Don’t Leave Me Broken Hearted
Persistent Right Aortic Arch in a Canine

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Introduction:

Vascular ring anomalies have been reported in most domestic veterinary species, as well as exotic species.[10] A persistent right aortic arch (PRAA) is the most common of these reported in canine and feline patients, representing ninety-five percent of all vascular ring anomalies[3]. In thirty-three percent of the PRAA cases reported, a second vascular anomaly has also been noted, an aberrant left subclavian artery.[3] Abnormal formation of the great vessels during embryonic growth results in the right fourth aortic arch and the left ligamentum arteriosum encircling the esophagus and trachea, leading to compression of the esophagus, and subsequent megaesophagus proximally.[5] The goal of surgical treatment is to alleviate the esophageal constriction(s), allowing for the resolution of clinical signs.[8, 10]

History and Presentation:

Patients with a PRAA commonly present to their referring veterinarian for persistent postprandial regurgitation shortly after being weaned (associated with being started on solid foods), stunted growth or unthriftiness despite having a ravenous appetite, and/or aspiration pneumonia.[4, 7, 9, 10]

Young, large, pure-breed dogs appear to be more commonly effected, with German Shepherds, Irish Setters, and Greyhounds being overrepresented.[2, 4, 6, 7, 8, 9] A previous study suggests that these three breeds also appear to be genetically predisposed to developing a PRAA, however the mode of inheritance is complex. One study suggests there may be a genetic predisposition in a particular line of German Pinchers with an unusual combination of vascular ring anomalies, but again, the mode of inheritance is complex, and not fully understood at this

time.[4] Vascular ring anomalies in cats are less common than in dogs, however, PRAA is the most frequently diagnosed. Persians and Siamese cats are overrepresented.

Pathophysiology:

All vascular ring anomalies result from abnormal embryonic development during the second and third week of gestation. During normal development, there are six pairs of aortic vessels/arches that correspond with each other. When these vessels come together, they form a ring around the "embryonic foregut", which will later develop into the esophagus and trachea. At a certain point in gestation, specific aortic arches will regress, allowing for the release of the esophagus and trachea from the vascular ring that has formed.[10]

In a normal patient, the left fourth aortic arch is retained, and is later joined by other vessels to form the aorta and descending aorta. The right fourth aortic arch joins other vessels to form the right subclavian artery. In the case of a PRAA, the right fourth aortic arch is retained and goes on to form the aorta, instead of the left fourth, which now becomes the left subclavian artery.[4, 10] These abnormalities in combination with a normal left ligamentum arteriosum create a ring, compressing the esophagus and trachea. In a normal patient, the esophagus and trachea lie just to the right of the aorta, thus a normal left ligamentum arteriosum does not entrap the structures of interest.[4, 9,10]

While the trachea is somewhat compressed with PRAA, it is not enough to result in respiratory signs. There are other reported cases, as in a double aortic arch anomaly, where there was significant enough narrowing of the trachea to cause dyspnea and upper airway noises.[7, 10]

Diagnostic Approach:

A presumptive diagnosis of a PRAA can be made based on signalment, history, clinical signs, and radiographic studies of the thorax, with or without contrast. A definitive diagnosis is best made with surgical exploration via a left thoracotomy at the level of the fourth intercostal space.[3, 4, 6, 7, 9, 10]

Coughing and dyspnea may be noted on physical exam of animals that have concurrent aspiration pneumonia as a result of the esophageal constriction. Laboratory findings are generally unremarkable, however, signs of malnourishment or aspiration pneumonia may be appreciated.[10] Cardiac auscultation is usually within normal limits in these cases, due to the patients still exhibiting normal cardiovascular function.[3, 9, 10]

Contrast radiographic evaluation is helpful in determining whether a patient has primary megaesophagus versus a PRAA, but may not necessarily be indicated for every patient.[3] Patients with primary megaesophagus will often have a generalized dilation of their esophagus throughout the thorax, on both the lateral and dorsovental/ventrodorsal projections, while a patient with a PRAA will have a dilated esophagus cranial to the base of the heart (level of the fourth intercostal space) only.[3, 10] With a PRAA, the lateral view may show mild to marked ventral deviation of the trachea at the cranial aspect of the heart, with mild to moderate tracheal narrowing. The ventrodorsal view may show moderate to marked, leftward deviation from midline of the cranial trachea. In a normal patient, the trachea is straight on midline, or mildly deviated to the right. One study showed that one-hundred percent, twenty-seven out of twenty-

seven dogs, had moderate to marked leftward deviation of the cranial portion of the trachea (near the cranial border of the cardiac silhouette), suggesting that this method is fairly reliable for differentiating primary megaesophagus from a PRAA.[3]

