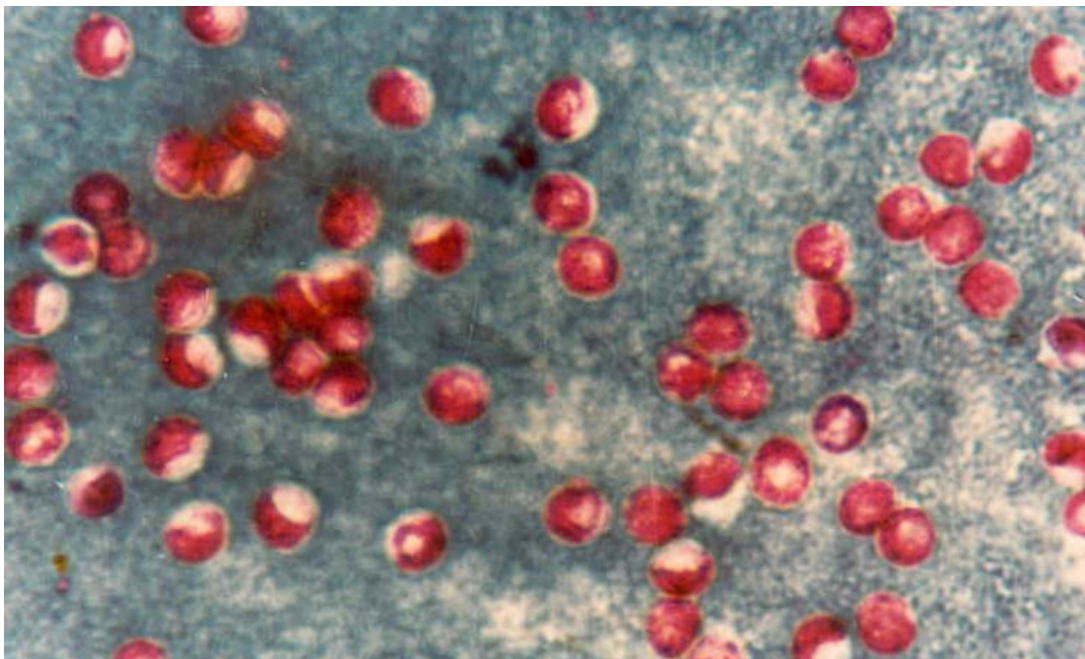


AIDS Cholangiopathy — Diagnosis and Treatment

[See online here](#)

AIDS cholangiopathy is an advanced, typically fatal, disease caused by biliary obstruction resulting from opportunistic infection-related biliary tract strictures. Cholangiopathy developed in 25 out of 100 patients with AIDS before the arrival of antiretroviral therapy, especially in patients with low CD4 counts (<100/mm³). Its symptoms are right upper quadrant and epigastric pain, fever, diarrhea, and sometimes, jaundice. The severity of pain depends on the lesions of the biliary tract. Endoscopic retrograde cholangiopancreatography establishes the diagnosis of cholangiographic abnormalities. Antimicrobial therapy is sometimes ineffective and highly active antiretroviral therapy is the best therapy for AIDS cholangiopathy. Surgical intervention is recommended to patients with a terminal disease and intractable pain in the abdomen.

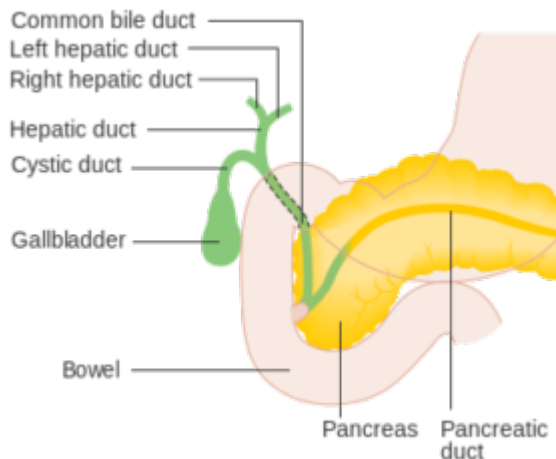


Definition of AIDS Cholangiopathy

Patients with **AIDS** can develop an abnormality of liver enzymes and **sclerosing cholangitis**. Alkaline phosphatase (ALP) level is usually elevated, while the CD4 count is always < 200/mm³. This form of sclerosing cholangitis is believed to be related to **opportunistic infections in AIDS** and is known as **AIDS cholangiopathy**.

