Congenital Peritoneo-Pericardial Diaphragmatic Hernia (PPDH) in A Dog with Thoracic Cavity Wall, Pericardium, and Ductal Plate Malformation

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ABSTRACT
Objective: To describe the clinical presentation, investigation findings, and outcome of a peritoneo-pericardial diaphragmatic hernia in a dog with thoracic cavity wall, pericardium, and ductal plate malformation.

Study Design
Case Report
Methods: A one-and-a-half-year-old female neutered Labradoodle was presented to a referral centre for acute distension of the abdomen and vomiting. The investigations confirmed herniation of the abdominal organs into the chest, sternal malformations, transitional cervical vertebral anomaly, and cervical ribs, alongside other findings described in detail in the article. A midline coeliotomy was performed to reposition the organs and suture the diaphragm. Sternotomy was also performed to fix the incomplete pericardial genesis, which made the peritoneal-pericardial diaphragmatic hernia (PPDH) more challenging. After the sternotomy, the pericardium edges were detached from the loose connective tissue of the mediastinum. A liver biopsy also revealed a ductal plate malformation, reported to be associated with the PPDH.

Results: To the author's knowledge, this is the first case report of a congenital PPDH in a dog associated with thoracic cavity wall, pericardium, and ductal plate malformation. The surgery was uneventful and the dog was discharged 5 days after presentation.

Conclusions: Congenital PPDH, incomplete pericardium genesis, abnormal chest wall conformation, and ductal plate malformation can all be concurrent in a single patient with mild non-specific clinical signs, and surgical procedures to address all of these conditions can be performed successfully under the same general anesthesia.

Introduction
The two main functions of the chest wall are protection of the thoracic organs and ventilation. The diaphragm is responsible for the ventilation process but also impedes the abdominal organs from getting into the thoracic cavity. However, the peritoneal and the pleural cavities can communicate in cases of congenital or acquired diaphragmatic hernias, or diaphragmatic tear, respectively. The abdominal organs get into the pleural cavity during the inspiratory phase of ventilation, due to a reduced pressure within the pleural space [1].

Traumatic (acquired) diaphragmatic hernia consists of displacement of abdominal viscera such as liver, stomach and small intestines, into the pleural cavity, generally due to a tear of the muscular portion of the costal part of the diaphragm and produces no hernial sac.

In the case of congenital diaphragmatic hernia, the most common site is at the esophageal hiatus. The abdominal viscera pass through the diaphragm in a hernial sac, which most times is the distended serosal cavity of the mediastinum [2].

Pericardial defects are a rare disorder that can be acquired or congenital. Acquired absence of the pericardium is found after pericardiectomy to treat constrictive or recurrent pericarditis [3]. In human medicine, the complete congenital absence of the pericardium is exceedingly rare with a reported incidence of <1 in 10 000 [4,5]. In dogs, congenital pericardial sac agenesis is uncommon and represents approximately 1% of the cardiac anomalies [6].

Peritoneo-pericardial diaphragmatic hernia (PPDH) is the most common congenital pericardial anomaly in dogs and cats. It consists of communication between the peritoneal and the pericardial cavities, the abdominal viscera sliding into the pericardial sac [1,2, 7-11]. This is thought to be due to incomplete genesis of septum transversum, as there is no continuity between the pericardial sac and the diaphragm after birth in dogs [12-15]. It can also be caused by injuries in the prenatal period, genetical defects, or physical malformations [7,8].

Dogs affected by PPDH can be asymptomatic for months to even years until they start showing gastro-intestinal, respiratory or cardiac signs [7,12,16]. Some dogs with PPDH can also have a malformation of the diaphragm, abdominal wall, sternum, heart, or ductal plate.

Congenital PPDH can be found in multiple individuals from the same litter [7, 11, 17-22].

To the author's knowledge, there is no report of a dog with PPDH, partial pericardial agenesis, chest wall and ductal plate malformation. All these together lead to a peritoneo-percardial diaphragmatic hernia (PPDH).

