

Severe Anemia and Coagulopathy in a Young Woman with Autoimmune Hepatitis: A Case Report

Aleena S Peter^[1], H S Pooja^[2], Dr. S. Vinod Naik^[3], Dr. Dumthi Namratha^[3]
*Pharm D Intern^[1], V Pharm D^[2] and Assistant Professors^[3], Department of Pharmacy Practice, Togari
Veeramallapa Memorial College of Pharmacy Ballari, Karnataka – 583104
Corresponding Author: Aleena S Peter***

Date of Submission: 07-03-2026

Date of Acceptance: 19-03-2026

Abstract

Background:

Autoimmune hepatitis (AIH) is a progressive immune-mediated liver disorder characterized by interface hepatitis, circulating autoantibodies, hyperglobulinemia, and potential progression to cirrhosis with portal hypertension complications like gastrointestinal bleeding in young adults.

Objective:

- To document therapeutic challenges and pharmacist interventions in managing AIH
- Rational treatment

Case Summary:

A 29-year-old female with established AIH presented with black tarry stools, epistaxis with icterus positive. Laboratory investigations showed profound anemia (hemoglobin 7.0 g/dL), hyperbilirubinemia (total bilirubin 19.8 mg/dL), markedly elevated alkaline phosphatase (1297 U/L), hypoalbuminemia, weak ANA positivity, splenomegaly, and positive direct Coombs test. The patient was started T. Prednisolone 10mg (BD), Inj. Vitamin K 10mg (OD), Inj. Tranexamic acid 500 mg (BD) and other supplements. Hemoglobin was improved to 9.1 g/dl over 16 days.

Methodology:

Prospective observational

Result:

- Diagnosis was confirmed as Autoimmune Hepatitis associated with Melena
- Progressive hemodynamic stability, hemoglobin improvement (9.1 g/dL), declining bilirubin and ALP levels, and resolution of acute symptoms was enabled on discharge.

Conclusion:

This AIH decompensation case from GI bleed in a young adult stresses prompt stabilization, anemia correction, immunosuppression, and pharmacist-led regimen reviews to prevent errors and optimize care.

Keywords: Autoimmune Hepatitis, Melena, Immunosuppression, Pharmacotherapy Optimization.

I. INTRODUCTION

Autoimmune hepatitis (AIH) is a rare immune-mediated liver disorder. It is a chronic and usually progressive form of hepatitis with an unknown cause. The condition can affect both children and adults of any age. In some cases, the disease may follow a fluctuating course, with periods of increased activity alternating with periods of reduced activity⁽¹⁾⁽⁷⁾. It is a necro-inflammatory liver disorder that typically presents with a fluctuating pattern of disease activity, the presence of circulating autoantibodies, elevated immunoglobulin G (IgG) levels, and/or a positive response to immunosuppressive therapy. The condition is characterized by significant heterogeneity, and no single clinical or biochemical test alone is sufficient to definitively diagnose AIH⁽²⁾.

There are two known types of autoimmune hepatitis⁽³⁾:

1. Type-1 is distinguished by the presence of anti-smooth muscle antibodies (ASMA) with or without anti-nuclear antibodies (ANA).
2. Type-2 autoimmune hepatitis presents with positive anti-liver/anti-kidney microsome (anti-LMK) type 1 antibodies or anti-liver cytosol (anti-LC) type 1 antibodies.

Autoimmune hepatitis (AIH) does not have a single pathognomonic feature for diagnosis. Instead, the diagnosis is established through a combination of characteristic histological findings, elevated liver enzymes, particularly aspartate aminotransferase (AST) and alanine aminotransferase (ALT), increased total IgG levels, and the presence of one or more circulating autoantibodies⁽⁶⁾.

Appropriate management can prolong survival, improve the quality of life, and avoid the need for liver transplantation, considerable therapeutic challenges remain in the treatment of this disorder⁽¹⁾.

Table 1. Classification of Autoimmune Hepatitis.