Epidemiology and Etiology of AIDS Cholangiopathy

AIDS cholangiopathy is a **fatal complication of AIDS** that has become very rare since the introduction of **highly active antiretroviral therapy (HAART)**. The median survival after the diagnosis of AIDS cholangiopathy is about 9 months. Patients who are HIV positive are expected to develop some liver abnormalities, but the diagnosis of AIDS cholangiopathy is reserved for patients with sclerosing cholangitis.



[Image](#): "Diagram showing the position of the bile ducts." by Cancer Research UK uploader. License: [CC BY-SA 4.0](#)

The most common etiology of AIDS cholangiopathy is **infectious**. Opportunistic infections of the **biliary system** are responsible for AIDS-related cholangitis and sclerosing cholangitis in this group of patients.

The most commonly identified organisms are **cryptosporidium** and **cytomegalovirus**, which can be isolated from the bile. **Enterocytozoon bieneusi** can also be identified in the bile of some patients with unexplained AIDS cholangiopathy.

Pathophysiology and Clinical Presentation of AIDS Cholangiopathy

Patients with AIDS cholangiopathy have a **very low CD4 count** that is less than 200/mm³, which puts them at risk of opportunistic infections. Opportunistic infections of the biliary system result in scarring and elevated levels of liver enzymes, such as **alkaline phosphatase**. Eventually, sclerosing cholangitis ensues, and patients can develop **chronic liver disease**.

No definite pathogen is identified in half of the patients. It affects AIDS patients with low CD4 counts (<135/mm³). **The possible causative organisms are:**

- Cytomegalovirus (CMV)
- Herpes simplex virus (HSV)
- *Cryptosporidium parvum*
- Microsporidium
- *Mycobacterium avium* complex

There are four pathological patterns:

- Combination of sclerosing cholangitis and papillary stenosis (50%)
- Isolated intrahepatic sclerosing cholangitis-like appearance (20%)
- Isolated papillary stenosis (15%)
- Long-segment extrahepatic duct stricture +/- concurrent intrahepatic disease (15%)

Symptoms of AIDS Cholangiopathy

Patients with AIDS cholangiopathy usually present with **right upper abdominal pain** and **elevated liver enzymes**. Abdominal pain is usually associated with nausea, vomiting, and fever. Jaundice is rarely seen in AIDS cholangiopathy.

Diagnostic Work-up for AIDS Cholangiopathy

The **incidental finding of an elevated alkaline phosphatase level** in a patient with AIDS can be a presenting feature of AIDS cholangiopathy. A **complete blood count** always reveals a CD4 count < 200 cells/mm³ and usually lower than 100 cells/mm³. Other hepatic enzyme levels can be abnormal in patients with AIDS cholangiopathy, but this finding is not specific. Bilirubin level is usually normal, hence there is typically no jaundice present on physical examination.



[Image](#): "Biliary stenosis" by Mbarrufet – Own work. License: [CC BY-SA 4.0](#)

Transabdominal ultrasound is indicated as an initial diagnostic tool in the evaluation of patients for whom the differential diagnosis of AIDS cholangitis is being considered.

Findings consistent with sclerosing cholangitis on transabdominal ultrasound include:

- **Dilated intrahepatic bile ducts**
- **Stenosis**
- **Thickening of the common bile duct wall**

If any of these findings are identified in a patient with AIDS, **endoscopic retrograde cholangiopancreatography (ERCP)** is indicated.

