Acquired Portosystemic Shunting in 2 Cats Secondary to Congenital Hepatic Fibrosis

Maurice M.J.M. Zandvliet, Viktor Szatmári, Ted van den Ingh, and Jan Rothuizen

Case 1. A 2-year-old female crossbred cat was referred to Utrecht University Companion Animal Clinic with a 2-month history of episodic vomiting, constipation, decreased appetite, apathy, and decreased activity. The vomiting was treated successfully with cisapride (0.25 mg/kg PO q8h).

Physical examination disclosed no abnormalities and a full CBC and serum biochemistry tests were performed. These evaluations showed a leukocytosis (18.6 × 10⁹/L; reference range, 4.3–16.2 × 10⁹/L), increased bile acid concentration (132 μmol/L [53.88 μg/mL]; reference range, 1–13 μmol/L) and a low BUN concentration (3.6 mmol/L [10.08 mg/dL]; reference range, 6.1–12.8 mmol/L). The combination of high bile-acid concentration and low BUN concentration indicated possible portosystemic shunting.

Fasting venous blood ammonia concentration was measured (202 μmol/L [53.88 mg/dL]; reference range, 1–13 μmol/L) and a low BUN concentration (3.6 mmol/L [10.08 mg/dL]; reference range, 6.1–12.8 mmol/L). The combination of high bile-acid concentration and low BUN concentration indicated possible portosystemic shunting.

Abdominal ultrasonography revealed several small cortical cysts in both kidneys compatible with polycystic kidney disease (PKD). Otherwise, the size and structure of both kidneys and the liver were considered normal. An anomalous tortuous vessel with hepatofugal flow was seen originating from the portal vein in the region where the splenic vein enters the trunk of the portal vein. It was not possible to follow the shunting vessel to its termination. The direction of blood flow was hepatofugal in the portal vein cranial to the origin of the anomalous vein and was hepatopetal caudal to it. The left ovarian vein was dilated. A very small amount of free abdominal fluid was seen. The spleen was ultrasonographically normal. The tortuous anomalous vein was suspected to be an acquired portosystemic collateral, therefore ultrasound-guided core biopsies of the liver were taken.

The liver biopsies were processed routinely and stained with hematoxylin-eosin, Von Gieson, and a reticulin stain according to the method of Gordon and Sweet. Histological examination identified extensive fibrosis in the portal areas with porto–portal bridging (Fig 1). In these fibrotic portal areas, an increased number of cross-sections of abnormally shaped bile ductules were found together with a few cross-sections of hepatic arteries and the absence of recognizable portal vein branches. Small focal infiltrates of lymphocytes were also seen. The liver parenchyma and hepatic veins showed no abnormalities. The histologic diagnosis was congenital hepatic fibrosis, a manifestation of congenital polycystic disease found in cats, which was compatible with the previously detected renal cysts. The histologic changes in the liver made the presence of portal hypertension a logical consequence and it was concluded that the shunting vessel was acquired. Supportive therapy consisting of a low-protein diet and lactulose was initiated.

Case 2. A 2½-year-old female Persian cat was presented at Utrecht University Companion Animal Clinic with a history of weight loss despite good appetite and episodic neurologic signs consisting of pyralism, decreased mentation, and ataxia. Results of general physical and neurologic examinations were normal. Based on the episodic signs of decreased mentation and ataxia, a central neurological problem was anticipated. Laboratory examinations were performed to identify metabolic causes. Results included increased plasma fasting bile acid (114 μmol/L [46.53 μg/mL]) and fasting ammonia (>286 μmol/L [>516 μg/dL]) concentrations and low BUN concentration (4.5 mmol/L [12.60 mg/dL]). A tentative diagnosis of hepatic encephalopathy was made. An ultrasonographic examination of the abdominal cavity and the liver was performed.

Ultrasonographic examination disclosed a small liver with irregular structure. The left kidney was slightly smaller (3.5 cm long) and the right kidney slightly larger (4.5 cm long) than normal. Multiple small cysts compatible with PKD were found in both kidneys. A connecting blood vessel was found between the portal vein and the hepatic region of the caudal vena cava. Due to the small size of the liver, it was impossible to determine whether the shunting vessel was located inside or outside of the liver. Ultrasonographic examination identified no other abnormalities, and the conclusion was that the cat presumably had both a congenital portosystemic shunt and PKD.

Because PKD had not resulted in measurable decompensation of renal function, surgical ligation of the shunt was considered the preferred treatment. Intraoperative ultrasonography confirmed the presence of an intrahepatic portosystemic shunt, which then was completely closed by ligation with polyester suture material.

Recovery was unremarkable and a postoperative evaluation was performed after 1 month. At that time, the blood ammonia concentration was high (202 μmol/L [364.62 μg/dL]). Ultrasonographic examination of the abdominal cav-
ity identified an extrahepatic portosystemic shunt arising from the portal vein at the point where the splenic vein enters the portal vein. The left ovarian vein was not dilated and a small amount of anechoic ascitic fluid was seen. Because this anomalous extrahepatic vein was not present before surgery, it was thought to be an acquired portosystemic collateral, and a percutaneous ultrasound-guided liver biopsy was taken to determine the underlying cause of the portosystemic shunting.

