What is a Portosystemic Shunt?

• A PSS is an abnormal blood vessel that delivers portal blood directly to the systemic circulation (caudal v. cava or azygos v. or their tributaries).

• Congenital PSS are birth defects and usually single.

• Multiple PSS are acquired from severe liver disease or high blood pressure and are NOT surgically correctable.

Breed Predisposition

• 90% of extrahepatic PSS develop in small breed dogs.
  – Risk for Yorkies is 36 times that of other breeds.
  – Other common small breeds: Maltese, Havanese, pugs, schnauzers, Jack Russells, dachshunds, shih tzus, poodles.

• Large breed dogs usually have intrahepatic shunts (known as patent ductus venosus if left sided).
  – Found in 2% of Irish wolfhounds in the Netherlands.
  – Labradors common in USA.
Clinical Signs

- Most dogs will have some signs:
  - “Poor doer”: small stature, underweight, poor hair coat.
  - Neurologic abnormalities: Quiet, confused, wobbly, drunk, blind, seizures.
  - Urinary: Pollakiuria or signs of urinary tract infection.
  - Gastrointestinal: Vomiting or diarrhea.

- No clinical signs noticed by owners in 5-10% of dogs.

Common Laboratory Findings

- Microcytosis.
- Decreased glucose.
- Decreased albumin, total protein, BUN.
- Decreased urine specific gravity.
- Ammonium biurate crystals.
  - Can progress to radiolucent calculi.
- Increased bile acids & ammonia.
- Decreased protein C activity.

Liver Biopsy Results: Hepatic microvascular dysplasia

- Microvascular dysplasia is not a disease but a pathologic finding.
  - Increased numbers of biliary ductules and arterioles, and small or absent portal venules.
- MVD occurs as a primary condition or as a result of reduced portal circulation.
Congenital Portal Vein Hypoplasia

- Underdevelopment of the microscopic portal system.
- Found in the same small breed dogs that get PSS.
  - Reported rarely in cats and large breed dogs.
- Hereditary.
  - Likely caused by the same genes as PSS.
- Sometimes called “microvascular dysplasia” (MVD or HMD) in the literature and on the internet.

Differentiating Congenital PSS and PVH

- Dogs with congenital PSS usually have classic signs; increased fasting and fed bile acids (>75 μmoles/L); microcytosis; and decreases in BUN, albumin, and protein C activity (88% of CPSS dogs).
- Dogs with CPVH often have normal fasting bile acids; fed bile acids ≤75 μmoles/L; and normal BUN, albumin, RBC size, and protein C activity (95% of CPVH dogs).

Definitive Diagnosis of Congenital PSS

- Ultrasonography
- Scintigraphy
- Mesenteric Portography
- Dual Phase CT
- MRA
- Exploratory Surgery
Ultrasonography

- Operator dependent.
- May require heavy sedation.
- Indications of EXPSS on ultrasound include increased kidney size, sediment in the bladder, small portal vein size, and abnormal direction or speed of portal vein flow.

Trans-splenic Scintigraphy

- Radioactive material is injected in the spleen, and the amount that shows up in the heart is compared to the amount in the liver.
- Animals must be “cleared” before release to owners.
- Spleenic scintigraphy can often tell location and number of shunts.
- Dogs with PVH-MVD are normal.

Portography

- The best images are obtained by injection of a splenic or mesenteric vein (usually in surgery).
- Sensitivity is 85%, 91%, and 100% in dorsal, right lateral, and left lateral recumbency, respectively.
CPSS: Medical Management

- **Protein restricted diet.**
  - 18-22% protein.
- **Lactulose or yogurt.**
  - Decrease ammonia production.
- **Antibiotics for urinary tract infection.**
- For severely affected dogs:
  - Intravenous fluids with dextrose.
  - Enemas.
  - Oral neomycin, metronidazole, or rifaximin ($$).

Diet

- Recommend liver specific:
  - Highly quality, highly digestible, moderately restricted protein.
  - Highly digestible complex carbohydrates.
  - Increased zinc, B-complex vitamins, antioxidants, fiber.
  - Restricted copper, sodium, and fat content.
- In 20 dogs with CPSS, soy diet resulted in lower HE score, decreased PT, lower NH3 compared with chicken diet.
- Apparent dietary requirement for experimental PSS was 2.11 g CP/kg/d (same as normal dogs.)
  - Greater restriction reduces ammonia metabolism within muscles.
- Dietary amino acid content less important that total protein intake.

Nutrition tailored to the patient....

Multiple acquired shunts and ascites 6 months after IHPSS ligation. Medically managed.

Appearance 2 weeks after owner added 20 eggs & 10 pieces of KFC per day!!

What about supplements for PSS or CPVH? 
**No one knows** if they help, but....

