



# 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 ptalism, 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.

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## 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 ptalism. No neurological abnormalities were noted at clinical examination. Serum biochemistry was normal apart from raised fasting bile acids (135  $\mu\text{mol}$ ; reference interval [RI] 0.1–5  $\mu\text{mol}$ ). A subsequent bile acid stimulation test demonstrated raised pre- and post-prandial bile acids (fasting 9.3  $\mu\text{mol}$ ; RI 0.1–5, post 92.4  $\mu\text{mol}$ ; RI 0.5–10  $\mu\text{mol}$ ). 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 hepatomegaly and cranial abdominal discomfort were evident on abdominal palpation. Profuse watery haemorrhagic diarrhea was produced over a period of 48 h during

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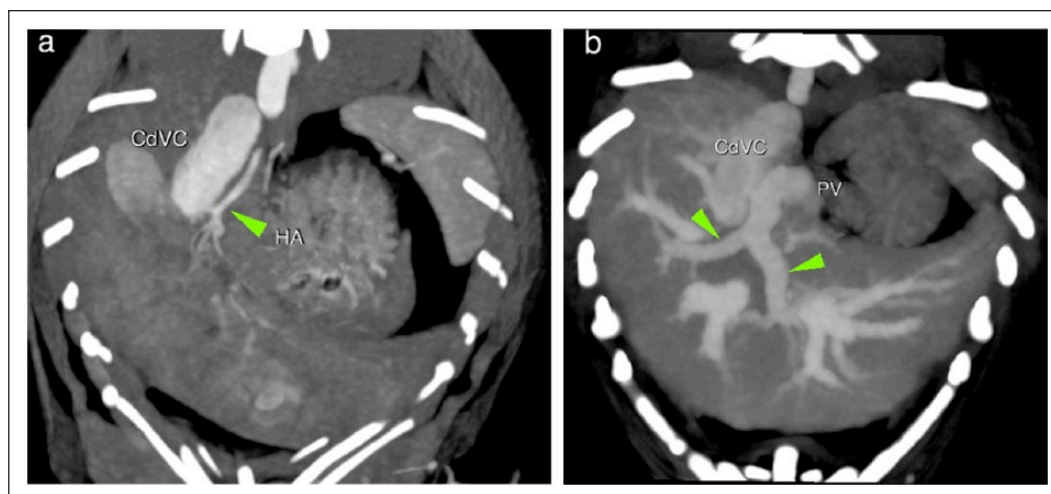
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**Figure 1** Abdominal CT angiography of a cat with congenital absence of the portal vein. (a) Oblique transverse multiplanar reformatted image through the porta hepatis. The portal vein, normally located dorsal to the hepatic artery branches, is absent and the hepatic artery (HA) is enlarged. There is patchy enhancement of the liver parenchyma and intrahepatic portal vein branches are absent. Contrast fills the intrahepatic caudal vena cava (CdVC). (b) Oblique transverse multiplanar reformatted image through the porta hepatis of a normal cat. The large-diameter terminal portal vein (PV) is seen ventral and slightly to the left of the CdVC. The PV spirals ventrally and to the right before dividing conspicuously into the lobar left and right portal vein branches (green arrowheads). The hepatic parenchyma enhances homogeneously

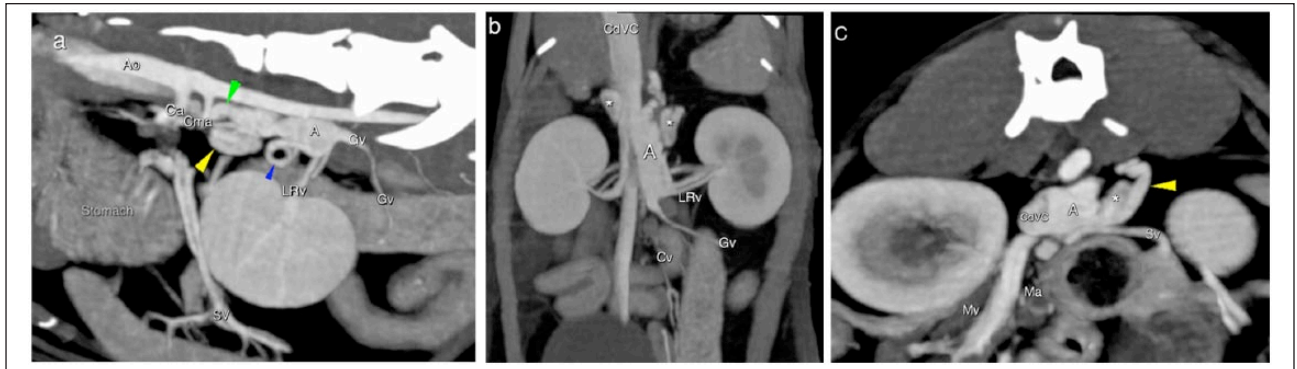
hospitalisation. Haematology and serum biochemistry demonstrated haemoconcentration (haematocrit 58%; RI 28.2–52.7%), mild neutropenia ( $2 \times 10^9/l$ ; RI 2.62–15.17  $\times 10^9/l$ ), mildly raised alanine aminotransferase (119 IU/l; RI 5–60 IU/l), aspartate transaminase (93 IU/l; RI 10–50 IU/l) and bile acids (fasting 11  $\mu\text{mol}$ ; RI 0.1–5  $\mu\text{mol}$ ).

