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NON CIRRHOTIC PORTAL VEIN THROMBOSIS

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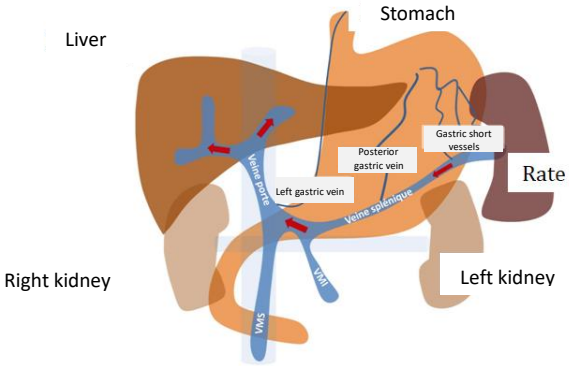
What is it?

This is the total or partial obstruction, by a blood clot, of the liver venous system. It can affect the portal veinous system from the right or left branch of the portal vein and/or its main trunk, and be extended to the superior mesenteric vein (SMV) or inferior mesenteric vein (IMV) and/or splenic vein. It may rarely be associated with a sus hepatic vein thrombosis (see brochure dedicated to Budd-Chiari syndrome) (figure



Is it frequent?

It occurs in only 0.7/100,000 people per year in Europe.

What are the clinical manifestations?

The majority of patients present with acute abdominal pain severe or not, which explains why the diagnosis may be delayed or incidental when the thrombosis has been totally asymptomatic. Liver tests are usually moderately and transiently disturbed. A biological inflammatory syndrome and moderate fever are currently observed in recent thrombosis. Peritoneal effusion so called ascites, is radiologically observed in half of patients. Rarely, this may be revealed by a hemorrhage with bloody vomit or blood in the feces. If the thrombosis extends to the mesenteric vein, there may be an intestinal infarction with bloody diarrhea or sometimes severe hypotension or shock.

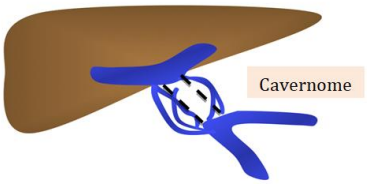
What are the causes?

Portal venous thrombosis usually occurs in patients with cirrhosis, but can also occur in the absence of liver disease. When cirrhosis is absent, several prothrombotic factors are associated in two out of 3 cases, responsible for a hypercoagulated state. Coagulation abnormalities must therefore be systematically screened by blood tests, and more rarely by bone marrow analysis. These abnormalities may be acquired with age, or constitutive. In this case there is often personal or family histories of other thrombotic events such as phlebitis or pulmonary embolism. In almost 30% of cases, an abdominal inflammation is present in the area of the portal vein, such as a gallbladder infection or an inflammation secondary to a recent abdominal surgery. Oestrogens contraception and pregnancy are considered at risk and must be stopped in case of portal vein thrombosis. Prematurity has also been incriminated as umbilical vein perfusion may be required. Finally, despite a meticulous screening, sometimes no cause can be found.

How is it diagnosed?

Doppler ultrasound is often performed as a first-line investigation of abdominal pain. The diagnosis of thrombosis must then be confirmed by an enhanced CT-scan which also allows to diagnose a local abdominal factor, to assess the extension of the thrombosis and to evaluate the condition of the intestine. If the thrombosis is chronic, a circulation may have developed around to shunt the obstruction. This circulation is called a cavernoma. The CT-scan can also rule out cirrhosis, or detect signs suggesting abnormalities of the small vessels of the liver (see the brochure dedicated to Porto-Sinusoidal Vascular Anomalies). The medical practitioner can

also measure the liver stiffness using elastometry (Fibroscan®). If liver disease is suspected, a liver biopsy may be required. A colonoscopy is also indicated at distance to screen a local inflammatory factor. Finally an upper endoscopy will be performed to screen varices in the upper digestive tract (see section on prognosis).



What is the prognosis?

A local abdominal infection can lead to portal vein infection or liver abscess. Intestinal infarction may lead to resection of the small intestine. Recanalisation is achieved in 6 months in 50% of cases under anticoagulant. If recanalisation is not achieved, a chronic portal thrombosis develops so called cavernoma. In this case, as pressure increases in the portal venous system (see brochure dedicated to portal hypertension), other veins may dilate and become varices, mainly around the oesophagus and stomach. These varices can bleed, causing hemorrhage. A cavernoma may be large and compress the common bile duct. The risk of thrombotic recurrence depends on the initial predisposing factor.

What are the treatments?

In emergency

In the absence of contraindication, curative anticoagulation either by subcutaneous injection or orally, depending on the situation, is administered to dissolve the clot and reduce the risk of extension. This treatment is recommended as soon as possible if the thrombosis is recent, and for at least 6 months. The decision will depend on the severity of the thrombosis and the level of risk of recurrence of the cause. Some procoagulant states require specific treatment. Oral oestrogens must be stopped permanently. Finally, local causes with abdominal bacterial infection have to be treated with antibiotics. In case of serious signs of intestinal damage, surgery may be required.

In the chronic phase

When oesophageal or gastric varices are present, a treatment with beta-blockers can prevent the risk of hemorrhage by reducing the pressure in the portal venous system. Varices can also be eradicated with elastic band ligations. In case of variceal rupture, these 2 treatments are combined. In case of severe complications of portal hypertension, radiologist can try to recanalise the portal venous system or place a stent between the sus hepatic veins and the portal vein (TIPS, see brochure dedicated to portal hypertension). Finally, in children, surgery to bypass the thrombosed area is sometimes possible.

Treating the cause

Once a cause has been identified, it must be treated, that's why monitoring usually requires the expertise of several medical practitioner all together (hepatologist, hematologist...).

How to monitor ?

An upper endoscopy is indicated at diagnosis to screen oesophageal or gastric varices and then regularly, in the absence of portal veinous system recanalisation. After 6 months of anticoagulation, a contrast enhanced imaging such as CT-scan can be used to assess recanalisation and must be repeated if symptoms recur. Regular blood tests are also required to ensure that there is no risk of bleeding or new thrombosis.

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