- Denosyl (Sam-e) improves function in dogs with hepatitis.
- Milk thistle (silymarin) improves hepatic function and regeneration in dogs with hepatitis.
  - Denamarin (chewable tablets) by Nutramax
  - HepatoSupport by Rx Vitamins for Pets
- Diets or supplements already have zinc and Vitamin E.

Hepatic Encephalopathy

- Precipitating factors:
  - Gastrointestinal bleeding
  - Dehydration
  - Constipation
  - Sedatives/anesthetics
  - Hypokalemic metabolic alkalosis e.g. diuretic therapy
- Seizures must be differentiated from HE.
  - HE is hard to recognize.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Consciousness</th>
<th>Intellectual and Behavior</th>
<th>Neurological Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal examination; if impaired psychomotor testing, consider MRH</td>
</tr>
<tr>
<td>1</td>
<td>Mild lack of awareness</td>
<td>Shortened attention span</td>
<td>Impaired addition or abstraction</td>
</tr>
<tr>
<td>2</td>
<td>Lethargic</td>
<td>Disoriented; inappropriate behavior</td>
<td>Obvious ataxic; slurred speech</td>
</tr>
<tr>
<td>3</td>
<td>Somnolent but arousable</td>
<td>Incoordination; bizarre behavior</td>
<td>Muscular rigidity and tonic hypertonia</td>
</tr>
<tr>
<td>4</td>
<td>Comatose</td>
<td>Comatose</td>
<td>Comatose progressing</td>
</tr>
</tbody>
</table>

Published studies on prognosis for dogs treated medically

- In one study, survival for surgically treated dogs was 87.9% and for medically treated dogs was 51.9% (median follow-up 597 and 1048 days, respectively).
- In another study, over half of medically treated dogs were euthanized an average of 10 months after diagnosis.

- Case series of 9 EHPSS, 17 IHPSS; no control group.
- Treatment: Diet, lactulose, antibiotics:
  - Hill's K/D, Hill's U/D, or homemade diet of cottage cheese, eggs, pasta, and rice.
- Ten dogs euthanized for uncontrolled neurologic signs (9) or persistent ascites (1).
- Weight and quality of life stabilized; liver enzymes improved; 6 dogs developed ascites.
- Survival time not correlated with bile acids, AP, ALT, MCV, TP, PCV.
- Predictors of survival: older age, higher BUN.

Greenhalgh et al. Comparison of survival after surgical or medical treatment in dogs with a congenital portosystemic shunt. JAVMA 2010;236:1215-20

- Nonrandomized prospective study comparing medical management to surgery (ligation, constrictor, or cellophane band).
  - Dogs that couldn’t be attenuated were euthanized.
  - Selection may have been biased based on owner, clinician.
- Treatment diet (16-24% protein) ± antibiotics, lactulose
  - i/d or l/d, Royal Canin Hepatic or Sensitivity, Eukanuba intestinal, homemade diet.
- Mortality from PSS related causes 30% for medically treated dogs and 10% for surgically treated dogs.
- Age at examination did not affect survival.
  - Could surgery be a second choice?

SA Center et al. Long-term survival of dogs with congenital or acquired PSS: 1980–2010. ACVIM 2012

- Unpublished, nonrandomized, retrospective study.
- EHPSS, 378 dogs; IHPSS, 105 dogs.
- Medical management (diet ± lactulose ± metronidazole), versus suture ligation to tolerance.
- Survival longer for dogs with EHPSS than IHPSS.
- No difference in outcome of medically and surgically treated dogs.
  - Survival curves did not exclude dogs that died of other causes.

<table>
<thead>
<tr>
<th></th>
<th>IHPSS Years (dogs)</th>
<th>EHPSS Years (dogs)</th>
<th>Yorkies Years (dogs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Surgery</td>
<td>8.0 (43)</td>
<td>10.5 (184)</td>
<td>11.8 (41)</td>
</tr>
<tr>
<td>Medical</td>
<td>5.5 (40)</td>
<td>10.5 (165)</td>
<td>11.8 (43)</td>
</tr>
</tbody>
</table>
CPSS Attenuation: Options

- Ameroid constrictor
- Cellophane band
- Partial or complete ligation
- Hydraulic occluder: IHPSS
- Coil embolization: IHPSS

Single extrahepatic gastrophrenic PSS emptying into left hepatic vein.

Ameroid Constrictors

- Hygroscopic inner ring absorbs water and causes swelling.
  - Secondary fibrous tissue reaction cause shunt closure within 4 weeks.
- Radio-opaque, so visible on x-rays.
Cellophane Banding

- Cellophane is folded into 3+ layers, wrapped around the shunt, and held in place with metal vascular clips.
- Gradual occlusion over 2-4 weeks.
- Degree of occlusion needed for eventual shunt fibrosis is thought to depend on type or thickness of cellophane. *Hunt ACVS 2011*
  - True cellophane is cellulose-derived.
  - Many other products are synthetic polymers that result in different types of postoperative behavior.

