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CASE REPORT

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Situs inversus totalis with single extrahepatic portosystemic shunt and azygos continuation of the caudal vena cava in a dog: a case report

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Abstract

Background A normal visceral arrangement is called situs solitus, whereas a state of visceral arrangement in a mirror-like positional relationship is called situs inversus (SI). Among the SI, the state in which the positions of only some thoracoabdominal organs are reversed is called situs inversus partialis, and the state in which the positions of all thoracoabdominal organs are reversed is called situs inversus totalis (SIT). Clinical information on dogs with SIT is limited.

Case presentation A 4-month-old Shiba Inu was referred with depression and neurological symptoms as the chief complaints. Computed tomography (CT) revealed the patient had SIT with an extrahepatic portosystemic shunt (EHPSS) and azygos continuation of the caudal vena cava. In addition, complete reversal of the lung lobes and cardiovascular system in the thoracic cavity was confirmed. The patient underwent surgery for partial attenuation of the EHPSS on day 8 after the initial examination. On day 124, after the initial examination, a second surgery was performed for complete attenuation. Under celiotomy, the positions of all abdominal organs, except for the rectum, were inverted; thus, SIT was confirmed via gross observation. In addition, the use of braided nylon sutures partially attenuated the concurrent portocaval shunt. At the conclusion of this study, approximately 6 years had passed since the second surgery, and the patient had a good general condition without any medications.

Conclusion In SIT, the complex anatomy of the abdominal organs and vessels is difficult to identify via gross observation. In contrast, CT is effective in detecting vascular abnormalities, confirming the anatomical position of each organ, and allowing for the definitive diagnosis of SIT.

Keywords Computed tomography, Dog, Portosystemic shunt, Situs inversus totalis, Surgery

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Background

The normal visceral arrangement is called situs solitus, whereas the state of visceral arrangement in a mirror-like positional relationship is called situs inversus (SI) [1, 2]. In SI, the state in which only the positions of some thoracoabdominal organs are reversed is called situs inversus partialis (SIP), and the state in which the positions of all thoracoabdominal organs are reversed is called situs inversus totalis (SIT). In addition, when the entire anatomical left–right axis is neither normal nor mirror image-inverted and shows an abnormal arrangement across the left–right axis, it is called heterotaxy (or situs ambiguus) [1, 2]. SIT is often associated with congenital malformations, such as vascular abnormalities [3, 4], and is extremely rare [4].

In veterinary medicine, only 11 dogs and one cat have been reported [5–15]; however, the accurate incidence rate in clinical settings remains unknown. Two reports described dogs showing SI concurrently with an extrahepatic portosystemic shunt (EHPSS) and azygos continuation of the caudal vena cava (CVC) [8, 16]. However, there is no veterinary literature on the long-term course of surgical treatment after a definitive diagnosis using preoperative computed tomography (CT) in dogs with SIT. This report aimed to describe the CT and surgical findings and postoperative prognosis of a dog with SIT, EHPSS, and azygos continuation of the CVC.

Case presentation

The patient was a 4-month-old female Shiba, weighing 3.7 kg. The patient was examined at the referring hospital with chief complaints of depression and neurological symptoms. The patient had high levels of total bile acid (TBA) and underwent dietary management and lactulose administration. Ten days later, the patient was referred to our hospital. The owner of the dog provided informed consent for all diagnostic and therapeutic procedures performed.

During the first evaluation, a general physical examination revealed no obvious neurological abnormalities. Hematology, serum chemistry, and coagulation test results are shown in Table 1. Serum chemistry showed abnormal levels of total serum protein (4.1 g/dL), albumin (2.0 g/dL), glucose (88 mg/dL), blood urea nitrogen (3 mg/dL), and total cholesterol (92 mg/dl). Blood coagulation tests also showed prolonged activated partial thromboplastin time (APTT; 23.3 s) and low antithrombin III activity (AT III; 51%). Radiography revealed microhepatitis and dextrocardia. Ultrasonography was not performed because the patient was restless. Electrocardiography revealed normal sinus rhythm and right-axis deviation. Contrast-enhanced CT under general anesthesia confirmed complete reversal of the lung lobes and cardiovascular system in the thoracic cavity (Fig. 1). In addition, complete inversion of the internal organs and vascular system in the abdomen, portocaval shunt (EHPSS), and azygos continuation of the CVC were confirmed (Fig. 2, Supplementary File S1). The owner was informed of the surgical treatment for attenuation of the EHPSS. Informed consent was obtained, and the patient underwent surgery eight days after the initial examination.

