Current Concepts in Congenital Portosystemic Shunts

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KEYWORDS

• Shunt • Extrahepatic • Intrahepatic • Surgical management • Embolization

KEY POINTS

- Protein C level testing may be a useful blood test to indicate liver dysfunction.
- Many methods of diagnostic imaging are available, including ultrasonography, nuclear scintigraphy, CT angiography, and MR angiography.
- Commonly performed open surgical procedures for congenital portosystemic shunt (CPSS) include the placement of ameroid constrictors or cellophane bands.
- Less invasive options for the treatment of intrahepatic and extrahepatic CPSS include laparoscopic placement of cellophane bands and interventional radiologic techniques such as coil embolization.

INTRODUCTION

This article focuses on current concepts in portosystemic shunts. A thorough review of portosystemic vascular anomalies was published in 2009.[1] Recent information on portosystemic shunts is presented.

Congenital portosystemic shunts (CPSS) are vascular anomalies that occur secondary to inappropriate closure of different portions of fetal vasculature, resulting in intrahepatic or extrahepatic CPSS. Typically, a single CPSS is present, although multiple CPSS have been reported.[2] The presence of a CPSS allows portal blood to bypass the liver and enter the systemic circulation. Operative intervention is often recommended with the goal being slow closure of the anomalous vessel to gradually accustom the liver to increased blood flow and prevent the development of portal hypertension. This goal is often accomplished through open surgical techniques, including ameroid constrictor or cellophane band placement. These surgical procedures should result in CPSS closure over approximately 2 to 5 weeks and good clinical results. More

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recently, minimally invasive methods of CPSS occlusion have been described.[3–8] Laparoscopic placement of cellophane bands has been described in dogs with extrahepatic CPSS, and endovascular occlusion has been described in dogs with either intrahepatic or extrahepatic CPSS.[3]

DIAGNOSTICS

Diagnosis and treatment of CPSS in dogs has evolved with technology. Although some aspects of the diagnostic workup and treatment of CPSS have remained similar for several years, other aspects have changed dramatically.

Clinicopathologic Findings

Laboratory testing is among the first steps recommended in the diagnostic workup of dogs suspected to have CPSS. A complete blood count (CBC), serum biochemistry profile, urinalysis, and preprandial and postprandial serum bile acids, and/or ammonia level are recommended. Recently, some veterinarians have begun testing protein C activity.

Protein C is a plasma anticoagulation protein. Along with antithrombin, protein S, and plasminogen, protein C is important for preventing thromboembolic disease. Protein C is a vitamin K–dependent serine protease enzyme that is synthesized by the liver. Once activated, protein C works to promote fibrinolysis, modulate inflammation, and inhibit apoptosis. In human patients, measurement of protein C levels has been used to assess liver function. Low protein C activity levels have been reported in people with a variety of liver diseases including inflammatory hepatopathy, cirrhosis, portal vein obstruction, and neoplastic infiltration.[9,10] Protein C activity levels may be used to assess hepatic function in a variety of liver diseases. In dogs, protein C may be useful in distinguishing CPSS from portal vein hypoplasia (also known as microvascular dysplasia).[11] Dogs with CPSS have significantly lower protein C activities than dogs with portal vein hypoplasia.[11] When protein C activity is considered with other laboratory findings, it may be useful to distinguish CPSS from portal vein hypoplasia. Further, dogs surgically treated for CPSS show postoperative improvement in protein C activity. Therefore, protein C activity levels may be a useful test, in addition to other blood tests, to aid in monitoring dogs after surgical treatment of CPSS.[11]

In humans, inflammation has been shown to be associated with hepatic encephalopathy (HE). For that reason, markers of inflammation, such as C-reactive protein, have been measured in dogs with CPSS.[12] A difference in C-reactive protein concentrations has been detected between dogs with CPSS exhibiting HE versus those not exhibiting HE and dogs without HE.[12]

Abnormalities in the CBC and serum biochemistry profile may be seen in dogs with CPSS. Changes in the CBC may include leukocytosis, microcytosis, and normocytic, normochromic, nonregenerative anemia. Leukocytosis may be present owing to increased antigenic stimulation from decreased hepatic endotoxin and bacterial clearance from the portal circulation. Anemia is seen commonly in dogs with CPSS and is associated with abnormalities in iron metabolism. The exact pathogenesis of CPSS-associated anemia has not been described. Recently, 1 study evaluated CPSS-associated anemia with abnormalities in hepcidin. Hepcidin is a hormone synthesized mainly by hepatocytes. It controls iron transport by binding and inhibiting ferroportin, an iron export protein. No evidence that dysregulated production of hepcidin was associated with anemia in dogs with CPSS was found.[13]

Changes seen on serum biochemistry profile are varied and may include decreased blood urea nitrogen, hypoalbuminemia, hypoglycemia, and hypocholesterolemia.

