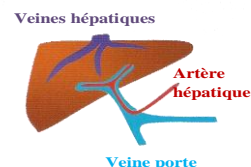


BUDD CHIARI SYNDROME

DEFINITION

It is a **liver disease** caused by the obstruction of hepatic veins. It is a **rare disorder**.



SYMPTOMS

Manifestations may vary a lot:

- no symptoms,
- fatigue,
- abdominal pain,
- progressive increase of the volume of the abdomen,
- leg oedema.

Less frequently:

- Gastro intestinal bleeding
- acute hepatitis,
- jaundice
- ascites.

DIAGNOSIS

Radiology procedures such as Doppler hepatic ultrasonography, Hepatic Magnetic Resonance Imaging (MRI) and hepatic computerised tomography usually visualize hepatic veins.

CAUSAL FACTORS

The combination of several predisposing **risk factors** is often necessary. The main one is a blood disorder: myeloproliferative disorder (overproduction of white or red blood-cells or of platelets). This disease is acquired, it is not transmitted by parents at birth.

Other predisposing risk factors are **congenital anomalies**. The main anomalies are the mutation of factorV Leiden (prothrombin gene mutation) and natural anticoagulant deficiency (protein C, protein S, antithrombin).

Other risk factors are **exogenous**: oral contraceptives containing oestrogen, pregnancy.

DIAGNOSIS OF THE CAUSES

A blood test is performed to make a diagnosis.

In the case of myeloproliferative disorder, thorough tests such as osteo-medullary biopsy or blood volume measurement may also be necessary

TREATMENT

Anticoagulation therapy:

It fluidifies the blood and thus helps to prevent the formation of new thrombosis at risk of new vein obstruction. There are two types of anticoagulants: those which are taken orally and those which are given by subcutaneous injections.

Dosage must be strictly adapted to avoid two risks: insufficient anticoagulation (risk of thrombosis) and excessive anticoagulation (risk of bleeding). The oral treatment is monitored by regular blood tests to measure your INR.

Thrombolysis :

It is an injectable pharmacologic therapy which is mainly used locally on the site of the thrombus so as to dissolve it. This therapy is only used in the case of recent thrombosis.

Angioplasty and insertion of a STENT:

Such procedure is used to widen (angioplasty) a narrowed vein ("stenosed") and to place a metal scaffold (stent) at the site of dilation to prevent restenosis.

Transjugular intrahepatic portal-systemic shunt (TIPS):

This procedure consists in creating a bypass between the portal vein in the liver and the hepatic veins or the inferior vena cava.

Transplantation: Transplantation is performed when angioplasty or TIPS have failed.

The above mentioned procedures are performed **one after the other, as needed**, starting from the simplest and less invasive: anticoagulants, angioplasty and placement of a stent, then TIPS, and finally liver transplantation. Going on to the next step will depend on the response to the previous treatment (if it was technically feasible).

SIDE-EFFECTS

Complications of Budd Chiari syndrome can be **due to the disease itself** (apparition of ascites, varices bleeding, liver failure, infection...).

Other side-effects may **result from some treatments** (anticoagulant-related bleeding, complications after angioplasty or TIPS).

RECURRENCES

Recurrences are unlikely to happen if the anticoagulant drugs are taken as prescribed, the treatment well-monitored, if the cause of the disease is treated. To avoid the recurrence of thrombosis, it is thus essential to **take medications as directed**. If any problems should arise, speak about them to your practitioner without delay.

PROGNOSIS

The first two years can be difficult and specialised treatments are required. After this period, liver condition ceases to be a problem for 80% of the patients. Future treatments will focus on the cause of the disorder. A near-normal family and professional life is absolutely possible.

Source : Hepatic Vascular Disorder Reference Centre . Hepatology Unit Beaujon Hospital.