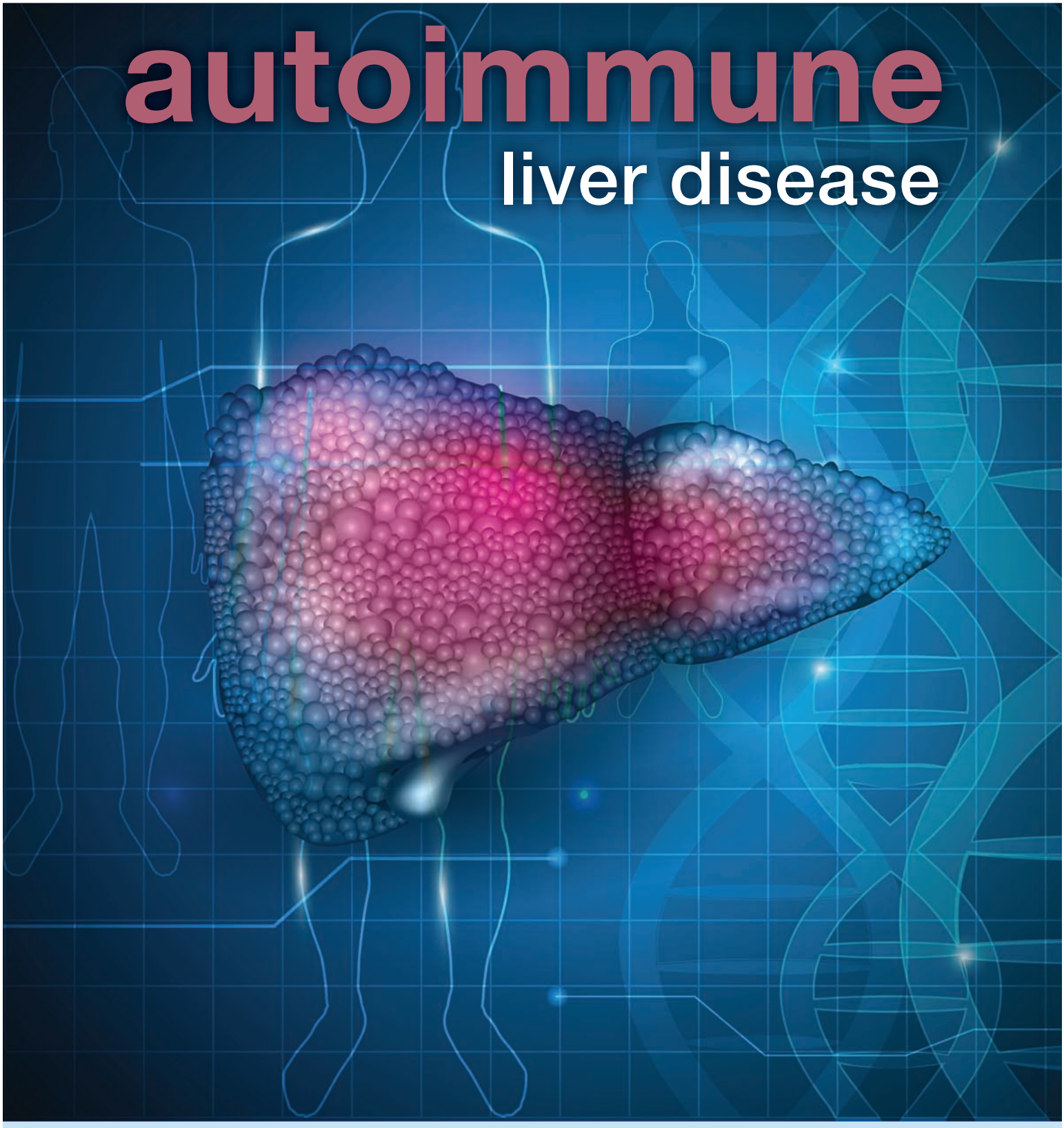
Inside autoimmune liver disease



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An overactive immune system can target any body tissue and cause damage. In AILD, the liver and bile ducts are under attack.

