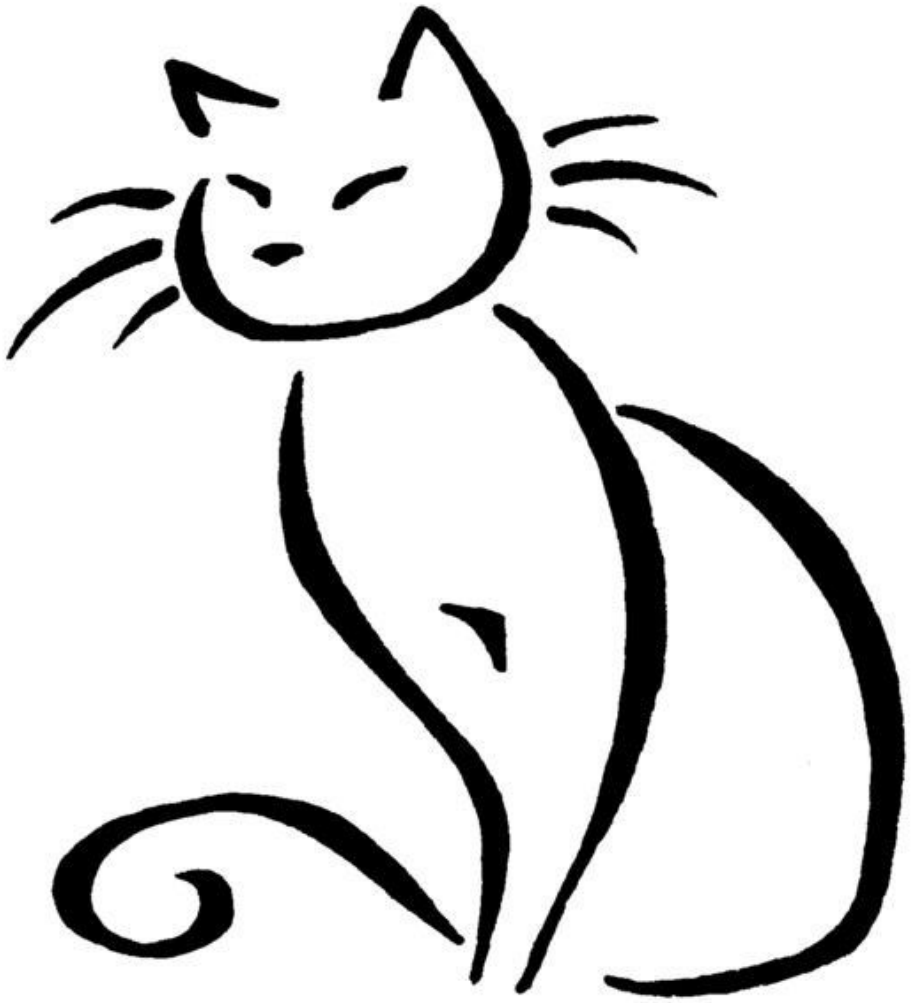
Peritoneopericardial Diaphragmatic Hernia In a Feline



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Introduction

Peritoneopericardial diaphragmatic hernia is one of the most common congenital defect of the diaphragm and pericardium in which there is a failure in the formation or fusion of the septum transversum during embryologic development. This abnormality allows for the passage of abdominal contents into the pericardial sac through the hernia. Clinical signs seen with this condition are attributed to the organs that are herniated into the pericardial sac and may include exercise intolerance, dyspnea, tachypnea, vomiting, coughing, and anorexia, with tachypnea and dyspnea being the most common. However in some patients, clinical signs may not be observed or may be severe enough to cause sudden death due to compromise of the cardiac, gastrointestinal, or respiratory tract. The main diagnostic modality used is diagnostic imaging such as radiographs and ultrasound, in which one may identify gas-filled intestines in the pericardium, loss of diaphragm border distinction, or absence of liver lobes in the abdomen. Treatment options include surgical repair via herniorrhaphy or a nonsurgical option of monitoring for worsening clinical signs.