Other abnormalities may include mild to moderate increases in serum liver enzyme activities. Determination of either bile acids and/or baseline ammonia level should be performed on dogs with suspected CPSS. Baseline ammonia level testing in fasting animals is nearly 100% sensitive, and ammonia tolerance testing is rarely necessary.[1,14]

Abnormalities in trace minerals also may be recognized. Manganese levels have been shown to be higher in dogs with CPSS than in healthy dogs or dogs with nonhepatic illness.[15] Manganese toxicity in humans and animals results in psychiatric disturbances, gait abnormalities, and cognitive deficits; therefore, elevated manganese levels may play a role in CPSS-associated HE.[15]

Diagnostic Imaging

Intraoperative mesenteric portography

Historically, intraoperative mesenteric portography was performed frequently to diagnose CPSS. This diagnostic modality is highly invasive but sensitive.[1] Findings on intraoperative mesenteric portography may be predictive of outcome after attenuation of CPSS.[16] The degree of intrahepatic portal vessel opacification during intraoperative mesenteric portography correlates with the ability to completely ligate the CPSS.[16] After CPSS occlusion, the degree of intrahepatic portal vascular opacification was greater in dogs without encephalopathy and in dogs with postoperative clinical improvement.[16] Another benefit of portovenography is the opportunity to measure portal venous pressures. The measurement of portal pressures may assist the surgeon in operative decision making.

Limitations of mesenteric portography include expense, difficulty in interpreting images based on patient positioning, and exposure of surgeon and staff to radiation. The major limitation of mesenteric portography is its invasive nature. Risks associated with laparotomy and general anesthesia may not be acceptable for owners, especially if a diagnosis of CPSS has not been made and the opportunity to treat concurrently is undetermined. Although this diagnostic modality may be used during laparotomy with treatment administered concurrently, the surgeon may wish to have a diagnosis before performing laparotomy. Less invasive methods (eg, abdominal ultrasonography, scintigraphy, MRI angiography, CT angiography) may be performed before surgery.

Abdominal ultrasonography

Abdominal ultrasonography is a noninvasive, accessible imaging modality that is used commonly to diagnose CPSS. Reported sensitivities and specificities associated with diagnosing CPSS using ultrasonography range from 80% to 95% and 67% to 100%, respectively.[17–20] Operator experience is important when diagnosing CPSS with abdominal ultrasonography; a marked decrease in false-negative results is reported with increasing operator experience.[18] Doppler ultrasonography has a reported sensitivity of 95% and specificity of 98%.[17] An experienced ultrasonographer may be able to determine the exact position and morphology of a CPSS. Ultrasound examination allows evaluation of the entire abdomen, including the urinary tract. Unfortunately, abdominal ultrasound examination of the abdominal vasculature is relatively time consuming and tedious.

Recently, the use of trans-splenic injection of agitated saline and heparinized blood has been evaluated to aid in the ultrasonographic diagnosis of CPSS in dogs.[21] Microbubbles are followed ultrasonographically through the portal system. Microbubbles cannot traverse the sinusoidal barrier of the liver in normal dogs; therefore, the presence of the microbubbles in the systemic circulation is the criterion used to diagnose CPSS. As described, the microbubble study is performed in a patient under mild to moderate sedation.[21] The spleen is identified by ultrasound and a needle connected

to an extension tube is introduced into the splenic parenchyma. The ultrasound transducer is repositioned to follow the microbubbles through the portal system. Agitated saline mixed with 1 mL of heparinized blood is injected into the spleen. The saline mixture is administered as a bolus over approximately 3 seconds and the ultrasound transducer monitors 3 locations in the abdomen. This technique is able to differentiate between intrahepatic and extrahepatic CPSS.[21] It is also able to differentiate between portoazygos and protocaval shunts and is useful to monitor dogs postoperatively for effective shunt attenuation.[21]