The pre-anesthesia medications were subcutaneously injected: 0.04 mg/kg atropine (Mitsubishi Tanabe Pharma Co., Osaka, Japan), 1.0 mg/kg prednisolone (Kyoritsu Seiyaku Co., Tokyo, Japan), 1.0 mg/kg maropitant (Cerenia®; Zoetis, Parsippany, NJ, USA), and 2.0 mg/kg ranitidine (Zantac®; GlaxoSmithKline K.K., London, UK). Anesthesia was induced by intravenous administration of 2.4 mg/kg propofol (Mylan; Mylan Seiyaku Ltd., Tokyo, Japan). Tracheal intubation was performed for mechanical ventilation using isoflurane (IsoFlo; Zoetis) and pure oxygen (2 L/min). For intraoperative and postoperative medical management, dopamine (2.5–6.0 μg/kg/min) (Teva Takeda Pharma Ltd., Nagoya, Japan), dobutamine (2.5–5.0 μg/kg/min; Kyowa Pharmaceutical Industry Co. Ltd., Osaka, Japan) and nafamostat (0.1–0.2 mg/kg/h; serine protease inhibitor; Nichi-Iko Pharmaceutical Co. Ltd., Toyama, Japan) was infused continuously. For analgesic management, continuous infusion of remifentanil (2.0–35 μg/kg/h) during and after surgery, and intramuscular administration of morphine (0.3 mg/kg each dose) before and after surgery were used. Furthermore, 20 mg/kg cefazolin (Nichi-Iko Pharmaceutical Co., Ltd.) was administered intravenously at the time of anesthesia induction. As abnormalities in APTT and AT III were revealed in the preoperative blood coagulation test, whole blood transfusion (total volume, 100 ml) was administered intraoperatively and postoperatively to enhance hemostatic function.

Under general anesthesia, the patient was placed in dorsal recumbency and prepared for abdominal surgery. A celiotomy was performed from the xiphoid to the middle of the umbilicus and pubis. The positions of the internal organs in the abdominal cavity were confirmed, and the positions of all abdominal organs, except for the rectum, were inverted (Fig. 3A). The hepatic lobe corresponding to the right lateral lobe was atrophied, and the hepatorenal ligament was absent (Fig. 3B). Polysplenia was identified based on gross examination. The gastric and intestinal tracts were pulled caudally to identify portocaval shunts. One greater omentum was manually separated, and the stomach was lifted to expose and detach the shunt vessels. A cannula was placed in the mesenteric vein using a 24–G indwelling needle (Surflow® indwelling needle, Terumo Co., Ltd., Tokyo, Japan) to measure portal pressure (10 mmHg). Iohexol (350 mg I/ml) was administered through the mesenteric vein,

Table 1 Hematology, serum chemistry, and coagulation test results of the patient

Parameter		Unit	Initial examination	POD 97*	POD 129†	Normal range
RBC		10^6/μL	6.0	8.1	6.9	5.5–8.5
PCV		%	35	46	41	37–55
WBC		/μL	8,900	11,000	9,500	6,000–17,000
	Stab	/μL	0	NA	NA	0–300
	Seg	/μL	5,207	NA	NA	3,000–11,500
	Lym	/μL	2,581	NA	NA	1,000–4,800
	Mono	/μL	845	NA	NA	150–1,350
	Eos	/μL	267	NA	NA	100–750
Plt		10^3/μL	126	188	184	200–500
TP		g/dL	4.1	5.6	5.8	4.8–7.2
Alb		g/dL	2.0	2.7	2.8	2.1–3.6
Glu		mg/dL	88	90	89	90–140
AST		U/L	86	26	30	0–50
ALT		U/L	190	19	24	8–75
ALP		U/L	226	129	112	46–337
GGT		U/L	6	10	4	0–2
BUN		mg/dL	3	10	14	7–29
Cr		mg/dL	0.3	0.6	0.7	0.3–1.2
TCho		mg/dL	92	231	210	100–400
NH_3		μmol/L	30	24	0	0–99
Na		mmol/L	145	144	144	134–153
K		mmol/L	4.1	4.4	3.9	3.4–4.6
Cl		mmol/L	113	110	110	105–118
CRP		mg/dL	0.61	0.27	0.09	0–1.0
APTT		s	23.3	NA	NA	10–16
PT		s	7.6	NA	NA	6–8
Fib		mg/dl	155.7	NA	NA	86–375
ATIII		%	51	NA	NA	102–156

