The effectiveness of intraoperative mesenteric portography for preventing misdiagnosis of congenital absence of the portal vein in dog with extrahepatic portosystemic shunt: a case report

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Abstract

A 5-month-old mixed-breed dog and a 10-month-old Welsh corgi dog were presented for evaluation with signs of congenital portosystemic shunt. In both dogs, computed tomography angiography revealed a single extrahepatic portosystemic shunt with the absence of the portal vein cranial to the shunt origin. Intraoperative mesenteric portography (IOMP) was performed after temporary complete occlusion of the shunt, revealing the portal branches faintly opacified with contrast medium. A cellophane band was applied around the shunt vessel without intraoperative attenuation. Postoperatively, both the dogs showed significant clinical improvement and the portal veins, not observed on preoperative ultrasonography, were visualized on postoperative ultrasonography. This study suggests the efficacy of IOMP, enabling the appropriate surgical procedure to be determined.

Cellophane band, gradual attenuation, mesenteric portovenography, portosystemic vascular anomaly

Portosystemic shunts (PSSs) are vascular anomalies that allow portal blood draining directly from the hepatic portal circulation to the systemic circulation. The portal venous system carries blood from the major abdominal organs (gastrointestinal tract, pancreas, and spleen), and delivers approximately 75% of the liver's blood supply, 50% of the oxygen requirements of the liver, and specific hepatotrophic factors (Vollmar et al. 2009; Eipel et al. 2010). In dogs with PSSs, the shunting vessel diverts portal blood away from the liver, resulting in underdevelopment of the liver and increased concentration of gastrointestinal-derived factors (primarily ammonia) within the systemic circulation (Greenhalgh et al. 2010). Portosystemic shunts can be classified into intrahepatic, extrahepatic, and multiple acquired shunts resulting from congenital or developmental anomalies of the liver or portal circulation (Hunt 2004).

Portal vein aplasia is a rare anomaly caused by defective development of the portal venous system in the embryo (Hu et al. 2008). In dogs with portal vein aplasia, the portal circulation to the liver is completely absent. The portal vein embryologically develops from the vitelline veins (Payne et al. 1990; Matsuoka et al. 1992; Hu et al. 2008). Excessive involution of the periduodenal vitelline veins and failure to form critical anastomosis with hepatic sinusoids leads to complete or partial absence of the portal venous system (Payne et al. 1990; Hu et al. 2008). The anatomical close proximity of the primitive portal and systemic venous systems allows alternative anastomosis when normal critical anastomosis fails to form (Bertolini 2019). Portal vein aplasia in dogs is associated with anomalous vascular connections between the portal and systemic venous systems (Zwingerberger et al. 2011). Imaging techniques, such as ultrasonography and computed tomography (CT), alone cannot distinguish true portal aplasia from hypoplasia and atresia. The definitive diagnosis of portal vein aplasia requires intraoperative mesenteric portography (IOMP) and additional histological analysis of the hepatic parenchyma, which demonstrates the

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absence of hepatic venules within the portal triad (Matsuoka et al. 1992; Van den Ingh et al. 2006). This case report presents a possible risk of misdiagnosis of portal vein aplasia based on CT scan alone and provides evidence for effectiveness of post-temporary ligation IOMP in preventing this risk.

Case description

A 4-month-old, 5.3 kg sexually intact female Welsh corgi dog (Dog 1) was presented for evaluation of persistent vomiting with lethargy. CT angiography performed at the referring hospital revealed a single extrahepatic PSS associated with the absence of the portal vein cranial to the shunt origin and hepatic portal vasculature. Empirical treatment was initiated with famotidine (1 mg/kg, *per os* [PO], twice per day [BID]), metronidazole (15 mg/kg, PO, BID), cefazoline (30 mg/kg, PO, BID), S-adenosyl-L-methionine (10 mg/kg, PO, BID), and lactulose (0.5 ml/kg, PO, BID) for 2 weeks prior to the referral. The dog was fed a low protein diet. The clinical signs were markedly alleviated after 2 weeks of medical treatment; however, intermittent vomiting (once per day) persisted.