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.

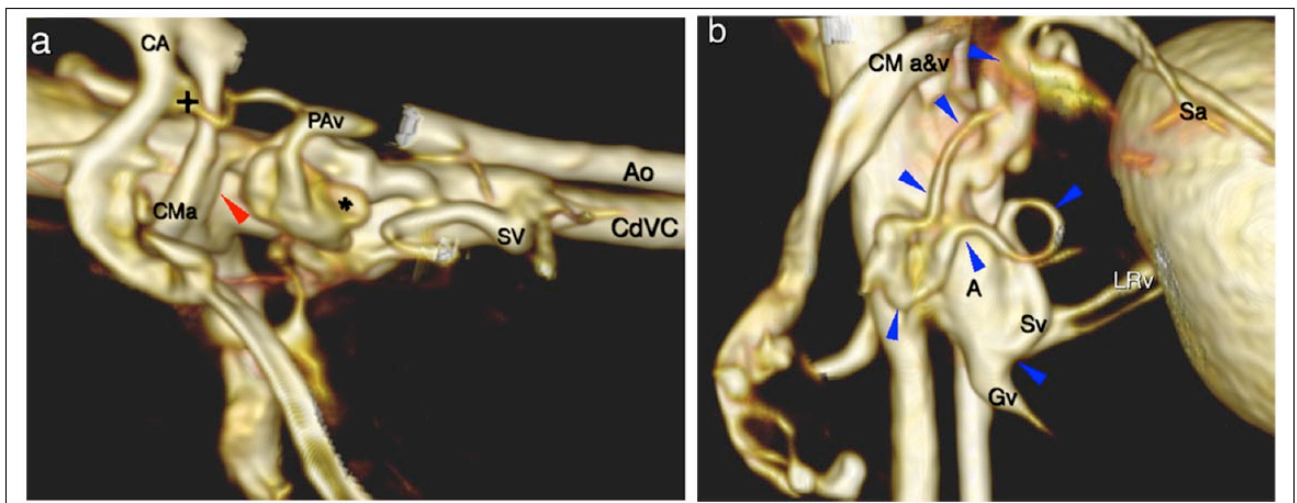
Dual-phase CTA of the abdomen was performed under general anaesthesia to confirm absence of the portal vein, demonstrate portal tributaries and exclude acquired portosystemic collaterals. CT images were acquired using a 16 slice scanner (Siemens Scope) using the following parameters: 120 kVP, 100 mA, 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. There was no portal vein at the level of the porta hepatis and intrahepatic portal branches were not recognised (Figure 1). Instead the extrahepatic portal tributaries

communicated separately with a large, 25 mm  $\times$  8 mm diameter, aneurysmal vessel located between the CdVC and left kidney. This paracaval vessel merged with the CdVC at the level of the second lumbar vertebra. The pre-hepatic CdVC cranial to the paracaval vessel was segmentally dilated. The cranial mesenteric vein was paired with the cranial mesenteric artery, returning to the mesenteric root. It entered the cranial aspect of the paracaval vessel (Figures 2 and 3). 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



**Figure 2** Abdominal CT angiography of a cat with congenital absence of the portal vein. (a) Oblique parasagittal multiplanar reformatted image at the level of the left kidney showing communications between portal tributaries and systemic veins and the prehepatic caudal vena cava (CdVC). The coeliac artery (Ca) is increased in diameter compared with the cranial mesenteric artery (CMA). An aberrant segmented paracaval vessel (A) lies to the left of the CdVC and ventral to the aorta (Ao). The cranial mesenteric vein (green arrowhead) enters the paracaval vessel cranially where it merges with the CdVC. The splenic vein (blue arrowhead) is tortuous and enters the ventrocaudal aspect of the aberrant paracaval vessel immediately ventral to the termination of the left renal vein (LRV) in the same vessel. The left gonadal vein (Gv) enters the most caudal aspect of the paracaval vessel. Yellow arrowhead – left adrenal. (b) Dorsal multiplanar reformatted image. Note the aneurysmal paracaval vessel (A) to the left midline and the segmental dilation of the prehepatic CdVC cranial to the point at which the paracaval vessel merges with the CdVC. The LRV and left Gv terminate in the caudolateral and caudal aspect of the paracaval vessel, respectively. Asterisks indicate the adrenals. Cv = colic vein. (c) Transverse oblique multiplanar reformatted image. The cranial mesenteric vein (Mv) remains paired with the CMA to the mesenteric root and enters the paracaval vessel (A) on the left where it merges with the CdVC. The phrenicoabdominal vein (yellow arrowhead) is enlarged and varicose and courses ventral to the left adrenal (asterisk) before entering the mid-segment of the paracaval vessel ventrally on the left. Sv = splenic vein



