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Unusual haemodynamics in two dogs and two cats with portosystemic shunt - implications for distinguishing between congenital and acquired conditions

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Abstract

Extrahepatic porto-systemic shunt (PSS) in small animals can be congenital (CPSS) or acquired (APSS) as a consequence of portal hypertension (PH), and are distinguished on the bases of their anatomical pattern. A precise morphologic imaging assessment, along with clinical and histopathologic findings, is important for distinguishing patients with PH from those with congenital PSSs, which require different therapeutic approach. Expected findings in patients with PH are presence of ascites, multiple APSS, and a confirmed cause of portal flow obstruction. On the other hand, a single PSS, absence of ascites and no evidence of portal vein, caudal vena cava or hepatic disorders are typical findings of CPSS patients. This paper describes four cases of PSSs in which the combination of the computed tomographic imaging findings did not match the standards for APSS nor for CPSS: one dog had chronic hepatitis causing PH and ascites and a splenoazygos PSS, to date considered a CPSS pattern. One dog showed a left splenogonadal PSS and porto-caval varices, to date considered an APSS pattern, without ascites, portal vein obstruction, primary structural hepatic disorders nor evidence of PH. Two cats, with and without diffuse hepatic structural disorders respectively, had a single left splenogonadal PSS without ascites. Possible interpretation of such unusual haemodynamic conditions and clinical repercussion, especially for orientation of treatment choice, are discussed.

Keywords: Cat, Computed tomography, Dog, Portal hypertension, Portosystemic shunt.

Introduction

Abnormal venous porto-caval connections are widely described in small animal veterinary literature, with majority of cases reported in dogs (Lamb, 1996; Bertolini *et al.*, 2006; Bertolini, 2010a, 2010b; Bruehschwein *et al.*, 2010; Nelson and Nelson, 2011; Fukushima *et al.*, 2014). These vascular anomalies are divided in two main categories according to their origin: congenital PSS (CPSS) deriving from embryogenetic errors in the development of vitelline and cardinal venous systems (Ferrell *et al.*, 2003), and acquired PSS (APSS) deriving from recanalization of pre-existing, vestigial embryonic vascular connections between portal and caval systems as a consequence of portal hypertension (PH) (Fossum, 2002; Sztatmari *et al.*, 2004; Bertolini, 2010a).

Ultrasound and computed tomography (CT) are reliable imaging techniques for the non-invasive diagnosis of PSSs and for distinction between acquired and congenital shunts (Sztatmari and Rothuizen, 2002; Sztatmari *et al.*, 2004; Ricciardi, 2016). Such imaging distinction is essential for differentiating hypertensive patients, which need further investigations in order to diagnose the cause of the underlying PH, from those that need surgical closure of a congenital vascular malformation. The most useful imaging findings which are taken into account for discriminating APSS and

CPSS are: the anatomical pattern of vascular anomalies (distinct vascular patterns have been observed within each category without any overlapping of anatomical pathway between APSS and CPSS), their number (more than one porto-caval shunt are typically found in dogs with PH and MAPSS) and presence/absence of ascites (which is typically absent in dogs with CPSS) (Ricciardi, 2016).

This paper describes four cases of PSSs in two dogs and two cats in which the combination of the computed tomographic imaging findings did not match the standards for APSS nor for CPSS. Possible interpretation and clinical repercussion, especially for orientation of treatment options, of such unusual haemodynamic conditions are discussed.

Case details

Dog 1

A 12-year-old intact male Yorkshire terrier dog was evaluated for a 2-week history of inappetence and abdominal distension. Clinical examination findings included thinness (Body Condition Score: 3/9) (Baldwin *et al.*, 2010) and free fluid in the abdomen. No clinical evidence of heart disease was present.

Complete blood count revealed neutrophilia with right shift (segmented neutrophils 7946/ μ l, reference interval [RI] 3601–6056) and monocytosis (702/ μ l, RI 172–462). Serum chemistry revealed marked increase in

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liver enzyme activity (aspartate aminotransferase 530 IU/l, RI 20–31; alanine aminotransferase 638 IU/l, RI 22–78), hypoalbuminemia (2.1 g/dl, RI 2.7–3.6) and increase in C-reactive protein (1.36 mg/dl, RI 0.01–0.09), and urinalysis revealed marked increase in urinary bile acids levels (249 mmol/l, RI 2.9–9.5). Abdominal fluid was consistent with pure transudate.

B-mode ultrasound abdominal examination (Esaote MyLab 30 Gold, Esaote SpA, Genoa, Italy) revealed abundant anechoic peritoneal effusion, with normoechoic and diffusely inhomogeneous hepatic parenchyma. Because of abundant ascites and overlying gastrointestinal content the portal vein was visualized with difficulty.

In order to clarify any possible abdominal or thoracic vascular disease that could explain the portal hypertension, thoracic and abdominal CT scan was performed immediately after the ultrasound examination using a 16-slice MDCT scanner (*Somatom Emotion, Siemens, Forchheim, Germany*). Computed tomography images were acquired before and after the intravenous injection of iodinate contrast medium (640 mg I/kg; *Iopamigita® Insight Agents GmbH, Heilbronn, Germany*). Scanning and reconstruction parameters were as follows: helical modality, 0.6 sec/gantry rotation, 1 mm slice thickness; 110 kV, 180 mAs; soft tissue reconstruction algorithm.