The dog was referred to the Konkuk Veterinary Medical Teaching Hospital for further evaluation and possible surgical treatment. Physical examination was unremarkable with no evidence of lethargy. Complete blood count and serum biochemistry tests revealed microcytic hypochromic anaemia (mean cell volume, 57.1 fl; reference range, 61.6–73.5 fl, mean corpuscular haemoglobin, 19.3 pg; reference range, 21.2–25.9, and haematocrit, 29.3%; reference range, 37.3–61.7%), hypoglycaemia (73 mg/dl; reference range, 77–150 mg/dl), hypoproteinaemia (4.2 g/dl; reference range, 4.8–7.2 g/dl), decreased blood urea nitrogen levels (4 mg/dl; reference range, 7–29 mg/dl), and elevated alkaline phosphatase (5.72 μkat/l; reference range, 0.77–5.62 μkat/l) and pre- and post-prandial bile acid levels (38.4 μmol/l and 81.4 μmol/l, respectively; reference range, 0–10 μmol/l and 0–25 μmol/l, respectively). The serum ammonia concentration was within the normal range (39 μmol/l; reference range, 0–99 μmol/l). All other laboratory values were within the reference ranges. Plain abdominal radiography revealed microhepatica, bilateral renomegaly, and a bladder calculus. On ultrasonography and CT, an anomalous vessel arising from the gastroduodenal vein and coursing to the caudal vena cava (CdVC) was identified (Plate VIII, Fig. 1). The shunt diameter of this vessel estimated by CT angiography was approximately 8 mm at the entrance point of the CdVC. The portal vein cranial to the shunt origin and portal branches within the hepatic parenchyma were not observed. A diagnosis of extrahepatic right-gastric caval shunt was established, and the shunting vessel was surgically managed using a cellophane band (CB) with the owner's consent.

General anaesthesia was induced and maintained with isoflurane in oxygen. A ventral midline celiotomy incision was made, and a large shunt vessel communicating with the CdVC was confirmed. After careful blunt dissection around the shunt at the insertion site into CdVC, a Rumel tourniquet using a segment of the 12-french silicone tube and 2-0 suture material was placed at the exposed parts of the shunt vessel. Intraoperative mesenteric portography was performed before (preligation) and after (postligation) temporary complete occlusion of the shunt vessel for assessment of portal venous system patency (Plate VIII, Fig. 2). Radiodense contrast medium was injected into the jejunal vein through a catheter and was imaged under fluoroscopy. Preligation images demonstrated that most of the contrast medium flowed through the shunt vessel to the CdVC without draining into the hepatic portal venous system. Postligation IOMP revealed that the portal vein cranial to the shunt origin and intrahepatic portal vasculature were faintly opacified. The presence and patency of the hepatic portal vasculature was confirmed; thus surgical intervention with gradual attenuation of the CB was performed. A three-layered CB, approximately 4 mm wide, was snugly placed circumferentially around the exposed parts of the shunt

vessel and secured using a surgical clip without intraoperative shunt attenuation (Plate IX, Fig. 3). Liver biopsy was performed for the histological assessment of the intrahepatic portal vasculature. The abdominal wall and skin were closed routinely. The dog recovered from anaesthesia without any complications. Analgesia was administered by a continuous infusion of butorphanol (0.1 mg/kg/h, intravenously [IV]) for 24 h and intermittent doses of butorphanol (0.1 mg/kg, IV) as needed for pain. Postoperative management included intravenous fluid therapy and maintenance of preoperative medications. Histopathological examination of the liver biopsy revealed lobular hypoplasia, hepatocellular atrophy, cytoplasmic vacuolization, sinusoidal dilation, and portal triads containing multiple tortuous arterioles (arteriolar hyperplasia), hyperplastic bile duct, and hypoplastic portal vein.

