

For a better knowledge of the illnesses of the blood vessels

From the Medical Service to Everyday Life



Hepatic portal vein

The Blood Vessels of the Liver, the Blood Vessels of Life

The liver plays a major part in permanently overhauling the blood, and in particular the blood coming from the digestive tract, so as to adapt it to the needs of the other organs. To do so, as for any other organ, it disposes of an artery (which enters the liver), and of hepatic veins (which leave the liver). However, as a unique exception in the organism, the liver also disposes of a vein which enters it (from the intestine, from the stomach and from the spleen): this is the portal vein. The total quantity of blood reaching the liver (and leaving it) represents at any given time one quarter of the amount which is pumped by the heart. Of this quantity, one third comes by the hepatic artery and two thirds by the portal vein.

The Assault on our Blood Vessels

The main damage is the obstruction of the hepatic vein (Budd-Chiari syndrome) or of the portal vein (pylephlebitis or cavernoma of the portal vein). In certain cases, the obstruction is subordinate to a compression due to a close injury. More often than not, the impairment is called primary: thrombosis (formation of a blood clot) triggering off a phlebitis (inflammation of the vein). This thrombosis happens in a situation where there is a tendency for an exces of blood to coagulate, or in a prothrombotic state. Certain states are hereditary: facteur V Leiden, mutation of factor 2, or a deficiency of protein C, of protein S or of anti thrombin. Others are acquired during the course of a lifetime: myeloproliferative disorder, antiphospholipid syndrome, paroxysmal nocturnal hemoglobinuria (PNH), oral contraception, or pregnancy.

As a general rule, it is necessary for there to be a combination of several causes. The reasons for which a thrombosis occurs in the hepatic vein, or the portal vein, and not elsewhere in the organism, are still not clarified.

Signs of our illnesses

When a thrombosis is taking place, the signs can be very different: either there are no signs, or they can be commonplace; or they can be quite outstanding like an extreme fatigue, pains, temperature, abdominal swelling due to liquid (ascites), or jaundice. Any or all of these symptoms may occur. The initial signs may lessen or disappear or worsen. Later, these signs show the difficulty in the bloodflow through the hepatic vessels (portal hypertension): numerous various parts in the abdomen of which some, the esophageal varices, may be the cause of a digestive haemorrhage. In the case of a thrombosis of the hepatic veins, a chronic ascites may set in as well as signs showing that certain functions of the liver have been affected (jaundice, sensitivity to bacterial infections, cerebral functions impaired, decrease of coagulation factors, and blood albumin). In the case of portal thrombosis, spreading of the thrombosis can cause serious intestinal pain. However, even if the onset was very pronounced, damage to the blood vessels may go completely unnoticed. Indeed, the organism disposes of very efficient means of compensation.

An early diagnosis is decisive for our future

An early recognition of a vascular injury enables us:

- To property identify the causes, which will be treated ;
- To put a stop to any deleterious consequences for the liver or for the intestine ;
- To avoid the spreading of the thrombosis to the undamaged reins ;
- Or even to obtain a regression of certain thrombosis by the treatment.

In 90% of cases, a diagnosis is obtained by non-agressive tests (Doppler ultrasound, Scanner or MRI) which give a view of the vessels. The quality of the tests is increased by having an experienced radiologist who is informed by the doctor of a possible vascular injury. Once the diagnosis of a hepatic vascular injury has been made, often an opinion should be given by a hemotologist who is familiar with the particular nature of hepatic thrombosis or portal vein thrombosis in order to identify the pro-thrombotic state by means of the appropriate examination

Existing treatments

Cooperation of experts of many disciplines is necessary: specialised laboratories, hematologists, radiologists, hepatic surgeons, anaesthetists, resuscitators, and of course hepatologists.

The first stage is to introduce the specific treatment for any existing causes: to administer anticoagulants to treat severe symptoms (acites, renal insufficiency, infections, and digestive haemorrhage). These treatments are codified today in the department of the Digestive Surgery Department (Services d'hépato-gastroentérologie).

The second stage is to consider restoring permanently the permeability of the vessels affected by thrombosis (percutaneous angioplasty of the hepatic veins or of the vena cava).

In case of failure, the third stage, for the hepatic veins, consists of creating a derivation between the portal system and the cava system which short-circuits the obstacle of the hepatic veins. At present this technique of TIPS is used (thus avoiding any surgery).

