CONDITION

Chiari-budd syndrome

Learn about Chiari-Budd syndrome, more commonly called Budd-Chiari syndrome, including symptoms, causes, diagnosis, treatment, complications, and follow-up care.

WHAT IT IS
Rare disorder blocking blood flow out of the liver

BODY SYSTEM
Digestive system

MAIN SYMPTOMS
Upper-right abdominal pain, abdominal swelling, leg swelling, jaundice

WHAT CAUSES IT
Usually blood clots, sometimes structural or unknown

HOW IT IS DIAGNOSED
Doppler ultrasound, contrast-enhanced CT or MRI

MAIN TREATMENT
Anticoagulation and procedures to restore blood flow

CAN IT BE CURED?
Sometimes - blocked veins can be reopened

IS IT AN EMERGENCY?
Sometimes, with vomiting blood or severe confusion

LONG-TERM OUTLOOK
Variable, with risks of cirrhosis, liver failure, and liver cancer

Overview

Chiari-Budd syndrome is more commonly called Budd-Chiari syndrome . It is a rare disorder in which blood cannot flow normally out of the liver. The blockage may affect one or more hepatic veins, which carry blood away from the liver, or the nearby part of the inferior vena cava, the large vein that returns blood to the heart.

When blood backs up in the liver, pressure rises inside its blood vessels. The liver may become enlarged and congested. Fluid can collect in the abdomen, and liver cells may be injured. The illness can develop suddenly, over several weeks, or gradually over months or years.

Budd-Chiari syndrome matters because untreated obstruction can cause portal hypertension, liver scarring, or liver failure. Portal hypertension means increased pressure in the vein system that carries blood from the digestive organs to the liver.

The condition is not the same as liver congestion caused by heart failure or disease of the sac around the heart. It is also different from sinusoidal obstruction syndrome, which affects the liver's smallest blood channels and may occur after certain cancer treatments.

Early recognition and treatment can restore or redirect blood flow, control complications, and protect liver function. Care usually involves a liver specialist and may also include blood-disorder and interventional radiology specialists.

Symptoms

Symptoms vary with how quickly the veins become blocked and how much of the liver is affected. Some people have few symptoms at first, especially when the obstruction develops slowly.

Possible symptoms include:

Long-standing disease may produce signs of portal hypertension. These can include enlarged veins in the esophagus or stomach, known as varices , and an enlarged spleen. Varices may bleed.

Symptoms alone cannot confirm Budd-Chiari syndrome because several liver, heart, and blood-vessel conditions can cause similar problems.

Seek emergency care for vomiting blood, black or bloody stools, fainting, severe confusion, marked difficulty breathing, or sudden severe abdominal pain with rapid swelling. These may indicate serious bleeding, liver failure, or another urgent condition.

Contact a clinician promptly for new jaundice, increasing abdominal or leg swelling, reduced urine output, fever with abdominal pain, or a clear decline in alertness or general health.

- Pain or discomfort in the upper right side of the abdomen
- Abdominal swelling caused by fluid, known as ascites
- An enlarged or tender liver
- Swelling in the legs or ankles
- Nausea, reduced appetite, or feeling full quickly
- Fatigue and weakness
- Yellow skin or eyes, called jaundice
- Shortness of breath from severe abdominal swelling or fluid around the lungs

Causes and risk factors

Budd-Chiari syndrome usually develops because a blood clot forms in the veins that drain the liver. Less often, a narrowed vein, membrane-like obstruction, tumor, or other structural problem blocks blood flow.

Many affected people have an underlying condition that makes the blood more likely to clot. More than one risk factor may be present.

Important associated conditions include:

Having one of these factors does not mean that a person has or will develop Budd-Chiari syndrome. In some cases, no clear cause is found even after a detailed evaluation.

The pattern of causes can differ by region. Obstruction involving the inferior vena cava is more common in some parts of Asia and Africa, while clotting within the hepatic veins is a frequent pattern in Western countries.

- Myeloproliferative neoplasms , which are bone marrow disorders that cause abnormal blood-cell production
- Inherited clotting tendencies, such as factor V Leiden or deficiencies of certain natural anticoagulant like warfarin or apixaban proteins
- Antiphospholipid syndrome, an immune disorder associated with blood clots
- Paroxysmal nocturnal hemoglobinuria, a rare acquired blood disorder
- Pregnancy and the period soon after delivery
- Estrogen-containing medicines, including some contraceptives
- Certain inflammatory or autoimmune diseases
- Cancer that increases clotting or directly compresses or enters a vein
- Abnormal narrowing or web-like tissue in the inferior vena cava or hepatic veins

Diagnosis

Diagnosis begins with a medical history and physical examination. A clinician may ask about the timing of abdominal swelling, previous blood clots, pregnancy, medicines, cancer, autoimmune disease, and family history of abnormal clotting.

Blood tests can assess liver injury, liver function, blood-cell counts, kidney function, and blood clotting. Results vary widely and may not show how severe the vein obstruction is. A person can have significant Budd-Chiari syndrome without extremely abnormal liver tests.

A Doppler ultrasound is usually the first imaging test. It uses sound waves to examine the liver and show the direction and speed of blood flow. It may reveal absent or reversed flow, narrowed veins, blood clots, or alternate vessels that have developed around an obstruction.