Postoperatively, the dog showed continuous improvement in clinical signs and rapidly gained body weight up to 6.5 kg within 3 weeks after the surgery. Although intermittent vomiting (once per week) persisted until 1 month postoperatively, the dog was in good health during the 2-year follow-up period without any symptoms. Pre- and post-prandial serum bile acid concentrations (5.2 μmol/l and 11.1 μmol/l, respectively; reference range, 0–10 μmol/l and 0–25 μmol/l, respectively) returned to normal at the 1-month postoperative follow-up, and other serum biochemical abnormalities returned to the normal limits at the 2-month postoperative follow-up. On follow-up ultrasonography, portal vein diameter was measured at the level of the porta hepatis. The portal vein cranial to the shunt origin, which was not visualized preoperatively, was observed as 2.7 mm in diameter at the 7-day postoperative follow-up and this diameter gradually increased up to 5.9 mm at the 3-month postoperative follow-up. The diameter of the shunt vessel was measured at the level where the CB was placed. The shunt vessel diameter was 3.2 mm at postoperative day 1 and attenuated to 0.3 mm (postoperative 6-month follow-up) without visualized blood flow within the vessel as identified on color Doppler ultrasonography. The blood flow turbulence in the CdVC near the shunt insertion site persisted on color Doppler ultrasonography until 2 months postoperatively; however, it was not detected at the 3-month postoperative follow-up.

A 11-month-old, 4.6 kg sexually intact female mixed-breed dog (Dog 2) was referred for evaluation of a 3-month history of lethargy, intermittent mental dullness with excessive salivation, delayed growth, and decreased appetite. On presentation, the dog was depressed, with a body condition score of 3/9. Complete blood count and serum biochemistry tests revealed hypochromic microcytic anaemia (mean cell volume, 58.2 fl; reference range, 61.6–73.5 fl, mean corpuscular haemoglobin, 19 pg; reference range, 21.2–25.9, and haematocrit, 34.6%; reference range, 37.3–61.7%), hypoproteinaemia (4.2 g/dl; reference range, 5.2–8.2 g/dl), hypocholesterolaemia (88 mg/dl; reference range, 110–330 mg/dl), hyperammonaemia (246 μmol/l; reference range, 0–98 μmol/l), and elevated aspartate aminotransferase (0.98 μkat/l; reference range, 0–0.85 μkat/l) and pre- and post-prandial bile acid concentrations (158.3 μmol/l and 597.9 μmol/l, respectively; reference range, 0–10 μmol/l and 0–25 μmol/l, respectively). Microscopic examination of urinary sediment confirmed the presence of ammonium urate and bilirubin crystals. On abdominal radiography, microhepatica was observed. Ultrasonography showed multiple tiny hyperechoic materials within the urinary bladder and a large tortuous vessel originating from a branch of the portal vein and coursing into the CdVC. The shunt vessel arising from the gastroduodenal vein and inserting into the CdVC was demonstrated on CT angiography. The diameter of the shunt vessel at the entrance point was approximately 13 mm. The portal vein cranial to the shunt origin and portal branches within the hepatic parenchyma were not observed. A diagnosis of extrahepatic right-gastric caval shunt was established.

The dog was fed a protein-restricted diet, and medical therapy with famotidine (1 mg/kg, PO, BID), metronidazole (15 mg/kg, PO, BID), amoxicillin-clavulanate (12.5 mg/kg,

PO, BID), S-Adenosyl-L-Methionine (10 mg/kg, PO, BID), and lactulose (0.3 ml/kg, PO, TID) was initiated for 10 days before surgical intervention. Laboratory tests performed on the day of surgery showed that the serum ammonia concentration had decreased to normal values (65 μmol/l; reference range 0–98 μmol/l). The dog underwent a similar surgical procedure as described for Dog 1 with the owner's consent. The portal vein cranial to the shunt origin and hepatic portal vasculature were faintly opacified on postligation IOMP. The hepatic portal vasculature was proved patent. A three-layered CB, approximately 4 mm wide, was snugly placed circumferentially around the exposed parts of the shunt vessel and secured using a surgical clip without intraoperative shunt attenuation. The abdominal wall and skin were closed in a routine manner. No major complications occurred, and the dog recovered uneventfully from anaesthesia. Postoperative analgesia was the same as that reported for Dog 1. Postoperative care included fluid therapy and maintenance of preoperative medications. The histopathological findings of the liver biopsy were consistent with those of Dog 1.