RBC, red blood cell count; PCV, packed cell volume; WBC, white blood cell count; Stab, stab neutrophil; Seg, segmented neutrophil; Lym, lymphocyte; Mono, monocyte; Eos, eosinophil; Plt, platelet; TP, total protein; Alb, albumin; Glu, glucose; AST, aspartate aminotransferase; ALT, alanine aminotransferase; ALP, alkaline phosphatase; GGT, gamma-glutamyltransferase; BUN, blood urea nitrogen; Cr, creatinine; TCho, total cholesterol; NH_3, ammonia; CRP, c-reactive protein; APTT, activated partial thromboplastin time; PT, prothrombin time; Fib, fibrinogen; ATIII, antithrombinIII

*POD 97, postoperative day 97; 97 days after the first operation, prior to the second operation

†POD 129, postoperative day 129; 129 days after the first operation, which was 13 days after the second operation

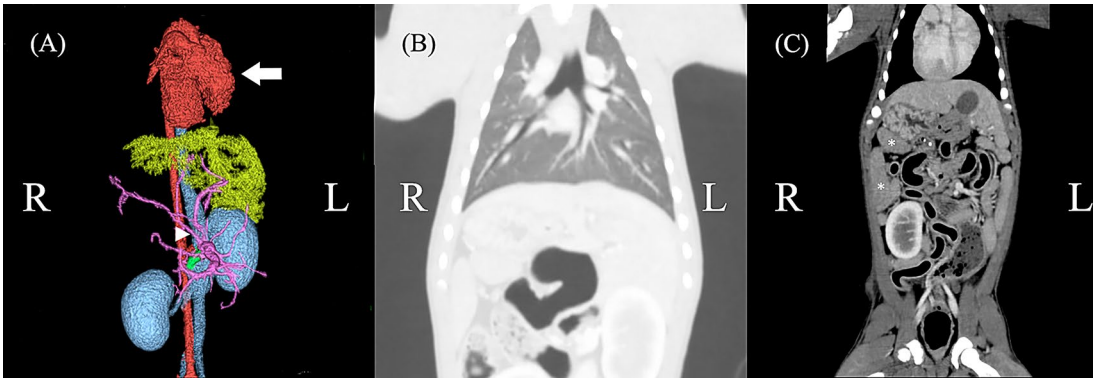

Fig. 1 Preoperative computed tomography images of the patient. **A**: Three-dimensional reconstruction image showing the heart (white arrow) inverted left to right, with the aorta (red) originating from the right aortic arch. The caudal vena cava (CVC; blue) runs on the left side of the aorta and connects to the azygos vein (white arrowhead) after merging with the renal vein. There is no CVC from the kidney to the liver. The left and right kidneys are reversed, and a shunt blood vessel (green) is present between the portal vein and CVC, confirming the presence of an extrahepatic portosystemic shunt. **B**: Coronal image of the lungs showing the caudal accessory lobe on the left side with the left caudal bronchus branching to the left accessory lobe. **C**: Coronal image of the heart and abdominal organs showing the apex of the heart located on the right side, gallbladder positioned on the left, and gastric fundus located on the right. Spleen (asterisk) indicates polysplenia