Variable	Type 1 Autoimmune Hepatitis	Type 2 Autoimmune Hepatitis
Characteristic autoantibodies	Antinuclear antibody* Smooth-muscle antibody* Antiactin antibody† Autoantibodies against soluble liver antigen and liver–pancreas antigen‡ Atypical perinuclear antineutrophil cytoplasmic antibody	Antibody against liver–kidney microsome 1* Antibody against liver cytosol 1*
Geographic variation	Worldwide	Worldwide; rare in North America
Age at presentation	Any age	Predominantly childhood and young adulthood
Sex of patients	Female in approximately 75% of cases	Female in approximately 95% of cases
Association with other autoimmune diseases	Common	Common§
Clinical severity	Broad range	Generally severe
Histopathologic features at presentation	Broad range	Generally advanced
Treatment failure	Infrequent	Frequent
Relapse after drug withdrawal	Variable	Common
Need for long-term maintenance	Variable	Approximately 100%

* The conventional method of detection is immunofluorescence.

† Tests for this antibody are rarely available in commercial laboratories.

‡ This antibody is detected by enzyme-linked immunosorbent assay.

§ Autoimmune polyendocrinopathy–candidiasis–ectodermal dystrophy is seen only in patients with type 2 disease.⁴⁷

Figure 01: Classification of Autoimmune Hepatitis ⁽¹⁾

EPIDEMIOLOGY

Autoimmune hepatitis (AIH) is a chronic necro-inflammatory liver disease of unknown etiology that can occur in both children and adults of any age ⁽²⁾. It is estimated to affect approximately 100,000–200,000 individuals in the United States, while the prevalence in India appears to be lower. Earlier reports even questioned the existence of autoimmune liver diseases. Despite advances in research, the exact prevalence, characteristics, and prognosis of autoimmune hepatitis remain incompletely understood ⁽⁵⁾. AIH is considered a global disease with varied clinical presentations, and its prevalence differs according to age, sex, ethnicity, and geographic location. The reported incidence ranges from 0.67 to 2.0 cases per 100,000 persons per year, while the prevalence varies between 4.0 and 24.5 cases per 100,000 people across different regions ⁽⁵⁾ ⁽⁶⁾. The condition predominantly affects women, with a male-to-female ratio ranging from 1:4 to 1:6 ⁽⁴⁾.

ETIOLOGY

There is no specific evidence of the cause. Sixty percent of patients have chronic hepatitis but without serologic evidence of a viral infection. The disease is associated with anti-smooth muscle autoantibodies⁽³⁾.

PATHOPHYSIOLOGY

- Autoimmune hepatitis postulates an environmental agent that triggers a cascade of T-cell-mediated events directed at liver antigens in a host genetically predisposed to this disease, leading to a progressive necro-inflammatory and fibrotic process in the liver ⁽¹⁾.
- Genetic predisposition, molecular mimicry and imbalance between effector and regulatory immunity are considered the primary pathogenic mechanisms for development of AIH.

- Several lines of evidence support the central role of T cell-mediated immunity in development of disease. Presentation of an autoantigenic peptide in conjunction with human leukocyte antigen (HLA) class II molecule to a naive CD4+ T helper cell (Th0) leads to differentiation into Th subsets, which produce cytokines involved in complex immunoregulation.
- In the presence of interleukin-12 (IL-12), Th0 differentiates into Th1 cells producing interferon (IFN)- γ and IL-2, the latter then binds to CD25 (important for function of regulatory T cells [Treg]).
- Th17 cells are involved in host defense and suppress Treg cells, and increased hepatic Th17 cells correlate with degree of inflammation and fibrosis in AIH. Activated Th1 and Th17 cells express C-X-C motif chemokine receptor-3 (CXCR3), which binds to its C-X-C motif chemokine ligands (CXCL) CXCL9 and CXCL10, and may influence progression of AIH via Th1-dependent mechanisms.
- CXCL10 has been suggested as a biomarker of hepatic inflammation and fibrosis and has been associated with disease severity. Furthermore, its serum level correlates with normalization of transaminases and decreased hepatic inflammation after successful steroid therapy in AIH patients⁽⁷⁾.

