What is Autoimmune Hepatitis?

Autoimmune Hepatitis (AIH) is a chronic disease in which one's immune system attacks normal liver cells because it mistakes them for foreign agents such as viruses or bacteria. The immune system creates antibodies, and these are what cause damage.

It is not caused by a virus, nor is it contagious.

The attack on the liver cells causes the liver to become inflamed.

AIH is usually treated with a combination of drugs to bring down the inflammation and swelling and a drug to suppress the immune system.

With proper treatment, most people can live well for many years with AIH. If untreated, AIH can lead to cirrhosis or liver failure. In these cases, a liver transplant maybe an option.

AIH can be a deadly disease that appears to affect individuals of color disproportionately, particularly females.

Not only is the disease more severe, but it also requires more intensive treatment and leads to liver transplantation more often than in other counterparts.

The incidence of AIH is approximately 1 per 100,000 people; thus, it is considered to be a rare disease.

AIH tends to affect females more than males and can occur at any age, in any ethnic group. There are two main forms of the disease. Type 1 is more common and affects mostly adults. It is often associated with other autoimmune diseases. Type 2 is less common and occurs more often in children.

What causes AIH?

While the cause is not clearly defined, field experts hypothesize that there are both genetic and an environmental element underlying autoimmune hepatitis.

The immune system is a very complex system of checks and balances, normally functioning to help protect our body from many types of infections.

When the balance is disrupted, cells within this system can attack and injure healthy cells in the same body.

The genetic susceptibility to autoimmune conditions is not from just one abnormal gene, but likely a combination of genetic alterations establishing the perfect environment for autoimmune conditions to occur.These genetic traits can be inherited; thus it is common to see autoimmune diseases in families. In autoimmune hepatitis, genetic risk factors in combination with an environmental trigger, such as extreme stress, some viruses, or various medications or chemical exposures occurs to trigger the immune system the body's white blood cells begin to attack the liver, causing inflammation and tissue damage. This uncontrolled inflammation is referred to as hepatitis, a general term which means "inflammation of the liver." Prolonged or severe inflammation can result in fibrosis or cirrhosis – scar tissue in the liver. Known environmental triggers of autoimmune hepatitis include medications such as statins, minocycline, and nitrofurantoin; and viruses including Epstein Barr, cytomegalovirus, and measles. Unfortunately, researchers believe we still only know some of the risk factors for autoimmune hepatitis.

What are the symptoms associated with AIH?

The initial and long-term symptoms associated with autoimmune hepatitis are commonly different among patients. There is a wide spectrum of complaints in patients with autoimmune hepatitis, as some will have no symptoms at all and others may have debilitating complaints. It may be normal to have variable symptoms as well, as patients with autoimmune hepatitis can have both "good and bad days". It remains unclear why these symptoms are present in some and not in others.

The following is a list of symptoms reported to be associated with AIH:

Fatigue and excessive sleepiness

Joint pain

Nausea and vomiting

Loss of appetite

Pain in the liver area (right upper quadrant of the abdomen) or in the abdomen

Itching (pruritus)

Skin rash

Dark colored urine

Light colored stools

Jaundice (yellowing of the skin and whites of the eyes)

Women may experience the loss of menstruation

How is AIH Diagnosed?

In general, a health provider diagnoses Autoimmune Hepatitis based on a patient's symptoms, a physical examination, blood tests, and a liver biopsy. During the examination a history of alcohol and drug use (both prescribed and other drugs) is done to rule out other possible causes of liver disease and to see if any drugs have been used that are toxic to the liver. Health history, during the exam, will help the provider learn about prior viral or bacterial infections, other autoimmune diseases the patient may have, and family health history.

The blood tests that are done help the provider determine if autoantibodies are present and if there are elevated liver enzyme numbers. The provider is attempting to rule out other liver diseases or reasons for the patient's symptoms. If the patient presented with no symptoms, additional blood testing may need to be done. Blood tests are usually done in the provider's office or in a lab.

A liver biopsy is performed to see if there is any scarring and/or cirrhosis of the liver. The liver biopsy is usually done under local anesthesia, with sedatives and pain medicine as necessary. During the biopsy procedure, a needle is placed into the liver that removes a small piece of liver tissue. This tissue is then examined by a pathologist to determine if any scarring and/or cirrhosis is present.

How is AIH treated?

The goals of treatment are to quickly reduce the inflammation and suppress the immune system to stop the damage to the liver and prevent further damage. Then in most cases, the liver will begin to regenerate, growing new healthy cells. Treatment typically begins with a fast-acting steroid such as prednisone or budesonide at a high dosage. As symptoms improve and once the liver enzymes approach normal levels, the steroid is reduced, and a second medication is usually added - an immunosuppressant such as azathioprine. The immunosuppressant allows the steroid to be reduced to a very low dosage or eliminated to avoid side effects.

While some patients can be weaned from both medications and remain in remission, most patients will eventually relapse, so Autoimmune Hepatitis is considered to be a chronic illness - one that must be monitored closely and treated long term, even for life. A small number of patients either do not respond adequately to standard treatments, or present with so much liver damage at the time of diagnosis that a liver transplant is necessary. Survival rates for liver transplantation are very high, and patients typically enjoy a good quality of life. There are also many alternative drugs that are being used for the treatment of AIH so if azathioprine and/or Prednisone don't seem to be working, your liver specialist will have others to try.

What outcomes may be expected?

AIH is a chronic disease, so at this point it can't be cured, but it can be controlled. Most of us will respond well to the treatment with a steroid and an immunosuppressant and will go into remission. We will have normal life spans. Once in remission, the steroid can often be eliminated or reduced to a very low dosage. Most will have to stay on an immunosuppressant because without it, relapse will occur. The great majority of us can live fairly normal lives while our AIH is controlled by medications. For those who don't respond to medications, their scarring may progress to cirrhosis and end-stage liver failure and for them, liver transplantation is an option. There is hope that through research, new treatments will be discovered, giving more options for treatment of AIH.