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Autoimmune liver disease (AILD)—primary biliary cholangitis (PBC), primary sclerosing cholangitis (PSC), and autoimmune hepatitis (AIH)—is comprised of three distinct pathologic processes in which a person's immune system doesn't recognize certain hepatic and biliary structures as belonging to the body. The immune system becomes overactive and targets healthy tissue, leading to inflammation, cirrhosis, and end-stage liver disease (ESLD). Some patients may require liver transplantation as a lifesaving measure.

This article presents the pathophysiology, epidemiology, risk factors, signs and symptoms, treatment strategies, and nursing care of patients with AILD.

Primary biliary cholangitis

PBC is a progressive inflammatory autoimmune disease that's manifested by a destruction of the small intrahepatic bile ducts.

Pathophysiology

Destruction of the bile ducts leads to cholestasis, which causes an impairment in the flow and retention of bile—a substance produced by the liver and stored in the gallbladder that flows through the bile ducts and into the small intestine to digest lipids. Bile is comprised of bile salts, cholesterol, and bilirubin—a waste product from damaged red blood cells that accumulates in the blood because of cholestasis in PBC.

Approximately 95% of patients with PBC possess the antimitochondrial antibody (AMA) and a PBC-specific antinuclear antibody (ANA) that has a rim-like staining pattern. These antibodies mediate

an immune response by causing a proliferation of T lymphocytes that attack and destroy the cholangiocytes of the interlobular and septal bile ducts. The T lymphocytes are both CD4 and CD8 cells and may be reactive to PDC-E2—another antibody in PBC. T-lymphocytes may contain granulomas that are identified as a "florid duct lesion" through liver biopsy. A florid duct lesion is an intense area of inflammation.

Over time, the damaged bile ducts disappear (ductopenia) even though the liver makes a futile attempt to transform hepatocytes and progenitor cells into new bile ducts. Progenitor cells normally replace damaged or dead cells in the liver. Because the ducts are no longer present, bile gets trapped in the liver and causes intense inflammation and destruction of hepatic structures, leading to cirrhosis (see *A brief cirrhosis primer*).

Epidemiology and risk factors

PBC affects approximately 35 out of 100,000 individuals. The typical patient is female between the ages of 40 and 60; 90% of patients are female and 10% are male. Research indicates that individuals at greater risk for PBC are those who have a family history of the disease and a coexisting autoimmune disease, such as Sjögren syndrome, thyroid disease, or rheumatoid arthritis.

Researchers postulate that environmental factors may trigger an immune response in the development of PBC, including, but not limited to, cigarette smoke; hair dye; hormonal therapy; toxic superfund sites; and infections, such as *Escherichia coli* infection, Epstein-Barr virus, herpes virus, retroviruses, and HIV-1. Some medications have been implicated in triggering an immune response in PBC, including interferon and chlorpromazine.

Signs and symptoms

Approximately 60% of patients are asymptomatic at the time of diagnosis. A diagnosis of PBC is suspected during a routine checkup with an analysis of lab tests. Liver enzymes show a cholestatic pattern, which means that the alkaline phosphatase (ALP) is disproportionately elevated to alanine aminotransferase (ALT), aspartate aminotransferase (AST), and gamma-glutamyl transferase (GGT). Patients also have elevated bilirubin and prolonged prothrombin time. These lab tests correlate with the presence of antibodies, specifically AMA. Most patients will become symptomatic within a few

A brief cirrhosis primer

Point #1

PBC, PSC, and AIH may lead to cirrhosis, which results from a progression of intense inflammation to a fibrotic or scarring process that causes ischemia in the liver. Ischemia leads to the liver being unable to carry out its vital functions, including, but not limited to, bile production, absorbing and metabolizing bilirubin, metabolizing carbohydrates, storing vitamins, protein synthesis, blood clotting, and metabolizing drugs and toxins.

