

# Cirrhosis

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## Background

### About Cirrhosis

With repeated episodes of inflammation and liver injury there is progressive scarring (fibrosis) and replacement of normal tissue. This impedes blood flow and normal synthetic liver function:

- Cirrhosis is usually diagnosed with ultrasound or liver biopsy.
- Cirrhosis can be asymptomatic and may or may not be accompanied by signs of chronic liver disease.
- It is a pre-malignant condition and screening for liver cancer is required. It can affect the functioning of the entire body.

Liver disease is an increasing cause of morbidity and death in the world:

- It is the fourteenth most common cause of death worldwide, and fourth in Central Europe.
- It has many causes – chronic alcoholism and [hepatitis C](#) are the most common.

Most chronic liver disease with cirrhosis is asymptomatic until clinical complications with subsequent decompensation occur. General practitioners are therefore encouraged to screen for viral hepatitis (hepatitis B and C), fatty liver, and investigate patients at risk of chronic liver disease.

## Assessment

1. Take a history to consider possible causes:

- **Alcoholic liver disease**

### Alcoholic liver disease

- *History of hazardous (heavy) alcohol intake*
- *This includes binge drinking*

- **Chronic hepatitis C**

### Chronic hepatitis C

*Risk factors:*

- *Injecting drug use (may have been only once)*
- *Blood products or organ donor recipients before 1990 in Australia, or any time overseas*
- *Tattoos and body piercing*
- *History of imprisonment or other custodial settings*

- *Medical or dental treatments in countries where equipment may not have been adequately sterilised*
- *Needle stick injury or occupational exposure to blood or body substances e.g., healthcare workers*
- *Sexual partners of people with HCV (higher risk of transmission for men who have sex with men and people with HIV infection)*
- *People born in countries with high HCV prevalence e.g., Southeast Asia, Middle East, Africa*
- *Aboriginal and Torres Strait Islander populations*
- *Children born to HCV-positive mothers (low risk of transmission)*
- *Evidence of liver disease*
- *Known HIV or hepatitis B infection*
- *History of HCV in a family member where blood-to-blood transmission may have occurred e.g., through sharing of toothbrushes or shavers*

See [Chronic Hepatitis C](#) pathway.

- **Chronic hepatitis B**

### Chronic hepatitis B

*Higher risk groups:*

- *People born in:*
  - *Asia or Pacific Islands*
  - *Africa*
  - *Middle East*
  - *Eastern, Southern Europe, and the Mediterranean*
  - *South and Central America*
  - *Caribbean*
- *Aboriginal and Torres Strait Islander people*
- *Men who have sex with men*
- *Inmates or past inmates of correctional facilities*
- *Patients infected with HCV or HIV*
- *All patients undergoing chemo- or immunosuppressive therapy*
- *Household contacts of hepatitis B positive patients*
- *Injecting drug users*
- *Patients with abnormal LFTs*
- *Patients undergoing renal dialysis*

*Routinely screen pregnant women.*

See [Chronic Hepatitis B](#) pathway.

- **Autoimmune hepatitis**

### Autoimmune hepatitis

- *Can present in a variety of ways.*
- *Diagnosis is by the presence of signs and symptoms and the exclusion of other potential causes.*

- **Inherited diseases**

## Inherited diseases

- Haemochromatosis
- Alpha-1 antitrypsin deficiency
- Wilson's disease
- Cystic fibrosis
- Galactosaemia
- Glycogen storage diseases

- **Non-alcoholic steatohepatitis (NASH)**

## Non-alcoholic steatohepatitis (NASH)

Fat build up in the liver associated with:

- Diabetes
- Obesity
- Coronary artery disease
- Treatment with corticosteroid medications
- See [Fatty Liver](#)

- **Blocked bile ducts**

## Blocked bile ducts

- Biliary atresia in babies
- Primary biliary cirrhosis in adults
- Secondary biliary cirrhosis if after gallbladder surgery, if the ducts are inadvertently tied off or injured.
- Primary sclerosing cholangitis (PSC)

- **Drugs, toxins, and infections**

## Drugs, toxins, and infections

- Severe reactions to prescription drugs.
- Prolonged exposure to environmental toxins.
- The parasitic infection schistosomiasis.
- Heart failure with resultant liver congestion.