Case Summary
A one-and-a-half-year-old female neutered Labradoodle weighing 16.0 kg presented to the Emergency and Critical Care (ECC) department of a veterinary referral hospital with a 2-day history of acute abdominal distension and vomiting.

On physical examination, the dog was bright and ambulatory. Tachycardia (150 beats per minute) was noted with regular rhythm and normal heart sounds. Peripheral pulses were considered fair and synchronous. The mucous membranes were pink and moist, with a capillary refill time of 1 second. The respiratory rate was 28 breaths per minute with intermittent panting, but no breathing effort. The upper respiratory tract noises were normal, whereas chest auscultation revealed borborygmi. The abdomen was distended and tense on palpation cranially, and the dog regurgitated immediately after this. Superficial lymph nodes were normal. The rectal temperature was 38.5°C.
The PCV was 60% and TS 85 g/L. Venous blood gas and electrolyte analysis revealed a marked metabolic alkalosis (pH 7.594 [RI 7.35-7.45], pCO2 38.2 mmHg [RI 26-36], [BE 13.7 mmol/L [RI -8--2]) with moderate hypochloremia (98 mmol/L [116-126]) and moderate hypokalemia (2.6 mmol/L [RI 3.5-4.8]). Serum biochemistry documented a mild increase in ALT (142 U/L, RI 10 – 125), otherwise it was unremarkable. Hematology showed mild lymphocytosis, mild monocytosis, otherwise was unremarkable. Pancreatic Lipase (DGGR) was checked at an external laboratory and was within normal ranges. Radiographs of the thorax and abdomen revealed:

- Enlarged cardiac silhouette, elevated trachea, and caudal vena cava
- Dorsal lung displacement/retraction, from left to right
- Gas-filled intestinal loops within the thoracic cavity, overlying cardiac silhouette
- Effaced ventral and left outline of the diaphragm, caudal border of cardiac silhouette
- Cranial displacement of abdominal viscera
- Increased soft tissue opacity in the left hemithorax
- Sternal malformations - absent sternal segments
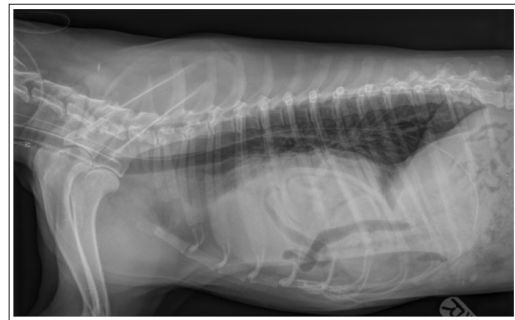- Transitional cervical vertebral anomaly - cervical ribs.

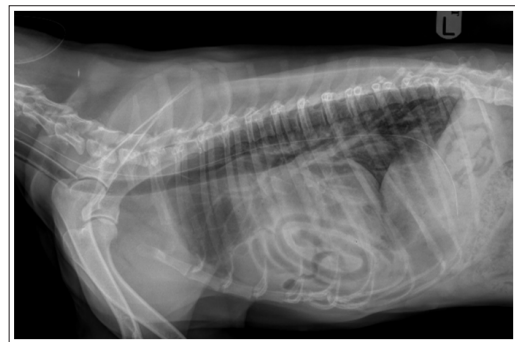**Figure 1:** Right Lateral Thoracic Radiograph Preoperatively.

Figure 2: Left Lateral Thoracic Radiograph Preoperatively.

Figure 3: Dorsoventral Thoracic Radiograph Preoperatively.

The radiographic changes were considered compatible with peritoneo-pericardial diaphragmatic hernia and possible ventral diaphragmatic hernia or rupture (the latter less likely). The sternum malformations were considered congenital as they commonly accompany peritoneo-pericardial diaphragmatic hernia. The transitional cervical vertebral anomaly consisting of cervical ribs was an incidental congenital malformation.