Scintigraphy

A nuclear portogram can be performed either trans-splenically or per rectum. Either method enables calculation of a shunt fraction, which represents the amount of blood that flows through the shunting vessel and bypasses the liver. Trans-splenic portal scintigraphy is able to provide information about shunt number and location, and requires less radionuclide than per-rectal scintigraphy, thus improving safety and allowing earlier operative intervention.[22] Sensitivity and specificity have been reported to be 88% and 67%, respectively for per-rectal scintigraphy[23] and 100% and 100%, respectively for trans-splenic scintigraphy.[22] Although trans-splenic portal scintigraphy has greater sensitivity and specificity and other benefits, complications can occur. Injection of the radionuclide into the spleen can lead to splenic hemorrhage. Similarly, injection of the radionuclide into the spleen may be difficult in smaller animals. Ultrasound guidance is often used for injection, but the injection may still occur outside the splenic parenchyma.

Computed tomographic angiography

Computed tomographic (CT) angiography is considered the best diagnostic imaging procedure for diagnosing portosystemic shunts (PSS) in humans.[24] CT angiography is gaining in popularity for use in diagnosing CPSS in dogs. It has been shown to be both sensitive (96%) and specific (89%) in dogs.[25] It is performed rapidly, provides 3-dimensional images, is noninvasive, and can provide excellent anatomic localization of shunt origin and insertion. CT angiography was shown to be 5.5 times more likely to correctly ascertain the presence or absence of CPSS compared with abdominal ultrasonography.[25]

Magnetic resonance angiography

MRI with angiography (MRA) is sporatically utilized as a diagnostic technique for CPSS. Magnetic resonance angiography provides 3-dimensional imaging of the shunt in good to excellent detail.[26] The exact anatomic position of the CPSS should be determined easily by MRA. Unfortunately, MRA can be time consuming and expensive. Reportedly, sensitivity and specificity of MRA for diagnosis of a single congenital shunt are 79% and 100%, respectively.[27] Although MRA is a promising new diagnostic modality for diagnosis of CPSS, CT angiography provides similar detail, is performed more quickly, and is often less expensive than MRA.

TREATMENT OF CONGENITAL PORTOSYSTEMIC SHUNTS

Surgical and medical treatments are available for dogs with CPSS. The goal of surgical treatment is to occlude blood flow through the shunt, thus directing portal blood through the available portal vasculature. The goal of medical management is to decrease the transport of factors absorbed from the gastrointestinal tract to the systemic circulation. One focus of medical management is reduction of ammonia absorption and systemic circulation. Both medical and surgical management play a role in the

treatment of dogs with CPSS and are often used in combination. A study found that the probability of survival for dogs with CPSS receiving medical treatment alone was lower than the probability of survival for dogs receiving surgical treatment.[28] A review of medical management of CPSS can be found elsewhere.[1]

Surgical treatment of CPSS in dogs and cats has evolved in the last 10 years. Most commonly used surgical options for attenuation of CPSS include placement of an ameroid constrictor (Research Instruments N.W., Inc, Lebanon, OR; researchinstrumentsnw.com) or cellophane band. Other minimally invasive options for identification and attenuation of CPSS include laparoscopy or interventional radiology.[1]

Preoperative Management

Proper patient management is appropriate before CPSS attenuation by ameroid constrictor or cellophane banding. Such management usually includes diagnostic workup and preoperative medical treatment. Once a diagnosis of CPSS has been made, medical therapy is usually instituted. Many surgeons recommend medical management of the CPSS patient to alleviate signs of HE before surgery, presumably to decrease blood ammonia levels and create a more stable anesthetic and surgical candidate. Preoperative medical management often consists of administration of oral lactulose, neomycin (or other non–orally absorbed antimicrobial), low-protein diet, and possibly anticonvulsant medication. One study has shown pretreatment with levetiracetam to be beneficial in preventing postoperative seizures.[29] Pretreatment with levetiracetam is instituted at least 24 hours before surgery and administered at 20 mg/kg by mouth every 8 hours. The study reports that no dogs pretreated with levetiracetam experienced postoperative seizures and 5% of dogs not treated with levetiracetam experienced postoperative seizures. All the dogs in this study underwent surgery for attenuation of an extrahepatic CPSS by ameroid ring constrictor placement.[29] Some surgeons elect to start proton pump inhibitors before surgical intervention, particularly for occlusion of intrahepatic CPSS.[1]

During the pretreatment period, the animal is prepared for surgery. Such preparation includes a preoperative fast, although a 12-hour fast in small patients with CPSS is not advised owing to the potential for hypoglycemia. Dextrose-containing fluids may be indicated to prevent hypoglycemia. Additionally, a small amount of easily digestible food may be administered until 4 to 6 hours preoperatively. Regardless, when fasting is instituted, the dog should be monitored for clinical signs of hypoglycemia and treated if appropriate.

