

Autoimmune Hepatitis:

Learning outcomes:

1. LO1: Definition of Autoimmune hepatitis (AIH)
2. LO2: Clinical presentation of AIH
3. LO3: Differential diagnosis of AIH
4. LO4: Diagnosis of AIH
5. LO5: Subclassification of AIH
6. LO6: Treatment of AIH

LO1: Definition:

- ▶ Self-perpetuating hepatocellular inflammation of unknown cause
- ▶ Characterized by the presence of:
 - periportal hepatitis
 - Hypergammaglobulinemia
 - Serum liver-associated autoantibodies
- ▶ Exclusion of other chronic liver diseases
- ▶ 40% of patients with untreated severe disease die within 6 mos of dx
- ▶ 40% develop cirrhosis
 - 54% develop esophageal varices
 - ▶ 20% die of hemorrhage
- ▶ An acute onset of illness is seen in 40% patients
- ▶ Prednisone and azathioprine are mainstay of treatment

LO2: Clinical Manifestations:

- ▶ Symptoms
 - Fatigue 85%
 - Jaundice 77%
 - Abdominal pain 48%
 - Pruritus 36%
 - Anorexia 30%

- Polymyalgia 30%
- Diarrhea 28%
- Fevers 18%

▶ Physical Findings

- Hepatomegaly 78%
- Jaundice 69%
- Splenomegaly 32%
- Spider nevi 58%
- Ascites 20%
- Encephalopathy 14%
- Concurrent immune disease 48%

▶ Laboratory features

- Elevated AST 100%
- Hypergammaglobulinemia 92%
- Increased immunoglobulin G level 91%
- Hyperbilirubinemia 83%
- Alkaline Phosphatase >2x 33%

LO3: Differential Diagnosis:

- ▶ **Wilson's disease**
- ▶ **A1AT deficiency**
- ▶ **Hemochromatosis**
- ▶ **Viral hepatitis**
- ▶ **Drug induced hepatitis**

LO4: Diagnostic Criteria:

- ▶ Clinical criteria
 - Presence of characteristic clinical features
 - Liver histology

- Exclusion of other diseases

LO5: Subclassification of AIH:

- ▶ Type I
- ▶ Type II
- ▶ Type III

LO4&LO5: Type 1 AIH:

- ▶ Diagnostic autoantibodies: ANA, ASMA
- ▶ Age: (10-20 and 45-70)
- ▶ Women
- ▶ Concurrent immune diseases
- ▶ Elevated gamma globulin: +++
- ▶ Steroid responsiveness: +++
- ▶ Progression to cirrhosis (%): 45

LO4&LO5: Type II AIH:

- ▶ Diagnostic autoantibodies: LKM1
- ▶ Age: Pediatric (2-14), rare adults (4%)
- ▶ Women
- ▶ Concurrent autoimmune disease:
- ▶ Elevated gamma-globulins: +
- ▶ Steroids responsive: ++
- ▶ % progression to cirrhosis: 82

LO4&LO5: Type III AIH:

- ▶ Diagnostic autoantibodies: SLA
- ▶ Age: adults (30-50)
- ▶ Women
- ▶ Concurrent autoimmune disease
- ▶ Elevated gamma-globulin: ++
- ▶ Steroid responsive: +++
- ▶ % progression to cirrhosis: 75

Recommendations:

- ▶ Diagnosis of AIH requires aminotransferase and globulin levels; detection of ANA +/- SMA, anti-LKM1; and histology
- ▶ Diagnostic criteria for AIH should be applied to all patients

LO6: Indications for Treatment:

Absolute	Relative
Serum AST > 10x uln	Symptoms (fatigue, arthralgia, jaundice)
Serum AST > 5x uln and globulin > 2x nl	Serum AST and globulin less than absolute criteria
Bridging necrosis or multiacinar necrosis	Interface hepatitis

	Prednisone only	Combination (Pred + AZA)
Week 1	60 mg	30 mg+50 mg
Week 2	40 mg	20 mg+50 mg
Week 3	30 mg	15 mg+50 mg
Week 4	30 mg	15 mg+50 mg
Maintenance until endpoint	20 mg	10mg+50 mg

LO6: Reasons for Preference:

- ▶ **Prednisone**
 - **Cytopenia**
 - **Pregnancy**
 - **Malignancy**
 - **Short course <6 months**

► **Combination**

- **Postmenopausal state**
- **Osteoporosis**
- **Brittle diabetes**
- **Obesity**
- **Acne**
- **Emotional Liability**
- **Hypertension**