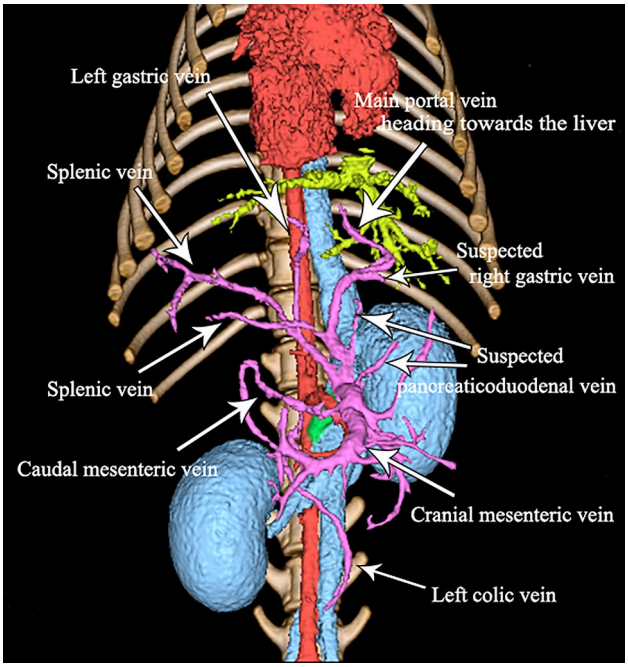

Fig. 2 Preoperative three-dimensional reconstruction computed tomography image of the patient. The right gastric and pancreatoduodenal veins were suspected because of unclear contrast enhancement, making accurate identification and delineation of these vessels challenging. Suboptimal contrast enhancement of the portal venous system in this region hinders comprehensive evaluation of these vessels

and intraoperative mesenteric portography (IOMP) was performed to visualize abnormal blood flow (see Supplementary file S2). Additionally, IOMP was performed again by temporarily closing the shunt vessel using atraumatic bulldog forceps. Portal vein blood flow toward the cranial side of the closed site was poor, and no abnormal blood flow other than that in the shunt blood vessel was observed, while the portal pressure increased to 44 mmHg (Fig. 3C). Partial ligation was performed using a coated braided nylon suture, size 0 (Surgilon; Medtronic Inc., Minneapolis, MN, USA), at a portal pressure of 12 mmHg. A portion of the margin of the left hepatic lateral lobe was obtained for histopathological examination. Subsequently, ovariohysterectomy was performed, and the abdominal wall, subcutaneous tissues, and skin were closed.

The operative time was 75 min. The patient recovered well after anesthesia. No abnormalities were observed in the general condition during hospitalization, and medical management was performed with cefalexin (35 mg/kg bid; Larixin®; FUJIFILM Toyama Chemical Co., Ltd., Tokyo, Japan) and oral administration of lactulose (2 mL/head bid; Kowa Co. Ltd., Osaka, Japan). The patient was discharged six days after surgery. Histopathological examination of the liver biopsy samples revealed poor vascular development of the portal system, irregular lobular structures, and mild atrophy of hepatocytes.

At 97 days after the first surgery, blood examination revealed that the abnormal values observed at the first visit had improved (Table 1). Contrast-enhanced CT under general anesthesia revealed narrowing of the shunt vessel and development of an intrahepatic portal vein branch (Fig. 4; Supplementary file S3). Therefore, 116 days after the first surgery, a second surgery was performed to completely attenuate the shunt vessel. The same anesthetic management was used as in the first surgery; however, no blood transfusions were performed. When the portal pressure was measured during celiotomy, as in the first surgery, the portal pressure was 11 mmHg. Although the portal pressure increased to 15 mmHg after temporary occlusion of the shunt vessel, cyanosis of the pancreas and intestinal tract and increased peristaltic motility of the gastrointestinal tract were not observed. In addition, IOMP revealed the development of intrahepatic portal branches (Fig. 5; Supplementary File S4). The shunt vessel was completely ligated using a size 0 coated braided nylon suture (Surgilon). The abdominal wall, subcutaneous tissue, and skin were closed.