**Figure 3** (a) Volume-rendered three-dimensional reformatted image from a left lateral perspective of a cat with congenital absence of the portal vein demonstrates enlargement of the coeliac artery (CA) vs the cranial mesenteric artery (CMA). The cranial mesenteric vein (red arrowhead) enters the left side of the paracaval vessel and caudal vena cava (CdVC) immediately caudal to the CMA. A cranial left adrenal artery branch (+) arises from the CA, communicating with the varicose left phrenicoabdominal vein (PAV). The PAV extends ventral to the left adrenal (asterisk) entering the mid-segment of the paracaval vessel on the left. The tortuous splenic vein (SV) terminates on the caudal margin of the paracaval vessel. A segment of the aorta (Ao) has been removed from the reconstruction to visualise the vessels terminating in the paracaval aneurysm better. (b) Volume-rendered three-dimensional reformatted image from a ventral perspective shows the tortuous splenic vein (blue arrowheads and SV) entering the left caudal aspect of the paracaval vessel ventral to the termination of the left renal vein (LRV). The left gonadal vein (Gv) terminates in the most caudal aspect of the paracaval vessel. The anatomy of the cranial mesenteric artery and vein (CM a&v) is abnormal. The vessels remain paired both returning to the mesenteric root. Sa = splenic artery

q6h). Lactulose was reinstated 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



signs associated with scavenging, but no further episodes of HE, were reported during the 18 months following discharge.

## Discussion

Congenital absence of the portal vein (CAPV) is a rare anomaly arising from defective development of the primitive venous system in the embryo.<sup>1</sup> 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.<sup>1,2</sup> In humans these anomalies are further categorised as type 1 or type 2 malformations.<sup>1,3</sup> In 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.<sup>1,3</sup> Only type 1a malformations are known as CAPV.<sup>1,3</sup> Fewer than 200 cases of CAPV have been reported in humans.<sup>1,4</sup> 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 has been reported previously in the dog.<sup>5,6</sup> The cranial mesenteric and splenic veins in these dogs formed a portal trunk draining directly into the prehepatic CdVC, hence the pattern reported in dogs reflects a type 1b malformation.

Although the factors causing CAPV are poorly understood, the events during embryogenesis that lead to CAPV are well established.<sup>7,8</sup> The intra- and extrahepatic portal vein develops from the paired left and right vitelline veins. The liver cords growing into the septum transversum interrupt the cranial portion of the vitelline veins and form the hepatic sinusoids whereas selective anastomosis between, and involution of, parts of the caudal portions of the vitelline veins around the duodenum forms the extrahepatic portal vein. In CAPV the portal vein fails to develop owing to excessive involution of the periduodenal vitelline veins and failure to develop critical anastomoses with the hepatic sinusoids.<sup>7,8</sup> Persistence of anastomoses between the vitelline veins and the right subcardinal vein, which forms the renal and prehepatic segment of the CdVC allow alternative communication between the vitelline (portal) and cardinal (systemic) venous systems.<sup>8,9</sup> The close proximity of the primitive portal and systemic venous systems in the rapidly developing embryo may allow development of alternative

anastomoses when normal critical anastomoses fail to develop.<sup>10–12</sup> The anomalous paracaval vessel in this cat is unusual as it drained both portal and systemic tributaries. The development of the CdVC is also complex and involution errors lead to specific segmental malformations.<sup>13,14</sup> In the embryo, the right supracardinal vein normally persists and forms the post-renal CdVC, whereas atrophy 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.<sup>14</sup> 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.<sup>1</sup> 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.<sup>1</sup> Portal hypertension is rare.<sup>4,15</sup> In comparison, all dogs in which portal vein aplasia has been reported presented with signs of HE at a young age.<sup>5,6</sup> Four out of five dogs in one study were euthanased following surgical exploration.<sup>5</sup> 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.<sup>1</sup> 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.<sup>15</sup> Ultrasonography is widely used to identify PSSs and has the advantage of being non-invasive and inexpensive.<sup>10,12,16</sup> The disadvantage of ultrasound is that an accurate diagnosis of a PSS is influenced by operator experience.<sup>17</sup> 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 spleno-gastric trunk and the intrahepatic portal system cannot be excluded except by intraoperative occlusive mesenteric portovenography (IMPV).<sup>6</sup> 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.<sup>5</sup> Furthermore, it has been demonstrated that CTA is a more reliable and sensitive technique for assessment of the extrahepatic portal tributaries than IMPV.<sup>18</sup>

In humans, and in the dog, type 1 shunts have also been associated with concurrent venous malformations, including interruption of the prehepatic CdVC.<sup>1,5,6,15</sup> 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 post-renal 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.

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