CLINICAL MANIFESTATION

Asymptomatic Disease

- The heterogenic nature of AIH causes a broad spectrum of clinical manifestations.
- It has been reported that about 25–34% of patients with AIH are asymptomatic; however, histological findings may be similar to symptomatic patients.
- A decreased 10-year survival has been reported in untreated asymptomatic patients compared to treated patients with more severe disease (67% versus 90%). The development of symptoms in this population can happen within 2–120 months (mean 32 months), but the non-appearance of symptoms should not delay treatment.
- Final diagnosis is often established after an investigation of unexplained elevated serum aminotransferases during routine testing. At diagnosis, about one third of adults and half

of children will have advanced liver disease, with the presence of cirrhosis⁽⁶⁾.

- The spectrum of presentation ranges from no symptoms to debilitating symptoms and even fulminant hepatic failure.
- Patients may present with nonspecific symptoms of varying severity, such as fatigue, lethargy, malaise, anorexia, nausea, abdominal pain, and itching. Arthralgia involving small joints is common.
- Physical examination may reveal no abnormalities, but it may also reveal hepatomegaly, splenomegaly, jaundice, and signs and symptoms of chronic liver disease⁽¹⁾.

Symptomatic Disease

- In symptomatic patients, easy fatigability is the main complaint (85%), and jaundice can be present.
- Chronic non-specific symptoms like malaise, arthralgias and amenorrhea often appear. As mentioned previously, in about one-third of patients, cirrhosis has developed by the time of diagnosis and physical signs of chronic liver disease may have appeared (splenomegaly, spider nevi, palmar erythema, caput medusa). In advanced stages, signs of portal hypertension such as ascites, esophageal varices, portal hypertensive gastropathy, cytopenias and hepatic encephalopathy can be seen.
- Although less common than in other chronic liver diseases, hepatocellular carcinoma (HCC) can develop in the context of cirrhosis, and surveillance is recommended. Symptoms of depression and anxiety are not uncommon and should be properly addressed to improve the health-related quality of life of these patients⁽⁶⁾.
- Patients with severe or fulminant symptoms accompanied by profound jaundice and a prolonged prothrombin time may have aminotransferase levels in the thousands. Many patients with an acute presentation have histologic evidence of chronic disease on liver biopsy, indicating that they probably have had subclinical disease for a long time. Long periods of subclinical disease may also occur after presentation.

One clue to diagnosing autoimmune hepatitis is the presence of other diseases with autoimmune

features, commonly thyroiditis, ulcerative colitis, type 1 diabetes, rheumatoid arthritis, and celiac disease. Occasionally, circulating anti-endomysial antibodies, anti-gluten antibodies, and anti-tissue transglutaminase antibodies may be found in patients with autoimmune hepatitis; this finding generally reflects the coexistence of celiac sprue and autoimmune hepatitis ⁽¹⁾.

DIAGNOSIS ⁽³⁾

As AIH displays a considerable heterogeneity, no single clinical or biochemical test may securely diagnose it. An exception may be the presence of anti-SLA/LP autoantibodies ⁽²⁾.

- A multi-pronged approach is used to make a diagnosis. This approach includes determining symptoms, laboratory tests, and biopsies, as no single diagnostic test is pathognomonic for autoimmune hepatitis.
- Marked elevation of serum transaminases (AST, ALT) and gamma-globulin is common; elevation in alkaline phosphatase is less common. The serum levels of AST, ALT, and gamma globulin reflect disease severity and immediate prognosis at presentation.
- The serologic markers required for the diagnosis of autoimmune hepatitis include antinuclear antibody (ANA), smooth muscle antibodies (SMA), and antibodies to liver-kidney microsome type 1 (anti-LKM1).
- Indirect immunofluorescence detects ANA, SMA, and anti-LKM1. The diagnostic

accuracy, specificity, and sensitivity of these markers are 74%, 99%, and 43%, respectively. Anti-liver cytosol type I, anti-soluble liver antigen (SLA) antibodies, and perinuclear antineutrophil cytoplasmic antibodies (pANCA) can also be associated with autoimmune hepatitis.