Point #2

The portal tracts are usually involved in the cirrhotic process. Portal tracts are connective tissues that join the portal vein and hepatic artery to supply the liver with blood and nutrients. Bridging necrosis may also be present in the portal tracts. Bridging necrosis is areas of confluent necrosis from the hepatic lobes to the portal tracts.

Point #3

The scarred or fibrotic liver causes a blockage of blood flow through the portal venous system. This blockage causes pressure in the blood vessels throughout the body, particularly those that line the esophagus and stomach, which places the patient at high risk for hemorrhage. This is called portal hypertension. Ascites—a leakage of fluids and proteins into the peritoneal space—may also occur. Third-space fluid shift may cause edema in the legs.

Point #4

Elevated serum ammonia may occur from a breakdown in protein and cause hepatic encephalopathy, which leads to changes in the patient's mental status from altered brain function.

Point #5

Fibrosis and bridging necrosis cause a collapse of the hepatic parenchyma, leading to ESLD.

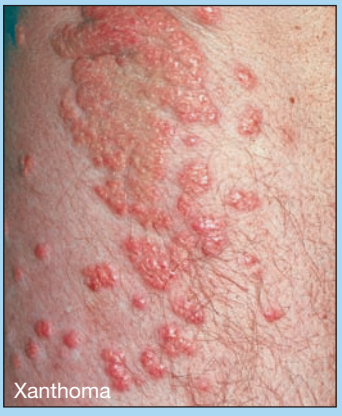
Point #6

Airway management, bleeding, anemia, fluid volume excess, infection, fatigue, nutrition, mobility, and safety are major problems for the ESLD patient that need to be addressed by the interprofessional team.

Point #7

Priority goal: Implement early and aggressive therapy in PBC, PSC, and AIH to prevent the development of cirrhosis and progression to ESLD.

Picturing xanthelasma and xanthoma



Sources: Ayala C, Spellberg B. *Boards and Wards for USLME Step 2*. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2017. DeLong L, Burkhart NW. *General and Oral Pathology for the Dental Hygienist.* 3rd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2018.

years of diagnosis; however, some patients with PBC may initially present with severe disease such as cirrhosis.

The most common symptoms of PBC are fatigue, jaundice, and pruritus. Researchers postulate that fatigue is caused by an increase in mitochondrial activity. Patients must also cope with chronic pruritus, which interrupts rest and sleep, leading to fatigue. Cholestasis causes an accumulation of bile acids in the systemic circulation and on the skin. The bile acid receptor cell TGR5 may affect neurotransmission from the cholestatic process to the skin, causing intense pruritus. Scratching from pruritus may cause the skin to bleed. Jaundice from an increase in bilirubin and hyperpigmentation of the skin from an increase in melanin deposits on the skin may accompany PBC. Jaundice may indicate hepatic decompensation, leading to ascites, peripheral edema, and gastrointestinal bleeding. Changes in mental status may indicate the onset of hepatic encephalopathy.

Two cutaneous features of PBC are xanthelasma and xanthoma. These yellow, fatty lesions are caused by a markedly elevated cholesterol level in PBC, although they may also occur in other disease states. Xanthelasma appears around the eyes and xanthoma appears on the hands, arms, and legs (see *Picturing xanthelasma and xanthoma*). Patients may also have hepatomegaly, splenomegaly, joint pain, and dry eyes and mouth (Sicca syndrome).

A diagnosis of PBC may be made by correlating the patient's physical symptoms with lab data. However, a liver biopsy is usually necessary to confirm the diagnosis, determine the extent of liver damage, and develop a treatment plan. The Ludwig Histological Staging System for PBC is commonly used to classify the disease:

- Stage 1: florid duct lesion
- Stage 2: loss of normal bile duct structure, an increase in the number of biliary epithelial cells, and inflammation of the hepatic parenchyma
- Stage 3: progressive loss of bile ducts and bridging necrosis
- Stage 4: cirrhosis with ESLD.