Three-dimensional (3D) multiplanar reformatted and volume-rendered images were obtained using a dedicated 3D software (*Pixmeo, OsiriX; OsiriX DICOM-viewer; Pixmeo, Geneva, Switzerland*). Computed tomography scans showed a large amount of free fluid in the abdomen, and a large, single tortuous vessel connecting the splenic vein with the azygos vein. The liver appeared slightly reduced in volume, with irregular margins and without macroscopic parenchymal abnormalities or attenuation changes. No compressions/thrombosis of the portal vein or caudal vena cava were present (Fig. 1).

Based on the imaging findings a diffuse microscopic hepatic disorder with secondary portal hypertension, associated with a spleno-azygos portosystemic shunt, was suspected.

Specimens of liver biopsy samples were sent for histopathologic evaluation in a certified veterinary laboratory (*San Marco Veterinary Laboratory, Padova, Italy*). Histopathologic evaluation of a liver biopsy sample revealed cytoplasmic expansion of hepatocytes with infiltration of lymphocytes, plasma cells and macrophages between hepatocyte layers. Sinusoidal congestion, dilation of centrilobular veins and surrounding lymphatic vessels and stromal expansion with centro-central bridging surrounded by inflammatory cells were evident in centrilobular tracts. These findings were consistent with subacute-chronic centrilobular and midzonal hepatitis.

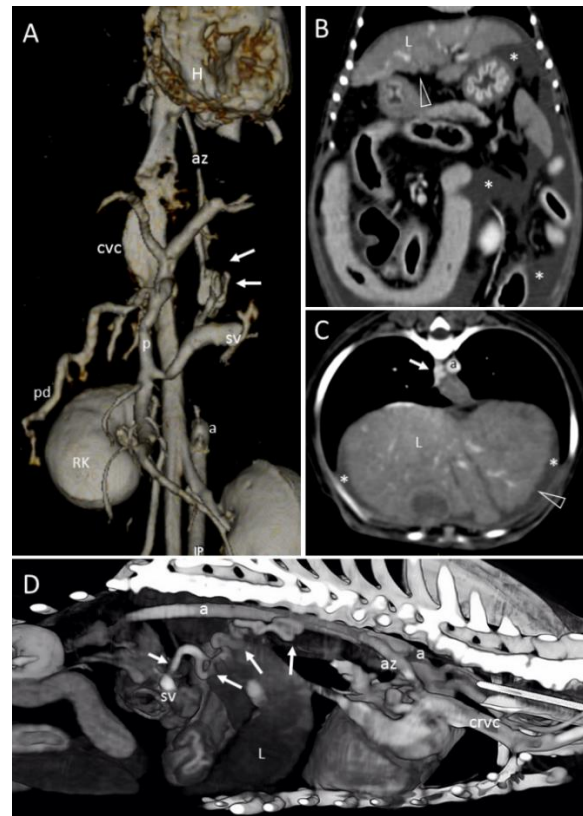


Fig. 1. Dog 1. (A): Ventral three-dimensional volume-rendered CT angiography of portal system, caudal vena cava, heart and azygos vein showing a splenoazygos PSS (arrows) connecting splenic vein (sv) and azygos vein (az). (B) Dorsal and (C) transverse multiplanar reformatted contrast-enhanced CT images of abdomen. The liver (L) appears slightly reduced in volume, with irregular margins (arrowheads) and without macroscopic parenchymal abnormalities or attenuation changes. A large amount of fluid (ascites) is evident in the background (asterisks). The section of the splenoazygos PSS is evident adjacent to Aorta (a), on the right (C-arrow). (D): Oblique right lateral thick-slab multiplanar reformatted contrast-enhanced CT image of abdomen and thorax showing in detail the tortuous splenoazygos PSS (arrows) and its connection with azygos vein (az). H: heart; p: portal vein; cvc: caudal vena cava; pd: pancreaticoduodenal vein; RK: right kidney; crvc: cranial vena cava.

In order to avoid worsening of portal hypertension surgical closure of the spleno-azygos PSS was not considered. A supportive therapy with spironolactone (2mg/kg q12h), amoxicilline-clavulanic acid (20 mg/kg q 12 h) and a commercial therapeutic diet for hepatic health (Prescription Diet l/d Canine, Hill's Pet Nutrition Inc, Topeka, Kan) were started. Three months later the dog was still alive.

Dog 2

An 8-year-old intact male mongrel dog was evaluated for a 1-month history of waxing and waning abnormal mentation and sporadic episodes of vomiting. At time of clinical examination the dog was normal. Complete

blood count was unremarkable. Serum chemistry revealed increase in liver enzyme activity (aspartate aminotransferase 95 IU/l, RI 16–38; alanine aminotransferase 463 IU/l, RI 16–123) and hypoalbuminemia (2.2 g/dl, RI 2.7–3.6), and urinalysis revealed increase in urinary bile acids levels (87 mmol/l, RI 2.9–9.5). Abdominal ultrasound and CT of thoracic and abdominal region were performed as described for dog 1. Imaging evaluation revealed microhepatia without macroscopic structural abnormalities of liver parenchyma, a large tortuous vessel connecting splenic vein and left gonadic vein and a network of small and tortuous vessels (varices) cranio-medially to the left kidney connecting left phrenicoabdominal vein and portal vein (Fig. 2).