Postoperatively, the clinical signs significantly improved and the body weight increased up to 6.5 kg within 2 months after the surgery. Pre- and post-prandial serum bile acid levels (4.7 μmol/l and 13.5 μmol/l, respectively; reference range, 0–10 μmol/l and 0–25 μmol/l, respectively) returned to normal at the 2-month postoperative follow-up, and other biochemical abnormalities in laboratory values were within the normal limits at the 3-month postoperative follow-up. The portal vein diameter at the level of the porta hepatis was measured on follow-up ultrasonograhy. The portal vein with a diameter of 0.3 mm was first visualized at the 1-month postoperative follow-up, and the diameter gradually increased up to 4.3 mm at the 6-month postoperative follow-up. The diameter of the shunt vessel at the level where CB was placed decreased from 9.1 mm (postoperative day 1) to 3.6 mm (postoperative 6 months follow-up). However, the blood flow within the shunt vessel persisted on color Doppler ultrasonography. Revision surgery using an ameroid ring constrictor (ARC) was suggested for completely blocking the shunt. The owner refused additional surgery owing to economic concerns and decided to continue the medical treatment alone. No evidence of clinical signs of PSS was found during the entire follow-up period.

Discussion

Surgical attenuation of the shunting vessel is the treatment of choice for dogs with PSS as continued diversion of portal blood flow may further deteriorate the liver function. Many surgical options, including acute occlusion with ligatures and gradual attenuation with CB, ARC, hydraulic occluder, or intravascular embolization, have been described (Hunt et al. 2014; Yoon et al. 2014; Traverson et al. 2017). Up to 68% of dogs with PSS develop portal hypertension after acute total ligation of the shunting vessel (Youmans and Hunt 1999). Gradual, rather than acute, occlusion has been advocated because gradual attenuation allows the development of the hepatic structure in response to increased blood supply, while avoiding severe portal hypertension (Vogt et al. 1996; Youmans and Hunt 1999). The CB and ARC are the most commonly described surgical devices for gradual attenuation, and they are considered safe and effective based on low morbidity and mortality rates (Traverson et al. 2017). In an original study by Traverson et al. (2017), the suspected residual shunting rate upon ultrasonography was higher after placing CB (31.6%) than after placing ARC (0%). Despite residual shunting, most dogs do not show any persistent symptoms associated with PSS (Hunt et al. 2014). The significance of a small amount of residual blood flow within the shunt vessel on long-term prognosis remains unknown. Therefore, further controlled studies are required to elucidate the extent to which residual shunting adversely affects liver function. Revision surgical procedures using ARC and complete ligation of the vessel are occasionally performed for complete

occlusion of residual shunting following primary surgical attenuation (Traverson et al. 2017).

In PSS with concurrent portal aplasia or hypoplasia, acute complete shunt closure will result in fatal portal hypertension, acute shock, and death, especially in cases of aplasia (Matsuoka et al. 1992; White et al. 2003; Bertolini 2019). In such dogs, differentiating the true portal aplasia from hypoplastic or hypoperfused portal veins is necessary for providing proper treatment. First-line imaging evaluation for PSS in small animals is commonly performed with colour Doppler ultrasonography. Nowadays, CT angiography is the method of choice for the diagnosis and monitoring of portal vascular anomalies in veterinary practice because CT angiography allows comprehensive assessment of the relationship between portal and systemic venous systems by visualizing portal vasculature and other non-vascular structures of the abdomen. However, CT angiography has the following limitation; the hypoplastic portal vein is occasionally not visualized, most commonly secondary to diminished portal flow in association with PSS (White et al. 2003; Parry and White 2016). Furthermore, the failure to demonstrate small or hypoperfused vasculature with CT angiography has been reported (Lee et al. 2006; Parry and White 2016). Since contrast medium is not administered under pressure into the portal circulation as in postligation IOMP; portal vasculature can be underestimated on CT angiography. Intraoperative mesenteric portography following temporary complete occlusion of the shunt vessel has advantages over CT angiography, as postligation IOMP can differentiate true portal aplasia from hypoplasia or atresia and increase the visibility of the portal vasculature (Lee et al. 2006; Parry and White 2018). In one study consisting of 62 dogs and cats that had hypoplastic or atretic portal vasculature on preligation IOMP, 50 of these cases had an improved hepatic portal circulation on postligation IOMP (White et al. 2003). Lee et al. demonstrated that a well-developed portal vasculature identified on postligation IOMP could be a positive prognostic indicator for surgical outcome (Lee et al. 2006). Similar findings were observed in cats (Lipcomb et al. 2009).