Treatment strategies

The first-line treatment in PBC is administering ursodeoxycholic acid (UDCA)—a bile acid normally found in the body that isn't toxic to hepatocytes, unlike other bile acids. Increasing the amount of UDCA in the body reduces biliary epithelial and hepatic cell damage from cholestasis and slows the progression of PBC by improving bile flow through the ducts. UDCA may

did you know?

Primary biliary cholangitis was previously called primary biliary cirrhosis. The name change more accurately describes what's happening in the disease, which is an inflammation of the intrahepatic bile ducts.

also reduce the immune-mediated response underpinning disease development, reduce cholesterol, and prevent gallstones.

UDCA is most effective in Stages 1 and 2 when the bile ducts are still present. Patients in Stages 3 and 4 aren't likely to have a therapeutic outcome because of ductopenia. UDCA is administered orally and generally well tolerated. Diarrhea, a decrease in white blood cells (WBCs), and an increase in blood glucose and creatinine are the most common adverse reactions. UDCA is effective in normalizing liver enzymes and reducing antibody titers in at least 70% of patients with PBC.

Additional medications may be used to treat the underlying pathology of PBC. Methotrexate (MTX) is an immunosuppression agent that reduces inflammation in the bile ducts and improves hepatic function in combination with UDCA. Patients should take folic acid to decrease the chances of anemia secondary to MTX treatment. Corticosteroids, such as budesonide, may also be used in PBC because of their immunosuppressive properties. Patients should have good body hygiene; avoid people who are sick; and report symptoms of infection, such as a fever or a sore throat. Colchicine is an antigout agent that may be used in combination with UDCA for its anti-inflammatory properties. The most common adverse reaction is diarrhea that resolves once the dosage is reduced. Major research studies indicate that colchicine also reduces fibrotic processes in the liver and delays the need for liver transplantation. Improvement in liver enzymes is the desired outcome of these agents.

Symptom management is important. The cholesterol-lowering agent cholestyramine is the first-line therapy for pruritus because it's a bile acid resin, which means it metabolizes and removes excessive bile from the blood. It reduces pruritus in 90% of patients with PBC. Patients shouldn't take this medication with other agents to avoid problems with absorption and have sufficient fluid and fiber in their diet to avoid constipation. The antituberculosis antibiotic rifampin is a second-line medication to reduce pruritus in PBC because it increases the rate of bile acid excretion in the body. Liver enzymes, bilirubin level, platelets, and WBC count should be monitored closely during therapy, although the medication has a low incidence of toxicity. Opioid receptor blockers, serotonin reuptake inhibitors, antihistamines, and corticosteroids may also be used to control pruritus. Plasmapheresis may be considered with intractable cases of pruritus.

Patients with PBC have a poor prognosis when they have elevated bilirubin or signs and symptoms of ESLD, such as ascites, bleeding esophageal varices, and hepatic encephalopathy. Some research findings indicate that one-third of patients die from ESLD within 1 to 2 years. These patients must have immediate liver transplantation to save their life. There's a rapid improvement in quality of life after transplantation. The survival rate is approximately 90% at 5 years and 70% at 10 years after transplantation. PBC may recur after transplantation because the AMA is still a part of the patient's immune system. Recurrence is between 20% and 40% after transplantation.

Primary sclerosing cholangitis

PSC is an autoimmune disease that causes progressive inflammation, strictures, fibrosis, and obliteration of the intrahepatic and extrahepatic bile ducts.

Pathophysiology

PSC is classified as an autoimmune disease because it's associated with the human leukocyte antigen (HLA) genes B8 and DR3. HLA is a cell surface protein responsible for immune system regulation. Unlike PBC, PSC isn't associated with specific antibodies. However, patients with PSC may have nonspecific antibodies, including ANA, anti-neutrophil cytoplasmic antibodies (ANCA), and smooth muscle antibody (SMA). Ductopenia

occurs secondary to obliterative cholangitis and cholestasis, leading to portal tract fibrosis, cirrhosis, portal hypertension, ESLD, and a risk of liver cancer. Patients with PSC are also prone to developing pancreatitis, colorectal cancer, and cancer of the bile ducts.

