Case report: diagnosis complications in an English Bulldog

Tom Mullany MVB Cert VC MRCVS, Nutgrove Veterinary Hospital, Rathfarnham and Louise O’Leary MVB, Veterinary Teaching Hospital, University College Dublin, present a case report on the peritoneopericardial diaphragmatic hernia complicating the diagnosis of additional congenital defects in a young English Bulldog

A seven-month-old, male English Bulldog presented with a five-month history of exertional cyanosis. Plain thoracic radiographs revealed the presence of gas-filled loops of small intestine in the pericardial sac which obscured the heart and prevented satisfactory echocardiography. An abdominal ultrasound confirmed small intestine imaged to the diaphragmatic line. A diagnosis of peritoneopericardial diaphragmatic hernia (PPDH) was made. The PPDH was successfully repaired using a midline coeliotomy approach. Twenty-four hours post-surgery, the puppy again developed exertional cyanosis, this time with tachypnoea. Iatrogenic pneumopericardium, diagnosed on thoracic radiographs with the heart clearly delineated within the air filled pericardial sac, was successfully treated by pericardiocentesis. Subsequent thoracic radiographs taken a few days later confirmed almost complete resolution of the pneumopericardium. Four-stage echocardiography was performed which revealed: right ventricular hypertrophy, pulmonic stenosis, a large muscular ventricular septal defect (VSD), and an over-riding aorta. These findings are consistent with a diagnosis of Tetralogy of Fallot (TOF). Surgical treatment was declined and the dog is being managed conservatively.

This article describes presentation, diagnosis and treatment of PPDH and demonstrates that coexisting congenital intracardiac defects may adversely affect the prognosis.

BACKGROUND
Peritoneopericardial diaphragmatic hernia is the most common congenital pericardial defects diagnosed in small animals accounting for 0.5 % of all reported congenital heart defects in dogs and cats. Congenital PPDH is a result of the failure of the lateral pleuropertitoneal folds and the ventromedial pars sternalis to unite or, as a result of a defect in the development of the dorsolateral septum transversum. Acquired PPDH has been reported in humans following trauma and it thought that acquired PPDH may also have been caused following postnatal injuries. It has been reported that male dogs are more likely to be affected than females. The Weimaraner and Persian cats may be predisposed to this condition. PPDH creates an abnormal communication between the peritoneal and pericardial cavities allowing herniation of abdominal viscera into the pericardial cavity. The organs most commonly herniated are the liver and gallbladder, omentum, intestines and spleen. Animals may be asymptomatic, or may have clinical signs relating to the gastrointestinal tract (vomiting, anorexia, failure to gain weight) or the respiratory system (dyspnoea, tachypnoea, exercise intolerance, coughing, cyanosis). Cats tend to present more commonly with respiratory signs, and dogs with gastrointestinal signs. Physical examination may be unremarkable with smaller defects. With larger defects, the most common findings on physical examination are muffled or displaced heart sounds, ascites, an inability to palpate abdominal organs (empty abdomen), and rarely, signs associated with cardiac tamponade and right-sided cardiac failure.

The diagnosis of PPDH is usually made by plain radiographs. Radiographic changes include dorsal displacement of the intra-thoracic trachea, silhouetting of the cardiac and diaphragmatic borders and enlargement of the cardiac silhouette. The cardiac silhouette may contain heterogeneous densities and gas patterns associated with the presence of abdominal contents within the pericardial sac.