In this case report, the portal vasculature, which was not identified on CT angiography and preligation IOMP, was revealed on postligation IOMP. This showed that hypoperfusion of the portal vein and hepatic portal branches was caused by the pressure gradient associated with PSS, and thereby, temporary complete occlusion of the shunt visualized and confirmed the patency of the intrahepatic portal vasculature. The decision whether to use CB or ARC for gradual attenuation was made based on the IOMP findings. Considering that the hepatic portal vasculature was faintly opacified on postligation IOMP, CB designed to induce slower attenuation than ARC was used in these cases (Youmans and Hunt 1999; Hunt et al. 2014). Cellophane band attenuation was expected to allow the hepatic and portal venous system to accommodate increased blood circulation, limiting the possibility of portal hypertension and secondary acquired portosystemic shunt development. In dog 1, attenuation by CB was enough to stop the blood flow within the shunt and considerably dilated portal vasculature. The serum biochemical abnormalities observed preoperatively in dog 1 returned to the normal ranges. In dog 2, the clinical signs improved and serum biochemical abnormalities returned to normal levels. However, residual shunting was present, with 60% occlusion of the original shunt vessel diameter. These results showed that although shunt attenuation could be incomplete, the CB could be used safely in dogs with PSS when postligation IOMP revealed patency of the hepatic portal vasculature.

In conclusion, congenital PSSs may accompany severe portal hypoperfusion that could mislead the diagnosis of portal vein aplasia followed by inappropriate treatment. An accurate assessment of the hepatic portal vasculature can be done through a thorough evaluation of postligation IOMP. In cases of PSS with hypoperfused portal vasculature, the rate of gradual attenuation by CB sufficiently allows the intrahepatic portal vasculature to develop and accommodate the increase in portal blood flow without the occurrence of portal hypertension.

Conflict of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

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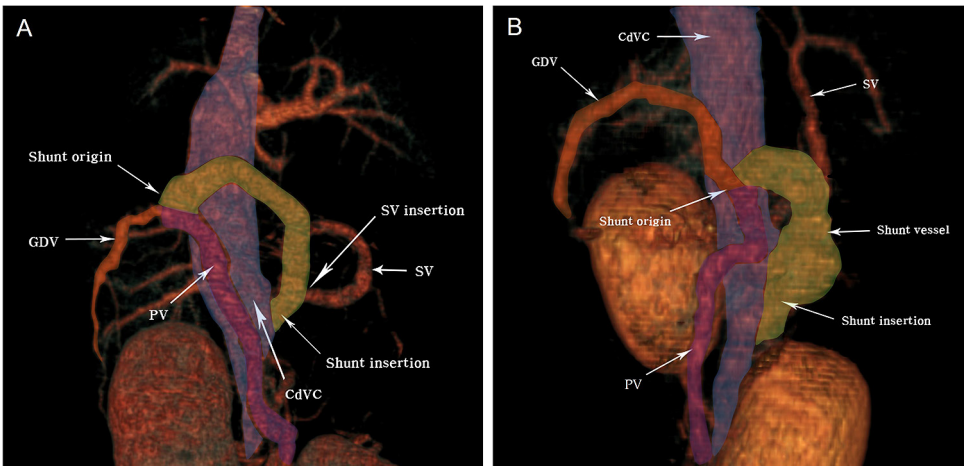

Fig. 1. Ventral view of the three-dimensional rendering of the portal vasculature of dog 1 (A) and dog 2 (B) during the portal phase computed tomographic scan. In both the dogs (A and B), the shunt vessel originates from the GDV (indicated as orange colour) and courses to the CdVC (indicated as blue colour). No portal vasculature cranial to the shunt origin is visualized. PV and shunt vessel are indicated, respectively, as purple and green colour.
CdVC, caudal vena cava; GDV, gastroduodenal vein; SV, splenic vein; PV, portal vein.