- Cryptogenic cirrhosis (no cause found, approximately 10% of cases)

2. Check for **symptoms**. Many patients with cirrhosis have no symptoms in the early stages.

## Symptoms

- Exhaustion/fatigue
- Loss of appetite
- Nausea
- Weakness
- Decreased mental functioning
- Weight loss
- Abdominal pain
- Itching

3. Clinically assess for liver disease:

- **signs of cirrhosis**

### Signs of cirrhosis

- Jaundice
- Leukonychia
- Spider naevi
- Palmar erythema
- Gynaecomastia
- Muscle wasting
- Liver enlargement
- Shrunken liver
- Hardened liver edge
- Ascites

- **acute liver failure.**

### Acute liver failure

Defined as a rapid decline in hepatic function, characterised by:

- jaundice.
- hepatic encephalopathy.
- associated with coagulopathy INR > 1.5

- **acute hepatitis.**

### Acute hepatitis

- Fever
- Gastrointestinal symptoms – abdominal pain, anorexia, nausea, vomiting
- Icteric features, jaundice, dark urine, pale faeces – may not be present (anicteric hepatitis)
- Enlarged tender liver, +/- spleen
- Urticaria, and joint pains (particularly in Hepatitis B)

- **signs of complication.** In some cases, they may be the first presentation of cirrhosis.

### Complications of cirrhosis

- Portal hypertension
- Splenomegaly
- Varices with risk of upper GI bleeding
- Insulin resistance and type 2 diabetes
- Liver cancer, hepatocellular carcinoma
- Encephalopathy with degrees of mental impairment
- Hepato-renal syndrome
- Sensitivity to medication and their side effects
- Immune system dysfunction, leading to infection
  - Ascites – this may become infected with bacteria normally present in the intestines
- Impotence
- Osteoporosis

4. Arrange **blood tests** and organise **abdominal ultrasound**.

### Abdominal ultrasound in liver disease

*This is used to assess:*

- *structure and integrity of the liver*
- *portal hypertension*
- *splenomegaly*
- *abdominal varices*
- *hepatocellular carcinoma.*

### Blood tests

- *Consider electrolytes, LFTs, INR, FBE, iron studies, HIV, hepatitis A, B, and C serology.*
- *Perform HBV DNA if hepatitis B surface antigen positive, HCV RNA (PCR) genotyping if hepatitis C serology positive.*
- *Any other tests related to the likely cause of the cirrhosis:*
  - *Antinuclear antibody (ANA) , smooth muscle antibody (SMA), liver kidney microsomal antibody (LKM), Ig G levels in autoimmune hepatitis*
  - *Antimitochondrial antibody in primary biliary cirrhosis*
  - *Serum ceruloplasmin in Wilson's disease*
  - *Coeliac serology as can be a cause for abnormal LFTs*
- *Liver function tests may be only mildly abnormal in severe disease. Elevated bilirubin, reduced albumin and raised INR are indicators of impaired liver function. Thrombocytopenia suggests cirrhosis and portal hypertension.*

5. Assess for liver fibrosis:

- **AST to platelet ratio index (APRI)** is a useful initial assessment.

### AST to platelet ratio index (APRI)

See [AST to platelet ratio index \(APRI\) Calculator](#).

- *If a patient score is < 1.0 and there is no concern about risk of cirrhosis, further fibrosis assessment is not necessary.*
  - *If the score is ≥ 1.0, request [FibroScan](#).*
  - *If the score is < 1.0, but there is an increased pre-test probability of cirrhosis (e.g. physical signs of chronic liver disease, history of heavy alcohol use), then cirrhosis is not ruled out – request [FibroScan](#).*
- Consider **FibroScan** – transient elastography (TE).