The recommended treatment for PPDH involves returning the abdominal viscera from their position in the pericardial cavity to their normal intra-abdominal location, and closure of the defect, via laparotomy or thoracotomy. TOF is a complex congenital cardiac defect, characterised by the presence of pulmonic stenosis, a large ventricular septal defect, varying degrees of dextroposition or overriding of the aorta, and secondary right ventricular hypertrophy. It is a rare congenital condition, with an estimated prevalence of one per 4,000 dogs. TOF is a genetically transmitted condition, propagated as a single major gene, interfering with myocardial growth. The Keeshond and English Bulldog are over-represented in reported cases. Increased right ventricular pressures arise as a result of increased right-sided resistance resulting from the presence of the pulmonic stenosis. When this pressure exceeds left ventricular pressures, blood flows from the right ventricle through the ventricular septal defect into the left ventricle. This means an increase in de-oxygenated blood being delivered to the systemic circulation. This, in turn, results in systemic hypoxemia, decreased haemoglobin oxygen saturation, cyanosis and a secondary polycythaemia. However, some juvenile animals may be hypoxaemic and not present with a severe polycythaemia. TOF is the most common cardiac defect causing cyanosis in small animals. Clinical signs can vary and depend on the severity of the pulmonic stenosis and the VSD size. Signs include: a failure to grow, cyanosis, exercise intolerance lethargy and syncope. A murmur may be auscultated in some cases. Because the VSD is large, flow through the defect is laminar
with low-flow velocity, therefore the right sternal holosystolic murmur associated with VSDs may not be audible. An ejection murmur associated with pulmonic stenosis may be audible over the left base or may radiate to the right hemithorax. Dogs presenting with polycythaemia may have no audible murmur due to the increased blood viscosity which dampens turbulence. Confirmation of the diagnosis is by echocardiography. Two-dimensional imaging will reveal the structural components of the condition: right ventricular hypertrophy, large VSD, a small left ventricle and an over-riding aorta. Doppler echocardiography will demonstrate the direction and magnitude of shunting, and can be used to determine the severity of the pulmonic stenosis. Bubble studies can be used to confirm the reversal of flow through the VSD.

Treatment may be medical or surgical. Medical treatment includes periodic phlebotomy to limit the increase in PCV to between 65-68%. In such cases, the volume of blood removed should be replaced by a corresponding volume of a crystalloid to preserve cardiac output and to maintain oxygen delivery. The use of the myelosuppressive agent has been reported in cases which require frequent phlebotomies. Beta-blockers (propranolol) have been reported to successfully control hypoxaemic episodes by reducing right ventricular contractility, reducing right ventricular outflow (muscular) obstruction and reducing myocardial oxygen consumption. Definitive surgical correction involves closure of the VSD and correction of the pulmonic stenosis under cardiopulmonary bypass. This procedure is rarely performed in dogs. Palliative surgical procedures involve the creation of a shunt between the systemic and the pulmonary circulation (essentially the creation of a PDA type shunt). Techniques including the Blalock-Taussig (subclavian to pulmonary artery), the Potts (ascending aorta to pulmonary artery) and the Watson-Cooley (aorta to right pulmonary artery). Increased right ventricular resistance can be addressed by reduction of the pulmonic stenosis with surgical or balloon valvuloplasty.

CASE PRESENTATION

A seven-month-old male English Bulldog was referred for further investigation of a five-month history of exertional cyanosis. The dog was considered small for his age when compared to his siblings. On initial exam, the puppy was bright, alert and responsive. Body condition score (BCS) was low at 2/5. Cardiac sounds were muffled bilaterally, but no abnormal cardiac sounds could be detected. Femoral pulses were palpable bilaterally, of normal quality and synchronous with the heartbeat. Cranial and caudal mucous membranes were moist and pink with a brisk capillary refill time (less than two seconds). There was no evidence of jugular distension, and there was a negative hepatosplenic reflex. An abdominal component to breathing was noted. During the initial part of the clinical examination, the pup’s respiration remained normal. Slightly increased adventitious lung sounds were auscultated across all lung fields. During the examination the dog became excited, and his mucous membranes became cyanotic.

The dog was sedated with intravenous acepromazine (ACP, Ceva) and butorphanol (Torbugesic, Zoetis) and orthogonal radiographs of the thorax were taken (see Figures 1 and 2). These radiographs demonstrated a grossly enlarged cardiac silhouette, with gas-filled loops of intestine visible within the pericardial cavity. There was no evidence of displacement of other abdominal structures on the radiographs. An abdominal ultrasound was performed, and apart from imaging small intestines to the diaphragmatic line, no abnormalities involving the abdominal viscera were detected. A provisional diagnosis of congenital peritoneopericardial diaphragmatic hernia was made. Efforts were made to perform a cardiac ultrasound, but the presence of gas-filled intestinal loops precluded satisfactory imaging.