Epidemiology and risk factors

PSC affects approximately 1 to 6 individuals out of 100,000. The disease predominately affects males of all ages and ethnicities. PSC may also be associated with other autoimmune diseases, such as rheumatoid arthritis, thyroiditis, celiac disease, diabetes mellitus, myasthenia gravis, and inflammatory bowel disease (IBD). Approximately two-thirds of patients have IBD (ulcerative colitis and Crohn disease). People with a Northern European heritage are at increased risk for developing the disease. Research studies indicate that the incidence of PSC is lower in nonsmokers and those who drink coffee.

Signs and symptoms

Most patients with PSC are asymptomatic at the time of diagnosis. Lab values follow a similar pattern to PBC. For example, ALP is disproportionately elevated to ALT, AST, and GGT, and the bilirubin level is elevated. Like PBC, patients with PSC may present with fatigue, jaundice, pruritus, hepatomegaly, and splenomegaly.

There's a major difference in the way that PBC and PSC are diagnosed. A diagnosis of PSC requires a special imaging study called endoscopic retrograde cholangiopancreatography (ERCP) in which a lighted endoscope is inserted into the patient's mouth and through the esophagus, stomach, and small intestine. The ampulla of Vater—a small opening in the small intestine that provides access to the extrahepatic bile ducts—is identified. A very small guide wire is inserted through the endoscope into the ducts. A contrast medium is injected through the endoscope and images are obtained to determine if there are bile duct strictures or damage. Another approach to determining bile duct strictures or damage is through a magnetic resonance cholangiopancreatography (MRCP)—a noninvasive imaging procedure of the intrahepatic and extrahepatic bile ducts. A contrast medium is injected into the patient's vein and images of the bile ducts are obtained.

A liver biopsy often complements a diagnosis of PSC by ERCP, lab analysis, and the patient's signs and symptoms. A characteristic histologic finding in PSC is a fibrous obliteration of the bile ducts with connective tissue in an "onion skin" pattern. This connective tissue is concentric and resembles the layers of an onion. PSC may be classified in the following manner:

- Stage 1: chronic inflammation of the portal tracts and bile duct proliferation
- Stage 2: chronic inflammation of the portal tracts and interface hepatitis (inflammation of the liver from destruction of hepatocytes between the parenchyma and the portal tracts)
- Stage 3: fibrosis of the portal tracts with bridging necrosis
- Stage 4: cirrhosis.

Treatment strategies

Some research studies support the use of UDCA, whereas others don't. Additional large studies are needed to further determine the efficacy of this medication in PSC. Immunosuppression medications, including corticosteroids, azathioprine, cyclosporine, and MTX, have been used to slow the progression of PSC with minimal success. Some research studies suggest that oral vancomycin may improve the patient's signs and symptoms and liver chemistries, especially in the pediatric population. Balloon dilation during ERCP may be helpful to reduce bile duct strictures and remove gallstones to improve the patient's symptoms. Pruritus is managed by using the same medications that are used in PBC.

The nurse and patient become partners in the care plan to achieve optimal outcomes.

key points

Priority nursing interventions

PBC

- Assess for osteoporosis due to malabsorption of vitamin D; calcium supplements should be prescribed.
- Assess for visual problems at night, neurologic deficits, and bleeding tendencies due to malabsorption of vitamins A, E, and K; vitamin supplements should be prescribed.
- Assess cholesterol levels; some research studies suggest that patients with hyperlipidemia in PBC are at risk for atherosclerosis.
- Assess for abdominal pain that may indicate acute cholecystitis.
- Assess ALT, AST, and ALP in response to UDCA, MTX, and colchicine therapy, and correlate with the patient's signs and symptoms.
- Assess the patient's responses to medications and other comfort measures to treat intense pruritus.
- Provide the patient with good oral hygiene, lubricating oral solutions, and eye drops due to dry mucous membranes (Sicca syndrome).

