

34. Portal Vein Thrombosis, Mesenteric Vein Thrombosis, Budd-Chiari Syndrome

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Q1: "I am 39 years old and have portal vein thrombosis and have been on coumadin® for 12 months. The thrombosis specialist that I have been seeing does not feel it is necessary for me to stay on coumadin since they have been unable to find a reason for the clots forming in my portal and splenic veins. I believe, that because they have not found the cause for my condition (other than the birth control pill) I should stay on coumadin®. Why would I want to take the chance of other clots forming? He seems quite optimistic that I am healthy and is open to me staying on coumadin®, until I am (mentally) ready to go off it...it is up to me."

A1: Unfortunately, in view of the lack of clinical studies it is not known what the best length of anticoagulant treatment is (see detailed discussion below).

Q2: "During my pregnancy I had numerous "abdominal attacks" that the doctors didn't know why I was having them. Eventually portal vein thrombosis was diagnosed. I had so much abdominal pain they had to take my spleen out. I do not fear FV Leiden, but wonder if I will loose any more organs. The concern is for my liver and bowels."

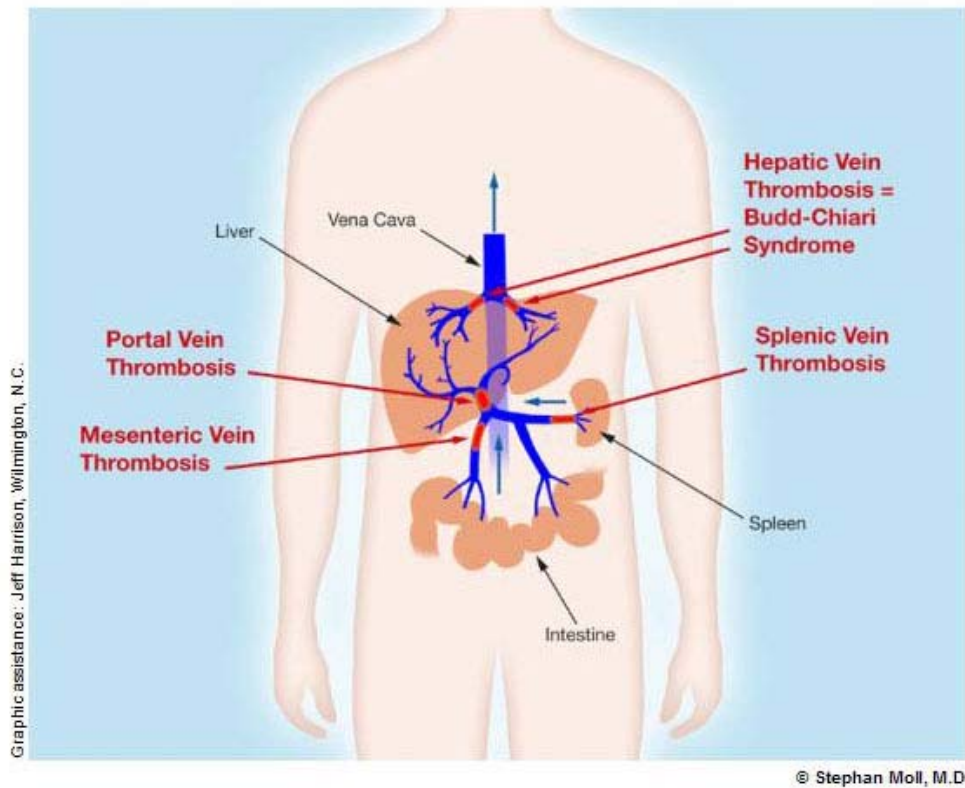
A2: In portal vein thrombosis the liver typically does not get damaged and functions normal, as opposed to hepatic vein thrombosis (= Budd-Chiari syndrome), which is a different disorder. The patient above should not be concerned about losing her liver. A recurrent clot in the portal vein could, however, lead to bowel damage that may make resection of part of the bowel necessary. Discussion about length of coumadin® therapy is essential for this patient (see discussion below).

Q3: "Thrombosis of the portal vein is called Budd-Chiari syndrome"

A3: This is not correct. In portal vein thrombosis the vein leading into the liver is clotted (the portal vein; see figure 2). In Budd-Chiari syndrome the veins leading out of the liver are clotted (the hepatic veins; see figure 3). The clinical problems that these 2 groups of patients experience are quite different.

Q4: "My mother is 58 years old. A CT scan of the abdomen showed extensive thrombosis of the portal vein and superior mesenteric veins. Reason for clots was not found yet. Urokinase was administered for 72 hours through IV, but clots were not dissolved. Heparin was administered through IV for some time and then warfarin was prescribed. What is the reason for the clot and how to dissolve?"