The following day, the dog was anaesthetised using standard protocols of acepromazine (ACP, Ceva) and buprenorphine (Vetgesic, Ceva) for premedication, propofol (Vetofol, Norbrook) for induction agent, and
isoflurane (Vetfluorane, Virbac) for maintenance. A standard midline colieotomy was performed. A 4cm defect was identified in the ventral aspect of the muscular portion of the diaphragm, through which approximately 60cm of small intestine was herniated into the pericardial cavity. This small intestine was replaced into the abdominal cavity. The diaphragmatic defect was closed with a double-layer of polydioxanone 3.5 metric (MonoPlus) sutures. Air was aspirated from the pericardial sac via the insertion of a small chest drain tube through the incision immediately after closure. Early post-operative recovery was unremarkable. Twenty-four hours post-operatively, the dog developed stress induced tachypnoea and cyanosis. Cardiac auscultation revealed a soft, grade II/VI right apical systolic murmur. Orthogonal thoracic radiographs were taken (see Figure 3) that revealed a well demarcated area of gas opacity surrounding the cardiac silhouette, within a defined, radiopaque line of the pericardium. This pneumopericardium had been iatrogenically produced following the repair of the hernia. The gas in the pericardial sac outlined a bulge in the outline of the cardiac silhouette in a position normally occupied by the main pulmonary artery.

Using a standard right-thoracic approach, a pericardiocentesis was performed, and the pneumopericardium aspirated. Following this procedure, during a period of post-operative hospitalisation, his oral mucous membranes became cyanotic whenever he became agitated or excited.

Some days later, a repeat thoracic radiograph (see Figure 4) confirmed a substantial reduction in the size of the pneumopericardium. It was expected that this degree of pneumopericardium would resolve spontaneously and no further pericardiocenteses were performed.

Haematology was also performed which revealed a moderate polycythemia: 62 I/L (37-55 I/L) and hyperhaemoglobinemia: 200 g/L (120-180 g/L). All other components of the tests were unremarkable. At this point, echocardiography was performed satisfactorily. The 2D study, spectral and colour flow Doppler studies confirmed the following:
- right ventricular hypertrophy (see Figure 5);
- an overriding aorta (see Figure 6);
- a large defect in the muscular portion of the ventricular septum with a right-to-left flow through the septal defect (see Figure 7);
Based on the results of this study, a diagnosis of TOF was made. It was decided to proceed with medical management. The dog was discharged on low-dose beta-blockers, (atenolol, Clonmel, [1mg/kg BID]) 10mg BID, with instructions to avoid any excitement or vigorous exercise. On discharge, the PCV was below 68g/L the figure above, which intervention is recommended and phlebotomy should be performed. Monthly follow-up PCVs were recommended to reassess the need for therapeutic phlebotomy.

To date, the patient remains comfortable at rest, but develops cyanosis with any exertion.

DISCUSSION
PPDHs and TOF are uncommon congenital cardiac conditions in the dog, making up 0.3% and 1% of congenital cardiac disease respectively, in one study of 967 dogs. PPDHs have been associated with concurrent intra-cardiac or congenital midline anomalies. The comorbidities of TOF and PPDH have been reported previously in the dog. In this reported case, echocardiographic diagnosis of the TOF was possible, as fat was the only abnormal abdominal content herniated into the pericardial sac. Angiography, has been used to diagnose other intracardiac defects, such as pulmonic stenosis, prior to surgery in dogs with concurrent PPDH.

In the case presented here, the presence of gas-filled small intestine in the pericardial sac initially prevented the use of ultrasound as a means of detecting the additional complex congenital cardiac defects present in the dog. Plain radiographs were suggestive of a PPDH, but the presence of intestinal loops obscured the cardiac silhouette and masked the radiographic changes in the cardiac silhouette (right-heart cardiomegaly, possible changes in the segment of the cardiac silhouette associated with the aorta and pulmonary artery) that may be noted in cases of TOF.