Angiography may be performed prior to surgery for identification of any other concurrent vascular anomalies or causes of vascular ring changes, other than PRAA. However, angiography is not commonly performed, as the vast majority of vascular ring cases can be corrected via a left fourth intercostal thoracotomy.[10]

Treatment/Management:

Medical management of a PRAA involves managing the secondary megaesophagus, and the sequelae to it, by feeding the patient a liquid or semi-solid food diet in an elevated position. Medical management alone is almost always unrewarding, and is associated with a poor long-term prognosis, thus it is not recommended.[8, 9, 10] Surgical dissection and ligation of the ligamentum arteriosum, along with any other fibrous connective tissue surrounding the esophagus, is the treatment of choice.[8, 10] The goal is to alleviate the esophageal stricture(s), which ultimately leads to the resolution of clinical signs. By removing this compression, the esophagus is allowed return to a normal diameter, and esophageal motility/function can be returned to the effected area.[8, 10]

A standard left fourth intercostal thoracotomy is performed.[3, 8, 10] Exposure of the ligamentum arteriosum is made by blunt dissection. Passing a rigid tube into the esophagus may assist with localizing the area(s) of esophageal compression. Once the ligamentum arteriosum is

isolated, it can be ligated, and transected. Using a double ligation technique is important, due to the potential for vascular patency through the ligamentum arteriosum. Again, the passage of a rigid tube through the esophagus will demonstrate any further strictures, and allow for any addition break down of fibrous connective tissue that may be surrounding the esophagus, as well as evaluate surgical success.[8, 10]

Post-operative management recommendations include feeding the patient a liquid or blenderized diet in an elevated position, for a minimum of three days to one week, followed by a more solid diet. Some clinicians reported feeding a liquid diet for up to two months, then slowing transitioning to a semi-solid diet.[1, 6, 7, 9, 10] Returning the patient to a solid food early on decreased the risk of aspiration as well.[10] Elevated feedings are almost always recommended post-operatively to assist with emptying of the esophagus, and encourages the return of esophageal function and motility. One study discussed the use of Metoclopromide for its effects on distal esophageal motility. However, it also increases lower esophageal tonicity, and its use is not well documented in these cases.[10]

Outcome/Prognosis:

Surgical correction is often successful, and associated with a good long-term prognosis, but may not result in complete resolution of clinical signs.[2,6] Most complications develop secondarily to unresolved megaesophagus, and include persistent episodes of regurgitation and aspiration pneumonia.[2, 8] One study that followed twenty-three dogs postoperatively reported that nineteen of the twenty-three dogs never regurgitated again after surgery, and the remaining four dogs had infrequent episodes of regurgitation, often associated with rapid eating or consuming a

food that was not in their normal diet.[2] These patients will need to be medically managed for the remainder of their lives with a modified diet and elevated feedings. Despite medical management, these patients go on to live fairly normal lives, and owner satisfaction is reported to be very high.[2]

Early surgical repair is recommended to reduce the irreversible esophageal dilation and concurrent loss of esophageal function, and motility.[6, 7, 9] It is also important to make owners aware of the potential for continued medical management throughout the remainder of their pet's life prior to surgery, as well as their role in the postoperative period, to ensure the best prognosis possible.[10]

Conclusion:

Persistent right aortic arch is a congenital vascular ring anomaly that is most commonly associated with young, large breed dogs. The most common clinical signs are persistent regurgitation shortly after a meal, along with unthriftiness, despite having a voracious appetite. These signs are often associated with the animal first consuming solid foods shortly after weaning. Early detection, diagnosis and surgical correction are associated with the best long-term prognosis. Aspiration pneumonia is of the greatest concern, with or without surgical correction, as it can be a fatal complication. Overall, the prognosis for this condition is good to excellent, even if the patient must be medically managed for life.

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