PSC

- Assess for symptoms related to malabsorption of vitamins A, D, E, and K.
- Assess for symptoms of infection, including fever, elevated WBC count, erythrocyte sedimentation rate, and C-reactive protein related to bacterial cholangitis.
- Assess pancreatic enzymes, including lipase and amylase, because chronic pancreatitis may coexist with PSC.
- Assess for colorectal and biliary tract cancer.
- Assess for abdominal pain that may indicate acute cholecystitis.
- Assess results of MRCP and/or ERCP that indicate bile strictures and gallstones.
- Assess ALT, AST, and ALP in response to UDCA therapy and correlate with the patient's signs and symptoms.
- Assess the patient's responses to medications and other comfort measures to treat intense pruritus.

AIH

- Assess ALT, AST, ALP, and immunoglobulins.
- Priority assessments and teaching are related to medication therapy to induce and maintain remission.

Prednisone

- Assess for bleeding and infection.
- Assess complete blood cell (CBC) count, blood glucose, BP, daily weight, and mental status.
- Teach the patient to practice good body hygiene and avoid people who are sick.
- Teach the patient to take medication as prescribed.
- Teach the patient to eat a low-sodium diet.
- Teach the patient to take vitamin D to avoid osteoporosis.
- Teach the patient to have regular eye exams.

Azathioprine

- Assess for bleeding and infection.
- Assess CBC count, ALT, AST, ALP, albumin, and proteins.
- Assess for enlarged lymph nodes; azathioprine places the patient at risk for lymphoma.
- Teach the patient to practice good body hygiene and avoid people who are sick.
- Teach the patient to report any skin lesions; azathioprine places the patient at risk for skin cancer.

Mortality in PSC may be as high as 30% at 5 years. Medical treatment and medications don't conclusively slow the progression of the disease. The best option for patients with PSC is liver transplantation. The 5-year survival rate after transplantation is favorable at 85%.

Autoimmune hepatitis

AIH is a chronic inflammatory autoimmune disease that causes damage to the liver because the liver is being attacked by the immune system.

Pathophysiology

The immune system loses tolerance and causes dysregulation in T and B lymphocytes, leading to inflammation and destruction of hepatocytes by cytotoxic T lymphocytes. AIH is sometimes referred to as a T-cell disease. Antibodies are produced in response to immunologic intolerance and considered markers in AIH, although they aren't specific to the disease and don't cause it. AIH is classified based on these antibodies.

Genetically linked to HLA-DR3 and HLA-DR4, type 1 AIH, or classic AIH,

constitutes 80% to 90% of cases and includes ANA, SMA, or both. Genetically linked to HLA-DR4, type 2 AIH occurs when patients have anti-liver kidney microsomal antibody (anti-LKM-1), anti-liver-soluble cytosol antigen (anti-ACL-1), or both. Type 2 AIH is more common in Europe and typically affects girls and young women.

Patients with AIH may also have other antibodies, including ANCA and asialoglycoprotein receptor antibodies. Some patients may be antibody negative and still have the signs, symptoms, and histologic features of the disease. Patients may or may not be symptomatic at the onset of the disease. However, some patients may present with decompensated cirrhosis with ascites, encephalopathy, and bleeding from esophageal varices.

Epidemiology and risk factors

Approximately 1 to 2 in 100,000 individuals are diagnosed with AIH each year. The prevalence of the disease is approximately 24 people in 100,000; 90% of patients are female and 10% are male. Patients with coexisting autoimmune diseases, such as thyroiditis, diabetes mellitus, rheumatoid arthritis, and systemic lupus erythematosus, are at increased risk for AIH. Research indicates that variables in genetically predisposed individuals trigger an immune system dysregulation and attack on the liver, which may include infections and medications, such as minocycline, methyldopa, and possibly herbs.