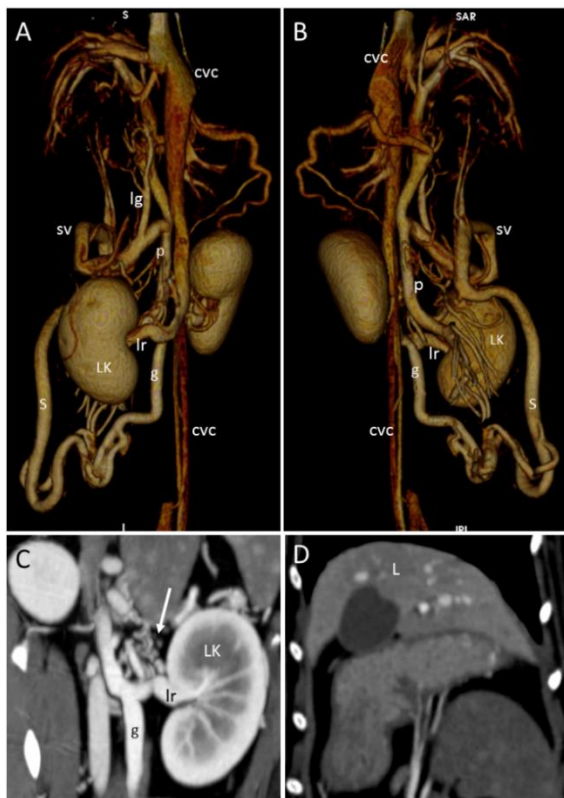


Fig. 2. Dog 2. (A) Left dorso-lateral and (B) right ventro-lateral three-dimensional volume-rendered CT angiography of portal system and caudal vena cava showing a large left splenogonadal PSS (s) connecting splenic vein (sv) and left gonadic vein (g) and entering the left renal vein (lr). (C): Dorsal maximum intensity projection (MIP) reformatted contrast-enhanced CT image of abdomen at level of kidneys. Porto-caval varices (arrow) are visible medially to the left kidney (LK). The enlarged left gonadic vein (g), which constitutes part of the left splenogonadal PSS, is visible at level of left renal vein (lr). (D): Dorsal multiplanar reformatted contrast-enhanced CT image of abdomen. The liver (L) appears slightly reduced in volume without macroscopic structural parenchymal abnormalities. P: portal vein; cvc: caudal vena cava; lg: left gastric vein.

Based on the imaging findings a diffuse microscopic hepatic disorder with secondary non-ascitic portal hypertension and MAPSS was suspected. In order to evaluate the presence and quantify eventual underlying portal hypertension, invasive portal pressure was measured during laparotomy for liver biopsy sampling. According to the described technique (Bojrab *et al.*, 2014) a 22-Gauge intravenous catheter was placed into a jejunal vein and was attached to an extension set, a 3-way stopcock, a syringe and a water manometer with a zero point at the level of the right atrium. A value of 11.5 cm H₂O (reference interval: 8-13 cm H₂O) was recorded.

Histopathologic evaluation of a liver biopsy sample revealed diffuse lobular hypoplasia, portal venous hypoplasia, arteriolar and biliary hyperplasia, non-specific hepatocellular degeneration and multifocal lipogranulomas. These findings were suggestive of parenchymal damage caused by portal hypoperfusion. Supportive therapy with lactulose (10 mg PO q8h), metronidazole (7.5 mg/kg q12h) and a commercial therapeutic diet for hepatic health (Prescription Diet l/d Canine, Hill's Pet Nutrition Inc, Topeka, Kan) was started.

Four months later the dog was in good clinical condition with significant reduction of neurologic signs.

Cat 1

An 11-year-old spayed female domestic short-hair cat was referred for CT staging of a histopathologically confirmed lumbar cutaneous sarcoma. Clinical examination confirmed a cutaneous mass at level of lumbar region and a palpable firm mass in the cranial abdomen.

Haematology, serum chemistry analyses and urinalysis results were within normal limits. CT of the whole body was performed as described for dog 1. CT findings included: two large irregularly marginated, inhomogeneous, hypervascularized lumbar cutaneous and subcutaneous masses isoattenuating to the soft tissue with inhomogeneous contrast-enhancement; multiple irregularly marginated rounded hepatic masses (maximum dimensions: 3 x 5 x 4 cm), hypoattenuating to soft tissue with irregular contrast-enhancement; multiple, regularly marginated rounded pulmonary nodules (maximum dimensions: 3 x 3 x 3 mm) and a large tortuous vessel connecting splenic vein and left gonadic vein. No free fluid in the abdomen was evident (Fig. 3). An imaging diagnosis of metastatic soft tissue sarcoma with a left splenogonadal PSS was made.

Cat 2

A 13-year-old spayed female domestic short-hair cat was referred for CT staging of a histopathologically confirmed soft tissue sarcoma at the level of right scapular region.



Fig. 3. Cat 1. (A) Dorsal thick-slab multiplanar reformatted contrast-enhanced CT image of lumbar region showing two large irregularly marginated, inhomogeneous, hypervascularized cutaneous and subcutaneous neoplasms (arrowheads). (B) Dorsal multiplanar reformatted contrast-enhanced CT image of abdomen. Multiple, irregularly marginated, rounded hepatic masses, hypoattenuating to soft tissue, with irregular contrast-enhancement, compatible with primary or metastatic diffuse hepatic neoplasia, are evident (asterisks). (C) Dorsal thick-slab multiplanar reformatted contrast-enhanced CT image of abdomen showing a large left splenogonadal PSS (arrows) connecting the splenic vein (sv) with the left gonadic vein (g) and entering the left renal vein (lr). No ascitic fluid is evident in the background (B,C). P, portal vein; a, aorta; cvc, caudal vena cava; pd, pancreaticoduodenal vein; LK, left kidney; sp, spleen.