A 10 Fr nasogastric tube was placed consciously, secured with a bullring suture in the middle nasal plane and with a simple interrupted suture to the left cheek. Afterwards, 350 ml of gastric residual were removed, with small blood clots at the end. The tube placement was confirmed with a right lateral chest radiograph.

Intravenous (IV) fluid therapy was instituted with a constant rate infusion (CRI) of isotonic crystalloidsa supplemented with potassium chlorideb 60 mmol/L at 4 ml/kg/h. The initial supportive treatment consisted of maropitantc 1 mg/kg IV q24, paracetamold 10 mg/kg IV q8, and ondansetrone 0.5 mg/kg IV q12.

Venous gases and electrolytes were rechecked 14 hours later, and the results were unremarkable (pH 7.346, pCO2 40.4 mmHg, BE –3.4, potassium 4.1 mmol/L, chloride 117 mmol/L). Therefore, the potassium supplementation was stopped, and the fluid rate was reduced to 2 ml/kg/h.

As the surgery was anticipated to be challenging and invasive due to its complexity, the dog was blood-typed, being a DEA 1 positive.

During the first attempt at general anaesthesia, the dog had an episode of cardiopulmonary arrest, but was resuscitated shortly thereafter.

An intravenous catheter was placed in the right cephalic vein, the dog was premedicated with methadonef 0.2 mg/kg IV, then anesthetized with midazolamg 0.2 mg/kg IV and alfaxaloneh 1 mg/kg IV to good effect the following day with the view to reposition the organs into the abdominal cavity. However, it gradually became bradycardic to a heart rate of 45 beats per minute, then received a 0.04 mg/kg dose of atropinei. The dog arrested, cardiopulmonary resuscitation (CPR) was instituted with chest compressions and endotracheal intubation, naloxonej and flumazenilk were administered intravenously at 0.04 mg/kg and 0.01 mg/kg, respectively, and return of spontaneous circulation (ROSC) was obtained within the first 2-minute cycle of CPR. The surgery was postponed until later on the same day, allowing the reversal agents to wear off from the body.

Several hours later, the dog was premedicated with fentanyll 3 mcg/kg IV and ketaminem 250 mcg/kg IV, then anesthetized with midazolam 0.2 mg/kg IV and alfaxalone 1 mg/kg to good effect. Fentanyl was continued as a CRI at a dose of 3 mcg/kg/h as a plain solution of 50 mcg/ml, run at 0.96 ml/h. Ketamine was also continued as a CRI at 0.6 mg/kg/h, diluted to a 10 mg/ml concentration, run at 1 ml/h. The general anesthesia was uneventful the second time.

The dog was placed in dorsal recumbency, the whole ventrum was clipped and prepared aseptically, and midline celiotomy was performed from the xiphisternum to the pubis. A thin patch of fibrous tissue was noted between the linea alba and the last sternebra, which revealed the heart with no visible pericardium when incised. An incomplete genesis of the diaphragm was also noted ventrally, allowing herniation of the duodenum, right limb of the pancreas, jejunum, left and central liver lobes, gallbladder, spleen, and omentum into the mediastinum. Cranially, the pericardium diverged laterally with attachments to the loose connective tissue of the mediastinum. The incision was extended cranially into the thoracic cavity via a midline sternotomy with an oscillating saw. The phrenic nerves were identified, the pericardial tissue was released from the mediastinum, and the edges of the viable diaphragmatic muscle were incised to be used for closure. The diaphragm was sutured in a simple continuous pattern with 2 metric PDS suture material. A chest drain was placed in the 9th intercostal space of the left hemithorax to manage post-operative fluid and air accumulation and for analgesic administration. The sternotomy incision was closed with a 5-metric stainless-steel wire in an alternating cruciate pattern. Pectoral muscles were sutured with 2 metric PDS. A liver biopsy was taken for histopathology as ductal plate malformation has reportedly been associated with PPDH. The dog was spayed a month prior, thus the skin around the previous wound was excised en bloc, and abdominal body wall was closed routinely in three layers: linea alba was sutured with 3.5 metric Ethilon in a simple continuous pattern, and the subcutaneous (simple continuous) and skin (intradermal) tissues were sutured with a 2 metric Monocryl. The pleural drain was secured with a 2 metric Ethilon. Postoperatively, the air was removed from the pleural space via the drain, chest radiographs were taken and there was no persistent pneumothorax.
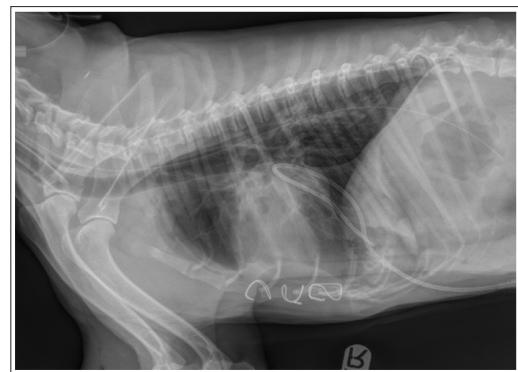