The second surgery lasted for 44 min. The patient's general condition was good during the second hospital stay, and she was discharged 6 days after surgery, following continued medical management with cefovecin sodium (8 mg/kg sc; Convenia®; Zoetis) and dietary management with a liver therapy diet. On the 13th day after the second surgery, the patient was in a good general condition without dietary control. No abnormal values were detected in the blood test (Table 1), and TBA level was within the normal range (fasting: 1.8 μmol/L; postprandial: 2.1 μmol/L).

At the conclusion of this study, approximately 6 years had passed since the second surgery, and the patient had a good general condition without any medications. The owner provided written informed consent for the publication of this case report, including the use of any accompanying images and clinical details.

Discussion and conclusions

SIT is extremely rare in humans. A study using human data from the National Birth Defects Prevention Study reported an estimated prevalence of 0.3/10,000 live births [4]. Although no epidemiological studies have been conducted in veterinary medicine, the incidence is likely to be as low in animals as in humans.

The portal vein develops embryologically from the right ventricle. The development of the venous system in the abdominal cavity has been reported to differ among animal species; however, it is generally recognized that the CVC and azygos veins are formed as a result of the appearance, anastomosis, and disappearance of the caudal cardinal, subcardinal, and supracardinal veins [17].

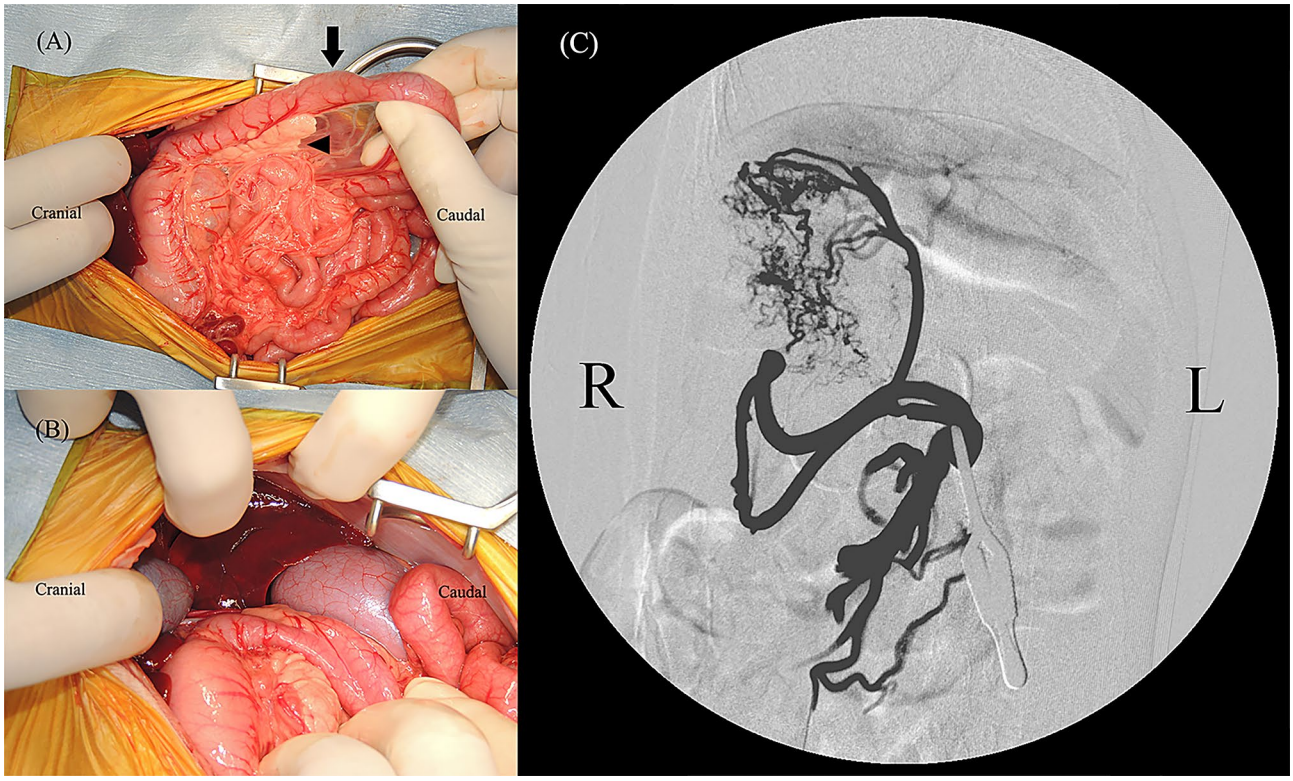