The liver biopsies were processed routinely and stained as described above. Histologic evaluation of the liver biopsy revealed marked portal fibrosis and dilated, irregularly formed bile ductules (Fig 2). No inflammatory cells were present in the portal areas, but an occasional lymphocyte and necrotic hepatocyte were observed in the limiting plate. The final diagnosis was congenital hepatic fibrosis compatible with congenital polycystic disease in cats as well as a mild degree of periportal hepatitis.

On the basis of these results, it was concluded that the cat had 2 different congenital abnormalities, a congenital portosystemic shunt and congenital hepatic fibrosis. Supportive therapy consisting of a low-protein diet and lactulose was started.

Portosystemic shunting can be due to a congenital portosystemic shunt with an intrahepatic or extrahepatic localization or to multiple acquired portosystemic collaterals resulting from portal hypertension, which in most cases is due to primary hepatic disease. In order to identify the cause of portosystemic shunting, a thorough ultrasonographic examination of the abdominal cavity, liver, and portal vein by an experienced radiologist is essential because these results determine therapy and prognosis.

A congenital portosystemic shunt is amenable to surgical ligation, whereas acquired portosystemic collaterals should be managed medically by dietary measures (low-protein diet) and lactulose. In the case of acquired shunting, it is important to diagnose the underlying hepatic or vascular lesion in order to identify whether it can be treated medically (e.g., cholangitis in cats) or surgically (e.g., arteriovenous fistula).

Portosystemic shunting results in portal blood bypassing the liver and decreased hepatic clearance of substances from the portal circulation. Increased concentrations of bile acids, ammonia, and endotoxins can be measured in the systemic circulation. Decreased urea concentration is due both to decreased hepatic uptake and metabolism of ammonia and increased renal losses due to polyuria, often associated with portosystemic shunting. Increased systemic endotoxin concentrations may result in a systemic inflammatory response and leukocytosis and induce vomiting by stimulation of the chemoreceptor trigger zone.

Polycystic kidney disease is a well-described disease in cats, especially in Persian cats. In this breed, it is an autosomal dominant trait due to a mutation in the PKD1 gene. The phenotype can be detected early in life by means of ultrasound examination of the kidneys from as young as 6–8 weeks of age, but cysts can arise at a later age. The cysts originate from both the proximal and distal tubules and can be found in both the renal cortex and medulla. The disease is progressive, and both the size and number of cysts can increase over time and may result in chronic renal failure later in life. PKD can be diagnosed by means of ultrasonographic examination of the kidneys once the cysts are of sufficient size or by kidney biopsy that may show lesions suggestive of PKD at an earlier stage. The pathogenesis of the disease is unknown, but in humans, partial or complete obstruction of tubules is hypothesized. Structural abnormalities of the tubular basement membrane is another possibility that has been suggested in the literature.

Less well known is that PKD in most cats is associated with congenital biliary cystic lesions, either as multiple large cysts (adult-type polycystic disease) or as congenital hepatic fibrosis, characterized by portoportal bridging fibrosis and abnormally structured cystic bile ductules. The hepatic lesions can be found both with or without lesions in the kidney both in humans and in cats. It is therefore more appropriate to refer to this disorder as polycystic disease and consider the kidney and liver lesions as 2 distinct manifestations. The renal form (PKD) is the more common
variant in cats and most commonly associated with clinical disease. The observed hepatic fibrosis causes portal hypertension and may result in the formation of acquired portosystemic collaterals.

In this case report, we describe 2 cats with polycystic disease and signs attributable to portosystemic shunting rather than to renal disease. Acquired portosystemic shunting is a rare event in cats, in contrast with dogs. To our knowledge, these cats represent the first report of acquired portosystemic collaterals due to congenital hepatic fibrosis as a manifestation of polycystic disease in cats. In accordance, the clinical signs in these cats were those of hepatic encephalopathy.

A second unusual observation is that cat 2 had 2 congenital disorders, a congenital intrahepatic portosystemic shunt and congenital polycystic kidney and liver disease. The presence of a congenital portosystemic shunt prevented development of portal hypertension, and ligation of the congenital shunt resulted in portal hypertension due to preexisting hepatic fibrosis and formation of an acquired portosystemic collateral circulation. The combination of a congenital and an acquired shunt in 1 animal has been reported in the dog, but ours is the first report in a cat. To the authors’ knowledge, no causative relation exists between these 2 congenital diseases, and the combination is thought to be purely coincidental.

On the basis of these 2 cases, it seems advisable to examine liver histology in Persian cats and crossbred cats in the presence of hepatic encephalopathy. Even if the only ultrasonographic lesion is a congenital portosystemic shunt, the cat should be evaluated for the potential presence of hepatic fibrosis to prevent unnecessary surgical intervention.

Footnotes
4 Pro-Mag 2.2L biopsy system, Manan Medical Products, Northbrook, IL, and ACN™ Biopsy Needle (16 G × 16 cm), Med-Tech Inc, Gainesville, FL
5 Ethibond Excel 2/0, Ethicon, Johnson & Johnson Intl, Hamburg, Germany

References