Contrast-enhanced CT or MRI can provide more detail. These tests may show which veins are affected, whether the liver has changed in size or shape, and whether there are blood clots, enlarged collateral vessels, or suspicious liver nodules.

Venography involves placing a thin tube into a vein and injecting contrast material to outline the vessels. It may be used when other imaging is unclear or when a procedure to open the vein is being planned. Pressure measurements may be taken during the same procedure.

A liver biopsy is not always needed. It may help when imaging is inconclusive or when another liver disease is possible. The biopsy can show congestion, liver-cell injury, or scarring, but these findings must be interpreted alongside imaging.

Testing for an underlying clotting disorder is an important part of the evaluation. This may include genetic tests, tests for antiphospholipid antibodies, and assessment for bone marrow disorders such as testing for a JAK2 gene variant. Some clotting-test results can be altered by acute illness, liver dysfunction, or anticoagulant treatment, so a specialist may need to repeat or interpret them carefully.

Treatment and management

Treatment has several goals: prevent additional clots, improve blood flow out of the liver, manage portal hypertension, and treat the underlying cause. The approach depends on the location and length of the obstruction, liver function, symptoms, and response to earlier treatment.

Most patients are considered for anticoagulation , which means medicine that reduces the blood's ability to form new clots. These medicines do not remove fixed scar tissue or every existing obstruction, but they can help prevent clot extension and new thrombosis. Many people require long-term treatment. The choice and duration must be individualized because liver disease, bleeding risk, pregnancy, kidney function, and planned procedures can affect which medicine is appropriate.

Ascites may be managed by reducing dietary sodium and using medicines that help the kidneys remove excess salt and water. Large or uncomfortable fluid collections may require paracentesis , a procedure in which fluid is drained through a small tube. Treatment plans should avoid excessive fluid loss that could affect the kidneys or circulation.

Other supportive care may include treatment for varices, nutrition support, management of

infections, and treatment of confusion caused by severe liver dysfunction. Alcohol may worsen liver injury, so the clinical team may advise avoiding it. Medicines and supplements should be reviewed because some can increase bleeding or harm the liver.

When there is a short, reachable narrowing, a specialist may use angioplasty to widen the vein with a small balloon. A stent may be placed to help keep it open. Selected recent clots may be treated with catheter-based clot removal or clot-dissolving therapy, but these procedures are not suitable for everyone and can cause serious bleeding.

If symptoms or liver injury continue, a transjugular intrahepatic portosystemic shunt , or TIPS, may be considered. During TIPS, an interventional radiologist creates a channel inside the liver that allows blood to bypass the blocked outflow pathway. This can reduce portal pressure and improve ascites. TIPS can also cause complications, including bleeding, blockage of the new channel, heart strain, or episodes of confusion.

Liver transplantation may be needed when other treatments cannot control the condition or when severe liver failure develops. Transplant evaluation considers overall health, the cause of the clotting disorder, and whether ongoing anticoagulation will be required.

An underlying blood or inflammatory disorder also needs treatment. For example, a hematologist may manage a myeloproliferative neoplasm or another clotting condition. Treating the associated disorder can lower the risk of further thrombosis, but it does not replace management of the liver-vein obstruction.

Complications

Budd-Chiari syndrome can cause both sudden and long-term complications. The risk depends on the extent of obstruction, liver damage, and response to treatment.

Possible complications include:

Not every liver nodule in Budd-Chiari syndrome is cancer. Altered blood flow can produce noncancerous nodules that may look unusual on imaging. Specialists may use repeat imaging, multiple imaging methods, or occasionally a biopsy to clarify the diagnosis.

- Persistent or recurrent ascites
- Portal hypertension and enlarged varices
- Bleeding from the esophagus or stomach
- Kidney dysfunction
- Bacterial infection of abdominal fluid

- Confusion from hepatic encephalopathy, which occurs when the liver cannot clear certain toxins
- Progressive liver scarring and cirrhosis
- Acute or chronic liver failure
- New or recurrent blood clots
- Narrowing or blockage of a stent or TIPS
- Liver nodules, including an increased risk of liver cancer in chronic disease

Follow up

Regular follow-up is important even when symptoms improve. The care team may monitor liver and kidney blood tests, blood-cell counts, clotting measurements, and the effects of anticoagulant treatment.

Doppler ultrasound or other imaging may be used to check hepatic-vein blood flow and confirm that an angioplasty site, stent, or TIPS remains open. People with chronic liver damage may also need monitoring for varices and liver cancer. The exact schedule depends on the individual's findings and treatment.

Patients should tell each clinician and dentist that they have Budd-Chiari syndrome and whether they take an anticoagulant. Anticoagulation should not be stopped or changed without guidance. Before surgery or an invasive procedure, the prescribing team may need to create a temporary plan that balances bleeding and clotting risks.

Routine appointments are appropriate for stable symptoms, medication review, test results, and planned imaging. Contact the care team sooner for increasing swelling, new jaundice, unusual bruising or bleeding, persistent fever, worsening fatigue, or problems taking prescribed medicine.

Emergency assessment is appropriate for vomiting blood, black stools, fainting, severe confusion, sudden severe abdominal pain, major uncontrolled bleeding, or serious breathing difficulty.

Sources

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