Fig. 3 Intraoperative findings in the first surgery. **A**: Gross findings during laparotomy. Most intra-abdominal organs, except the rectum, were inverted. The pylorus of the stomach is located on the patient's left side, and the duodenum (black arrow) descends on the left side. The pancreas (black arrowhead) is similarly inverted. **B**: Gross findings upon lifting the duodenum from left to right. In normal dogs, the liver is inverted, with the left hepatic division corresponding to the usual location of the right hepatic division. The left kidney, positioned where the right kidney normally resides, is in contact with the left hepatic lobe, and no hepatorenal ligament is formed. **C**: Intraoperative mesenteric portography during temporary occlusion of the shunt vessel. Poor portal vein blood flow to the liver and underdeveloped intrahepatic portal veins were found, and no other shunt vessels are identified

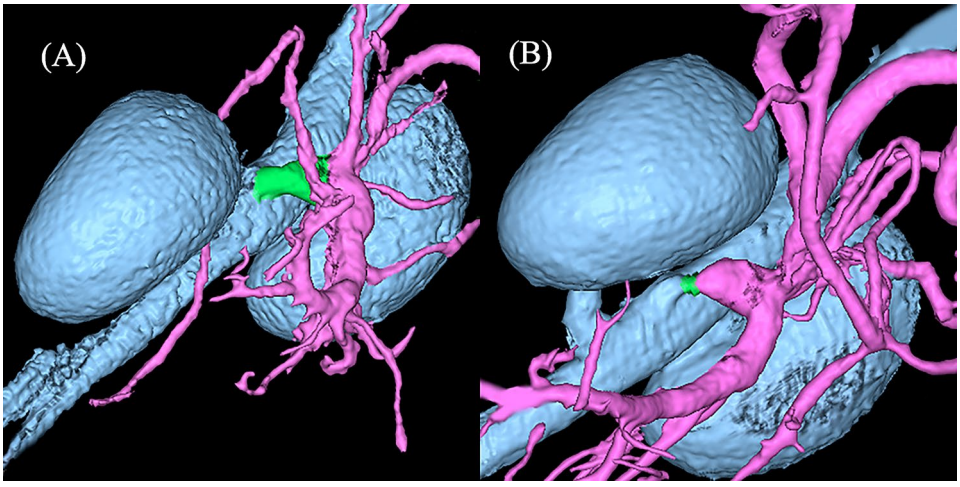

Fig. 4 Comparison of three-dimensional reconstruction computed tomography (3D-CT) images before the first and second surgeries. (**A**) 3D-CT image prior to the first surgery, showing the shunt vessel (green) without evidence of narrowing. (**B**) 3D-CT image prior to the second surgery, demonstrating narrowing of the shunt vessel (green) caused by partial ligation in the first surgery

During interruption of the CVC and azygos continuation, the right subcardinal vein, which forms the suprarenal segment of the CVC during development, fails to connect with the right vitelline vein, which forms the intrahepatic segment. In this malformation, blood in the caudal region flows through the azygos vein and cranial vena cava to the heart because of the switching of the blood passage to the right supracardinal vein [8, 18–20].