Clinical examination confirmed a cutaneous mass at the level of right scapular region and did not reveal any other abnormalities. Haematology, serum chemistry analyses and urinalysis results were within normal limits. CT of the whole body was performed as described for dog 1. CT findings included: a large, irregularly marginated, multilobulated, inhomogeneous, hypervascularized cutaneous and subcutaneous masse isoattenuating to the soft tissue with inhomogeneous contrast-enhancement latero-caudal to the right scapula. Mild enlargement of ipsilateral axillary lymph node was present. No volume abnormalities of the liver nor macroscopic structural or attenuation changes were evident. A large tortuous

vessel connecting splenic vein and left gonadic vein was observed with no evidence of free fluid in the abdomen (Fig. 4). An imaging diagnosis of soft tissue sarcoma associated with a left splenogonadal PSS was made. Table (1) shows the main clinical, imaging and histopathologic findings for each patient.

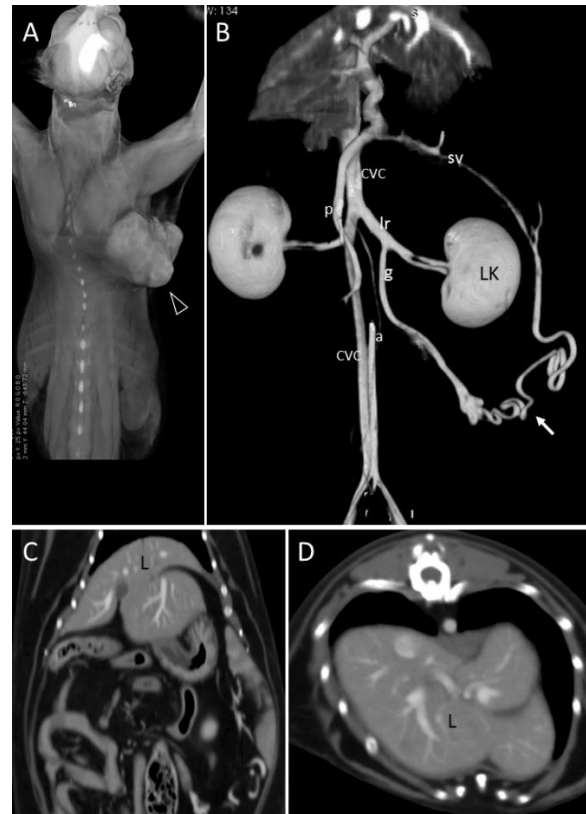


Fig. 4. Cat 2. (A) Dorsal thick-slab multiplanar reformatted contrast-enhanced CT image of thoracic region showing a large, irregularly marginated, multilobulated, hypervascularized cutaneous mass isoattenuating to the soft tissue with inhomogeneous contrast-enhancement latero-caudal to the right scapula (arrowhead). (B) Ventral three-dimensional volume-rendered CT angiography of portal system and caudal vena cava showing a large left splenogonadal PSS (arrow) connecting splenic vein (sv) with left gonadic vein (g) and entering the left renal vein (lr). (C) Dorsal and (D) transverse multiplanar reformatted contrast-enhanced CT images of abdomen at level of liver (L) showing absence of hepatic macroscopic structural abnormalities or attenuation/volume changes. Also note absence of ascites in the background (C). P, portal vein; a, aorta; cvc, caudal vena cava; LK, left kidney; sv, splenic vein.

Discussion

In veterinary medical literature the classical expected clinical and imaging findings in patients with portal hypertension are ascites, multiple APSS (either varices and large shunts) and a confirmed cause of portal flow obstruction at level of extrahepatic portal vein, liver, hepatic veins, posthepatic caudal vena cava, or right atrium (Buob *et al.*, 2011).

Table 1. Major clinical, imaging and histopatologic findings of each patient.

Case	Signalment	Clinical signs/clinical findings	Abdominal effusion	Pattern of porto-caval connections	invasive portal pressure measurement (cm H ₂ O)	CT appearance of the liver	Histopatologic diagnosis from liver biopsy
Dog 1	12-year-old intact male, Yorkshire Terrier	Inappetence, abdominal distension	Pure transudate	Single splenoazygos PSS	N/A	Mild decrease in liver volume with irregular margins	subacute-chronic centrolobular and midzonal hepatitis
Dog 2	8-year-old intact male mongrel	waxing and waning abnormal mentation; sporadic vomiting	absent	Left splenogonadal PSS; Gastro-phrenic varices;	11,5 (RI in normal dogs: 8-13; RI in dogs with PSS: 0-12)	microhepatia without parenchymal abnormalities	portal hypoperfusion
Cat 1	11-year-old spayed female DSH	lumbar cutaneous neoplasm (sarcoma)	absent	Single left splenogonadal PSS	N/A	multiple hepatic masses of presumptive metastatic origin	N/A
Cat 2	13-year-old spayed female DSH	Scapular cutaneous neoplasm (soft tissue sarcoma)	absent	Single left splenogonadal PSS	N/A	normal appearance	N/A

N/A: Not available; RI: Reference interval.

On the other hand patients with congenital PSS are traditionally described as having a single porto-caval connection without ascites or portal flow obstruction (Szatmari *et al.*, 2004; Bertolini *et al.*, 2006; Nelson and Nelson, 2011; Fukushima *et al.*, 2014). Furthermore, current classification of canine PSS differentiate repeatable and distinct vascular patterns of CPSS and APSS without any overlapping of anatomical pathway between each category (Ricciardi, 2016). These differences in the course of CPSS and APSS allow for their categorization during an angiographic study and help, in addition to other clinical and imaging findings, in the distinction between patients with PH from other with congenital porto-caval connection (Ricciardi, 2016). In the cases reported herein, some overlapping between clinical, histopathologic and imaging findings of PH and CPSS were observed, making the categorization of patients challenging.

