

AUTOIMMUNE HEPATITIS

What is Autoimmune Hepatitis?

Autoimmune hepatitis or autoimmune chronic hepatitis is a progressive inflammation of the liver that has been identified by a number of different names, including autoimmune chronic active hepatitis (CAH), idiopathic chronic active hepatitis, and lupoid hepatitis. The reason for this inflammation is not certain, but it is associated with an abnormality of the body's immune system and is often related to the production of antibodies that can be detected by blood tests.

Autoimmune hepatitis was first described in 1950 as a disease of young women, associated with increased gamma globulin in the blood and chronic hepatitis on liver biopsy. The presence of antinuclear antibodies (ANA) and the resemblance of some symptoms to "systemic lupus erythematosus" (SLE) led to the label "lupoid hepatitis." It later became evident that this disease was not related to SLE. The disease is now called autoimmune hepatitis.

What are the Symptoms?

The typical patient with autoimmune hepatitis is female (70%). The disease may start at any age, but is most common in adolescence or early adulthood. Blood tests identify ANA or smooth muscle antibodies (SMA) in the majority of patients (60%). More than 80% of affected individuals have increased gamma globulin in the blood. Some patients have other autoimmune disorders such as thyroiditis, ulcerative colitis, diabetes mellitus, vitiligo (patchy loss of skin pigmentation), or Sjogren's syndrome (a syndrome that causes dry eyes and dry mouth). Other liver diseases such as viral hepatitis, Wilson's disease, hemochromatosis, and alpha-1-antitrypsin deficiency should be excluded by appropriate blood tests, and the possibility of drug-induced hepatitis is ruled out by careful questioning.

The most common symptoms of autoimmune hepatitis are fatigue, abdominal discomfort, aching joints, itching, jaundice, enlarged liver, and spider angiomas (tumors) on the skin. Patients may also have complications of more advanced chronic hepatitis with cirrhosis, such as ascites (abdominal fluid) or mental confusion called encephalopathy. A liver biopsy is important to confirm the diagnosis and provide a prognosis. Liver biopsy may show mild chronic active hepatitis, more advanced chronic active hepatitis with scarring (fibrosis), or a fully developed cirrhosis.

How is Autoimmune Hepatitis Treated?

The 10-year survival rate in untreated patients is approximately 10%. The treatment of autoimmune hepatitis is immunosuppression with prednisone alone or prednisone and azathioprine (Imuran). This medical therapy has been shown to decrease symptoms, improve liver tests, and prolong survival in the majority of patients. Therapy is usually begun with prednisone 30 to 40 mg per day and then this dosage is reduced after a response is achieved.

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The standard dosage used in the majority of patients is prednisone 10-15 mg per day, either alone or with azathioprine 50 mg per day. Higher doses of prednisone given long-term are associated with an increase in serious side effects, including: hypertension, diabetes, peptic ulcer, bone thinning, and cataracts. Lower doses of prednisone may be used when combined with azathioprine.

The goal of treatment of autoimmune hepatitis is to cure or control the disease. In two thirds to three quarters of the patients, liver tests fall to within the normal range. Long-term follow-up studies show that autoimmune hepatitis appears more often to be a controllable rather than a curable disease, because the majority of patients relapse within six months after therapy is ended. Therefore, most patients need long-term maintenance therapy.

Not all patients with autoimmune hepatitis respond to prednisone treatment. Approximately 15-20% of patients with severe disease continue to deteriorate despite initiation of appropriate therapy. This is most common in patients with advanced cirrhosis on initial liver biopsy. Such patients are unlikely to respond to further medical therapy, and liver transplantation should be considered.

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