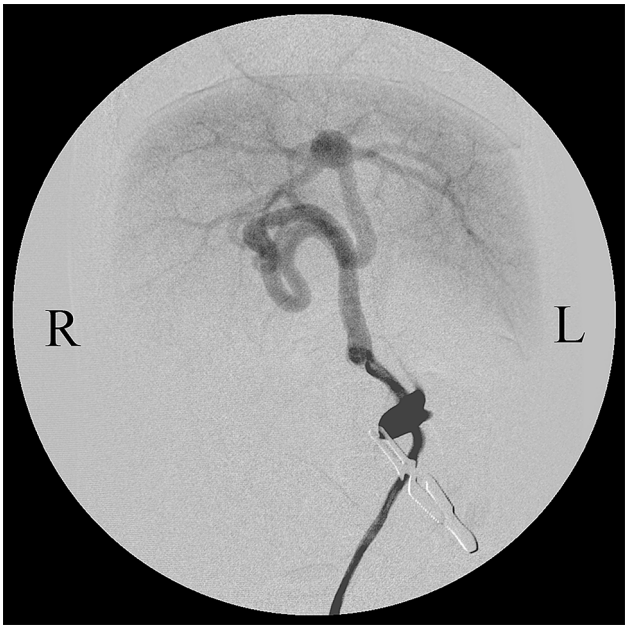

Fig. 5 Intraoperative mesenteric portography during temporary closure of the shunt vessel at the second surgery. The intrahepatic portal vein branches are visualized up to the tertiary branches, confirming improved portal vein development in the second surgery compared to the first surgery

EHPSS is an abnormal congenital anastomosis thought to be caused by the influx of the right vitelline veins from the caudal site into the right cardinal vein [21]. The association between SI and vascular malformations is unclear because the placement of internal organs is determined prior to developmental changes in the cardiovascular system [8]. However, an abnormal visceral placement can predispose individuals to errors during the development of complex cardiovascular systems.

In addition to SIT, this case exhibited several abnormalities in portal venous morphology. Owing to polysplenia, the splenic veins were found to connect to the individual spleens. The right gastric vein and right gastroepiploic vein were challenging to identify on CT imaging. Tortuosity of the portal vein on the cephalic side of the gastroduodenal vein was also observed in the CT images taken prior to the second surgery. Notably, the left gastric vein, which courses along the lesser curvature of the stomach, was anatomically connected to the tortuous segment of the portal vein, rather than to the splenic vein. These abnormalities may occur concurrently with SIT. While CT is a useful diagnostic tool for SIT and extrahepatic PSS, concurrent abnormalities with SIT may not always be identifiable on CT alone. Thus, IOMP and intraoperative gross findings are essential for identifying abnormalities beyond SIT.

SIT is usually asymptomatic and is often accidentally diagnosed [1, 2]. However, SI may be associated with airway abnormalities, such as chronic rhinosinusitis and bronchiectasis, that is, Kartagener syndrome (KS), which is recognized as a subtype of primary ciliary dysfunction (PCD) [3, 9, 22]. In human medicine, 20–25% of patients with SIT are estimated to have KS [3]. Although respiratory symptoms were expected, no clinical symptoms indicative of KS were observed in this case.

In humans, laterality defects are associated with various congenital malformations, including those of the cardiovascular system [3, 4], which result in clinical symptoms. Previous reports of SIT patients in veterinary medicine have shown that hydrocephalus [9, 23], congenital heart disease [9, 14], vascular malformation [8], diaphragmatic hernia [13], and respiratory diseases, including KS [5, 7, 9, 11, 12, 23], may occur. There have also been a few cases [10, 15] with no comorbidities. Additionally, there have been previous reports of the co-occurrence of EHPSS in patients with SI or heterotaxy [8, 16, 24]. In these reports, clinical symptoms caused by congenital malformations emerged and led to therapeutic interventions. In human medicine, interruption of the CVC and azygos continuation is generally considered asymptomatic [25, 26]. In veterinary medicine, there are case reports of vascular malformations, such as EHPSS [8, 19, 25], deep vein thrombosis [18], and pulmonary thromboembolism [26], associated with azygos continuation in CVC. Based on these previous reports, although the causal relationship between SIT, EHPSS, and azygos continuation of the CVC remains unclear in the present case, it is likely that these conditions may have contributed to the development of one another. Furthermore, therapeutic intervention was initiated in this case due to the clinical symptoms caused by EHPSS.