In dog 1, a single splenoazygos PSS was observed in a patient with diffuse subacute-chronic hepatitis, which caused PH and ascitis. In major classifications of porto-caval connections in dogs the splenoazygos PSS pattern has been described among CPSS and never in the APSS group (Szatmari *et al.*, 2004; Bertolini *et al.*, 2006; Nelson and Nelson, 2011; Fukushima *et al.*, 2014). In the author's opinion, the presence of such PSS pattern in a dog with PH would corroborate two hypotheses:

1) Pre-existence of CPSS of splenoazygos pattern in a patient which has developed PH in adulthood due to an acquired liver disease. In this case, a CPSS may be not

sufficient to alleviate PH caused by primary diffuse intrahepatic parenchymal disease. Consequently, APSS and ascites develop. Interestingly a similar condition has been already reported in two dogs with PH in which a splenophrenic PSS, traditionally described as CPSS, was found in addition to other MAPSS (Ricciardi, 2016). This hypothesis would disavow the theory that the presence of a CPSS, in a dog with portal flow obstruction at level of the liver, would make development of PH unlikely bypassing portal blood in the systemic (caval) circulation, that presents the lowest resistance, (Szatmari, 2003). This eventuality would help refine imaging evaluation of patients with PH and CPSS reconsidering their haemodynamics.

2) The splenoazygos PSS may also be a pattern of APSS until now unreported, which regains patency if PH develops. The portal vein in the normal dog has at least three embryonic connections with the systemic venous system, which usually are not, or only minimally perfused. One of these connects the cardiac branches of the left gastric vein with the oesophageal branches of the azygos vein (Huntington and McClure, 1920; Bertolini, 2010a; Moubarak *et al.*, 2012). Such embryonic pathways of porto-caval connection may develop in splenoazygos APSS when PH occurs. Such hypothesis has already been considered for the splenophrenic PSS pattern - traditionally described as CPSS but also found in dogs with PH (Ricciardi, 2016) - since embryonic connections between phrenic vein and small branches of portal vein have also been

described in dogs (Huntington and McClure, 1920; Bertolini, 2010a; Moubarak *et al.*, 2012).

Interestingly, in human medical literature, a portoazygos shunt associated with multiple paraesophageal, retroperitoneal, parasplenic and gastric varices has been reported in an adult patient with liver cirrhosis and portal hypertension (Gebrael *et al.*, 2013). In this case, the authors considered the portoazygos PSS an acquired portal collateral developed as a consequence of PH. If splenoazygos and splenophrenic PSS represent pattern shared by both APSS and CPSS, categorization of the vascular anomaly based only on imaging findings (anatomical pathway) may be challenging.

In this case, the distinction between PH with APSS and CPSS may be influenced by the presence or absence of abdominal effusion as a frequent distinctive hallmark between these two conditions. However, as reported in humans (Gines *et al.*, 1987; Sarin and Kapoor, 2002; Sarin *et al.*, 2007) and dogs (Adam *et al.*, 2012; Ricciardi, 2016) ascites may be absent in some PH cases especially if the APSS are completely effective in alleviating portal pressure.

Hence, as previously reported for the splenophrenic PSS pattern (Ricciardi, 2016), in cases of primary microvascular or parenchymal hepatic disorders macroscopically undetectable on imaging evaluation (such as PHPV or hepatitis like in dog 1 of this series), the possible absence of ascites and the presence of a single acquired splenoazygos shunt (without other MAPSS) may disorient the presumptive diagnosis of PH (acquired splenoazygos PSS misinterpreted as CPSS). However, the number of PSSs may be helpful in such distinction since presence of more than one porto-caval connections has been considered a constant imaging finding shared among dogs with PH (Bertolini, 2010a; Ricciardi, 2016).

In dog 2, a large left splenogonadal PSS and varices near to the left kidney, also known as gastrophrenic varices (Bertolini, 2010a; Ricciardi *et al.*, 2014; Ricciardi, 2016), were found without ascites. These patterns of porto-caval connection have been reported as two of the most consistently observed route of APSSs in dogs (Szatmari *et al.*, 2004; Bertolini, 2010a; Ricciardi *et al.*, 2014; Ricciardi, 2016) suggesting non-ascitic PH in this patient. Regarding the portal vein pressure, in normal dogs it is between 8 and 13 cm H₂O, while patients with a PSS, because of the diversion of portal blood flow into the systemic circulation, the portal pressure is usually lower, ranging between 0 and 12 cm H₂O (Johnson *et al.*, 1987; Slatter, 2003). In our patient the portal pressure was 11.5 cm H₂O resulting not very helpful in discriminating between a PH and a normal portal pressure, even if a much lower value could be expected in the presence of such large splenogonadal PSS.