- Conversely, anti-mitochondrial antibodies are more commonly seen with primary biliary cirrhosis and are usually absent in autoimmune hepatitis; however, they can be present in those with overlapping syndromes.
- Atypical perinuclear antineutrophil cytoplasmic antibodies are commonly associated with type-1 autoimmune hepatitis and primary sclerosing cholangitis.
- Anti-LKM1 is common in type 2 AH autoimmune hepatitis and is mainly observed in children.
- Anti-SLA antibodies are more useful from a prognostic standpoint as these are associated with more severe disease, treatment failure, and a higher relapse rate. Liver biopsy is required for both diagnosis and staging of autoimmune hepatitis.

Differential Diagnosis ⁽³⁾

- Primary biliary cirrhosis
- Primary sclerosing cholangitis
- Hepatitis A
- Hepatitis B
- Hepatitis C
- Hepatitis D
- Hepatitis E

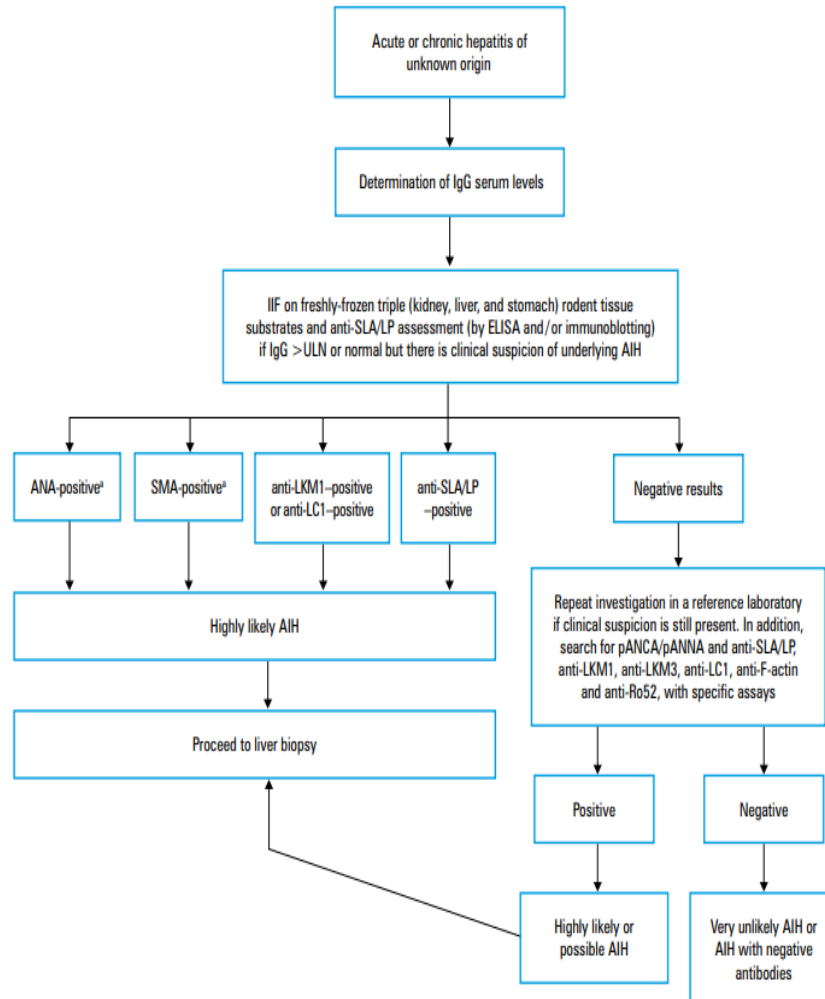


Figure 02: Diagnosis of AIH ⁽⁴⁾

TREATMENT

Standard treatment: Steroid induction therapy is the first-line treatment option, followed by maintenance therapy using a steroid-sparing agent. In this context, prednisolone alone, or more frequently in association with azathioprine, has been considered for more than 40 years as the first-line treatment for AIH. Initial nonresponse should immediately raise doubts about AIH diagnosis or indicate problems with adherence to treatment ⁽⁴⁾.