History and Presentation

A peritoneopericardial diaphragmatic hernia is an abnormal opening between the pericardial sac and the peritoneal cavity. This abnormality is considered one of the most common congenital cardiac, pericardial, and diaphragmatic defects in cats 2 years of age or older. [5,7,8] The prevalence of this condition in cats ranges from 0.06% to 1.45%.[3] The most common breeds implicated with this abnormality were domestic longhair cats, Persians, Himalayans, and Maine Coons.[1,3,8] No statistically significant sex predisposition or median age at time of diagnosis has been identified.[1] The most common clinical signs reported are weight

loss, febrile episodes, abdominal pain, ascites, shock, collapse, exercise intolerance, dyspnea, wheezing, tachypnea, vomiting, polyphagia, diarrhea, lethargy, coughing, and anorexia.[1,4,8] In patients with hepatic incarceration, hepatic encephalopathy may manifest as head pressing, blindness, and convulsions.[4] The animals whose PPDH are discovered incidentally are usually evaluated for other disease processes such as vehicular trauma, hypertrophic cardiomyopathy, seizures, renal insufficiency, orthopedic disease, upper respiratory tract infection, neurologic disease, pericardial cyst/effusion, bronchitis, unclassified cardiomyopathy, polycystic kidney disease, pancytopenia, urinary tract obstruction, hyperthyroidism, chylothorax, and mastocytosis.[1,8] During these evaluations, radiographs or other imaging modalities are performed which reveal the PPDH.[1] This defect is usually present at birth, although an animal may be asymptomatic and therefore undiagnosed for years.[4,8] It has also been reported that mature adult and geriatric cats may have an increased incidence of PPDH as an incidental finding as compared to young cats.[1]

Pathophysiology/Anatomical Considerations

The diaphragm is formed in utero from diverse tissues including the paraxial mesoderm, somatic and splanchnic mesodermal layers, and the dorsal mesentery of the caudal part of the esophagus.[7] The septum transversum is the ventral component of the diaphragm which develops early in cardiogenesis and becomes the central tendon.[7] The caudal mediastinum is the second component of the diaphragm and is the tissue through which the esophagus and caudal vena cava pass.[7] The final component is the pleuroperitoneal membranes/folds which expand and fuse with the septum transversum ventrally and the caudal mediastinum medially.[7] This closure seals the pleural cavity from the peritoneal cavity.[7]

The exact cause of the diaphragmatic defect causing a peritoneopericardial diaphragmatic hernia has not been identified, but various mechanisms have been proposed which include abnormal development or prenatal trauma to the septum transversum or the site of fusion at the pleuroperitoneal folds, failure of fusion of the lateral aspects of the pleuroperitoneal folds and ventromedial aspect of the pars sternalis, defective closure of the midline of the abdominal wall near the diaphragm, or aberrant development of the dorsolateral aspects of the septum transversum.[1,4,5,7,8] These abnormalities may result in a gap or an unusually thin ventral diaphragmatic tissue that ruptures and allows communication between the two cavities.[4,7] This defect allows passage of the liver, falciform ligament, spleen, omentum, stomach, or intestines into the pericardial sac which may result in symptoms such as cardiac tamponade, compromised respiratory function due to indirect pulmonary compression, obstruction of the gastrointestinal tract, gallbladder torsion, or entrapment of the liver or spleen.[1,4,7,8]

While it is not known whether PPDH is genetically inherited or is the result of in utero conditions in which the diaphragm does not develop properly, congenital agenesis of the diaphragm has an autosomal-recessive mode of inheritance in cats.[1] Also, patients with PPDH and a portosystemic shunt have been reported which suggests a possible genetic relationship between the two congenital abnormalities.[1] A teratogenic agent was hypothesized to cause congenital PPDH in a litter of Collie puppies.[2,4]

Diagnostic Approach

PPDH is usually diagnosed in patient evaluation for cardiorespiratory difficulty but may be also discovered incidentally when radiographic evaluation is performed for other reasons such as foreign body ingestion.[1,4,5,8] PPDH is diagnosed as an incidental finding in 40% of affected

cats.[4] Common findings on physical examination include a heart murmur due to concomitant cardiac defects, pyrexia, tachypnea, dyspnea, thin body condition, abnormally positioned or muffled heart sounds, sternal click, palpable pectus excavatum, thoracic borborygmi, expiratory wheeze, micropthalmia, lid atresia, increased respiratory effort, and decreased/abnormal lung sounds, with muffled heart sounds being the most common.[1,4,7,8] The presence of a PPDH may also be accompanied by other genetic physical abnormalities such as umbilical or ventral abdominal hernias, stenotic nares, abnormal swirling of hair in the sternal region, cryptorchidism, cleft palate, and persistent pupillary membranes.[1,4,7,8] Abnormalities may also be identified during diagnostic testing and imaging and may include intrahepatic/extrahepatic portosystemic shunts, pulmonary vascular disease, cardiovascular defects, or sternal/vertebral abnormalities.[1,4,8] Unlike dogs in which ALT and calcium elevations may be seen, CBC and chemistry in cats is often within normal limits.[1,8]