Liver histopathology was suggestive of parenchymal damage caused by portal hypoperfusion, as typically reported in patients with CPSS (Borrows, 2003; Isobe *et al.*, 2008). However, histopathologic liver changes may be identical in case of primary diseases causing PH (idiopathic non-cirrhotic PH (NCPH), portal venous hypoplasia, hepatic microvascular dysplasia and congenital arterioportal fistula) or in the case of reduced portal perfusion (congenital PSS). Thus, differentiation between PH with MAPSS due to primary liver disease and CPSS may not be possible based only on liver histopathologic findings (Van den Ingh *et al.*, 1995a, 1995b; Center, 1996; Bunch *et al.*, 2001). Hence, based on the overall histopathologic and imaging findings in this patient, three diagnostic hypotheses may be reasonably considered:

- 1) A PH due to primary liver diseases (idiopathic NCPH, portal venous hypoplasia, or hepatic microvascular dysplasia) with completely efficient MAPSS which avoided development of ascites. In this case, surgical closure of the PSS, as therapeutic approach for hepatic encephalopathy of this dog, would not be recommendable.
- 2) Transient PH developed in adulthood due to a self-limiting hepatic or portal disease (such as portal vein thrombosis or inflammatory liver disease), subsequently regressed, with opening of MAPSS. In this case, the multiple acquired PSS may represent the remnant of a transient episode of PH and, as responsible for the occurrence of hepatic encephalopathy without any other hemodynamic utility, may need surgical closure.
- 3) Multiple congenital porto-caval connections in a patient without PH. Although considered a rare phenomenon, multiple CPSS have also been described in dogs (Johnson *et al.*, 1987; Wilson *et al.*, 1997; Morandi *et al.*, 2005; Leeman *et al.*, 2013); in all cases however, large porto-caval or porto-azygous vascular connections were reported without varices nor splenogonadal pattern. Hence, this last hypothesis would redefine the classifications of CPSSs including among these even the left splenogonadal PSS and the porto-caval varices close to the left kidney. This implies that the finding of a single left splenogonadal PSS in a non-ascitic patient without macroscopic evidence of liver disease on imaging evaluation would be a diagnostic challenge – for discrimination between underlying PH and CPSS – especially in the case of unspecific liver histopathologic results (as those of dog 2) and doubt/border-line portal pressure values.

Regarding the hypothesis no. 1, NCPH is one of the most reported cause of presinusoidal intrahepatic PH in humans and dogs and, from histopathological point of view, it resembles primary hypoplasia of the portal vasculature (PHPV) so that the latter term is recommended by the World Small Animal Veterinary Association liver study group (Buob *et al.*, 2011). It is unclear, however, if the lesion in NCPH-PHPV patients is in all cases a primary hypoplasia of the intrahepatic portal vasculature or a consequence of a primary congenital or acquired disorder in hepatic perfusion (Buob *et al.*, 2011).

In humans the basic criteria to diagnose NCPH are: 1) presence of unequivocal signs of portal hypertension, 2) absence of diffuse liver diseases that can cause PH and 3) absence of occlusive disorders of the hepatic veins or of the portal vein (Schouten *et al.*, 2015). In our dog, the portal pressure measurement failed to testify an unequivocal condition of PH so that the hypothesis of an underlying NCPH-PHPV and MAPSS seems unlikely and a condition of multiple CPSS, of both large and small calibre, without PH, could not be ruled out.

Moreover, the hypothesis of a congenital origin of the left splenogonadal PSS has been previously reported in veterinary literature (Valentine and Carpenter, 1990). Interestingly a single left splenogonadal PSS without ascites was found in the two cats described in this series.

In cat 1 the finding of diffuse hepatic structural disorder (suggestive of diffuse hepatic metastases) made it reasonable to consider the splenogonadal PSS as a classical APSS recanalized as a consequence of raised intrahepatic portal pressure. In this cat, as previously described (Ricciardi, 2016), the lack of ascites may be explained by a complete effectiveness of the acquired splenogonadal PSS in alleviating portal pressure.

On the contrary, in cat 2, the left splenogonadal PSS was found as an incidental finding not associated with attenuation changes, nor volume abnormalities nor macroscopic structural alterations of the liver. Ascites was also absent.

In this case, two hypotheses may be reasonably considered:

- 1) as discussed in the third hypothesis of dog 2, the left splenogonadal PSS may be considered a congenital porto-caval connection, until now unreported among classification of CPSSs, causing subclinical, unrecognized or misinterpreted clinical signs (this cat had no history of clinical sign attributable to PSS).
- 2) as discussed in the second hypothesis of dog 2, transient PH developed in adulthood because of a self-limiting hepatic or portal disease with a single remnant APSS.

Unfortunately liver histopathology and ultrasonographic assessment of portal flow direction were not available in this patient.

In a series of 33 cats with single PSS connecting the splenic vein and the left renal vein or the adjacent segment of caudal vena cava, only 14 patients had a hepatopathy with the potential for associated PH. Adult spayed females were significantly overrepresented, as the cats of this report, and the portal blood flow, available in a limited number of patients, resulted to be hepatopetal in the majority of cases.

In this study, the authors concluded that the aetiology of the splenosystemic PSS in cats could not be definitely determined but, as hypothesized for the cat 2 reported herein, it may represent an acquired shunts secondary to past or present portal hypertension or it can be a congenital shunt of unknown clinical significance (Palermo *et al.*, 2013).

In the author's opinion, findings from these cases should prompt to reconsider porto-caval haemodynamic in dogs and cats with PSS, in the light of shunt phenotype and number, presence/absence of ascites, presence of macroscopic liver structural disorders, liver histopathologic results and, when possible, portal pressure measurement.

The splenoazygos PSS pattern, may be considered a classical CPSS pattern (Szatmari *et al.*, 2004; Bertolini *et al.*, 2006; Nelson and Nelson, 2011; Fukushima *et al.*, 2014) or an APSS until now unreported.