ERCP is the gold-standard diagnostic modality for AIDS cholangiopathy. Sclerosing cholangitis, intrahepatic sclerosing cholangitis, and extrahepatic bile duct strictures are usually evident on ERCP in patients with AIDS cholangiopathy. It enables clinicians to extract a sample for small-bowel biopsy to identify the causative organism. An **abnormal ultrasonography study** is highly predictive of an abnormal ERCP result in patients with AIDS cholangiopathy.

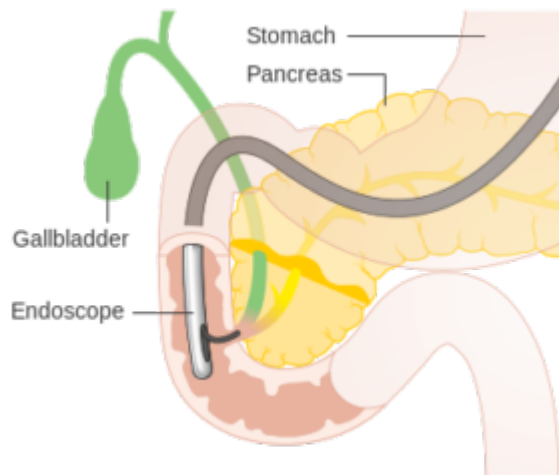


Image: "Detailed diagram of an endoscopic retrograde cholangiopancreatography (ERCP)." by Cancer Research UK - Original email from CRUK. License: [CC BY-SA 4.0](https://creativecommons.org/licenses/by-sa/4.0/)

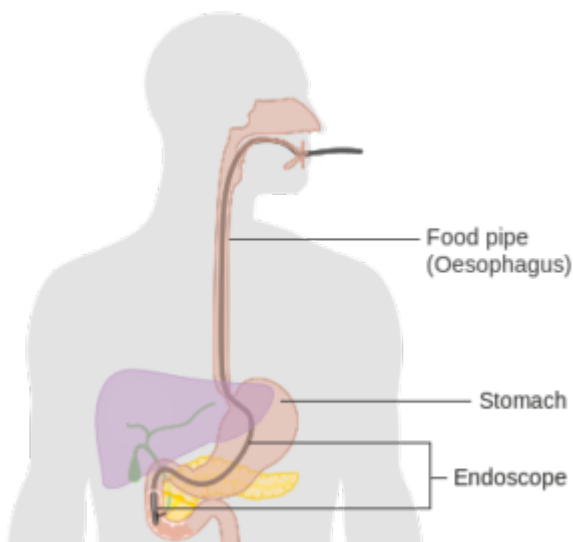


Image: "Diagram showing an endoscopic retrograde cholangiopancreatography (ERCP)." by Cancer Research UK - Original email from CRUK. License: [CC BY-SA 4.0](https://creativecommons.org/licenses/by-sa/4.0/)

Other less commonly used imaging modalities include **computerized tomography (CT) scanning** and **hepatobiliary scintigraphy**. CT scanning of the biliary system can reveal intrahepatic ductal dilation, but it is not sensitive enough to diagnose increased wall thickness of the extrahepatic bile duct. Hepatobiliary scintigraphy can identify intrahepatic and extrahepatic sclerosing, stenotic, and dilatation abnormalities.

Treatment of AIDS Cholangiopathy

A **holistic approach** to treating AIDS cholangiopathy is critical and includes:

- Diet and lifestyle modifications
- Antimicrobial and antiviral therapy

- ERCP
- Palliative pain control, when appropriate

Current consensus is that AIDS cholangiopathy is infectious in origin, hence diet modifications are not very beneficial. As a preventive measure, sexual education about **safe-sex practices** can lower the risk of HIV infection and AIDS.

Unfortunately, patients with AIDS cholangiopathy do not benefit from antimicrobial therapy unless the etiology is related to cryptosporidium infection. A few studies have reported clinical improvement and bile clearance of oocysts in patients with AIDS cholangiopathy due to cryptosporidium infection when **intravenous paromomycin** followed by **oral letrazuril** was given.

