIC ACTIVE HEPATITIS (AUTOIMMUNE HEPATITIS)?

Autoimmune Hepatitis (AIH) is a rare disorder which predominantly affects females. The old name for this disease was lupoid hepatitis (immune system attacking like a 'wolf'). The majority of cases can be controlled by immunosuppressive agents such as steroids. It is thought to be due to the immune system mistakenly attacking liver cells- resulting in inflammation and damage within the liver (hepatitis). If this is untreated it is progressive and can lead to end-stage liver failure and death; however it must be emphasized that this liver condition, once recognised and assessed can be well-treated. Most patients with this condition require lifelong treatment. The estimated prevalence in Northern Europe is approximately 170 cases per million, with a female to male ratio of 4 to 1. Most often it occurs around the ages of 15-25 years and 45-55 years. It may occur in conjunction with other 'autoimmune' conditions such as thyroid disease (like Graves disease).

WHY OR HOW DO I GET AIH AND WHAT IS THE COURSE OF AIH?

AIH results from a breakdown of self-tolerance (ie the body's immune system thinks the liver is 'foreign' (not self) and directs attack against various parts of the liver cell as if they were an infection. The cause of AIH and other 'autoimmune-mediated' diseases is not fully understood. One theory is that a 'triggering event' (such as a minor infection or exposure) in a 'primed' individual (genetically susceptible person- someone who may have set of genes that interact to cause the immune system to perhaps over-react) may then lead to disease. Perhaps differing triggers in different circumstances lead to different autoimmune conditions in a susceptible person or family. This is an attractive though simplistic idea. AIH is not due to a virus and is not transmissible. Genetic associations have been reported, the strongest association by far is to female sex (ie chromosome X); interestingly, this applies to most autoimmune diseases. Other genes which control the type, specificity and

magnitude of the immune system response (tissue typing genes called the HLN MHC) have been shown to be more common in AIH patients. It must be emphasised that these are not 'bad' genes but certain sets of genes that everyone has to control their immune system. Rarely drugs can precipitate abnormal liver tests with 'inflammatory' immunoreaction. Best characterised is the autoimmune 'lupus-like' syndrome induced rarely in young people taking minocycline (an antibiotic for acne). This resolves after withdrawal but presents with a very similar constellation of signs.

In most cases AIH presents over several months with non-specific symptoms (see below), however occasionally it presents acutely. Untreated AIH has a poor prognosis- with reported five and ten year survival rates of only 50% and 10% respectively. With treatment, albeit life-long, patients can lead a normal life with virtually normal life expectancy. Some patients (a minority) require assessment and consideration for liver transplantation if inflammation has resulted in end-stage cirrhosis.

What are the symptoms?

The symptoms depend on the inflammatory activity of the disease. The most common symptom is fatigue and a general feeling of ill-health (malaise). Fatigue may be the only symptom and can be indolent and difficult to quantify. It can manifest in many ways and there are many contributing factors to an individual's perception of the symptom of fatigue. Some patients complain of joint pains (arthralgia) or muscular tenderness (myalgia). Other symptoms are less common, are manifest in severe disease, and are a result of significant liver damage (cirrhosis). These symptoms are nausea, abdominal pain or bloating symptoms, decreased appetite and peripheral swelling. There may be muscle wasting, easy bruising and the development of fluid in the abdomen (ascites) or varices ('varicose veins' in the upper stomach which can bleed profusely). Jaundice (yellowing of the sclera of the eye) is a rare though important manifestation of severe disease. Abdominal pain is not a common feature in AIH.

How is AIH diagnosed?

General fatigue can have many causes, your doctor should ask some questions to ascertain the cause and severity of fatigue. If cirrhosis has occurred from inflammation these signs will be apparent. Blood tests such as liver function tests are crucial in demonstrating inflammation (particularly the ALT or AST). As AIH is an autoimmune condition, autoantibodies are often present- most commonly those termed SMA (smooth muscle autoantibody) and ANA (anti-nuclear autoantibody). There are different sub-types of AIH depending on the type of molecule the immune system is reacting abnormally against (types I to 3); however these classifications serve only minor clinical importance.

On many occasions abnormalities of liver function and or positive autoantibodies are found 'in passing'- if these remain so they require assessment by a liver specialist (hepatologist). Signs on clinical examination consistent with liver disease always require referral and assessment by a specialist.

A hepatologist will take a full clinical history, examination and further blood tests. An ultrasound scan excludes conditions such as gallstones. A liver biopsy is the gold standard for determining the cause of abnormal liver tests and diagnosis of AIH.

What else could it be?

1. Infectious hepatitis if acute presentation (minority)
2. Chronic Hepatitis C / B: (until specific /sensitive tests for these viruses became widely available many cases were mis-diagnosed)
3. Any cause of cirrhosis (link).
4. Drugs: prescription/ non-prescription.
5. Part of a multi-system disease.
6. Primary Biliary Cirrhosis (PBC) is another probable autoimmune disease that results in damage to the cells that line the drainage ducts of the liver (bile ducts). A very small proportion of patients can 'crossover' ie have features of both conditions.