Pass folded cellophane around the shunt.

Hold ends together with a hemoclip.
Acute Suture Ligation

• Degree of ligation based on objective and subjective measurements of portal hypertension.
  – Portal pressure change
  – CVP and AP changes
  – Visceral color

• Partial ligation most common.
  – Required in 60% to 75% of animals because of portal hypertension.
  – Postop euthanasia of 35% at some point because of signs.
  – Clinical signs in 11%.

Transvenous Coil Embolization

• Primarily for IHPSS.
• Requires advanced training
  – Drs. Chick Weisse & Allyson Berent at Animal Medical Center, NY.
• A stent is placed in the CVC and coils placed through it into the PSS.
• Postoperative complications 10-15% but survival 98%.
• Lifelong omeprazole required.
• 10-25% require future coils.

Hydraulic Occluders

• Vascular access port placed subcutaneously; occluder cuff inflated gradually over an 8 week period.
• Revision required in 3/10 dogs.
  – Cuff rupture
  – Port detachment
  – Subsequent reinforcement reduced complications
Complications of Surgery

- Hypoglycemia
- Prolonged recovery
- Coagulopathy
- Gastrointestinal perforation
- Aspiration pneumonia
- Neurologic dysfunction
- Seizures in 3-18%
- Portal hypertension (most commonly with ligation).
  - Shock/death from over-attenuation
  - Portal vein thrombosis
  - Ascites from mild hypertension
- Persistent flow (most common with ligation).

Pretreatment with Anticonvulsants

- Pretreatment with KBr does not decrease seizure rate.
  - Seizures in 4/48 (8.3%) on KBr and 4/120 (3%) not on KBr
- Pre (IM) and postop (2 week) treatment with phenobarbitone may reduce risk of grand mal seizures but does not significantly reduce neurologic episodes in animals undergoing acute attenuation (ligation or tight cellophane).
  - Neurologic signs in 6% on phenobarb, 16% not on phenobarb.
  - Seizures in 0/31 on phenobarb, 4/58 not on phenobarb.
- Seizures in 0/42 pretreated with Keppra and 4/84 not treated with Keppra
  - 20 mg/kg PO q8h for minimum of 24 hours preop; 30 days postop.
  - Keppra more commonly used in dogs with encephalopathy.
  - Dogs on Keppra were more likely to have postop vomiting and diarrhea.

Canine Surgical Attenuation: Outcome Summary

<table>
<thead>
<tr>
<th>Method</th>
<th>Poor Short Term Mortality (%)</th>
<th>Morbidity (%)</th>
<th>Poor Long Term Outcome (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PL N=225</td>
<td>11.7</td>
<td>12</td>
<td>32</td>
</tr>
<tr>
<td>CL N=108</td>
<td>7.1</td>
<td>8.6</td>
<td>9.5</td>
</tr>
<tr>
<td>AC N=200</td>
<td>7.0</td>
<td>11.5</td>
<td>7.0</td>
</tr>
<tr>
<td>CB N=132</td>
<td>6.0</td>
<td>12.6</td>
<td>13</td>
</tr>
</tbody>
</table>

For review see Tivers MS et. al. J Small Anim Pract 2012;53:3-11
### Mortality Rates

<table>
<thead>
<tr>
<th>Modality</th>
<th>EHPSS mortality</th>
<th>IHPSS mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ligation</td>
<td>2-32%</td>
<td>6-23%</td>
</tr>
<tr>
<td>Ameroid constrictor</td>
<td>7%</td>
<td>0-9%</td>
</tr>
<tr>
<td>Cellophane Band</td>
<td>6-9%</td>
<td>27%</td>
</tr>
</tbody>
</table>

### Survivors with Good/Excellent Outcome

<table>
<thead>
<tr>
<th>Modality</th>
<th>EHPSS outcome</th>
<th>IHPSS outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ligation</td>
<td>50-93% (median, 75%)</td>
<td>75-100%</td>
</tr>
<tr>
<td>Ameroid constrictor</td>
<td>90-94% (median, 92%)</td>
<td>76-100%</td>
</tr>
<tr>
<td>Cellophane Band</td>
<td>79-100% (median, 87%)</td>
<td>50%</td>
</tr>
</tbody>
</table>

Tivers et al (Evidence Based Review), JSAP 2012;53:3-11

### Predictors of Outcomes

- Poorer outcome:
  - Low body weight (for IHPSS)
  - Preoperative encephalopathy
  - Preoperative hypoalbuminemia
    - Albumin of recovered dogs, 2.25 g/dL; nonrecovered, 1.9 g/dL.
  - Preoperative anemia
  - High preoperative WBC
  - High portal pressure after test ligation
  - Postop seizures, continued shunting
- Age not associated with outcome.
- Better in dogs with more hepatic opacification on portography.

