EXTRAHEPATIC PORTOSYSTEMIC SHUNTS

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Congenital portosystemic shunts (CPS) occur as a result of an error in embryonic development where primordial vessels either develop abnormal connections, or fail to undergo atresia. Many breeds have been reported as predisposed around the world, the most notable being a number of terriers (Yorkshire, Cairn, Maltese, Jack Russell), Chihuahuas, miniature schnauzers, pugs, toy poodles, Australian cattle dogs, Labrador and Golden retrievers and Irish Wolfhounds. In general, extrahepatic shunts are more common in terriers and other small breed dogs, whereas intrahepatic shunts are more common in medium to large-breed dogs. However, exceptions occur (eg Toy and Miniature poodles) and some form of shunt imaging is recommended prior to surgical exploration. Patients from breeds that are not known to be predisposed to CPS are more likely to “break the rules”, and should be evaluated especially carefully.

Surgical Therapy

The goal of surgery is to identify the shunt and attenuate it. As most patients have poorly developed hepatic portal circulation, and relatively high portal vascular resistance, total immediate occlusion of the shunt is highly likely to result in life-threatening portal hypertension. Therefore, surgical methods aim either to provide partial occlusion (which may be increased in stages by subsequent procedures) or application of a device to promote slow occlusion. The goal of slow or stage occlusion is to allow the intrahepatic portal circulation time to regenerate, with reduction in vascular resistance, at the same time as the shunt is being slowly or incrementally occluded. The main techniques used for slow occlusion of extrahepatic shunts are cellophane bands and ameroid constrictors. Cellophane bands promote occlusion by stimulating a foreign body reaction and fibrosis between the band and the shunt wall. Ameroid constrictors produce some mechanical occlusion as the ameroid clay takes up fluid, but also cause local inflammation with fibrosis and thrombosis of the shunt vessel. Recent reports have described the use of a variety of other clear films for shunt occlusion in dogs and cats, including polypropylene and polyethylene. These seem to be as effective as cellophane, probably due to the chemicals incorporated during the manufacturing process.

Peri-operative Monitoring and Treatment

Serum glucose concentration should be monitored regularly due to the tendency for hypoglycemia. Albumin levels may drop markedly when intravenous crystalloid is administered at surgical rates, and plasma transfusion is often required. Hemodynamic parameters should be measured regularly during and following surgery to gauge trends, and allow early detection of developing portal hypertension. Patients can be severely polyuric prior to surgery and may require increased maintenance fluid rates to avoid dehydration postoperatively. Potassium supplementation may be required.
Complications of surgery

The major life threatening complications are portal hypertension and postligation neurological dysfunction. The incidence of portal hypertension has been greatly reduced by adoption of the slow occlusion techniques, and publication of guidelines for safe shunt attenuation. Mild to moderate portal hypertension may not cause life-threatening problem; affected patients may have inappetance or diarrhea for a short period of time, or develop ascites. Assuming they are able to compensate hemodynamically for the reduced central venous return, and the bowel does not become critically hypoxic, they will eventually develop acquired shunts and a consequent return of portal pressure towards normal. Early published reports suggest that up to 20% of patients can be expected to develop acquired shunts, but more recent studies, in which patients underwent postoperative evaluation using DPCTA, showed that the incidence is probably less than 10%, with an additional 10% of patients experiencing ongoing shunting as a result of malplacement, or failure of the attenuating device to promote complete occlusion. Postoperative neurological sequelae range from blindness or muscle twitching, through disorientation, to generalized motor seizures and status epilepticus. Hypoglycemia and hepatic encephalopathy may also produce signs that can be difficult to distinguish from true postligation neurological dysfunction (PLND). In addition, patients experiencing seizures preoperatively may continue to have seizures postoperatively if the seizures were caused by something other than their CPS.

Hypoglycemia, hepatic encephalopathy and the seizures of idiopathic epilepsy should be manageable by evaluating the patient and relevant blood work, and treating accordingly. Post-ligation neurological dysfunction seizures and status epilepticus constitute a much more serious problem with a guarded prognosis. Pre-operative treatment with phenobarbital and levetiracetam (Keppra, UCB pharmaceuticals Inc, GA) has been shown to significantly reduce the incidence of seizures, although published reports and anecdotal experience suggest that the risk is not completely abolished.

Hepatic disease is a risk factor for abnormal wound healing, with a delay in development of wound strength. Skin sutures should be left if for at least 2 weeks following major surgery in these patients to reduce the risk of late dehiscence.

Therapeutic Decision-making

In light of the fact that many patients have mild clinical signs, and surgical shunt attenuation has a significant morbidity/mortality rate, owners are sometimes faced with a difficult choice between long-term medical management and surgical correction. Some dogs live apparently normal lives with no management at all, whereas others develop severe clinical signs within the first few months of life. On risk-benefit analysis, a young puppy with obvious hepatic encephalopathy is more clearly a candidate for surgery than an 8 year old dog in which CPS is detected on a routine wellness check. An exception would be the older male dog in which CPS is diagnosed following urethral obstruction with ammonium urate calculi. Medical management in these patients may not control stone formation, and as urethral obstruction is, in itself, potentially life-threatening, many owners will opt for surgical correction of the CPS. Likewise, patients with severe polyuria/polydipsia may be generally happy, but present unacceptable management issues, making surgery an attractive option.

Many dogs with severe clinical signs will become near-normal on medical management, prompting the owners to question the necessity for surgery. The choice then is between lifelong dietary management and other therapies, weighed against a high probability for return of liver function to normal following CPS attenuation.
**Prognosis**

The mortality rate following correction of CPS is less than 5%. Up to 80% of patients receiving slow occlusion devices for extrahepatic CPS can be expected to regain normal liver function, with a median survival of 152 months. (Falls et al). The mortality and morbidity rate are higher for surgical correction of intrahepatic shunts, and for cats with CPS of any anatomical type, and the chance of return to liver function is lower than for dogs with extrahepatic CPS.

The prognosis for patients undergoing PTCE has not yet been established definitively, however, early reports suggest that the mortality and morbidity rate is low, and patients are highly likely to show substantial clinical improvement, although complete closure of the shunt is unlikely.

Regardless of the technique used, the clinical impact of residual flow through the original shunt has yet to be determined. Intuitively, a reduction in shunt fraction should improve hepatic function, however, the actual shunt fraction at which a patient might resume a normal diet and not show clinical signs is not known.

Approximately 10% of patients will never return to normal liver function even following complete shunt occlusion and will likely require lifetime medical management, although interestingly they are often less severely affected clinically once acquired shunts have developed than they were with a single shunt.

**Reference**