Gastrointestinal symptoms related to cryptosporidium infection, such as [diarrhea](#) and abdominal pain, may be alleviated with antimicrobial therapy.

There is no current treatment for patients with cytomegalovirus-related AIDS cholangiopathy. Patients who develop **cholecystitis** as a complication of AIDS cholangiopathy may benefit from treatment with **trimethoprim-sulfamethoxazole** because the most commonly implicated organism is *Cyclospora cayentanensis*, which is sensitive to this antibiotic combination.

The best treatment to lower the risk of AIDS cholangiopathy is **HAART**, which improves [immune function](#) in patients with HIV infection. Additionally, patients with cryptosporidiosis benefit from HAART as it was shown to eradicate the organism oocysts in the stool.

HAART can also eradicate cryptosporidiosis oocysts from the bile in patients with AIDS cholangiopathy. HAART, especially if a **protease inhibitor** is included, is also effective in patients with AIDS cholangiopathy who did not respond to antimicrobial therapy as it cannot reduce biliary tract damage.

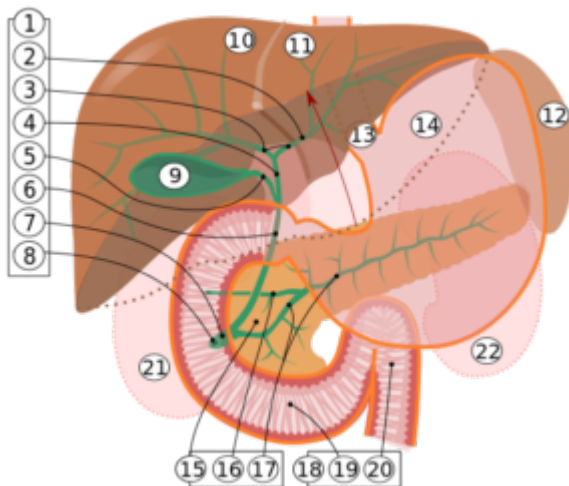


Image: "Bile duct and pancreas 1. Bile ducts: 2. Intrahepatic bile ducts 3. Left and right hepatic ducts 4. Common hepatic duct 5. Cystic duct 6. Common bile duct **7. Ampulla of Vater** 8. Major duodenal papilla 9. Gallbladder 10-11. Right and left lobes of liver 12. Spleen 13. Esophagus 14. Stomach 15. Pancreas: 16. Accessory pancreatic duct 17. Pancreatic duct 18. Small intestine 19. Duodenum 20. Jejunum 21-22: Right and left kidneys" by Jmarchn - Own work. License: [CC BY-SA 3.0](#)

HAART is not only effective in treating AIDS cholangiopathy but can also lower the risk of

AIDS and AIDS-defining disease. HAART also results in an **improvement in liver function**, as manifested by a decrease in alkaline phosphatase level and a reduction in the diameter of the intrahepatic and extrahepatic [bile duct system](#).

Therapeutic ERCP is indicated for patients who have **stenosis in the papilla of Vater** as urgent biliary decompression and drainage is usually needed. Unfortunately, ERCP does not affect survival in patients with AIDS cholangiopathy, and HAART is the only treatment that was found to directly increase survival.

ERCP usually results in significant improvement in symptoms attributed to AIDS cholangiopathies, such as abdominal pain and fever, but it is not known to affect liver chemistry or decrease alkaline phosphatase levels.

Finally, patients with AIDS cholangiopathy may develop medically intractable abdominal pain. In these situations, a **celiac plexus block** may be beneficial for pain relief. However, this interventional treatment is typically reserved for patients with AIDS-related sclerosing cholangitis associated with papilla of Vater stenosis that is unresponsive to **narcotics** or ERCP treatment.

Ursodeoxycholic acid may have a role in treating intrahepatic ductal sclerosis and cholestasis.

Patients with acalculous cholecystitis related to AIDS cholangiopathy might benefit from **laparoscopic cholecystectomy**, particularly when ERCP, HAART, and antimicrobial therapy are ineffective.

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