Case description
A 4-month-old neutered male, Russian Blue cat was investigated for a 4 week history of hypersalivation, failure to thrive. In addition, there was a 2 week history of soft tissue swelling on the ventral abdomen, which had failed to improve with antimicrobial therapy. There were no significant physical examination or neurological deficits on examination; however, the cat had a quiet demeanor for its age. Postprandial bile acids were increased (32 µmol/l; reference interval <25 µmol/l). An abdominal CT scan revealed changes consistent with an extrahepatic portosystemic shunt and inflammation of fat of the ventral abdominal body wall. Surgical biopsy and culture of the subcutaneous swelling identified non-infectious steatitis. Ten weeks following initial presentation, surgical exploration, liver biopsy and ligation of the portosystemic shunt were performed. Liver biopsy was submitted to the Anatomical Pathology Laboratory of Cornell University Animal Health Diagnostic Center, New York, USA. Histopathology revealed a ductal plate malformation (Caroli’s type), as well as changes consistent with a portosystemic shunt.

Relevance and novel information
Ductal plate malformations are rarely described in the veterinary literature. To our knowledge this is the first reported case of Caroli’s-type malformation in a cat. There are no biochemical changes that allow for differentiation of ductal plate malformations from other hepatopathies. Liver biopsy is required for a definitive diagnosis.

Keywords: Liver; feline ductal plate malformation; Caroli’s disease; cystic liver disease; portosystemic shunt

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On physical examination, the cat was lethargic with hypersalivation with a poor body condition score (3/9). Vital signs were normal. There was a firm nodular thickening on the ventral abdomen. A dual-phase (portal and delayed-venous) abdominal CT scan was performed under general anesthesia with a six-slice helical unit (Brilliance 6; Philips). The left gastric vein was dilated (5 mm vs 1.5 mm for the main portal vein) and communicated directly with the left phrenic vein, terminating in the caudal vena cava (Figures 1 and 2). Intrahepatic portal vein branches were subjectively small and
difficult to identify. There was subjective microhepatia. These changes were supportive of an extrahepatic portosystemic shunt. In addition, soft tissue attenuating streaking of the ventral subcutis was identified, suggestive of steatitis. Surgical tissue biopsies from the ventral abdomen were obtained for histopathology and tissue culture.

The patient was discharged with oral lactulose (0.5 ml PO q12h) for interim medical management of hepatic encephalopathy. Doxycycline (11.1 mg/kg PO q12h [12.5 mg]) was started for possible mycobacterial panniculitis, at the clinician’s discretion. Histopathology, including Ziehl–Neelsen staining for mycobacteria of the subcutaneous tissue, identified steatitis with no pathogenic infectious agent identified. There was no microorganism growth on prolonged tissue culture. A lower protein diet was recommended, such as Hill’s k/d.

Ten weeks after initial presentation, the patient represented for exploratory laparotomy for ligation of a suspected portosystemic shunt. The patient was receiving oral lactulose (0.5 ml PO q12h) and had been commenced on levetiracetam (22 mg/kg PO q8h) 7 days prior as per hospital policy for portosystemic shunt patients. On examination, the patient was alert and responsive but considered quieter than expected given its age. Body condition score had improved (4/9) and vital signs were normal. The steatitis had resolved.

Preanaesthetic haematology and biochemistry indicated a mild hypochromic microcytic non-regenerative anaemia (haematocrit 27.7% [RI: 30.3–52.3%]; mean cell volume 34.4 fl [RI 34.3–35.9 fl]; mean cell haemoglobin 11.5 pg [RI 11.8–17.3]) and moderate lymphocytosis (7.36 × 10⁹/l; RI 0.92–6.88 × 10⁹/l). A low creatinine (65 µmol/l; RI 71–212 µmol/l) and mild hyperphosphataemia (2.72 mmol/l; RI 1.00–2.42 mmol/l) were attributed to suboptimal body condition score and age-related change, respectively. Electrolytes were within normal limits.