### FibroScan

- *A non-invasive alternative to liver biopsy and easily detects risk of severe liver disease.*
- *Assesses the degree of liver fibrosis.*
- *Excludes advanced liver disease in patients with long-standing abnormal LFTs.*
- *Not MBS subsidised. Access to [FibroScan](#) can be facilitated through tertiary hospital services and specialist liver clinics by referral.*

6. Estimate cirrhosis severity by calculating the [Child-Pugh score](#) (low albumin, high INR, high bilirubin, encephalopathy, and presence of ascites are poor prognostic markers).

## Management

Treatment aims to stop or delay further progression and reduce complications. It depends on the cause of cirrhosis and any complications a patient is experiencing. Coordinated care is the optimal management involving general practitioner and specialist services.

1. If any **serious hepatic complications**, refer to [Emergency Department](#).

### Serious hepatic complications

- [Acute liver failure](#)
- *Sepsis in a patient with cirrhosis*
- *Severe hepatic encephalopathy*
- *Severe ascites restricting movement and breathing*

2. Refer for [urgent or routine gastroenterology referral](#) if suspected cirrhosis suggested by one or more of:

- *Evidence of cirrhosis on imaging*
- *Platelet count < 120 x 10<sup>9</sup>/L*
- *Ascites*
- *Hepatic encephalopathy*
- [AST to platelet ratio index \(APRI\)](#) > 2.0

3. Offer **lifestyle** and **nutritional advice** to all patients and support them to make changes.

### Nutritional advice

- *Attend to obesity and healthy living with metabolic syndrome.*
- *Maintain adequate nutrition to avoid loss of muscle mass and reduce risk of encephalopathy. Dietary management is a cornerstone of cirrhosis management. See [Nutrition in Chronic Liver Disease, Table 3: Recommendations of the 1997 ESPEN Consensus Group](#).*
- *Consider referral to a dietitian.*

### Lifestyle advice

- *Cease alcohol intake. Any alcohol will lead to more liver damage.*
- *Cease [smoking](#) (nicotine or cannabis).*
- *[Vaccinate](#) against hepatitis A and B viruses, influenza virus, and check if eligible for pneumococcus as early as possible, as antigenic response becomes weaker as cirrhosis progresses.*
- *Monitor drug interactions and the possible need for dose reductions when prescribing for patients with cirrhosis.*

4. Address **underlying causes** of the liver disease, to stop disease progression.

### Treatment of underlying causes

- *Immunosuppression for autoimmune hepatitis.*
- *Venesection for haemochromatosis.*

- *Copper chelators or zinc for Wilson's disease.*
- *Patients with viral hepatitis should be assessed for antiviral treatment. See [Chronic Hepatitis B](#) and [Hepatitis C](#).*
  - *All patients with cirrhosis who are positive for HBsAg should receive oral antiviral therapy with a potent antiviral (entecavir or tenofovir) irrespective of viral load.*
  - *Patients with cirrhosis who respond to antiviral treatment still need regular surveillance for hepatocellular carcinoma, because the risk, although reduced, is not eliminated.*
- *Manage non-viral hepatitis patients in partnership with a [gastroenterologist](#).*

5. Ensure management of complications of cirrhosis are in place through medication and monitoring:

- **Portal hypertension**, which is the underlying cause of most of the complications of cirrhosis and subsequent mortality.

### Portal hypertension

- *All patients with cirrhosis should be screened for varices as the risk of developing varices with subsequent acute variceal bleeds in the first year are 7% and 12% respectively.*
- *Any development of ascites which is directly related to the degree of portal hypertension and splenic enlargement.*
- *Monitor for possible renal impairment and the development of hepato-renal syndrome.*

- **Ascites**

### Ascites

- *Educate patient about limiting dietary sodium to 80 to 120 millimoles daily (4.0 to 6.9 g per day)*
- *Diuretic therapy should start with a morning dose of spironolactone 50 mg with or without frusemide 40 mg*
- *Monitor renal function and serum electrolytes weekly before incremental dose changes:*
  - *Weight loss should not exceed 1 kg per day in patients with peripheral oedema or 0.5 kg per day in those without.*
  - *Maximum doses of 400 mg spironolactone and 160 mg frusemide.*
- *Refractory ascites is when there is no response to maximum tolerated diuretic doses (or cannot be tolerated e.g., due to renal disease) and requires consideration of paracentesis. Contact on-call Gastroenterologist for advice.*

6. Monitor for any signs of **deteriorating mental function**, considering that signs may be very subtle.

### Deteriorating mental function

- *Confusion*
- *Forgetfulness*
- *Personality or mood changes*
- *Impaired judgement*
- *Impairment of concentration*
- *Incoordination*
- *Change in intellectual function*

- *Change in sleep patterns*

7. **Manage encephalopathy** which has a high (64%) one-year mortality rate.