Thoracic radiographs commonly display cardiomegaly, dorsal displacement of the trachea, gas-filled/feces-filled abdominal organs present in the pericardial sac, presence of a mesothelial remnant, microhepatia, and inability to discern the diaphragmatic borders.[1,3,4] The cardiac silhouette may have a double density appearance of either soft tissue and fat or soft tissue and gas opacities which may cause a loss of distinct cardiac silhouette margins.[1,4] Changes in the positions of abdominal viscera or gastric axis are unreliable signs of a PPDH.[4] Pleural effusion is not commonly present, but may be seen due to compression of the right heart by the herniated organs.[4,6] Pericardial effusion may be present which may lead to cardiac tamponade and signs of right-heart failure due to interference with venous return.[4] Other diagnostic modalities that may be performed include upper GI contrast studies, pneumoperitoneography, positive-contrast peritoneography, and nonselective angiography, but each of these have their

limitations and are rarely necessary.[4,7] Ultrasonography is the most useful diagnostic tool for confirming PPDH due to its ease of use, reliability, noninvasiveness, and ability to detect structural differences of the contents of the pericardial sac.[4] These images may be obtained via the right fifth intercostal space or transabdominally.[4] Also, with this modality, pericardial effusion may be differentiated from extracardiac intrapericardial masses, such as the liver, gallbladder, fat, and intestine.[4]

Complete cardiac evaluation via echocardiography may be difficult in some cases due to abnormal heart position or herniated abdominal organs/tissues compressing the heart muscle; however, abnormalities which may be identified in cats include hypertrophic cardiomyopathy, double-chambered right ventricle with pulmonic stenosis, tricuspid/mitral regurgitation, mitral stenosis with left atrial thrombus, thickening of the left ventricular wall, and dynamic right ventricular outflow obstruction.[1] Dorsal displacement of the heart as well as presence of peristaltic intestinal loops in the pericardial sac may be seen on echocardiography.[3] Cardiac abnormalities identified include ventricular/atrial septal defects, mitral regurgitation, tricuspid dysplasia, persistent left cranial vena cava, pseudotruncus arteriosus, and subaortic/pulmonic stenosis.[1,8] Electrocardiography may reveal low-amplitude complexes and electrical alternans due to pericardial effusion or axis deviation from cardiac displacement.[4]

Treatment and Management

Treatment for a peritoneopericardial diaphragmatic hernia is based on the severity of clinical signs.[1,4] There are two options for treatment of a PPDH: surgical repair and medical management.[1,4,7] Animals with clinical signs attributed to PPDH typically receive surgical repair while those whose PPDH was an incidental finding, typically receive conservative treatment via

medical management and monitoring unless treatment for another underlying condition requires a celiotomy at which time the hernia may be repaired.[1,4,5,8] Owners may also opt for conservative treatment if the patient is of advanced age, financial difficulty is present, or concerns are present about the morbidity and mortality rates associated with anesthesia and surgery.[8] Young cats are more likely to have a primary diagnosis of this condition and receive surgical correction of the PPDH.[1] This may be due to the size of the hernia or the volume of abdominal contents present in the pericardial sac.[1] The larger defects allow organs to pass freely into the pericardium and form adhesions to the pericardium as well as organ entrapment in the pericardium leading to respiratory, cardiac, and gastrointestinal tract abnormalities.[1,7,8] Tissues most frequently removed from the pericardial sac include omentum, stomach, colon, falciform ligament, spleen, liver lobes, gallbladder, and sections of small intestine.[1,8] Adhesions may be present between the herniated intestine/liver and the pericardium.[1]