Before and after induction of general anesthesia, monitor the patient for hypoproteinemia, hypotension, hypothermia, and hypoglycemia. Hypoproteinemia is common in dogs with CPSS. Depending on the severity of hypoproteinemia, colloidal support may be indicated. Hypotension and hypothermia are also common and are a serious concern for the anesthetic team.

Technique/Procedure

Open operative technique

With ameroid constrictor or cellophane band placement through a routine celiotomy, position the patient in dorsal recumbency, and perform a ventral midline abdominal approach. Take care while opening the abdomen to not transect a CPSS within the falciform fat. Identify the anomalous vessel. Different anatomic descriptions have been made for extrahepatic CPSS. Historically, CPSS have been categorized as either portoazygos or portocaval. Introduction of more advanced diagnostic imaging techniques (eg, MRA, CT angiography) has allowed more specific characterization of 6 types of CPSS: splenocaval, splenoazygos, splenophrenic, right gastric–caval, right

gastric–caval with caudal shunt loop, and right gastric–azygos with caudal shunt loop.[30] Splenophrenic and right gastric-azygos shunt with caudal loop are newly described anatomic variants of CPSS.[30] Splenophrenic shunts originate from the splenic vein and pass cranial to the liver to terminate in the caudal vena cava.[30] The right gastric–azygos shunt with caudal loop seems to have a component of right gastric–azygos and splenoazygos shunts, and it is likely that the right gastric vein is the dominant contributor to the shunting blood.[30]

Surgical treatment of all types of CPSS is similar when applying a cellophane band or ameroid constrictor. Identify the portal vein, because some dogs have portal agenesis or atresia, making attenuation of CPSS contraindicated. Inspect the caudal vena cava to identify the anomalous vessel. This process is usually accomplished by direct inspection of the vasculature, although intraoperative mesenteric portography may be performed occasionally. No large veins normally enter the caudal vena cava between the renal and hepatic veins. A shunt entering the caudal vena cava often produces turbulent blood flow with dilation of the caudal vena cava. These features may assist in the detection of the shunt. Inspect the area of the epiploic foramen. Also inspect other potential locations of extrahepatic CPSS, because multiple congenital CPSS may be present.[2] Other reported locations for CPSS include the falciform fat, gastrophrenic extrahepatic portocaval shunt, caudal abdominal CPSS, and portoazygous shunts. Gastrophrenic shunts are identified near the lesser curvature of the stomach, joining the phrenic vein at the level of the diaphragm. Caudal abdominal CPSS enter the caudal vena cava caudal to the renal veins. Portoazygous shunts cross the diaphragm and are found most commonly at the aortic or esophageal hiatus.

Place the occluding device (ameroid or cellophane band) as close to the caudal vena cava or diaphragm as possible. Create a small window in the tissues surrounding the shunt, and place the ameroid constrictor or cellophane band around the shunting vessel. Select an ameroid constrictor such that the internal diameter of the constrictor is equal to or slightly larger than the CPSS. With a cellophane band, fold the segment of cellophane in thirds longitudinally and pass it around the shunt. Loop the cellophane band back on itself, creating a ring around the CPSS. Compress the CPSS to a diameter of 3.0 mm or less with the cellophane band.[31] Alternatively, placement of the cellophane band without CPSS occlusion has been shown to be effective.[32] Secure the ends of the cellophane band together with hemoclips in an alternating pattern.[33] Placement of the constrictor as distal on the shunting vessel as possible enables inclusion of all shunt tributaries. Minimal dissection around the CPSS provides some stability to the ameroid constrictor or cellophane band to help prevent twisting or kinking of the shunting vessel.

