Double aortic arch in a dog

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- Double aortic arch is a rare congenital heart defect resulting from improper development of the embryonic arches.
- Naturally occurring double aortic arches have been rarely reported, and animals with double aortic arches generally have a poor prognosis for survival.
- Most dogs and cats that undergo surgical correction of double aortic arches die during or a few hours after surgery.

A 3-month-old 1.86-kg (4.1-lb) female mixed-breed dog was evaluated for regurgitation during and after eating of 2 months' duration. On physical examination, the dog was bright and alert, had a rectal temperature of 38.2°C (100.8°F), had a heart rate of 150 beats/min, and was panting. Results of auscultation of the heart and lungs were unremarkable. Results of a CBC indicated leukocytosis (19.7×10^3 WBCs/μL; reference range, 6.0 to 17.0×10^3 WBCs/μL), a low PCV (36%; reference range, 37% to 55%), and a low RBC count (5.38×10^6 cells/μL; reference range, 5.5 to 8.5×10^6 cells/μL). Results of serum biochemical analyses indicated low creatinine (0.2 mg/dL; reference range, 0.5 to 1.3 mg/dL) and high phosphorus (8.7 dL; reference range, 3.1 to 5.8 mg/dL) concentrations.

Radiography of the thorax revealed dilation of the esophagus cranial to the fourth rib and sacculation of the esophagus around the trachea (**Figure 1**). Prominence of the right ventricular border of the heart was also seen. There was no evidence of aspiration pneumonia. Results of an esophagram using barium sulfate (15 mL) confirmed the esophageal dilation cranially with narrowing of the esophageal lumen at the level of the fourth rib (**Figure 2**). Dilation of the distal esophagus was also seen; however, primary and secondary peristaltic activity was present. There was also evidence of gastroesophageal reflux.

Esophageal dilation due to a vascular ring anomaly was determined to be the cause of the dog's regurgitation. A persistent right aortic arch was suspected. Surgical correction via a left lateral thoracotomy was planned. The dog was premedicated with acepromazine (0.1 mg/kg [0.05 mg/lb], SC), atropine (0.05 mg/kg [0.02 mg/lb], SC), and morphine (0.55 mg/kg [0.25 mg/lb], SC). Anesthesia was induced with propofol (4.4 mg/kg [2 mg/lb], IV) and maintained using sevoflurane. Cefazolin (22 mg/kg [10 mg/lb], IV, q 2 h) was given perioperatively.

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The dog was positioned in right lateral recumbency, and a left lateral thoracotomy was performed at the fourth intercostal space. Exploration of the mediastinal structures revealed that the left aortic arch was normal. The esophagus was dilated and filled most of the cranial portion of the mediastinum. Further dissection of the right side of the dilated esophagus revealed a second slightly smaller aortic arch. The brachycephalic and left subclavian arteries originated from the left aortic arch normally. The right aortic arch was dissected from the mediastinum, double ligated with 2-0 silk, and transected between the ligatures (**Figure 3**). Additional dissection of the mediastinum around the transected right aortic arch completely freed the entrapped esophagus. An inflated 5-mL Foley catheter[a]

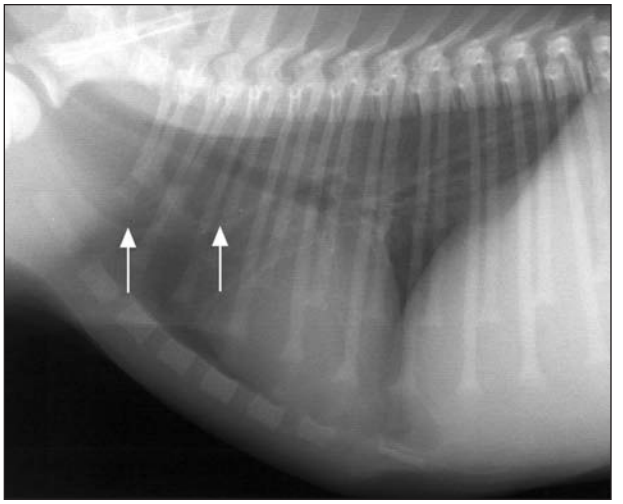

Figure 1—Left lateral radiographic view of the thorax of a 3-month-old dog evaluated for regurgitation of 2 months' duration during and after eating. Notice dilation and sacculation of the esophagus cranial to the fourth rib (arrows).

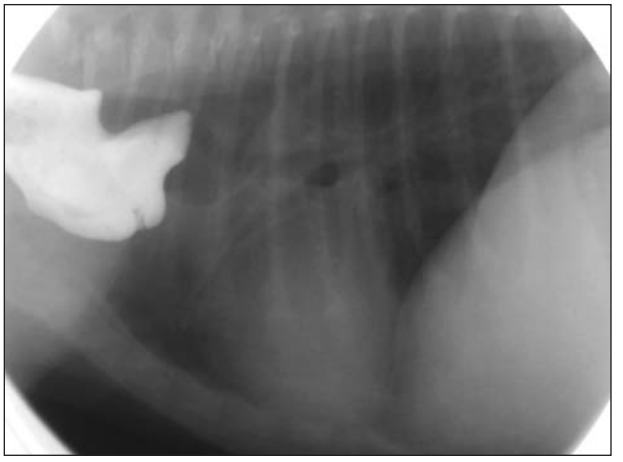

Figure 2—Left lateral esophagram of the dog in Figure 1. Notice marked dilation of the esophagus cranial to the fourth rib and a prominent narrowing of the esophageal lumen.