The left splenogonadal PSS and porto-caval varices near to the left kidney may be considered, as traditionally described, classical APSSs when found in patients with ascites and a confirmed cause of portal flow obstruction (Ricciardi *et al.*, 2014; Ricciardi, 2016), however, in non-ascitic patients without macroscopic evidence of liver disease, without evidence of hepatofugal portal blood flow or portal pressure measurement suggestive of PH, such PSS patterns may be also considered as CPSSs (encountered either as single CPSS, like in cat 2, or as multiple CPSSs, like in dog 2). In this last circumstance, the univocal presumptive diagnosis of NCPH with MAPSS may be questionable. Hence, as previously described for the splenophrenic PSS pathway (Ricciardi, 2016), acquired and congenital large PSSs may share the same anatomical pathway of porto-caval connection (here described for splenoazygos and left splenogonadal pattern). Such preliminar observations may help in reconsidering the distinction between CPSS and APSS, traditionally based on the shunt phenotype assessed by imaging evaluations (Bertolini *et al.*, 2006; Bertolini, 2010a, 2010b; Nelson and Nelson, 2011; Fukushima *et al.*, 2014; Ricciardi *et al.*, 2014; Ricciardi, 2016) and consequently the distinction between patient with PH from those with congenital PSS(s).

These findings, however, need to be enriched by further data regarding the direction of portal blood flow in patients with and without signs of PH and presence of left splenogonadal or splenoazygos PSS.

Finally, this case series emphasizes the importance of the precise imaging assessment of the PSS pattern, along with clinical and histopathologic findings, during diagnostic workup of patients with and without PH.

Conflict of interests

The Author declare that there is no conflict of interest.

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References

- Adam, F.H., German, A.J., McConnell, J.F., Trehly, M.R., Whitley, N., Collings, A., Watson, P.J. and Burrow, R.D. 2012. Clinical and clinicopathologic abnormalities in young dogs with acquired and congenital portosystemic shunts: 93 cases (2003-2008). *J. Am. Vet. Med. Assoc.* 241, 760-765.
- Baldwin, K., Bartges, J., Buffington, T., Freeman, L.M., Grabow, M., Legred, J. and Ostwald, D.Jr. 2010. AAHA nutritional assessment guidelines for dogs and cats. *J. Am. Anim. Hosp. Assoc.* 46(4), 285-296.
- Bertolini, G. 2010a. Acquired portal collateral circulation in the dog and cat. *Vet. Radiol. Ultrasound.* 51, 25-33.
- Bertolini, G. 2010b. MDCT for abdominal vascular assessment in dogs: MDCT basics, CT angiography, normal anatomy and congenital anomalies. LAMBERT Academic Publishing, Saarbrücken, Germany.
- Bertolini, G., Rolla, E.C., Zotti, A. and Caldin, M. 2006. Three-dimensional multislice helical computed tomography techniques for canine extrahepatic portosystemic shunt assessment. *Vet. Radiol. Ultrasound.* 47, 439-443.
- Bojrab, M.J., Waldron, D.R. and Toombs, J.P. 2014. *Current Techniques in Small Animal Surgery*. 5th ed. Jackson, W.Y.: Teton New Media, pp: 334.
- Borrows, C.F. 2003. Liver disorders. In: *Clinical Medicine of the Dog and Cat*, 1st ed, (Schaer, M. ed.), Veterinary Press, London, England, pp: 337-349.
- Bruehschwein, A., Foltin, I., Flatz, K., Zoellner, M. and Matis, U. 2010. Contrast-enhanced magnetic resonance angiography for diagnosis of portosystemic shunts in 10 dogs. *Vet. Radiol. Ultrasound.* 51, 116-121.
- Bunch, S.E., Johnson, S.E. and Cullen, J.M. 2001. Idiopathic noncirrhotic portal hypertension in dogs: 33 cases (1982-1998). *J. Am. Vet. Med. Assoc.* 218, 392-399.
- Buob, S., Johnston, A.N. and Webster, C.R. 2011. Portal hypertension: pathophysiology, diagnosis, and treatment. *J. Vet. Intern. Med.* 25, 169-186.
- Center, S.A. 1996. Hepatic Vascular Diseases, In: *Strombeck's Small Animal Gastroenterology*. 3rd ed. W.B. Saunders Company, Philadelphia, pp: 802-846.
- Ferrell, E.A., Graham, J.P., Hanel, R.S., Randell, S., Farese, J.P. and Castleman, W.L. 2003. Simultaneous congenital and acquired extrahepatic portosystemic shunts in two dogs. *Vet. Radiol. Ultrasound.* 44, 38-42.
- Fossum, T.W. 2002. *Small Animal Surgery*, 2nd ed., Mosby, St. Louis, pp: 457-468.
- Fukushima, K., Kanemoto, H., Ohno, K.M., Takahashi, R., Fujiwara, R., Nishimura, R. and Tsujimoto, H. 2014. Computed tomographic morphology and clinical features of extrahepatic portosystemic shunts in 172 dogs in Japan. *Vet. J.* 199(3), 376-381.
- Gebrael, J., Yu, H. and Hyslop, W.B. 2013. Spontaneous Portoazygos Shunt in a Patient with Portal Hypertension. *J. Radiol. Case Rep.* 7(7), 32-36.
- Gines, P., Quintero, E., Arroyo, V., Terés, J., Bruguera, M., Rimola, A., Caballería, J., Rodés, J. and Rozman, C. 1987. Compensated cirrhosis: natural history and prognostic factors. *Hepatology* 7(1), 122-128.
- Huntington, G.S. and McClure, C.F.W. 1920. The development of the veins in the domestic cat (*Felis domestica*) with especial reference, (1) to the share taken by the supracardinal veins in the development of the postcava and azygos veins and (2) to the interpretation of the variant conditions of the postcava and its tributaries as found in the adult. *Anat. Rec.* 20, 1-30.
- Isobe, K., Matsunaga, S., Nakayama, H. and Uetsuka, K. 2008. Histopathological Characteristics of Hepatic Lipogranulomas with Portosystemic Shunt in Dogs. *J. Vet. Med. Sci.* 70, 133-138.
- Johnson, C.A., Armstrong, P.J. and Hauptman, J.G. 1987. Congenital portosystemic shunts in dogs: 46 cases (1979-1986). *J. Am. Vet. Med. Assoc.* 191, 1478-1483.
- Lamb, C.R. 1996. Ultrasonographic diagnosis of congenital portosystemic shunts in dogs: Results of

