Congenital absence of the portal vein in a cat

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Abstract
Case summary  A 9-month-old female neutered domestic shorthair cat presented with a history of episodic ptyalism, lethargy and abnormal behaviour. The clinical signs together with elevated pre- and post-prandial bile acid concentrations were consistent with hepatic encephalopathy (HE). In the absence of a portosystemic shunt (PSS) on abdominal ultrasound, medical management of HE was established with a protein-restricted diet and lactulose and the neurological signs resolved. Following an episode of acute vomiting and haemorrhagic diarrhoea at 19 months of age abdominal ultrasonography was repeated. The portal vein could not be demonstrated ultrasonographically; instead, portal vein tributaries were tortuous and communicated with the caudal vena cava (CdVC) at the level of the left kidney. CT angiography (CTA) confirmed the absence of the portal vein. CTA demonstrated the tortuous terminations of the portal tributaries, and several systemic veins, draining into the CdVC via a large-diameter paracaval vessel at the level of the left kidney. Gastrointestinal signs were stabilised and medical management for HE of a protein-restricted diet and lactulose was re-established.

Relevance and novel information  Congenital absence of the portal vein has not been described previously in the cat and should be considered in cats presenting with signs suggestive of a PSS and HE. The portal vein in the cat can be demonstrated using ultrasound, but complex congenital vascular malformations of the portal or systemic abdominal veins should be characterised using CTA and further distinguished from other vascular anomalies that may present with similar ultrasonographic features.

Accepted: 11 November 2017

Case description
A 9-month-old female neutered domestic shorthair cat presented with a recent history of post-prandial episodic behavioural change characterised by apathy, weakness and ptyalism. No neurological abnormalities were noted at clinical examination. Serum biochemistry was normal apart from raised fasting bile acids (135 µmol; reference interval [RI] 0.1–5 µmol). A subsequent bile acid stimulation test demonstrated raised pre- and post-prandial bile acids (fasting 9.3 µmol; RI 0.1–5, post 92.4 µmol; RI 0.5–10 µmol). The reported neurological signs, together with evidence of hepatic dysfunction, were considered consistent with hepatic encephalopathy (HE). The cat was stabilised using a combination of a proprietary liver diet (Prescription Diet L/D Feline; Hill’s) and a home-cooked, protein-restricted diet, lactulose (0.75 ml orally q12h) and metronidazole (10 mg/kg [22 mg/lb] orally q12h for 14 days) and episodic signs resolved. At abdominal ultrasonography at a referral centre a congenital portosystemic shunt (PSS) could not be identified. Further investigation of the hepatic dysfunction by CT angiography (CTA) and liver biopsy were declined on financial grounds.

At 19 months of age the cat presented for vomiting and diarrhoea of 24 h duration. At physical examination the cat weighed 3.3 kg with a body condition score 7/9 and was considered to be 5% dehydrated. Mild hepato-megaly and cranial abdominal discomfort were evident on abdominal palpation. Profuse watery haemorrhagic diarrhoea was produced over a period of 48 h during
hospitalisation. Haematology and serum biochemistry demonstrated haemoconcentration (haematocrit 58%; RI 28.2–52.7%), mild neutropenia (2 × 10^9/l; RI 2.62–15.17 × 10^9/l), mildly raised alanine aminotransferase (119 IU/l; RI 5–60 IU/l), aspartate transaminase (93 IU/l; RI 10–50 IU/l) and bile acids (fasting 11 µmol; RI 0.1–5 µmol).

Two days after hospitalisation abdominal ultrasound demonstrated that the liver was subjectively normal in size, but an extrahepatic portal vein could not be identified. The mesenteric and splenic veins were tortuous and appeared to communicate with the caudal vena cava (CdVC) at the level of the left kidney together with multiple tortuous retroperitoneal vessels. The stomach was hypomotile containing a strongly shadowing, non-obstructing, 15 mm diameter foreign body. The bladder contained a small amount of shadowing crystalline sediment. Small intestinal ileus was present, but there was no evidence of mechanical obstruction.

Dual-phase CTA of the abdomen was performed under general anaesthesia to confirm absence of the portal vein, demonstrate portal tributaries and exclude acquired portosystemic collaterals. CT images were acquired using a 16 slice scanner (Siemens Scope) using the following parameters: 120 kVP, 100 mA, 0.6 mm slice thickness, spiral pitch of 0.8 and 0.8 s rotation. In the arterial phase the coeliac artery and hepatic arteries were increased in diameter and there was conspicuous patchy enhancement of the hepatic parenchyma. In the portal phase the splenogastric and mesenteric trunks did not converge to form a portal trunk. The proximal splenic vein was paired with the splenic artery to the level of the left limb of the pancreas, there received a tributary from the pancreas and was then joined by the left gastric vein. The splenic vein distal to these tributaries looped ventral to the CdVC to enter the left side of the caudal aspect of the paracaval vessel. Several systemic veins also drained into the aberrant paracaval vessel. The left gonadal vein drained into the most caudal aspect of the paracaval vessel and the left renal vein entered its left side immediately dorsal to the termination of the splenic vein. The left phrenicoabdominal vein was a large varicose vessel draining a tortuous left adrenal artery arising directly from the coeliac artery, as well as draining a caudal adrenal branch arising directly from the aorta. The enlarged phrenicoabdominal vein passed ventral to the left adrenal entering the mid-segment of the paracaval vessel on the left.