Figure 4: Right Lateral Thoracic Radiograph Postoperatively.

Bupivacainen 0.5% was administered via the chest drain at 1 mg/kg q6, diluted in 6.8 ml of 0.9% saline. This was discontinued 42 hours after the chest drain placement. Initial fluid production (modified transudate) from the chest drain was 1.1 ml/kg/hr. This reduced to 0.4 ml/kg/hr over 48 hours, at which time the chest drain was removed.

Three hours postoperatively, the fentanyl CRI was discontinued, methadone was instituted at 0.3 mg/kg IV q4 for 2 administrations, which was then reduced to 0.1 mg/kg IV q4 for 3 administrations, then changed to buprenorphine for one single administration.

By the following day, a mild bradycardia was noted at 56-60 beats per minute; otherwise, the repeat physical examinations were unremarkable.

Once the opioids were discontinued, analgesia consisted only of paracetamol 15 mg/kg IV q12, switched to oral administration[o] at 72 hours postoperatively as the dog started presenting a good appetite.

The dog was tube-fed for 48 hours after the surgery, then the nasogastric tube was removed due to consistent appetite.

Trazodone[p] was also administered as an anxiolytic at 3.12 mg/kg PO q8. Venous blood gas and electrolytes run at 13 hours postoperatively were within normal limits. The dog was discharged after 4 days of hospitalization (3 days after the surgery) with a 5-day course of paracetamol 15 mg/kg PO BID.

The histopathology results indicated the von Meyenburg complex, a subtype of ductal plate malformation, prompting a recommendation for regular liver enzyme assessment (yearly or more frequently if indicated by abnormal behaviour or reduced appetite).

Three months after the surgery, the dog was reported back to its normal self with no concerns or complications following the surgical procedure.

Discussion

This study describes a case of peritoneo-pericardial diaphragmatic hernia, which was diagnosed with thoracic radiography. It is well-known that radiography is an effective tool in diagnosing diaphragm discontinuities. However, plain radiographs can sometimes be inconclusive, as normal variations of the dog's diaphragm can be challenging [12,23]. Contrast radiography, including cholecystography, intravenous hepatography, angiography, pneumoperi cardiography, and gastrointestinal barium study have been reported in the diagnostic workup for PPDH and can be considered in order to obtain an accurate diagnosis [12,17,24,25]. Pneumoperitoneography26 and positive contrast pleurography27 have also been reported in the literature.

Peritoneo-pericardial diaphragmatic hernia can be congenital or acquired and is reported in both dogs and cats. In another study, the cats were overrepresented compared to dogs, and Weimaraners and domestic short-hair cats had a higher prevalence [28]. In another study, Weimaraner was the over-represented breed in dogs with PPDH [11].

Diaphragmatic hernia may be an incidental finding, but more often the animal presents with some or all of the following signs: dyspnea, tachypnea, cyanosis, increased respiratory effort, vomiting, and fluid distension of the abdomen [12]. In the current study, the dog presented with vomiting and acute abdominal distension.