Table 4. Drugs Used in the Treatment of Autoimmune Hepatitis in Adults and Children.

Drug	Initial Therapy	Maintenance Therapy	Comments
Prednisone or prednisolone	Used as monotherapy in adults (20–60 mg/day) and children (1–2 mg per kilogram of body weight/day); also used in combination therapy in adults (15–30 mg/day) and children (1–2 mg/kg/day) with azathioprine or mercaptopurine	Used as monotherapy in adults (5–15 mg/day) and children (1 mg/kg/day); also used in combination therapy in adults (5–10 mg/day) and children (0.5–1.0 mg/kg/day) with azathioprine or mercaptopurine	Relatively contraindicated in patients with osteoporosis, diabetes mellitus, glaucoma, cataracts, arterial hypertension, major depression, and femoral avascular necrosis; reduced doses may work; use of budesonide under investigation ⁷⁷
Azathioprine	Used in combination with prednisone or prednisolone in adults (50–100 mg/day) and children (1.5–2.0 mg/kg/day)	Used as monotherapy in adults (50–200 mg/day) and children (1.5–2.0 mg/kg/day); also used in combination therapy in adults (50–150 mg/day) and children (1.5–2.0 mg/kg/day)	Contraindicated in patients with homozygous thiopurine methyltransferase deficiency; relatively contraindicated in patients with heterozygous thiopurine methyltransferase deficiency, cancer, or cytopenia, and pregnant patients
6-Mercaptopurine	May be substituted for azathioprine in combination therapy in adults (25–100 mg/day) and children (0.75–1.0 mg/kg/day)	Used as monotherapy in adults (25–100 mg/day) and children (0.75–1.0 mg/kg/day); also used in combination therapy in adults (25–100 mg/day) and children (0.5–1.0 mg/kg/day)	Contraindicated in patients with homozygous thiopurine methyltransferase deficiency; relatively contraindicated in patients with heterozygous thiopurine methyltransferase deficiency, cancer, or cytopenia, and pregnant patients
Cyclosporine	Sometimes used as monotherapy in children ⁷⁸ ; sometimes used as an alternative drug in adults with treatment-refractory disease	Sometimes used as an alternative drug in adults with treatment-refractory disease	Once remission achieved in children, maintenance therapy initiated with a combination of prednisone and azathioprine ⁷⁸ ; role of tacrolimus in place of cyclosporine not established
Mycophenolate mofetil	Sometimes used in patients with treatment-refractory disease or in patients with adverse drug reactions to or intolerance of azathioprine, mercaptopurine, or both	Sometimes used in patients with treatment-refractory disease or in patients with adverse drug reactions to or intolerance of azathioprine, mercaptopurine, or both	Role of mycophenolate mofetil, methotrexate, and cyclophosphamide not established
Ursodiol	Sometimes used in combination with prednisone, azathioprine, or both	Sometimes used in combination with prednisone, azathioprine, or both	Role of ursodiol not established

Figure 03: Treatment of Autoimmune Hepatitis in Children and Adult⁽¹⁾

II. CASE PRESENTATION

A 29 years old female patient was admitted to Ballari Medical College and Research Centre (BMC&RC), Ballari, Karnataka with chief complaints of blackish discoloration of stool since 1 day, foul smelling and watery and bleeding from nose since 1 day. On day 4 the patient was advised to check for Direct Coombs Test were it was found to be positive. SOCIAL HISTORY- No habits

PAST HISTORY- K/C/O Autoimmune hepatitis since 1 year.