Exploratory laparotomy under routine general anaesthesia confirmed the presence of a left gastrophrenic portosystemic shunt, in addition to a double vena cava, which joined at the level of the renal veins. A partial ligation of the portosystemic shunt with cellophane banding was performed. A 6 mm punch biopsy from the quadrate lobe of the liver was obtained. The patient had an unremarkable anaesthetic recovery. Liver biopsy was submitted for external histopathological review to Cornell University Animal Health Diagnostic Center. The patient was discharged 2 days after shunt ligation with phenobarbitone (1.9 mg/kg PO q12h for 2 weeks), levetiracetam (19 mg/kg PO q8h for 2 weeks) and buprenorphine (0.02 mg/kg PO q8h) as required for analgesia. The patient developed upper respiratory tract signs with mucoid nasal discharge and was commenced on doxycycline (9.6 mg/kg PO q12h for 7 days). The previous low-protein diet was advised until liver function was reassessed.
Histopathology of the liver biopsy revealed large bile ducts with irregular, sacculated contours (Figure 3). There was increased profile of abnormal bile ducts that occasionally breached the limiting plate. The proliferations extended out a short distance blindly into the hepatic parenchyma. Both the small and medium portal tracts contained thick-walled arterioles and mild serpiginous, plexiform proliferations of arterioles. The portal veins were frequently inconspicuous and portal lymphatics were frequently moderately dilated (Figure 4). The sinusoids were irregularly branching (Figure 5). Portal tracts contained small numbers of lymphocytes and plasma cells primarily around bile ducts. There was portal vein hypoperfusion. These findings are consistent with Caroli’s-type ductal plate malformation and congenital portosystemic shunt.

Two months after surgery, the cat was bright and clinically well. Repeat haematology and biochemistry indicated a mild microcytosis and a mild elevation in alkaline phosphatase (72 IU/l; RI 5–50 IU/l). Resting ammonia was 87 µmol/l (RI 0–95 µmol/l) and fasting bile acid remained elevated at 18 µmol/l (RI <11 µmol/l), attributed to incomplete attenuation of the portosystemic shunt.

Discussion
To our knowledge, this is the first reported case of a Caroli’s-type ductal plate malformation (DPM) in a feline patient.

The ductal plate is an embryonic precursor of intrahepatic bile ducts. Initially formed from transformation of hepatoblasts, the plate consists of a double layer of cuboidal biliary-style epithelium that surrounds the developing portal vein. Development and remodeling of the ductal plate is orchestrated by a range of regulatory factors in a time- and context-dependent manner, commencing in a centripetal fashion from the larger portal vein branches at the hilum.
DPMs represent any malformation, persistence or lack of remodelling of the ductal plate. Varying degrees of cystic dilation of intrahepatic bile ducts are observed in DPMs.\textsuperscript{1,4} The pathomechanisms resulting in DPMs remain elusive, and the associated syndromes in people are genetically and phenotypically heterogeneous. Genetic abnormalities that affect the structure and function of the primary cilia of the biliary epithelium have been identified in people with DPM, leading to the consideration that DPM may be a primary congenital cholangiociliopathy.\textsuperscript{5,5} In veterinary patients, congenital cystic liver disease has been categorised as:\textsuperscript{6} (i) congenital dilation of the large and segmental bile ducts (morphologically identical to Caroli’s disease); (ii) juvenile polycystic disease/congenital hepatic fibrosis; and (iii) adult polycystic disease, including Von Meyenburg complexes.

In Caroli’s disease, insufficient involution of the ductal structures at the level of the segmental bile duct leads to formation of large, cystic ducts. In the medical literature, concurrent congenital hepatic fibrosis is included with the histopathological description of Caroli’s syndrome, which is not included in the World Small Animal Veterinary Association (WSAVA) categorisation.\textsuperscript{5,7}

Only a small number of dogs have been diagnosed with Caroli’s-like disease in the veterinary literature. The most recent publication describes the clinical and histological features of ductal plate malformation in 30 Boxer dogs. Of these dogs two had histological change consistent with Caroli’s malformation. One of these dogs had concurrent bridging portal fibrosis.\textsuperscript{1} A small case series identified hepatic cystic and fibrotic changes in two Golden Retriever littermates.\textsuperscript{5} Clinical signs described in these cases were non-specific and included vomiting, polyuria, polydipsia, anorexia, lethargy and reduced activity.\textsuperscript{1,4,5,8}

A well-documented autosomal dominant polycystic kidney disease in Persian cats is also recognised to have concurrent hepatic cystic changes.\textsuperscript{5,9–11} Cystic disease of the liver and kidney has been described in a range of canine breeds, including Cairn Terriers, Collies, Frisian Stabyhouns, Jack Russell Terriers and a mixed-breed dog.\textsuperscript{5,8} Several case reports suggest that Caroli’s disease without cystic kidney disease occurs in both dogs and people, as was also the case in this patient.

In this cat, Caroli’s disease was present with an extrahepatic portosystemic shunt with a histopathological diagnosis of portal hypoperfusion and Caroli’s disease. In the absence of fibrosis that could contribute to portal hypertension, we propose the portosystemic shunt is the potential sequelae of the partially attenuated portosystemic shunt and Caroli’s disease, a proactive approach towards diagnostic investigations and management is therefore encouraged for this patient.

Conclusions
To our knowledge, this is the first case of Caroli’s-type DPM in a cat. There are no identified clinicopathological abnormalities that allow for distinction of DPM from other hepatopathies. Therefore, diagnosis requires liver biopsy for histopathology.
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