A4: There is no way telling from this CT report what the reason for the clot is. Q/A 49 lists reasons for clot formation that should be considered in this patient. Urokinase is a clot buster and was an appropriate attempt to dissolve the blood clots; heparin followed by warfarin is appropriate long-term treatment. Now the body just needs time to form collaterals (little veins that bypass the clot and enlarge over time to drain the intestine).



Anatomy

The portal vein is the major vein in the abdomen that collects the blood from the intestine and the spleen and channels it into the liver (see figure). Within the liver the portal vein splits into 2 major branches, the left and the right portal vein, and then into a fine meshwork of very small vessels that bring the blood in close contact with the liver cells. The liver cells absorb some of the toxic substances from the blood, and excrete into the blood essential proteins and lipids, which they have synthesized. The blood then collects into 4 major veins (the hepatic veins), which leave the liver and bring the blood into the big abdominal vein (the vena cava), which transports the blood to the heart.

Portal vein thrombosis, splenic vein thrombosis, mesenteric vein thrombosis

Portal vein thrombosis is a rare condition, affecting both children and adults. If the portal vein clots (= portal vein thrombosis), blood can not flow from the intestine and spleen into the liver and therefore backs up, leading to abdominal pain and enlargement of the spleen (= splenomegaly). The clot can also extend into the splenic vein, or the superior or inferior mesenteric vein; alternatively, the clot can be limited to those veins. The patient may have nausea, vomiting, diarrhea, and blood in the stool, and may notice increasing abdominal girth. The clot can occur acutely over a few hours, and be associated with severe acute symptoms. It can also occur slowly over several weeks or months and then lead to either mild or only intermittent symptoms. Since the blood cannot get into the liver, it finds ways to bypass the liver; small veins connecting the intestinal veins with the esophagus get bigger. These enlarged veins (= esophageal varices) can rupture and bleed, and the patient may vomit blood or pass blood in the stool.

Causes for portal vein thrombosis are

1. liver cirrhosis,
2. abdominal infections (appendicitis, inflammation of the bowel),
3. abdominal surgeries, such as spleen removal,
4. severe abdominal trauma,
5. blood clotting disorders,
6. umbilical vein catheterization in newborns,
7. no obvious cause, i.e. spontaneous (= idiopathic).

As with deep vein thrombosis of the legs, contraceptives, pregnancy, and hormone replacement therapy increase the risk for portal vein thrombosis.

Therapy consists of

1. surgery if the bowel has died due to severe pressure secondary to the back up of blood,
2. "clot-busters" (= thrombolytic therapy) in the case of an acute clot,
3. blood thinner (heparin and coumadin).

Taking coumadin not only protects people from further clots, but also puts them at risk for bleeding. Serious bleeds in people on warfarin occur in 3 % of people per year, life threatening bleeds in 0.25-0.5 % per year. One therefore always has to weigh the risks and benefits of long-term warfarin. Unfortunately, there is a lack of studies examining the risk/benefit ratio of warfarin in portal vein thrombosis. I am only aware of one study on this topic: *Gastroenterology* 2001;120:490-497. In view of the lack of data there is no right or wrong regarding length of warfarin therapy in patients with portal vein thrombosis. The 3 treatment options are:

1. discontinuation of warfarin after 6 months of treatment or more,
2. indefinite warfarin therapy, target INR 1.5-2.0,
3. indefinite warfarin therapy, target INR 1.5-2.0.

Personal comment: I usually keep patients who have had one episode of portal vein thrombosis on long-term warfarin, if they have tolerated warfarin well and if upper endoscopy does not show any significant varicose veins in esophagus or stomach. If varicose veins are found, then the risk of bleeding associated with them usually makes me suggest to the patient to stop taking warfarin.

Budd-Chiari Syndrome

Thrombosis of the liver veins is called Budd-Chiari syndrome (see figure). Blood can flow into the liver, but cannot flow out. It therefore backs up in the liver, leading to liver swelling and thus damage. Liver failure is the consequence and patients may need liver transplantation. Risk factors are the inherited and acquired thrombophilias, and also the blood disorders (a) PNH (= paroxysmal nocturnal hemoglobinuria) and (b) myeloproliferative syndromes (polycythemia vera, essential thrombocythemia). Whether patients should be on long-term warfarin after liver transplant has not been studied and is not clear. An individualized decision is necessary, depending on the risk factors that led to the Budd-Chiari syndrome.

References:

1. Condat B et al.: "Current outcome of portal vein thrombosis in adults: risk and benefit of anticoagulant therapy". *Gastroenterology* 2001;120:490-7.
2. Berney T et al.: "Risk factors influencing the outcome of portal and mesenteric vein thrombosis". *Hepato-Gastroenterology* 1998;45:2275-81.
3. "Current Concepts: The Budd-Chiari Syndrome". *New England Journal of Medicine*, Feb 5, 2004;350:578-85.