In case of a new set-back, a liver transplant must be considered.

The evolution has been transformed by these different treatments since, over 30 years of constant improvement, the picture of potentially lethal illnesses after a few months or years has changed to illnesses which, in 85% of cases, have become inconspicuous, thanks to continuous treatment and regular supervision.

Downtown Reference Center for Liver Diseases

This is a multidisciplinary team which includes hepatologists, radiologists, surgeons, hematologists, and anatomical pathologists. This team has a particular experience in several fields :

- The medical care of the patients suffering from an illness of the hepatic vessels.
- Research on the diagnostic and therapeutic techniques, and the identification of the causes. Laboratory research on animals regarding liver diseases, or on the taking of blood samples or biopsies of patients.

The Center's mission is to facilitate patient care as close as possible to their home, and at the highest standards, by offering advice and logistical or technical help to the local doctors. The team also has a mission to increase its knowledge regarding these illnesses through its work on research, and to disseminate such information.

The Center can be contacted at Hôpital BEAUJON, Clichy (tel : +331 40 87 51 60

Numerous problems to be overcome

Immediate medical requirements are to :

- Identify the factors of thrombosis yet unknown.
- Have simpler tests at our disposal to identify the factors known more particularly the myeloproliferative disorder.
- Have at our disposal anticoagulants more easy to handle by a greater interval between the therapeutic effect and overdose.
- Have at our disposal more efficient treatments which are better tolerated for certain causes, notably for blood disorders.
- Be able to judge more accurately the perfect time for the various therapeutic procedures (sooner or later).
- Be able to raise the level of successes and diminish the complications due to therapeutic procedures (angioplasty and TIPS).

Regarding the patients' problems:

- Assure a medical follow-up wherever we live.
- Relieve the patients (suffering caused by ascites) and support them.
- Make available a more humane and modern hospitalisation which respects both the patient and relatives.
- Increase the information to be received by patients and their families.
- Take into account the geographical distances between the specialised medical centers.
- Reduce our difficulties in continuing our professional activities.

- Take into account the impact that the illness has on the families of the diseased.
- Improve any financial aid and reduce the reimbursement period.

Regarding the development of medical research

- Understand why a thrombosis arises in the hepatic veins or the portal vein, and not elsewhere in the organism.
- Understand by which mechanisms certain blood diseases lead to a thrombosis.
- Understand how the irreversible after-effects (or sequelae) of thrombosis arise, and how to prevent them.
- Understand how ascites arise during the Budd-Chiari syndrome.
- Understand how the naturel mechanisms, for compensation after a veinal thrombosis develop, so as to be able to stimulate them.
- Understand what part the hepatic vascular obstructions play in the aggravation of frequent liver diseases, like cirrhosis of the liver due to a viral hepatitis or to an intake of alcohol.

Our Association

Created in October of 2005, AMVF has its Head Office at Hôpital BEAUJON, Department of Hepatology, 100 bd du Général LECLERC, CLICHY 92110.

The Association gathers together the sicks, their families and friends confronted by their illnesses. Its objectives are :

- To increase the knowledge of these diseases amongst the public concerned (doctors, the sick, the general public, government representatives and the public authorities).
- To create a bond between the patients to avoid loneliness, to inform them and to help and support them.
- To promote medical research to fight against the disease of liver vessels and to promote the creation of reference centers on these patholologies.
- To assure the financial support of the specific characteristics of the illnesses of the vessels of the liver, and to develop it.
- To reenforce the link between patients and medical practitioners, to insure a better all-round patient care.

WE, as well as the sicks and their relations, can act

To become a member of the Association means:

- To allow the Association to expand in terms of numbers, and thus be recognised by the public authorities and the medical profession.
- To participate in the accomplishment of our actions, through your help, your suggestions or your acts.
- To have the opportunity of sharing your difficulties, to formulate any questions you may have, to find a listening ear from people who, like you, have suffered from the various degrees of the illness.

• To be kept regularly informed, thanks to the news-letters issued with our Scientific Advisory Board and at the general meeting, regarding the different actions and projects carried out within our association, and the impact they have on patients' lives.

Together, we can help each other to fight the disease, so JOIN US

We are here to help you for any advice or information:

On Internet : <u>contact@amvf.asso.fr</u> Site : <u>www.amvf.asso.fr</u>

On Facebook AMVF

Our hotline at Beaujon (consult the dates on site)

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

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