Providing the best quality care and service for the patient, the client, and the referring veterinarian.

James Y Kim DVM, DACVS

Introduction

- Anomalous connection between the portal and systemic venous systems

- Bypass of the liver
  - Normal in utero (ductus venosus)
  - Abnormal postpartum
    - Patent ductus venosus
    - Intrahepatic
    - Extrahepatic
      - Single
      - Multiple

- Portal vein hypoplasia – Microvascular dysplasia
  - Abnormal hepatic bloodflow
  - Histopathologic finding
    - Small intrahepatic portal vessels
  - Communication between portal & systemic circulation at microvascular level
  - Occur concurrently w/ PSS
    - 58% of dogs & 87% of cats
  - PVH-MVD alone have similar signs to PSS
  - Cairn & Yorkshire Terriers
Liver physiology

- Protein metabolism
  - Albumin
  - Coagulation proteins
- Carbohydrate metabolism
  - Gluconeogenesis
  - Glycogenolysis
- Fat metabolism
  - Bile acids
  - Cholesterol production

Immune/Clearance
- Kupffer cells
- Toxic metabolites
- Bacterial endotoxins
- Drug metabolism

Storage
- Vitamins
- Fats
- Glycogen
- Trace minerals

Signalment

Extrahepatic
- Small/toy breeds
  - Yorkies
  - Maltese
  - Havanese
  - Pugs
  - Terriers

Intrahepatic
- Large breeds
  - Irish Wolfhounds
  - Retrievers (labs & goldens)
  - Australian cattle dogs
  - Australian shepherds

Clinical signs

- Neurologic
  - Lethargy, ataxia, obtundation, pacing, circling, blindness, seizures, coma
- Gastrointestinal
  - Vomiting, diarrhea, anorexia, pica, melena, hematemesis

Urinary
- Hematuria, stranguria, pollakiuria, urethral obstruction

Hepatic Encephalopathy (HE)

- Pathophysiology
  - largely unknown & complex
- >20 compounds in excess w/ PSS
  - Ammonia, aromatic AAs, benzodiazepines, GABA, glutamine, short chain FAs, tryptophan

- Ammonia
  - Excitotoxic
  - ↑ glutamate
  - Excitatory neurotransmitter
  - Chronically leads to ↑ inhibitory factors
  - GABA
  - Endogenous
  - Benzodiazepines
Diagnostics

Complete blood count
- Microcytosis
  - +/- normochromic, nonregenerative anemia
- Target cells
- Poikilocytes
- Leukocytosis
  - associated with a poor prognosis

Serum chemistry
- Hypoalbuminemia
- Reduced BUN
- Hypocholesterolemia
- Hypoglycemia

Urinalysis
- Hyposthenuria to isosthenuria
  - Hepatic encephalopathy (psychogenic PD)
  - Poor renal medullary gradient (decreased BUN)
- Crystaluria
  - Ammonium biurates
- Proteinuria
  - Glomerular sclerosis secondary to antigen exposure

Bile acids
- Paired samples
  - 12 hour fasting
  - 2 hour postprandial
- Normal values
  - Fasting 1.7 ± 0.3µM
  - Postprandial 8.3 ± 2.2µM
- Some normal Maltese dogs have increased serum bile acids
Diagnostics

- Baseline Ammonia
  - Abnormal in 62-88% of animals w/ PSS
- Ammonia tolerance test (ATT)
  - Administration of ammonium chloride per os or per rectum
  - Samples taken at prior administration & 30 minutes post
  - Contra-indicated in animals exhibiting HE

Protein C

- Vit K dependent serine protease enzyme
- Test to differentiate PSS from PVH-MVD
  - Normal >70%
  - PSS < 70%
  - PVH-MVD >70%

Imaging Diagnostics

- Nuclear Scintigraphy
  - Great screening tool
  - Highly sensitive
  - Noninvasive, requiring no anesthesia
  - Technetium pertechnetate

Imaging Diagnostics

- Transcolonic scintigraphy
  - Isotope infused per rectum into the colon
  - Radioactivity is visualized sooner and in higher concentration at the heart
Imaging Diagnostics

- Transsplenic scintigraphy
  - Sedation required
  - Ultrasound guided injection
  - Identify location and number of shunts


Imaging Diagnostics

- Abdominal Ultrasound
  - Highly operator dependent
  - Sensitivity and specificity increases with sedated or anesthetized patients
  - Findings
    - Shunting vessel
    - Microhepatica
    - Renomegaly
    - Cystic calculi