Overall, the use of ameroid constrictors and cellophane bands for attenuation of extrahepatic CPSS results in good outcomes.[34] Long-term clinical outcomes for 206 dogs with an extrahepatic CPSS treated with an ameroid constrictor revealed that 7% of the dogs died within a month of surgery, 24% had continued shunting, and 92% had no clinical signs. Overall, 75% of dogs had successful outcomes, with 25% having unsuccessful outcomes. The median survival time was 153 months. In 22 dogs with extrahepatic CPSS treated by ameroid ring placement, CT angiography at least 8 weeks postoperatively revealed that, although none of the ameroid constrictors closed completely, only 18% of dogs had residual shunt flow.[35] Owners of dogs with residual flow reported no impact on the clinical condition of the dog, and reoperation was not recommended.

Testing of mechanical properties of cellophane film commonly used for banding CPSS revealed that only 1 of 4 films was cellophane.[36] Saline immersion and 3 methods of sterilization (ethylene oxide, gamma irradiation, and hydrogen peroxide)

decreased the strength of the cellophane bands.[36] Only autoclave sterilization did not weaken the wet cellophane.[36]

Minimally invasive technique

Vessel attenuation devices can be placed in patients with congenital extrahepatic CPSS by minimally invasive techniques, including laparoscopy and interventional radiology. The technique for laparoscopic surgery is similar to that described for placement of a cellophane band in an open surgical technique. Place the first scope portal just caudal to the umbilicus. Place 2 instrument portals (to the left and right) caudal to the first portal. Place 2 sutures through the body wall and into the ventral stomach to retract the stomach and improve visibility. Inspect the epiploic foramen and other areas in the description carefully. Dissect the CPSS from the surrounding tissues. Prepare a strip of cellophane by folding it longitudinally and securing each end to a piece of silk suture. Pass the cellophane into the abdomen and place it around the CPSS with the help of the silk sutures. Secure the cellophane with hemoclips placed in an alternating pattern while lifting the attached silk sutures. A more detailed description of laparoscopic placement of cellophane bands for attenuation of a CPSS is available.[3]

Interventional radiologic methods, using fluoroscopy, can be used to attenuate CPSS. Only a small incision is made to allow access to the vasculature (eg, femoral, saphenous, jugular vein) for introduction of instruments and implements.

Although interventional radiology is used more often in dogs with intrahepatic CPSS, it can also be used in dogs with extrahepatic CPSS.[4] The Amplatzer vascular plug has been used to achieve acute occlusion of the CPSS. To assess tolerance to complete occlusion of the CPSS, monitor heart rate and systemic arterial blood pressure during complete, temporary occlusion of the CPSS. An increase in heart rate or decrease in systemic arterial blood pressure during complete temporary occlusion of the CPSS was interpreted as the patient's intolerance of complete occlusion owing to portal hypertension. Only 1 of 7 dogs in the study was intolerant to acute occlusion.[4] No complications were observed in 6 dogs acutely occluded by an Amplatzer vascular plug.[4]

Coils have been used to attenuate blood flow through the CPSS.[5–7] Coil migration was more likely to occur in dogs with extrahepatic versus intrahepatic CPSS when a vena caval stent was not used. The technique of coil embolization of intrahepatic CPSS has been refined, including placement of a stent in the caudal vena cava to minimize coil migration.[6] Use of a vena caval stent in the coil embolization of extrahepatic CPSS in dogs has been described.[7] Confirm shunt location by cavography, and introduce a catheter into the shunt via the CPSS. Coils are deployed into the CPSS until they occupy more than 75% of the shunt diameter. Closure of the CPSS without development of portal hypertension results in most cases.[7]

Because many intrahepatic shunts are difficult to treat with an open surgical procedure, interventional radiology techniques may provide a better option. The endovascular treatment of intrahepatic shunts in dogs resulted in an excellent, fair, or poor outcome in 66%, 15%, and 19%, respectively. Dogs with an excellent outcome had absence of clinical signs without administration of low-protein diet or medications. Fair outcomes were seen in dogs with an absence of clinical signs on a low-protein diet or medications, whereas poor outcomes were dogs with continued or worsening clinical signs despite low-protein diet or medications, lack of response to surgery, or surgery-related death.

