Association des Malades des Vaisseaux du Foie

AMVF

PORTAL HYPERTENSION

THE PORTAL VENOUS SYSTEM

All the organs of the body receive their blood via an artery coming from the aorta. For most of the organs, the blood coming from the vena caval system drains into the inferior or superior vena cava. However, for the digestive system located in the abdomen (stomach, pancreas, small and large intestine) as well as for the spleen, the blood is collected by a particular venous system, the portal venous system, which drains into the liver (Fig. 1). The portal venous system brings most of the blood from the digestive tract to the liver. This blood carries around all food absorbed by the intestine, medicines taken orally, but also many hormones and substances secreted by the digestive organs, for example insulin from the pancreas.

In the liver, the portal vein branches out in many little veins similar to the arteries in order to irrigate liver capillaries. The level of blood flow in the portal vein is very high; it equals to about 20% of the cardiac output and corresponds to 2/3 rd of blood flow received by the liver, the liver artery bringing the remaining 1/3. (fig.2).

Fig 1 : System of the portal vein

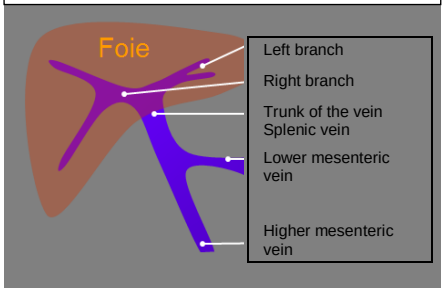
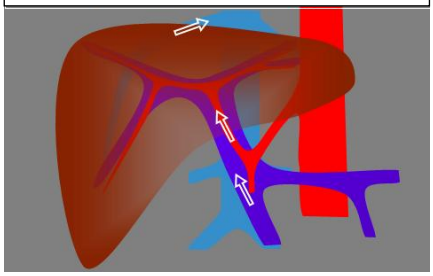


Fig 2 : Liver vessels and direction of blood flows



PORTAL HYPERTENSION

Portal hypertension is characterised by an increase in the blood pressure in the portal vein system**.**

CAUSES OF PORTAL HYPERTENSION

There must be an obstacle to the blood flow, in a strategic place, to cause portal hypertension. This strategic obstacle can be:

1. An obstacle of the portal vein trunk which is mainly represented by the thrombosis of the

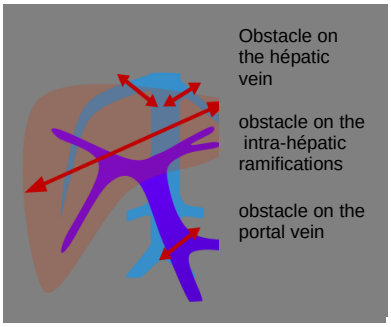


Fig 3. Different types of causes to portal hypertension

portal vein or its consequence, the entity called a cavernoma or cavernous trnasformation. Such a thrombosis is often due to a blood disorder which increases the risk of venous thrombosis in general.

2. The most frequent obstacle on the branches of the portal vein or liver capillaries is cirrhosis which is the result of a long lasting disease of the liver. These long lasting diseases of the liver are generally associated to drinking alcohol, diabetes or obesity, a chronic viral hepatitis B or C. However, there exists also damages to the portal vein ramifications or liver capillaries causing a portal hypertension without being due to a cirrhosis. These diseases proper to the small vessels have been called with different names which shows our lack of knowledge in regards to them. They were named as idiopathic portal hypertension, portal liver sclerosis**,** intra-hepatic portal hypertension non cirrhotic, occlusive portal venous disorder, or regenerative nodular hyperplasia.
3. The obstacle on the hepatic veins is represented by the syndrome of Budd-Chiari which is generally caused by a thrombosis. Thrombosis of the hepatic veins is often due to a blood disorder which increases the risk of thrombosis.

Blood disorders increasing the risk of thrombosis of the portal vein or the liver veins include mainly the myeloproliferative disorders, the antiphospholipid syndrome, inherited coagulation defects (V factor Leiden, factor II Leiden, deficiency in antithrombin, in protein C, or in protein S), or chronic inflammatory diseases, in particular those related to the digestive track.

CONSEQUENCES OF PORTAL HYPERTENSION AND ITS TREATMENTS

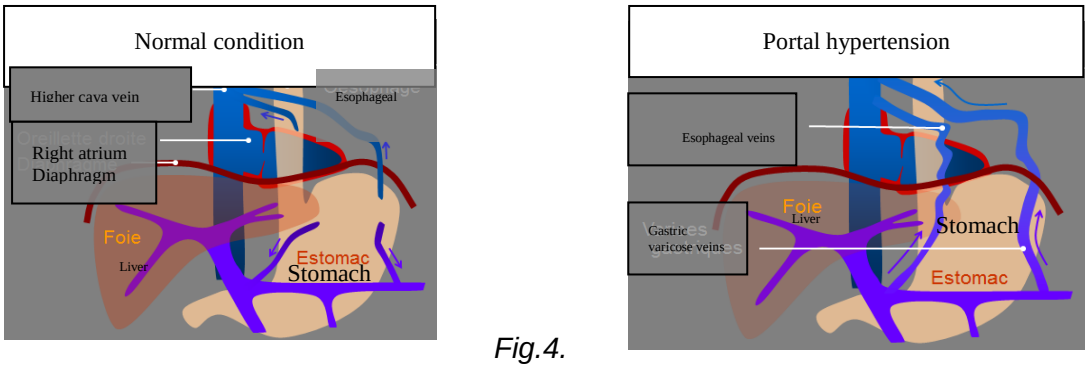


Fig.4.