Imaging Diagnostics

- Computed Tomographic Angiography
  - Gold standard in human medicine
  - Dual-phase CT angiography superior
  - Identify non-surgical PSSs
**Portovenography**
- Performed during laparotomy w/ fluoroscopy
- Injection into splenic or jejunal vein w/ iohexol
- Sensitivity:
  - 85% in dorsal
  - 91% in right lateral
  - 100% in left lateral

**MRI**
- Extended anesthesia
- Cost prohibitive
- CT provides superior detail
- CT is extremely quick w/ multi-sliced scanners

**Medical treatment**
- **Antibiotics**
  - Treat bacteremia
  - Decrease bacterial ammonia production
  - Metronidazole:
    - 7.5mg/kg
  - Ampicillin
    - 22mg/kg
  - Neomycin
    - 22mg/kg

- **Lactulose**
  - Convert ammonia (NH₃) to ammonium (NH₄⁺)
  - Decrease intestinal transit time
  - 0.5-1.0 ml/kg PO q 6-8hrs
Medical treatment

- **Gastroprotectants**
  - Gastrointestinal ulcerations
  - Famotidine
    - 0.5-1.0mg/kg
  - Omeprazole
    - 1.0-2.0mg/kg
  - Misoprostol
    - 2-3µg/kg
  - Sucralfate
    - 40mg/kg

- **Anti-convulsants**
  - Phenobarbital
    - Loading 16mg/kg divided over 4 doses during a 12-24hr period
    - Maintenance 2-4mg/kg PO divided into 2-3 doses/day
  - Potassium bromide
    - Loading 400-600mg/kg divided over 1-5 days
    - Maintenance 20-30mg/kg/day
  - Keppra (levetiracetam)
    - 20mg/kg PO q 8hrs

- **Protein-restricted diet**
  - Moderate protein restriction
    - 18-22% dogs
    - 30-35% cats
  - Dairy & vegetable protein sources

- **Hepatosupport**
  - SAMe
    - 17-22mg/kg/d
  - Ursodeoxycholic acid (ursodiol)
    - 10-15mg/kg/d
  - Vit E
    - 15 IU/kg/d
  - Milk thistle (silymarin)
    - 8-20mg/kg divided q 8hrs
Medical treatment outcomes

- Long term survival\(^1\)
  - 51.9% for medically treated patients
  - 87.9% for surgically treated patients
- Age\(^1\)
  - Did not affect survival
- Intrahepatic vs Extrahepatic\(^2\)
  - 64.7% vs 33% were euthanized due to uncontrolled signs

Predictors of outcome

- Poor prognostic indicators
  - Low albumin
  - Anemia
  - Leukocytosis
  - Low body weight (IHPSS)
  - Neurologic signs (HE)

Surgical treatment

- Suture ligation
  - 2-0 silk
  - Complete attenuation
    - 86% cannot tolerate
  - Partial attenuation

Surgical treatment

- Ameroid ring
  - Ring of casein surrounded by stainless steel
  - Hygroscopic substance that swells after absorbing fluid
  - Incites fibrous tissue reaction


Surgical treatment

- Cellophane banding
  - Clear nonmedical grade cellophane
  - 1 x 10cm strips folded into thirds
  - Hemoclips used to secure the band
  - Fibrous tissue reaction leading to gradual occlusion

Surgical video

Surgical treatment

- Percutaneous transjugular coil embolization
  - Fluoroscopic guidance
  - Caval stent placed via jugular vein
  - Embolization coils inserted through a vascular catheter

Surgical treatment

- Liver biopsies
  - Identify PVH-MVD
  - 58% of canine cases
  - 87% of feline cases
Surgical complications

- Hypoglycemia
  - 44% of cases
- Portal hypertension
  - 2-14% of cases usually w/ suture ligation
- Seizures
  - 3-18% dogs, 8-22% cats
  - w/in 80hrs post-op
  - Concurrent disease
    - GME, hypoxia, epilepsy

- Recurrence
  - Failure of attenuation
  - Second shunt
  - Development of multiple extrahepatic shunts
  - PVH-MVD

Post-operative management

- Continued medical management
  - Antibiotics, lactulose, protein restricted diet
- Recheck technesium scan
  - @ 6 weeks

- Recheck bloodwork
  - @ 3 months
  - Bile acids
  - Albumin
  - BUN

Questions?

Thank you!