- a prospective study. *Vet. Radiol. Ultrasound.* 37, 281-288.
- Leeman, J.J., Kim, S.E., Reese, D.J., Risselada, M. and Ellison, G.W. 2013. Multiple congenital PSS in a dog: case report and literature review. *J. Am. Anim. Hosp. Assoc.* 49, 281-285.
- Morandi, F., Cole, R.C., Tobias, K.M., Berry, C.R., Avenell, J. and Daniel, G.B. 2005. Use of ^{99m}TcO₄(-) trans-splenic portal scintigraphy for diagnosis of portosystemic shunts in 28 dogs. *Vet. Radiol. Ultrasound.* 46, 153-161.
- Moubarak, E., Bouvier, A., Boursier, J., Lebigot, J., Ridereau-Zins, C., Thouveny, F., Willoteaux, S. and Aubé, C. 2012. Portosystemic collateral vessels in liver cirrhosis: a three-dimensional MDCT pictorial review. *Abdom Imaging.* 37(5), 746-766.
- Nelson, N.C. and Nelson, L.L. 2011. Anatomy of extrahepatic portosystemic shunts in dogs as determined by computed tomography angiography. *Vet. Radiol. Ultrasound.* 52, 498-506.
- Palerme, J.S., Brown, J.C., Marks, S.L. and Birkenheuer, A.J. 2013. Splenosystemic shunts in cats: a retrospective of 33 cases (2004-2011). *J. Vet. Intern. Med.* 27, 1347-1353.
- Ricciardi, M. 2016. Splenophrenic portosystemic shunt in dogs with and without portal hypertension: can acquired and congenital porto-caval connections coexist? *Open Vet. J.* 6, 185-193.
- Ricciardi, M., Martino, R. and Assad, E.A. 2014. Imaging diagnosis--celiacomesenteric trunk and portal vein hypoplasia in a pit bull terrier. *Vet. Radiol. Ultrasound.* 55, 190-194.
- Sarin, S.K. and Kapoor, D. 2002. Non-cirrhotic portal fibrosis: current concepts and management. *J. Gastroenterol. Hepatol.* 17, 526-534.
- Sarin, S.K., Kumar, A., Chawla, Y.K., Bajjal, S.S., Dhiman, R.K., Jafri, W., Lesmana, L.A., Guha Mazumder, D., Omata, M., Qureshi, H., Raza, R.M., Sahni, P., Sakhuja, P., Salih, M., Santra, A., Sharma, B.C., Sharma, P., Shiha, G. and Sollano, J. 2007. Noncirrhotic portal fibrosis/idiopathic portal hypertension: APASL recommendations for diagnosis and treatment. *Hepatol. Int.* 1(3), 398-413.
- Schouten, J.N., Verheij, J. and Seijo, S. 2015. Idiopathic non-cirrhotic portal hypertension: a review. *Orphanet. J. Rare Dis.* 10, 67.
- Slatter, D.H. 2003. *Textbook of Small Animal Surgery*, 3rd ed. Philadelphia: Saunders, pp: 738.
- Szatmari, V. 2003. Simultaneous congenital and acquired extrahepatic portosystemic shunts in two dogs. *Vet. Radiol. Ultrasound.* 44, 486-487.
- Szatmari, V. and Rothuizen, J. 2002. How can you tell with ultrasound that a patient with high blood ammonia has a congenital or acquired portosystemic shunt or no shunt at all? In the Proceedings of the 27th Congress of the World Small Animal Veterinary Association, October 3-6, Granada, Spain, pp: 42.
- Szatmari, V., Rothuizen, J., van den Ingh, T.S., van Sluijs, F. and Voorhout, G. 2004. Ultrasonographic findings in dogs with hyperammonemia: 90 cases (2000-2002). *J. Am. Vet. Med. Assoc.* 224(5), 717-727.
- Valentine, R.W. and Carpenter, J.L. 1990. Spleno-mesenteric-renal venous shunt in two dogs. *Vet. Pathol.* 27, 58-60.
- Van den Ingh, T.S., Rothuizen, J. and Meyer, H.P. 1995a. Circulatory disorders of the liver in dogs and cats. *Vet. Q.* 17, 70-76.
- Van den Ingh, T.S., Rothuizen, J. and Meyer, H.P. 1995b. Portal hypertension associated with primary hypoplasia of the hepatic portal vein in dogs. *Vet. Rec.* 137, 424-427.
- Wilson, K., Scrivani, P. and Léveillé, R. 1997. What is your diagnosis? Portocaval and portoazygous shunts and microhepatia in a dog. *J. Am. Vet. Med. Assoc.* 211(4), 415-416.