PORTOSYSTEMIC SHUNTS

Introduction

Portosystemic shunts (PSS) are abnormal pathways of the vascular system in which blood draining the intestines bypasses the liver and enters the main circulation. Because the liver does not have the opportunity to detoxify substances absorbed from the intestines, these accumulated substances can affect the brain and cause seizures and abnormal behavior.

Diagnosis

The diagnosis of portosystemic shunts can sometimes be challenging. A bile acid blood test is usually the first step to determine liver dysfunction. Patients with a portosystemic shunt usually have very high (over 100) values but they can also be only slightly elevated or even normal. Further diagnostic tests would include a transcolonic portal scintigraphy study (liver scan) and venous portogram (a specialized x-ray study). Scintigraphy has the advantage of not requiring anesthesia and is non-invasive. The disadvantage is that it does not give a precise location of the shunt. If a shunt is suspected based on scintigraphy but cannot be found at the time of surgery, a venous portogram is needed. The venous portogram requires anesthesia and open abdominal surgery to place a catheter in a portal vein (vein that should go to the liver.) A contrast material (x-ray dye) is injected at radiographs taken to trace the course of the dye. A shunt may be associated with a rectal vein and thus not show on a portogram or be found at surgery. Because the portogram increases anesthesia time and many of the patients are small, the definitive surgery is often done at a later date.

Treatment

Surgery is required to treat single portosystemic shunts. At surgery, a single extrahepatic shunt is either partially or totally ligated depending on theamount of pressure occurring in the portal system. Patients with total ligation seem to do better long term, however total ligation is not always possible. If there is too much venous stasis, shock and death can occur. There is controversy over the course of action if partial ligation is performed. Some surgeons re-operate one month later to ligate the shunt further. Some wait to see if fibrous reaction will further constrict the shunt with time. Scintigraphy or a portogram can be repeated and if shunting is still present, further ligation is performed. Another option for the surgeon is an ameroid constrictor. This is a stainless steel ring with ameroid (a casein material) that absorbs fluid, swells and thus slowly compresses and constricts the vessel.

The constriction (ligation) takes 30 to 40 days and the patient will continue to be treated medically during this time.

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Complications

The mortality rate with PSS and surgery is about 10%. Potential intraoperative complications include hemorrhaging and also tearing and bleeding from the shunt. Both can be life threatening. Potential complications after surgical ligation of the shunt include portal hypertension, bleeding, ascites (fluid accumulation in the abdomen) and uncontrollable seizures. The cause of the seizures is still unknown. There is a high mortality (death) associated with these seizures. Researchers are continuing to investigate the cause of seizures post surgical ligation.Removing the ligature does not resolve the problem.

Thirty percent (30%) of patients with ameroid constrictors have incomplete constriction/ligation and may require additional surgery.

Ascites (fluid leakage in the abdomen) may accumulate postoperatively anywhere from a few days to 10 days after surgery. This usually resolves on its own, but sometimes a diuretic is needed to decrease the fluid. Even after surgery, a special Prescription Diet or a homemade diet for liver disease will need to be fed until the liver has had a chance to improve its function.

Bile acid tests are usually rechecked periodically after surgery. Some patients clinically improve yet don't have a significant decrease in bile acids. VMSG