Signs and symptoms

Patients are likely to have fatigue, malaise, anorexia, jaundice, abdominal discomfort, and a low-grade fever. A lab evaluation indicates that the ALT and AST are significantly elevated at least 10 times the upper level of normal out of proportion to ALP, which may be normal or mildly elevated. A mildly elevated or near-normal ALP generally excludes biliary tract disease, such as PBC and PSC. Immunoglobulins are also elevated in AIH.

In AIH, an elevation of liver enzymes indicates that the immune system is attacking the liver. Declining serum albumin and proteins, prolonged prothrombin time, and elevated serum bilirubin indicate that liver failure is occurring. Differential diagnoses must also be excluded as a cause of elevated liver enzymes, including viral hepatitis, drug-induced liver injury, alcohol-induced liver injury, liver tumor, heart disease, and ischemic injury to the heart.

An urgent liver biopsy should be performed to determine the extent of inflammation and fibrosis. Interface hepatitis and bridging necrosis are defining features of AIH, and there are portal tract lymphocytic cells and an abundance of plasma cells. These cells cross the connective tissue surrounding the portal tracts (called the limiting plate) and invade the surrounding parenchyma. Hepatocytes become engulfed in these inflammatory cells and undergo a process of cell death called apoptosis.

Histologic changes suggestive of biliary tract disease, such as granulomas, aren't usually present. The degree of fibrosis after intense inflammation in AIH is classified in the following manner from the results of the liver biopsy:

- Stage 0: no fibrosis
- Stage 1: mild fibrosis
- Stage 2: moderate fibrosis
- Stage 3: severe fibrosis (precirrhosis)
- Stage 4: cirrhosis.

Research indicates that the prognosis in AIH is favorable when fibrosis hasn't extended to the cirrhosis stage and immediate immunosuppression therapy is instituted.

Treatment strategies

The priority goal in AIH is to stop the immune attack on the liver through immunosuppression therapy. The induction

Support groups are helpful to patients and families coping with the chronicity of AILD.

Providing the patient with emotional support is crucial following a diagnosis of AILD.

of remission is achieved by administering corticosteroids, primarily prednisone at 40 to 60 mg/day with a subsequent taper as liver enzymes dramatically decrease over weeks to months after starting therapy. Budesonide is another corticosteroid that may be considered. A standard approach is to combine prednisone with the steroid-sparing immunosuppression agent azathioprine to induce remission, which is maintained with either a low-dose of prednisone or azathioprine. Research indicates that azathioprine alone is highly effective in maintaining remission.

Other immunosuppression agents may be considered in the treatment plan, including mycophenolate mofetil, cyclosporine, tacrolimus, and rituximab. Remission is achieved when there's a normalization of liver enzymes and immunoglobulins, improvement in the patient's symptoms, and histologic improvement in the patient's liver. Immunosuppression can reduce and reverse the fibrotic process in AIH. A repeat liver biopsy should be explored.

Approximately 80% of patients achieve full remission in 18 to 24 months after starting immunosuppression therapy. Even patients with cirrhosis may experience some histologic improvement. Approximately 10% of patients achieve partial remission and 10% fail to respond to therapy and progress to ESLD requiring liver transplantation. The relapse rate is 80% when immunosuppression therapy is stopped. It's crucial for patients to be adherent to medication therapy and report any adverse reactions to the healthcare provider.

Nursing care

The nurse should have a general understanding of AILD features (see *Selected features of AILD*). Nursing care begins with establishing a caring rapport with the patient. Providing the patient with emotional support is crucial following a diagnosis of AILD. Correlate the patient's health history, head-to-toe assessment, and signs and symptoms to develop a care plan.

Assess the patient's mental status and degree of joint pain, abdominal pain, or fatigue. Pay close attention to skin symptoms. The patient may have jaundice manifested by a yellow discoloration of the sclera and skin, with pale oral mucous membranes. Correlate jaundice with an elevated bilirubin level and pruritus, which may be a major source of discomfort, especially for a patient with PBC or PSC. In addition to medication administration, pruritus can be managed by bathing in cool, tepid water; taking a colloidal oatmeal bath; and applying moisturizers to the skin. A high bilirubin level will also cause the patient's stool to be a pale color.