Postoperative Treatment

Monitoring of the postoperative patient is critical after surgical management of CPSS. Dogs undergoing treatment (either by open or minimally invasive techniques) may

exhibit portal hypertension, hypoglycemia, hypotension, or seizures. Portal hypertension is a serious complication and can be fatal. Monitoring for clinical signs of portal hypertension, including abdominal distension, abdominal pain, systemic hypotension, prolonged capillary refill time, pale mucous membranes, and gastrointestinal hemorrhage (usually evidenced by bloody diarrhea), is performed.[1] Mild portal hypertension resulting in ascites is often self-limiting.[1] Postoperative laboratory and imaging studies may be indicated. In the presence of severe clinical signs, disseminated intravascular coagulopathy, and hypotension unresponsive to fluid therapy, removal of the occlusion device may be required.

Hypoglycemia and hypotension are common postoperative complications. In 1 study, 44% of dogs had a blood glucose level of 60 g/dL or less postoperatively.[37] Hypoglycemia may be avoided in some cases by feeding small, frequent meals once the animal is awake from anesthesia. Additionally, intravenous administration of a dextrose solution is useful in preventing hypoglycemia. Regardless of preventative measures, blood glucose is monitored postoperatively. Monitoring for clinical signs of hypoglycemia, including lethargy, dull mentation, and seizures, is recommended. If indicated, dextrose may be administered as a bolus or added to the intravenous fluids. In some cases of refractory hypoglycemia, administration of dexamethasone (0.1–0.2 mg/kg intravenously) has been used successfully as treatment.[37] Hypotension may also occur postoperatively. Blood pressure is monitored regularly in postoperative patients, and treatment is administered if necessary. Treatment of hypotension may include administration of intravenous fluids, colloids and/or pressor drugs.

Postoperative seizures occur in 5% to 12% of dogs.[38,39] Postoperative seizures are often refractory to standard anticonvulsant drug treatment, and progress to status epilepticus.[39] Status epilepticus develops typically within 2 to 3 days postoperatively.[40] Pretreatment with levetiracetam is reported to decrease the incidence of postoperative seizures, as discussed.[29] Other treatments such as benzodiazepines, barbiturates, and propofol have been attempted with varying results.[41,42] Despite attempts at different treatments, a high mortality rate is associated with the development of postoperative status epilepticus.[38,39,42]

OUTCOMES

Dogs undergoing surgical treatment for CPSS often experience 1 of 3 long-term outcomes: closure of the shunt with improved portal blood flow, closure or partial closure of the shunt with improved portal blood flow and persistence of abnormal laboratory findings, or development of portal hypertension resulting in multiple acquired portosystemic shunts.

Ideally, surgery results in complete closure of the portosystemic shunt, resolution of clinical signs, and normalization of laboratory findings. Once the shunting vessel has been occluded, dogs experience increased liver volume,[43,44] presumably from hepatic regeneration.[45] In 1 study, all dogs undergoing surgery for CPSS attenuation had resolution of clinical signs, but 16% of dogs continued to have abnormal laboratory findings.[31]

In 18% to 21% of dogs, the portosystemic shunt does not close completely, resulting in residual blood flow.[35,38] Factors predictive of continued shunting are low preoperative plasma albumin levels, high portal pressure after complete shunt occlusion, and high portal pressure difference.[38] In dogs with persistent shunting, abnormalities in laboratory and imaging studies are present.[35,38] Residual shunting could occur secondary to failure of the primary shunt to close, or surgical error or presence of additional hepatic abnormalities.[35] Treatment recommended depends on the presence

of clinical signs and the cause of residual shunting. If blood flow is present in the primary CPSS and clinical signs are present, or if an additional CPSS is identified that was missed during the first surgery, operative intervention is recommended.[1] If no blood flow is detected in the primary shunt and no additional shunts are found, a liver biopsy may be considered to diagnose portal vein hypoplasia.

An additional cause of shunting in a postoperative dog is the development of multiple acquired PSS. Multiple acquired PSS may occur in nearly 10% to 20% of cases undergoing surgical treatment for a single CPSS and occur when the liver is unable to tolerate increased portal blood flow.[46] Such vessels open in response to elevated portal pressures. Treatment is focused on controlling clinical signs of HE and slowing progression of the liver disease.[1]

SUMMARY

Protein C level testing may be a useful blood test to indicate liver dysfunction. Many methods of diagnostic imaging are available including ultrasound, nuclear scintigraphy, CT angiography, and MRA. Commonly performed open surgical procedures for CPSS include the placement of ameroid constrictors or cellophane bands. Less invasive options are available for the treatment of both intrahepatic and extrahepatic CPSS and include laparoscopic placement of cellophane bands and interventional radiology techniques, such as coil embolization.

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