In cases where there is cardiac tamponade secondary to hepatic lobe effusion, emergency pericardiocentesis may be necessary before herniorrhaphy.[4] While a midline celiotomy is the most common method to repairing this defect, a partial caudal sternotomy may also be performed.[1,4] The midline approach allows for inspection of all abdominal viscera and causes less pain postoperatively when compared to a thoracotomy.[4] If at this point any organs appear to be necrotic or infiltrated with adipose tissue or there are any lesions of herniated omentum such as cysts, hematomas, or myelolipomas, excision of the tissue may be necessary.[1,4,5] The major disadvantage to this approach is that the diaphragm is viewed from the concave surface which may lead to difficulty while suturing the defect.[4] Another disadvantage is if adhesions are present in the pericardial sac, the diaphragm defect may need to be enlarged or the celiotomy may need to be extended to a median sternotomy in order to break them down.[4] A lateral

thoracotomy is contraindicated in cases of PPDH.[4] During this procedure, precaution should be taken to not disrupt the pericardium as it is conjoined to the borders of the diaphragmatic defect and is continuous with the transverse fascia and peritoneum.[4] Disrupting this membrane may lead to pneumothorax and pleural effusion; if opening the pericardial sac is necessary, controlled ventilation will be essential to the survival of the patient.[4]

In cases of large defects which would cause tension on the hernia closure, the pericardium may be transected cranially to the diaphragmatic attachments, and this extra tissue may be used as a flap or free graft for defect closure.[4] However, the phrenic nerves should be isolated before the transection of the pericardium.[4] Closure of the remaining amount of pericardium is unnecessary.[4] Sternal or abdominal wall defects that are also present may be repaired during this procedure as well to prevent future surgeries.[4]

Peritoneopericardial diaphragmatic hernias may be repaired using slowly absorbable or nonabsorbable monofilament suture material in an interrupted or continuous suture pattern.[1,4] Closure of the pericardial sac in which air is entrapped may impede pulmonary expansion and lead to cardiac tamponade.[4] This air may be removed via pericardiocentesis which may be performed during closure with a red rubber catheter or placement of a thoracostomy tube through the diaphragm after hernia closure.[1,4] Most patients do not require a thoracostomy tube and recovery quickly if no concurrent issues are present.[4] Complications that may occur after/during this procedure include severe hemorrhage, esophageal strictures, hyperthermia, tachypnea, dyspnea, wheezing, hypoventilation, persistent pericardial/pleural effusion, chylothorax, persistent acidemia/respiratory acidosis, loss of palpable pulses, partial blindness, hypoxia, respiratory arrest, pericardial steatitis, regurgitation/vomiting, incisional dehiscence/inflammation, lethargy, hyporrhexia, hypotension, malignant cardiac arrhythmias

such as multifocal premature ventricular contractions, hernia recurrence, and tracheitis.[1,4,5,8] Other less common complications seen with PPDH include development of hepatic cysts, gallbladder torsion and rupture, and myelolipomatosis, or portal hypertension secondary to liver lobe incarceration.[4,5] A pericardial cyst following surgical repair has also been reported.[3,4] This cyst was found via echocardiography as a large anechoic structure associated with the pericardium.[3] On thoracic radiographs, the soft tissue opacity structure caused elevation of the cardiac silhouette from the sternum as well as dorsal displacement of the trachea.[3] Another report has been made of a hepatic cyst that underwent neoplastic transformation to an hepatic fibrosarcoma which is common in sites of inflammation.[5] It has been speculated that the incarceration of hepatic tissue in the hernia may lead to vascular and lymphatic congestion and subsequent hypoxia.[5] This predisposes the tissue to formation of cysts of myelolipomas.[5] Another complication seen with this surgical procedure, although rare, is constrictive pericarditis which is characterized by the encasement of the heart by a dense, rigid pericardial tissue which leads to restriction of diastolic filling and signs of right-sided congestive heart failure.[6]

The nonsurgical method of treatment for a PPDH may be chosen in patients with fewer to no clinical signs or patients with major underlying diseases that make surgery a higher risk than the current clinical signs.[1] These conditions may include severe renal disease or pancytopenia as seen in the retrospective study by Burns et al.[1] In these cases, it is recommended for the owners to continue monitoring the patient for signs of respiratory distress or gastrointestinal tract disorders. [1]

Prognosis

Prognosis of the surgical patients is very good with only an 8.8-14% short term mortality post-operatively with a success rate of 86% in cats.[1,4,8] However, of the cats who received surgical repair, 78% encountered complications.[4] Of the 24 nonsurgical patients in the Burns et al study, 16 of those died due to reasons unrelated to PPDH and sudden death or natural causes of death.[1]

Conclusion

When faced with a patient with a PPDH, the evaluation of the presence and severity of clinical signs is the major deciding factor on the pursuit of treatment. Those with no or minimal clinical signs seem to do very well with medical management while those patients with severe clinical signs require surgery but live normal lives post-operatively.

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