Following CTA the signs of haemorrhagic diarrhoea gradually resolved over a 72 h period with continued supportive care of intravenous Hartmann’s solution (Aquapharm 11), maropitant (1 mg/kg [2.2 mg/lb] IV q24h), famotidine (1 mg/kg [2.2 mg/lb] PO q24h) metronidazole (10 mg/kg [22 mg/lb] PO q12h) and analgesia (buprenorphine 0.03 mg/kg [0.066 mg/lb] SC
Lactulose was reinstituted once faecal consistency firmed. The owner declined liver biopsy owing to financial limitations. The cat was discharged and managed on lactulose and a combination of a proprietary liver diet (Prescription Diet L/D Feline; Hill’s) and home-cooked, protein-restricted diet. Episodic gastrointestinal...
Congenital absence of the portal vein (CAPV) is a rare anomaly arising from defective development of the primitive venous system in the embryo. 1 CAPV was first described in 1793 and in humans the term ‘Abernethy malformation’ is used to describe portosystemic vascular anomalies in which there is complete or partial diversion of portal blood into systemic veins, usually via the renal, hepatic or iliac veins. 2 In humans these anomalies are further categorised as type 1 or type 2 malformations. 3, 4 A type 1 malformation shunting is complete with no perfusion of the liver by the portal system, whereas in a type 2 malformation partial perfusion is present. Most congenital PSSs in the cat and dog are consistent with a type 2 malformation. A further distinction is made between a type 1a malformation, in which the splenic vein and common mesenteric vein drain separately into the inferior vena cava (IVC) or systemic vein and a type 1b malformation in which these vessels form a common trunk draining into the IVC or systemic vein. 3, 13 Only type 1a malformations are known as CAPV. 1, 3 Fewer than 200 cases of CAPV have been reported in humans. 1, 4 A type 1a shunt or CAPV has not been reported previously in the cat. In this cat the cranial mesenteric and splenic veins did not form a portal trunk but drained separately into the CdVC via an anomalous paracaval vessel. Aplasia of the portal vein fails to develop owing to excessive involution of the left supracardinal vein and left renal collar (anastomoses between the sub-, post- and supracardinal veins) results in blood from the left supracardinal vein reaching the prerenal division of the CdVC via the right side of the renal collar. 14 The paracaval vessel in this cat probably reflects incomplete involution of the left side of the renal collar and left supracardinal vein resulting in a truncated persistent left CdVC.

In humans, CAPV is often an incidental finding or is identified during investigation of associated more serious cardiac malformations. 1 If clinically significant CAPV usually presents later in life associated with hepatic masses, with metabolic derangements such as hyperinsulinaemia and hyperandrogenism secondary to hepatic dysfunction or with signs of HE. 1 Portal hypertension is rare. 4, 15 In comparison, all dogs in which portal vein aplasia has been reported presented with signs of HE at a young age. 5, 6 Four out of five dogs in one study were euthanased following surgical exploration. 5 The cat in this report initially presented with a history suggestive of a metabolic encephalopathy and evidence of hepatic dysfunction considered consistent with HE, and stabilised on medical management. The cause of the signs of haemorrhagic gastroenteritis at presentation 11 months later is unknown but dietary indiscretion was suspected as the cat tended to scavenge following the change to the less palatable, protein-restricted diet. Following this episode of gastroenteritis the cat has remained stable on a protein-restricted diet and lactulose, suggesting that the prognosis in the cat with CAPV could be similar to that in humans if signs of HE can be managed, but additional reports on clinical outcomes are required to determine the prognosis in the dog and the cat.

Although liver biopsy may provide evidence supportive of a hepatovascular malformation biopsy was declined by the owner on financial grounds. Histopathological changes in CAPV demonstrate a reduced number or absence of hepatic portal venules within the portal triad on histopathology. 1 As other conditions share similar histopathological changes diagnostic imaging plays a key role in the non-invasive diagnosis and differentiation of CAPV from other portosystemic anomalies. 15 Ultrasonography is widely used to identify PSSs and has the advantage of being non-invasive and inexpensive. 10, 12, 16 The disadvantage of ultrasound is an accurate diagnosis of a PSS is influenced by operator experience. 17 In the cat, the portal vein is recognised ultrasonographically as a large-diameter vessel in the
central abdomen, ventral to the CdVC that spirals loosely to the right before entering the liver. In our patient, the absence of the portal vein was recognised but characterising the complex morphology of the aberrant portal tributaries and differentiating these from concurrent vascular malformations was technically challenging. CTA allowed comprehensive assessment of the relationship between the anomalous portal and systemic venous systems by demonstrating vascularisation of the liver by enlarged hepatic arteries, the absence of intrahepatic portal veins and drainage of the abdominal viscera via the prehepatic CdVC in the absence of a portal trunk. It has been suggested that a limitation of CTA when portal vein aplasia is suspected is that a remnant or non-perfused vessel connecting either the mesenteric or splenogastric trunk and the intrahepatic portal system cannot be excluded except by intraoperative occlusive mesenteric portovenography (IMPV). However, non-perfused vessels have not been demonstrated in the small number of dogs with portal vein aplasia investigated using IMPV at surgery or by creation of corrosion casts of the vascular tree. Furthermore, it has been demonstrated that CTA is a more reliable and sensitive technique for assessment of the extrahepatic portal tributaries than IMPV. In humans, and in the dog, type 1 shunts have also been associated with concurrent venous malformations, including interruption of the prehepatic CdVC. In this cat, the anomalous paracaval vessel suggestive of truncation of a persistent left CdVC emphasises the value of CTA to allow complete assessment for concurrent venous malformations, including those of the postrenal CdVC.

Conclusions
Congenital PSSs in the cat are uncommon. Accurate, non-invasive demonstration of shunt morphology using ultrasonography is usually possible, but where ultrasonographic assessment demonstrates atypical PSS anatomy, CTA is recommended to differentiate congenital PSSs from other vascular malformations. This case demonstrates that CAPV, although extremely rare, should be included as a differential for congenital PSSs in the cat.

Conflict of interest
The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding
The authors received no financial support for the research, authorship, and/or publication of this article.

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