The signalment, clinical signs, and physical exam findings in this patient have been previously reported in cases of peritoneopericardial diaphragmatic hernia. However, these findings could also have been attributed to TOF. Similarities exist between the presentation and clinical findings in both.
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1. Signalment: English Bulldogs have a breed predisposition for TOF. As both PPDH and TOF are congenital conditions, young animals are more commonly presented for evaluation. The median age of presentation of PPDH in the dog is 30 months, although animals can present at any age. The median age of presentation of TOF in the dog is 3.6 months, with a range from 1.7 up to 49.4 months.

2. Clinical signs (cyanosis and failure to thrive), has been reported in animals with TOF as well as animals with a PPDH. 

3. Physical examination findings – the presence of cyanosis is associated with a number of congenital cardiac conditions such as:
   - TOF;
   - Eisenmengers physiology;
   - Pulmonic stenosis with patent foramen ovale;
   - Tricuspid dysplasia with patent foramen ovale;
   - Tricuspid atresia; and
   - Pulmonary atresia.

However, in this case, given the absence of detectable cardiac murmurs on initial examination, the presence of such a large cardiac silhouette compressing the lung fields, and an inability to perform a complete ultrasound examination, these conditions were not given priority during the initial clinical workup. Resolution of the PPDH allowed subsequent detection of cardiac murmurs and persistent cyanosis which lead to consideration of a comorbid, right-to-left shunt and initiation of further investigations.

In this case, routine haematology tests, performed after the repair of the PPDH, revealed a mild polycythemia vera (PCV 62). Polycythaemia can be related to neoplastic disease, to abnormally high production of erythropoietin hormone from neoplasia or renal disease, from lung disease or as in this case, from cyanotic heart disease due to the right-to-left shunt. The findings of polycythaemia and hyperhaemoglobinemia were suggestive of a right-to-left shunt. Polycythaemia has not been reported as associated with PPDH. Had these tests been performed prior to hernia repair, these findings would have alerted the veterinarian to the possibility of other co-existing congenital defect in addition to the PPDH.

The pneumopericardium in this case both aided and delayed the diagnosis of TOF. The air trapped within the pericardial sac highlighted the abnormal bulge on the cardiac silhouette, alerting the veterinarian to the fact that a congenital cardiac abnormality might be present. Iatrogenic pneumopericardium had been historically used in humans prior to the advent of echocardiography as a diagnostic aid for structural cardiac disease. The air enhances the detail of the cardiac outline. However, the presence of the air also interfered with echocardiography in this case. Pneumopericardium is a rare condition seen in small animals. Reported causes have been spontaneous, traumatic, and iatrogenic. Pneumopericardium has been reported as a complication of PPDH repair. The condition is usually asymptomatic and resolves spontaneously. The partial alleviation of the dogs respiratory distress after pericardiocentesis suggests that the pneumopericardium was affecting cardiac and respiratory function to some extent, though these clinical signs were also contributed to by the, as yet undiagnosed, TOF.

After an initial good prognosis following the diagnosis of the PPDH, the detection of the TOF greatly altered the prognosis in this patient. The prognosis for an uncomplicated peritoneopericardial diaphragmatic hernia is excellent. However, the prognosis of animals with TOF is less favourable and depends on the degree of pulmonic stenosis and the size of the septal defect. Mean survival time after diagnosis of TOF is estimated to be 14.5 months.

CONCLUSION

In cases where a diagnosis of peritoneopericardial diaphragmatic hernia is initially made on plain radiographs, but where physical exam findings include cyanosis, a high index of suspicion should be maintained by the veterinarian for concurrent congenital cyanotic cardiac disease. It may be prudent to warn owners that surgical correction of the PPDH may not resolve the clinical signs if other congenital defects co-exist. It may also be advisable to inform owners of the potential difficulties in diagnosing these conditions in the presence of a PPDH and that definitive diagnosis may not be possible until after the repair of the PPDH when techniques such as cardiac ultrasound can be utilised. Owners should be warned that diagnosis in these complex congenital cases is difficult and treatment may not be successful.

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