Assess liver enzymes because an elevation indicates that the hepatobiliary system is under attack by the patient's immune system. For example, a markedly elevated ALP out of proportion to ALT and AST may point to biliary pathology in PBC and PSC. Conversely, a markedly elevated ALT and AST out of proportion to ALP may point to AIH. Liver enzymes should be correlated with the presence of antibodies as indicated; findings from a liver biopsy; and the results from liver function tests, such as albumin, proteins, and prothrombin time. Decreasing serum albumin and proteins, along with a prolonged prothrombin time, may indicate decompensated cirrhosis and that liver failure with portal hypertension is underway.

Assess for easy bruising and bleeding, ascites, and peripheral edema. Also assess the patient's weight and abdominal girth daily. Patients are generally placed on a low-protein or no-protein diet when they have ascites. Sodium intake is also reduced. Potassium-sparing and loop diuretics may be prescribed to manage fluid volume excess. Fluid restriction is often necessary. Cardiopulmonary status should be assessed for fluid volume overload. Monitor serum creatinine because liver failure often leads to renal failure.

Patients should be adherent to medication therapy; report medication adverse reactions to healthcare providers; avoid

Selected features of AILD

PBC

- Mostly affects female patients
- History of autoimmune disease is a risk factor
- Associated with AMA and/or ANA; it's possible to have antibody-negative PBC
- ALP disproportionately elevated to ALT and AST
- Damage only to the intrahepatic bile ducts
- Associated with a florid duct tissue pattern by liver biopsy
- Fatigue, jaundice, abdominal pain, joint pain, dry mouth and eyes, xanthelasma, and xanthoma
- Pruritus is a major source of discomfort
- The use of UDCA is associated with an improved disease course and prognosis in the early stages
- Liver transplantation for ESLD
- May overlap with AIH

PSC

- Mostly affects male patients
- History of autoimmune disease is a risk factor
- Most patients have IBD
- Not associated with specific antibodies
- ALP disproportionately elevated to ALT and AST
- Damage to intrahepatic and extrahepatic bile ducts
- Associated with an onion skin tissue pattern by liver biopsy
- Fatigue, jaundice, abdominal pain, and joint pain
- Pruritus is a major source of discomfort
- UDCA and other medications and treatments haven't conclusively been proven to slow disease progression
- ERCP with balloon dilation may reduce bile duct strictures and remove gallstones to improve the patient's symptoms
- Risk of chronic pancreatitis, colorectal cancer, and cancer of the bile ducts
- Liver transplantation is the best option
- May overlap with AIH

AIH

- Mostly affects female patients
- History of autoimmune disease is a risk factor
- Associated with SMA, ANA, anti-LKM-1, and anti-ALC-1; it's possible to have antibody-negative AIH
- ALT and AST elevated at least 10 times the upper level of normal; ALP mildly elevated
- Interface hepatitis with prominent lymphocytic and plasma cells; bile ducts are generally spared
- Fatigue, jaundice, pruritus, abdominal discomfort, and joint pain
- Rapid reduction in ALT and AST in response to prednisone
- Remission maintained through prednisone and/or azathioprine
- 80% of patients achieve remission within 18 to 24 months of the start of therapy; some patients take longer
- 80% of patients relapse within 6 months of stopping immunosuppression therapy
- Liver transplantation for ESLD
- May overlap with PBC or PSC

taking hepatotoxic medications such as acetaminophen, alcohol, and smoking; and get plenty of rest. Patients receiving immunosuppression medications should receive influenza and pneumonia vaccines because they're at increased risk for infection.

Support groups are helpful to patients and families coping with the chronicity of AILD.

Partners for positive outcomes

Early recognition and prompt treatment are crucial in AILD. Nursing assessment and patient adherence are integral to this process. The nurse and patient become partners in the care plan to achieve optimal outcomes. ■

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