FAMILY HISTORY – Nothing significant
 ON EXAMINATION-

- BP- 80/50mmHg
- PR- 90bpm
- SPO2- 95% decreased at RA
- RR- 16cpm
- PICCLE- Icterus positive
- Patient was conscious and oriented

- S1 S2 heard
- RS B/L NVBS positive
- P/A For soft and non-tender

LABORATORY INVESTIGATIONS

PARAMETER	RESULT	RESULT	RESULT	RESULT	REFERENCE RANGE
Haemoglobin	7.0	7.9	7.8	9.1	12-16g/dl
RBC	2.71	2.77	2.91	2.80	3.5-5.0 x 10 ⁶ /mm ³
PCV	22.7	24.4	25.6	26.7	33-43%
MCV	83.8	88.1	68.0	69.0	76-100fl
MCH	25.9	28.5	26.7	24.9	27-33pg/cell
MCHC	30.9	32.4	30.4	34.1	33-37g/dl
WBC	12280	9200	13350	12500	4500-11000cells/cumm
Polymorphs	85	80	85	81	40-75%
Lymphocytes	10	16	10	10	20-50%
Platelets	1.04	1.14	0.87	1.25	1.5-4.5lacs/cumm
RDW-CV	18.7	21.0	19.0	20.7	11.5-14.5%
Serum Urea	50	12	20	21	20-50 mg/dl
AST	101	68	37	36	0-35 U/L
ALT	43	16	13	14	0-35 U/L
ALP	1297	729	458	324	30-120 U/L
Total Bilirubin	19.8	14.9	19.7	17.3	0.1-1 mg/dl
Direct Bilirubin	11.7	10.1	12.0	10.2	0-0.2 mg/dl
Indirect Bilirubin	8.1	11.7	7.7	7.1	0.1-0.8 mg/dl
Total Protein	5.7	4.9	5.0	-	6-8.3 g/dl
Albumin	2.0	1.5	3.0	-	3.2-5.4 g/dl
Globulin	3.7	3.4	2.0	-	2.5-3 g/dl
A/G Ratio	0.5	4.0	1.3	-	1.2-1.5

OTHER TEST ORDERED:-

LDH:- 725 IU/L (225-450 IU/L).

Peripheral Smear:- Dimorphic Anemia.

ANA:- Weak Positive.

Serum Ferritin:- >2000 ng/ml (11-307 ng/ml).

Serum Folic Acid :- 1267ng/ml (2.5-20 ng/ml).

CRP:- 69.7mg/L (0-6 mg/L).

Prothrombin time:- 13.4 sec (10-12 sec).

HBs AG:- Negative.

HCV:- Negative.

HIV-Antibody:- Non Reactive.

Direct Coombs Test:- Positive.

USG - Abdomen:- Mild Splenomegaly

Diffuse Liver Disease

MRI Cholangiopancreatography:- Marked Splenomegaly

Mild Hepatomegaly

ECG:- Sinus Trachycardia.

TREATMENT-

SL No.	NAME OF DRUG	DOSE	ROUTE	FREQUENCY	DAYS
1.	Inj. Ceftriaxone	1g	IV	1-0-1	D1-D18
2.	Inj. Pantoprazole	40mg	IV	1-0-1	D1-D18
3.	Inj. Vitamin K	10mg	IV	1-0-0	D1-D3
4.	Inj. Tranexamic Acid	500mg	IV	1-0-1	D1-D18

5.	T. Ursodeoxycholic Acid	300mg	PO	1-0-1	D11-D18
6.	Syp. Lactulose	10ml	PO	1-1-1	D1-D18
7.	Inj. 25% Dextrose	100ml	IV	1-1-1	D1-D18
8.	Inj. Levetiracetam	500mg	IV	1-0-1	D1-D18
9.	T. Prednisolone	10mg	PO	1-0-1	D1-D18
10.	1 Pint PRBC		IV	Every 4 hour	D1-D3
11.	Inj. MVI+ NS	100ml NS	IV		D4 & D5
12.	Inj. Noradrenaline	2amp in 100ml NS	IV	1-0-1	D1-D18
13.	Fresh Frozen Plasma	4 pint	IV		D8
14.	Inj. Cefotaxime	1gm	IV	1-0-1	D11-D18
15.	T. Rifaximin	550mg	PO	1-0-1	D11-D18
16.	T. Spironolactone	25mg	PO	1-1-0	D11-D18
17.	Inj. Cefoperazone + Sulbactam	1.5gm	IV	1-0-1	D11-D18
18.	Inj. Metronidazole	100ml	IV	1-1-1	D11-D18
19.	Syp. Ambroxol	10ml	PO	1-0-1	D5-D10