### Manage encephalopathy

- *Minimal hepatic encephalopathy is more common than overt encephalopathy, and influences complex cognitive or coordination skills (e.g. driving), leading to increased risks of accidents, and increases the risk of falls.*
- *Overt encephalopathy is generally transient and linked with a precipitating event, such as use of sedatives, constipation, dehydration, infection, or gastrointestinal bleeding.*
- *Lactulose is the first-choice drug for prevention of recurrent encephalopathy, titrating to 2 loose bowel motions a day.*
- *Rifaximin, a non-absorbable antibiotic, is effective when added to lactulose if encephalopathy recurs. It reduces the risk of further recurrence from 46% to 21%. Rifaximin can only be prescribed by a specialist.*

8. Control any **infection in a patient with cirrhosis**.

### Control any infection in patients with cirrhosis

- *The risk of infection worsens with worsening liver function e.g., UTIs, pneumonia, skin infections.*
- *In patients with ascites, spontaneous bacterial peritonitis (SBP), infection of the ascitic fluid, has a poor prognosis.*
- *All patients with suspected SBP should be hospitalised.*
- *If a patient survives an episode of SBP, prescribe long-term norfloxacin (or trimethoprim/sulfamethoxazole) prophylaxis.*

9. A liver transplant may be considered by the specialist team as a therapeutic option in a patient who develops decompensation or hepatocellular carcinoma with cirrhosis.

## Routine monitoring

Arrange:

- **Blood tests**

*Every 6 months:*

- *Electrolytes*
- *LFTs, including AST if need to calculate the APRI score*
- *INR*
- *FBE*
- *Iron studies*
- *Alpha-fetoprotein*

*Monitor cirrhosis grade and prognosis by calculating:*

- [\*Child-Pugh score\*](#), and
- [\*Model For End-Stage Liver Disease \(MELD\) score\*](#).

- **Ultrasound** – screen for hepatocellular carcinoma (HCC) every 6 months. This can develop in all stages of cirrhosis.
- **Gastroscopy** – every 2 to 3 years, with or without banding of varices.

- Bone mineral density scan every 2 years. A patient with cirrhosis is 2.5 times more likely to have osteoporosis compared with the normal population.
- Serum testosterone – check for gonadal insufficiency in male patients every 2 years.

## Referral

- If any **serious hepatic complications**, refer to [Emergency Department](#).
- Refer for [urgent or routine gastroenterology referral](#) if suspected cirrhosis suggested by one or more of:
  - Evidence of cirrhosis on imaging
  - Platelet count < 120 x 10<sup>9</sup>/L
  - Ascites
  - Hepatic encephalopathy
  - [AST to platelet ratio index \(APRI\)](#) > 2.0

## Information

### For health professionals

#### Further information

- ASHM:
  - [Hepatitis B Resources](#)
  - [Hepatitis C Resources](#)
- Australasian Hepatology Association – [Decision-Making in Viral Hepatitis Related Advance Liver Disease](#)
- Cancer Council:
  - [Optimal Cancer Care Pathway for People with Hepatocellular Carcinoma: Quick Reference Guide](#)
  - [Optimal Cancer Care Pathway for People with Hepatocellular Carcinoma](#)
- Therapeutic Guidelines (eTG) – [Gastroenterology: Advanced Liver Disease](#) [subscription required]

### For patients

- Better Health Channel – [Cirrhosis of the Liver](#)
- GESA:
  - [High Protein, High Energy Diet: For Advanced Liver Disease](#)
  - [No Added Salt Diet](#)

## References

#### References

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