PPDH in dogs has previously been associated with pulmonic stenosis, ventricular septal defect, umbilical hernia, sternal or costochondral deformities, dysplasia of the right atrioventricular valve, tetralogy of Fallot, as well as pulmonary arterial medial hypertrophy, necrotizing pulmonary arteritis and right ventricular hypertrophy [15,29-31].

To the author's knowledge, this is the first case report of PPDH in a dog with abnormal genesis of the thoracic wall, pericardium, and ductal plate malformation.

As other studies have reported, PPDH can be corrected surgically [7,8,19]. In this case, the dog's surgery was uneventful and consisted of ventral midline celiotomy and sternotomy. There were complications related to the general anesthetic drugs preoperatively. Following the reversal of the pre-medication and a short cardio-pulmonary resuscitation procedure, the dog recovered and was managed as per RECOVER post-CPA guidelines [23-32]. Later on the same day, the anaesthesia and surgery proceeded without further complication.

A ventral thoracostomy was performed on this dog to facilitate the replacement of abdominal organs into the peritoneal cavity. Following this, a thoracostomy tube was placed to monitor the amount of fluid in the pleural cavity and to administer analgesic drugs. Studies have shown complications associated with the thoracostomy tube placement which can prolong the hospitalization period and increase morbidity in animals and costs to clients. In this case, the chest drain was removed 48 hours after its placement due to reduced pleural fluid production, and no complications were encountered [33].

In most cases of PPDH that undergo surgery, the diaphragmatic defects are closed by apposition of the muscle edges. In cases where the large diaphragmatic defects can't be closed or their apposition would create increased tension, omental, muscle, pericardial flaps, autologous fascia grafts or polypropylene meshes can be used [22]. In this case, a simple continuous suture of the medial ventral aspect of the diaphragm was completed successfully.

One study showed that animals with PPDH with no clinical signs at the time of diagnosis can develop clinical signs at a later time [8]. In the same study, the short-term (2 weeks) mortality rate after the surgical repair of the PPDH was reported at 8.8%; the long-term (329.5 ± 596.1 days; mean $\pm$ SD; range 2 to 2,657 days) mortality rate in dogs and cats who had undergone surgery for PPDH was 9.6%.

Conclusion

Peritoneo-pericardial diaphragmatic hernia (PPDH) is a rare congenital defect that can cause a vast range of clinical signs, most commonly including respiratory and/or gastrointestinal changes, depending on the affected tissues. The blood test results can be unremarkable or show mild abnormalities, depending on the herniated organ(s) and the extent of the herniation. PPDH can also be found incidentally while investigating a patient for various reasons. It can be diagnosed with thoracic radiographs or CT scans and fixed surgically or laparoscopically. Patients affected by PPDH can also have other congenital abnormalities.

Disclosure of interest
The author declares no conflicts of interest.

Footnotes
- Vetivex 1 0.9% NaCl, Dechra, UK
- Potassium Chloride Concentrate 20%, Hameln, UK
- Vetemex 10 mg/ml, Virbac, UK
- Paracetamol, Braun, Germany
- Ondansetron 2 mg/ml, Hameln, UK
- Comfortan 10 mg/ml, Dechra, UK
- Midazolam 5 mg/ml, Hameln, UK
- Alfaxan 10 mg/ml, Zoetis, UK

- Atropine Sulphate 600 mcg/ml, Martindale Pharmaceuticals, UK
- Naloxone 400 mcg/ml, Hameln, UK
- Flumazenil 0.1 mg/ml, Hameln, UK
- Fentadon 50 mcg/ml, Dechra, UK
- Anesketin 100 mg/ml, Dechra, UK
- Marcain Polyamp Steripack 0.5%, Astra Zeneca, UK
- Pardale V 400mg/9mg, Dechra, UK
- Trazodone Hydrochloride 50 mg, Flamingo Pharma, UK

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