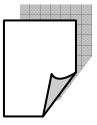
Summary and Conclusion

Autoimmune liver diseases include; autoimmune hepatitis, primary biliary cirrhosis and primary sclerosing cholangitis.

The exact etiology and pathogenesis of these diseases is still unknown. However , the most plausible explanations are that these autoimmune diseases develop in individuals with genetic predisposition when exposed to the appropriate environmental triggers which could be bacterial, viral, chemicals or drugs. This hypothesis is supported by several evidences including association of these diseases with HLA haplotypes, presence of humeral and cellular immune response abnormalities, impaired regulation of both B &T lymphocytes, association with other autoimmune diseases. AILDs have varying symptoms in different individuals.

Autoimmune hepatitisis one of the few liver diseases with excellent response to therapy. It is classified into 2 types according to serum autoantibody profile, type1 is the most common worldwide, type 2 is the variant common in children with more severe form of the disease and worse prognosis.

There is significant heterogeneity in its presentation that may mask its identity, affect its clinical behavior, and confound its management. It may start with a fulminant course, and the diagnosis should not be overlooked when dealing with patientswith acute liver failure. Alternatively, it may behave as a slowly progressing disease, and it is still controversial whether those patients need immunosuppressive treatment at all.



Diagnosis of AIH is made by a combination of clinical, biochemical, serological, and histological criteria and exclusion of other forms ofchronic liver disease.The scoring system using simplified criteria can also be helpful for establishing the diagnosis.

Most patients are well controlled with corticosteroids and zathioprine. here is no prescribed minimum or maximum duration of treatment. Some patients however, require alternative therapy because of refractory disease, poor compliance,or drug intolerance; the most promising are cyclosporine, mycophenolate , budesonide and tacrolimus.

Management, however, still faces several other important issues, such as in children, the elderly, in males, and during the preconception period, pregnancy, and lactation.

The key to successful management is recognizing the non-classical presentations and individualizing therapy.

Liver transplantation is the treatment of choice for AIH patients

who had end-stage decompensated liver diseases or HCC.

Recurrence of AIH is common in the graft after liver transplantation, and these patients normally require low-dose steroids.

The increased incidence of **Primary biliary cirrhosis** over recent decades can be attributed to augmented testing of liver biochemistry rather than a rise in disease incidence.

The diagnosis of PBC should be considered in any patient, particularly a woman, Who complains of unexplained itching, fatigue, jaundice, or

Unexplained Weight loss with right-upper-quadrant discomfort, and/or whose serum ALP is highly elevated. Such patients should be asked about diseases frequently associated with PBC.

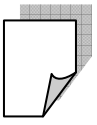
AMAs remain the hallmark of diagnosis in most cases and allow detection of asymptomatic patients. AMA test should beconsidered once the diagnosis of PBC is suspected. Anti-gp210 and anti-sp100 are highly specific for PBC.

UDCA at early stages of the disease is the only widely accepted medical treatment aiming at modifying the natural history of the Disease.

Methotrexate and Colchicine have not demonstrated a benefit in altering the natural history of PBC in most studies, but as they are generally safe, they are continued to be added to UDCA in patients not responding to UDCA alone. The prognosis of PBC has improved because of early diagnosis and use of UDCA.

Liver transplantation remains the only effective treatment option for patients with advanced liver disease due to PBC. PBC may recur in the allograft following liver transplantation.

Primary sclerosing cholangitis is strongly associated with IBD, mostly UC and the disease should be considered in patients who had otherwise unexplained abnormal liver biochemical tests, particularly an elevation in serum ALP.



The diagnosis of PSC is established by the demonstration of characteristic multifocal stricturing and dilation of intrahepatic and/or extrahepatic bile ducts on cholangiography and exclusion of other disorders that can produce a similar radiologic picture.

CCA is the most feared complication of PSC which is currently the leading cause of death in 7 to 15% of patients with a mean survival 5-7 months.

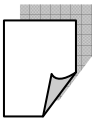
Routine screening for CCA in patients with PSC is not recommended as no benefits in patient outcomes have been demonstrated in different studies.

Patients with PSC and IBD have an increased risk of colon cancer, and should be enrolled in colonoscopic surveillance programs.

There is still no evidence supporting a specific medical therapy capable of halting disease progression in PSC.Furthermore, a 2010 guideline issued by the AASLD recommends against use of UDCA at any dose. Thus, a role for any medical therapy is unproven.

At present, the management of PSC remains focused on treating the symptoms and complications that are associated with disease progression as well as on close surveillance of those patients with advanced liver disease.

Liver transplantation is currently the only definitive form of therapy for patients with end-stage liver disease due to PSC. However disease recurrence can be a source of morbidity and mortality as transplanted patients survive longer.



Autoimmune pancreatitis is a rare disease of presumed autoimmune etiology. As T-regseem to take important roles in progression as well as induction of the disease, further studies are necessary to clarify the pathogenesis including genetic background , disease specific antigens, and the role of IgG4.

AIP should be considered in the differential diagnosis of patients presenting with varied pancreatic and hepatobiliary manifestations.

AIP mimics pancreatic cancer and it is critically important to distinguish between the two diseases.

A pancreatic biopsy is required to establish the diagnosis, although it is also supported by elevated serum levels of IgG4, characteristic radiologic findings and response to corticosteroids.

The diagnostic criteria proposed by the Mayo Clinic (the "HISORT" criteria) are most commonly used in USA to establish the diagnosis of AIP.

There is often a dramatic response to steroids although there are regional practice variations.

Relapses are quiet common and may need a prolonged course of steroids.