III. DISCUSSION

- Autoimmune hepatitis (AIH) is a chronic immune-mediated inflammatory liver disease that predominantly affects young females and may progress silently until cirrhosis or decompensation occurs.
- In this case, a 29-year-old woman with known AIH presented with upper gastrointestinal bleeding manifested by melena and epistaxis, along with jaundice and hypoalbuminemia, indicating advanced hepatic dysfunction and portal hypertension.
- Although transaminase elevations were modest, the presence of cholestatic features, hyperbilirubinemia, splenomegaly, and coagulation abnormalities suggested significant underlying fibrosis.
- Laboratory findings showed borderline antinuclear antibody positivity, elevated inflammatory markers, and negative viral hepatitis screening, supporting an autoimmune etiology. The patient also exhibited anemia with mixed morphology and elevated ferritin levels with positive Coombs testing, suggesting possible hemolysis and altered iron metabolism during ongoing transfusion therapy. Imaging findings of hepatosplenomegaly were consistent with chronic liver disease and increased bleeding risk.
- Standard AIH management involves corticosteroid induction followed by maintenance immunosuppression with agents such as azathioprine to prevent disease progression.
- In this case, temporary interruption of corticosteroid therapy during hospitalization deviated from continuous immunosuppressive management and may have increased the risk of disease flare.
- Additionally, multiple antibiotics and repeated transfusions without strict monitoring posed risks of fluid overload and infection in the setting of limited hepatic reserve.
- Pharmacotherapeutic review identified prescribing discrepancies, including an aldosterone antagonist without clear indication, vasopressor use despite persistent hypotension, and iron supplementation despite elevated iron stores.
- This case underscores the importance of careful medication review, coagulation monitoring, and multidisciplinary management.
- Clinical pharmacist involvement is particularly valuable in identifying drug-related problems and optimizing therapy in complex AIH cases complicated by gastrointestinal bleeding.

IV. CONCLUSION:

- This case of autoimmune hepatitis (AIH) decompensation triggered by gastrointestinal hemorrhage in a young adult highlights the silent progression of uncontrolled immune-mediated liver injury and its potential to impose significant multiorgan stress.
- Effective management relies on prompt hemodynamic stabilization, correction of anemia, and consistent immunosuppressive therapy to prevent relapse and preserve long-term health.
- The prescribing gaps identified in this case underscore the importance of appropriate therapeutic regimens, regular indication reviews and active pharmacist involvement within multidisciplinary teams to minimize medication-related errors and enhance the overall quality of patient care.

REFERENCES

- [1]. Krawitt EL. Autoimmune hepatitis. *N Engl J Med.* 2006;354:54-66.
- [2]. Bischoff S, Yesmembetov K, Antoni C, Sollors J, Evert M, Ebert M, et al. Autoimmune hepatitis: a review of established and evolving treatments. *J Clin Transl Hepatol.*
- [3]. Autoimmune Hepatitis (StatPearls). StatPearls Publishing; Treasure Island (FL).
- [4]. Dalekos GN, Samakidou A, Lyberopoulou A, Banakou E, Banakou E, Gatselis NK. Recent advances in the diagnosis and management of autoimmune hepatitis. *J Gastroenterol Hepatol.* 2018.
- [5]. Choudhuri G, Somani SK, Baba CS, Alexander G. Autoimmune hepatitis in India: profile of an uncommon disease. *BMC Gastroenterol.* 2005;5:27. doi:10.1186/1471-230X-5-27
- [6]. Mercado LA, Gil-Lopez F, Chirila RM, Harnois DM. Autoimmune hepatitis: a diagnostic and therapeutic overview. *Diagnostics (Basel).* 2023.
- [7]. Mercado LA, Gil-Lopez F, Chirila RM, Harnois DM. Autoimmune hepatitis: a diagnostic and therapeutic overview. *Diagnostics (Basel).* 2023.