In humans, SIT can be accidentally detected when CT is performed before surgery. Additionally, if the altered anatomical positional relationship is understood to be the result of CT, surgical treatment can be performed in the same manner as in patients with normal anatomical structures [27]. CT is also been shown to be effective in detecting congenital malformations of the superior and inferior vena cava [28]. CT is effective in detecting cardiovascular malformations associated with heterotaxy patients [29]. Furthermore, CT examinations of PCD patients have been reportedly effective in determining the presence or absence of bronchiectasis and its severity [30]. In veterinary medicine, recent studies have described the anatomical findings of EHPSS using CT angiography and intraoperative mesenteric portography, demonstrating their significant utility as diagnostic tools [31–34]. Therefore, this case illustrates that preoperative CT is very effective in detecting SIT and EHPSS, which require therapeutic intervention. CT is essential for assessing the presence of anatomical abnormalities that may interfere with surgical procedures. Due to the unusual anatomical position of the abdominal organs in

this case, surgery after laparotomy was expected to be difficult. However, the CT findings made it possible to safely perform the operation.

Laterality defects, also known as defects in left–right-axis patterning or malpositioned complexes of embryos, have been recognized as pathological conditions, including SIT and heterotaxy [2, 4]. Previous reports have revealed that laterality defects are caused by defects in signal transduction at the nodes, including in the structure and function of cilia and asymmetrically expressed genes [1–3, 35]. PCD, which can be associated with SI in 41–48% of cases [36, 37], is a rare hereditary disease caused by mutations in genes related to cilia, such as DNAH5 and DNAI1, and exhibits autosomal recessive or X-linked modes of inheritance [35, 36]. Although reports suggest autosomal recessive inheritance in dogs with PCD [22, 23], no extensive genetic studies have been conducted in dogs. Because our canine case showed the same pathological condition as that found in humans, the genetic or embryological mechanisms reported in humans may have occurred.

In summary, the complex anatomy of the abdominal organs and vessels is difficult to identify grossly in cases of SIT, whereas CT is effective for detecting vascular abnormalities, confirming the anatomical position of each organ, and for the definitive diagnosis of SIT.

Abbreviations

APTT	Activated partial thromboplastin time
AT III	Antithrombin III
CT	Computed tomography
CVC	Caudal vena cava
EHPSS	Extrahepatic portosystemic shunt
IOMP	Intraoperative mesenteric portography
KS	Kartagener syndrome
PCD	Primary ciliary dysfunction
SI	Situs inversus
SIP	Situs inversus partialis
SIT	Situs inversus totalis
TBA	Total bile acid

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12917-025-04565-7.

Supplementary Material 1: Additional file 1: Movie of three-dimensional (3D) reconstruction of computed tomography angiography (CTA) before the first surgery. CTA 3D reconstruction showed complete inversion of the vascular system in the abdomen, a portocaval shunt (EHPSS), and azygos continuation of the caudal vena cava.

Supplementary Material 2: Additional file 2: Movie of intraoperative mesenteric portography (IOMP) with digital subtraction angiography in the first surgery. During temporary occlusion of the shunt vessel using atraumatic bulldog forceps, portal venous flow was indicated to enter the inverted liver via IOMP.

Supplementary Material 3: Additional file 3: Movie of three-dimensional (3D) reconstruction of computed tomography angiography (CTA) before the second surgery. CTA 3D reconstruction revealed a reduction in shunt flow because of partial surgical ligation and the development of intrahepatic portal vascularity.

Supplementary Material 4: Additional file 4: Movie of intraoperative mesenteric portography (IOMP) with digital subtraction angiography in the second surgery. During the temporary occlusion of the narrowed shunt vessel using atraumatic bulldog forceps, IOMP revealed no shunt vessels or the development of intrahepatic portal vascularity; therefore, complete surgical ligation was achieved.

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Author contributions
RT, KI, HK, TF, YS, MS, and KA contributed to the study design and planning. All authors were involved in perioperative management and collection of data from all the dogs. All authors participated in writing the manuscript and read and approved the final manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
Informed consent was obtained from the owner before all procedures were performed. All methods in this study were performed in accordance with the relevant guidelines and regulations in compliance with the ARRIVE guidelines. This study was approved by our Institutional Ethical Committee.

Consent for publication
Not applicable.

Competing interests
The authors declare no competing interests.

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