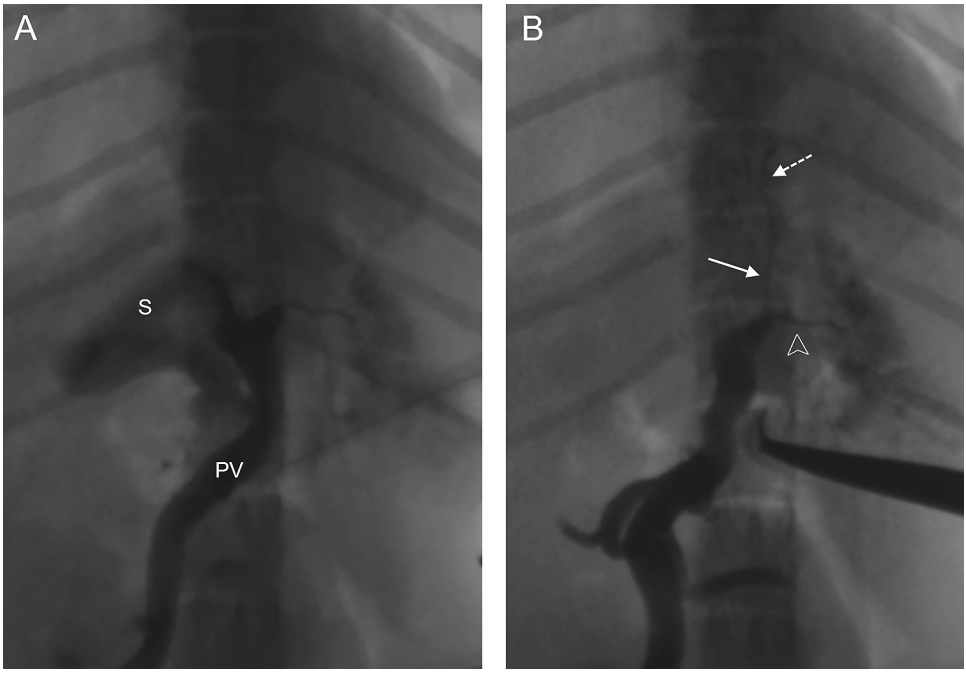

Fig. 2. Intraoperative mesenteric portography of dog 1 before (A) and after (B) temporary complete occlusion of the shunt vessel. (A) Most of the contrast medium flows through the shunt vessel to the caudal vena cava and the apparent lack of visualization of hepatic portal vasculature is seen. (B) The portal vein (arrow) cranial to the shunt origin from gastroduodenal vein (arrow head) and intrahepatic portal branches (dotted arrow) are faintly opacified.
S, shunt vessel; PV, portal vein.

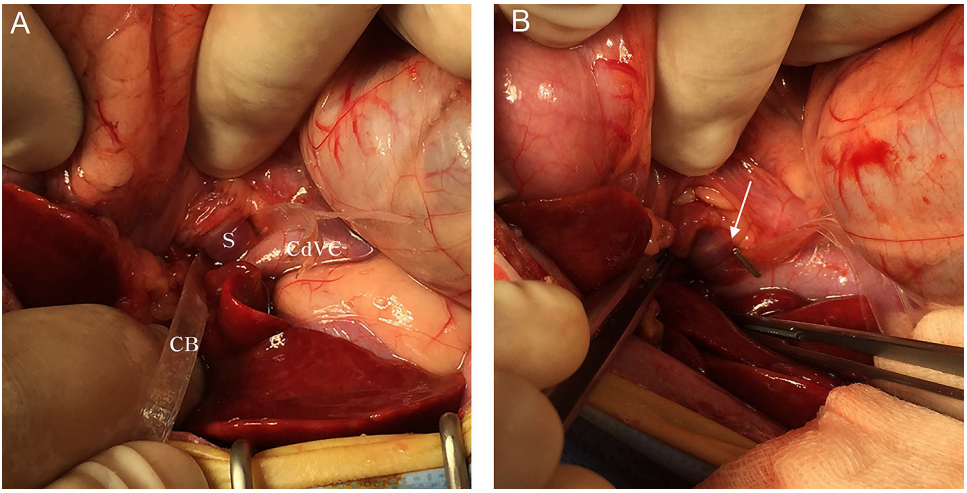

Fig. 3. Intraoperative image of dog 1 demonstrating cellophane band (CB) placement. (A) A three-layered CB, approximately 4 mm wide, is snugly placed circumferentially around exposed parts of the shunt vessel. (B) The CB is secured using a surgical clip without intraoperative shunt attenuation. The shunt vessel fits snugly into the ring of the CB (arrow).
S, shunt vessel; CB, cellophane band; CdVC, caudal vena cava.