### Postoperative Management

- Continue the protein restricted diet until liver function improves.
- Lactulose, yogurt, or antibiotics may be recommended for some dogs.
  - No research or standards of care.
- Recheck bile acids, protein C, and blood work in 3 months.
- Switch to adult maintenance or intestinal diet when blood work is normal.
What if the bile acids are still high after 3 months?

- If bile acids are abnormal but clinical signs, albumin and BUN are improving, continue protein restricted diet and retest in 3 months.
  - Other medications?
- If bile acids are very high and the dog has clinical signs or low BUN and albumin, either work up or retest in 3 months.
- If the bile acids are almost normal, try weaning off medical management?
  - Monitor NH3 and chemistry?

What if the bile acids are still high at 6 months after surgery?

- A scintigraphy, portogram, and/or liver biopsy needed to determine the cause.
  - Yorkies can have PSS and CPVH concurrently; this cannot be predicted before surgery.
  - Other differentials: Acquired shunts, second shunt, original shunt still open, other liver diseases.
- If the bile acids are only mildly increased, most likely the dog has PVH-MVD.
  - If the owners do not want further work up, stop checking bile acids.
  - If the owners do not want to do medical management, tell them not to give extra protein.

What is the outcome for dogs with congenital portal vein hypoplasia?

- No one knows for sure, because so many “normal” Yorkies and Maltese have PVH-MVD.
- Outcome is likely poorer in dogs with severe clinical signs or blood work abnormalities.
  - Mortality rate 18% over 5 years, with 79% of survivors in good to excellent condition.
- Lifespan also depends on development of other diseases that may affect the liver.
  - Steroid or barbiturate administration, Cushings.
  - Dogs with CPVH commonly have IBD (LP enteritis).
Useful Brochure for Clients

Summary:
Comparison Surgical Outcome in Dog and Cat

<table>
<thead>
<tr>
<th>Method (n)</th>
<th>Short Term Mortality</th>
<th>Morbidity</th>
<th>Long Term Outcome (Good)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dog (&gt;700)</td>
<td>8%</td>
<td>11.3%</td>
<td>85%</td>
</tr>
<tr>
<td>Cat (119)</td>
<td>7.6%</td>
<td>53%</td>
<td>46%</td>
</tr>
</tbody>
</table>


Three cats medically managed by K. Tobias:

- One had to be euthanized for recurrent urinary tract obstructions (requiring surgery) within 18 months of diagnosis.
- One had to be euthanized for progressive neurologic dysfunction and recurrent urinary tract obstructions within 2 years of diagnosis.
- One is alive more than 7 years after diagnosis; still on medical management.

Case presentation:
1 year old toy Havanese terrier

- Presented for 5 seizures over a 4 week period; no other clinical signs.
- Increased bile acids (60 and 80 µmol/L).
- Hypoalbuminemia (1.8 g/dl).
- Shunt possible based on ultrasound.
- Seizures less severe on lactulose, neomycin, and phenobarbital.

Differential Diagnoses

- Portosystemic shunt
- Portal hypoplasia (MVD/HMD)
- Liver hypoxia secondary to seizures
- Epilepsy
- Hydrocephalus
- Other causes of seizures
- Neuro exam WNL

Scintigraphy

Shunt Fraction 70%
Surgery

• Exploratory performed; no shunt located. Liver biopsy taken.
• Intraoperative mesenteric portography performed:

Options?

1. Repeat scintigraphy and recover until safe to handle.
2. Roll dog in lateral position and repeat portogram.
3. Recover and monitor bile acids and albumin and wait for liver biopsy results.
Outcome

- 3 mm vessel found under peritoneum/serosa of intra-abdominal portion of esophagus ("portophrenic shunt").
- 5.0 mm ameroid constrictor placed.

Small portophrenic shunt in another dog.
Postoperative Outcome

• Reduced mentation and weakness 2 days after surgery immediately before scheduled release.
  – Magnesium 1.1 mEq/L (normal 1.5-2).
• Recovered after magnesium infusion.
• Discharged on KBr, Hill’s l/d, lactulose.

Follow-up

• Pre- and postprandial bile acids and albumin normal 3 months after surgery (still on potassium bromide)
  – When do you discontinue KBr?
    • No seizures at 3 months postop
    • KBr at sub-therapeutic levels
• Weight gain and normal activity at 9 months; blood work normal.
• Seizures recurred 10 months postop
  – Screamed, went rigid, urinated, recovered quickly
  – Has been off KBr for months