Difference between normal condition (on the left) and portal hypertension (on the right) as regards the oesophagus and stomach venous drainage and leading to the formation of gastric and oesophagus varicose due to the obstacle on the portal vein, the liver or the liver veins.

a) **The increase in the portal pressure leads to a blood congestion** upstream of the obstacle, which can manifest in two ways;

1. **Ascites**: This is a watery liquid produced by the congested liver which accumulates in the abdomen. The congestion of the liver is due to the obstacle on the liver veins or the small ramifications of the vessels in the liver. However, in case of thrombosis on the portal vein there is no liver congestion and no ascites. The first treatment implemented for ascites is the intake of diuretics and a low-salt diet.
2. **Splenomegaly:** This is the increase of the size of the spleen, which is partly due to the congestion and partly to other more complicated mechanisms. In

the case of portal hypertension, the splenomegaly is accompanied by a decrease of white cells and platelets in the blood, which mechanism is once more complicated. We call hypersplenism, the association of a splenomegaly and the decrease of white cells and platelets. This decrease, when it is specifically due to the portal hypertension is never dangerous even when very noticeable.

b) **The difference of pressure between the portal vein system and the lower and higher cava veins system fosters the formation of veins between these two systems.** These veins are called shunts (or collateral veins, portacaval or portasystemic derivations)

These shunts can be seen as means used by the body to maintain the blood return towards the heart despite the obstacle on the portal system. These veins divert, direct away from the liver and into the general bloodstream, a part of the blood that should have gone first through the liver. These shunts can cause 2 types of complications: gastrointestinal bleeding and changes in the metabolism of some substances.

1. Gastrointestinal bleeding; some collateral veins run within the wall of the oesophagus or the stomach where they form varicose veins (Fig. 4 & Fig. 5). Varicose veins can crack and cause bleeding in the digestive track which can translate in fainting due to a strong decrease of the blood pressure or simply in an anaemia, or just with black stools (called melena) but sometimes with a more frightening vomiting of gastric liquid strongly coloured in red by red blood (called hematemesis)

Most of these bleedings stop by themselves. However they require a quick admission to hospital to take medicine to reduce the portal pressure and to have an endoscopy which will prevent a recurrence of the bleeding using simple mechanicals means. It may be necessary to perform a red cells transfusion.

These bleedings or their recurrence can be prevented, to a large extent, because they only affect the varicose veins that are large in size or look fragile, or those which already bled. An endoscopy done regularly (every 1 to 3 years) allows to detect them when no bleeding has occurred. Two types of treatments can then be implemented: medicinal or endoscopic.

The medicinal treatment: consists in the daily and permanent intake of Propranolol or nadolol without any risk in the absence of asthma, Raynaud syndrome, or some other heart problems. We think these medicines work in decreasing the portal pressure. They can affect some patients with a discomfort leading to stop the treatment for 15 to 20% of the patients.

The endoscopic treatment consists in fixing on the varicose veins small rubber bands which strangle them and make them disappear but this treatment is rarely definitive and a regular endoscopic check up of the oesophagus and stomach remains necessary. These treatments, (eventually combined with one another), prevent haemorrhages provided the program is properly followed.

3. **A** major **metabolic change:** Portacaval shunts carry blood from the gastrointestinal track to the general bloodstream without having been modified by the liver. Substances that are totally captured by the liver the first time they go through it, can then reach other organs, something which would

not have happened in a normal situation. The mechanism can disrupt the elimination of medicines but also the elimination of the substance produced by the digestive track. This is how we explain the drowsiness of the brain, reversible, called hepatic encephalopathy, due to the lack of elimination of ammoniac and other substances. This serious complication is thankfully rare in its permanent or recurrent form. The treatment of the encephalopathy consists of 2 medicines: lactulose or/and rifaximin are very efficient and very well tolerated. The mechanism of their beneficial action on the hepatic encephalopathy is still not well known.

A drastic way of correcting portal hypertension and to prevent bleeding is to connect directly the portal vein with the lower cava vein (anastomosis portacava) in building a bypass in the liver using a stent (fig.6). This method called TIPS is done with radiology reaching the venous systems by puncturing the right jugular vein at the neck level. The difficulty is to divert all the portal blood towards the general bloodstream with the metabolic consequences described above and the high risk of hepatic encephalopathy. The decision for this type of chirurgical intervention requires a careful analysis of the situation in a specialised centre.

Fig 5. Endoscopic view of oesophagus with varicose veins.

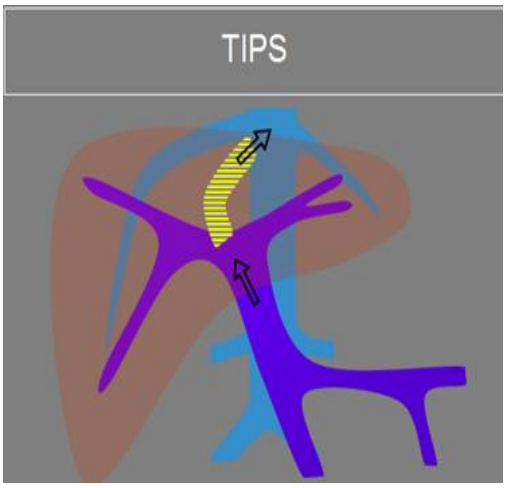


Fig 6: schema of connecting the portal vein system with the cava veins system with TIPS

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