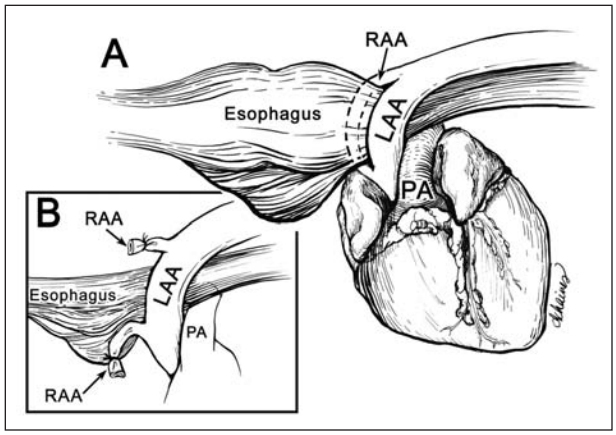

Figure 3—Illustration of a surgical approach for division and ligation of the right aortic arch as viewed from a left thoracotomy at the fourth intercostal space. Illustration courtesy of the University of Tennessee College of Veterinary Medicine. All rights reserved.

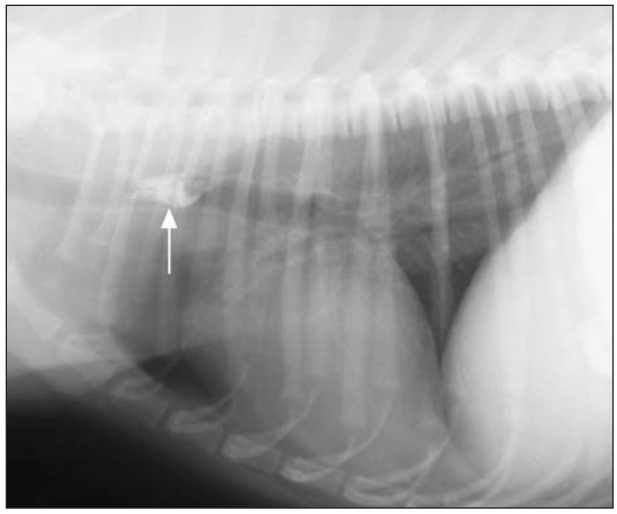

Figure 4—Left lateral esophagram of the dog in Figure 1 obtained 6 months after surgery for correction of a double aortic arch. Notice that the esophagus is no longer dilated cranial to the fourth rib (arrow).

was passed down the esophagus to ensure that there was no additional obstruction. The thoracic duct was torn during the procedure, but no attempt was made to ligate the duct. An 8-F thoracic catheter[b] was placed in the thorax at the sixth intercostal space. The catheter was sutured in place with 3-0 nylon stay suture. The ribs were opposed with 4 ligatures of 4-0 polydioxanone. The serratus ventralis muscle, scalenus muscle, and subcutaneous tissue were closed with 4-0 polydioxanone in simple continuous patterns. The skin was closed with 3-0 poliglecaprone in a continuous subcuticular pattern and 3-0 nylon in a simple continuous pattern.

Metoclopramide (0.2 mg/kg [0.1 mg/lb], PO, q 8 h), ranitidine (2 mg/kg [0.9 mg/lb] IV, q 8 h), amoxicillin-clavulanic acid (20 mg/kg [9 mg/lb], PO, q 12 h), sucralfate (0.5 g, PO, q 8 h), and morphine (0.2 mg/kg, IV, q 4 h) were administered after surgery. The thorax was aspirated every 2 hours, and the catheter was removed 2 days after surgery. Morphine was discontinued the day after surgery. The dog was fed a slurry of a growth diet in an elevated position. The dog was discharged 3 days after surgery.

The owners were instructed to continue feeding a gruel diet in an elevated position and to slowly decrease the elevation and the amount of water in the diet during the next month. Because of esophageal reflux, administration of ranitidine, metoclopramide, and sucralfate was continued as previously prescribed for 1 month. Complications due to hypertension caused by ligation of the aortic arch were not observed.

Conversations with the owner 6 months after surgery indicated that the dog was no longer regurgitating and had gained weight. The dog was eating a dry diet from a bowl at floor level. Results of an esophagram performed 6 months after surgery indicated that the size of the esophagus was normal, and it had normal peristaltic activity (**Figure 4**).

Vascular rings are developmental anomalies of the thoracic great vessels encircling the esophagus and the trachea by a complete or partial ring formation. There are 3 types of vascular ring anomalies: persistent aortic arch, aberrant subclavian artery, and double aortic arch. Of these, persistent right aortic arch with left ligamentum arteriosum accounts for 95% of the clinical cases of vascular ring anomalies in dogs.[1] The remaining 5% encompass all other types of vascular ring anomalies. The actual percentage of double aortic arch is unknown but is assumed to be low on the basis of the few reports in the literature. Double aortic arch is a congenital heart defect resulting from the improper development of the embryonic arches. During normal embryonic development, 6 pairs of aortic arches connect the aortic sac to the dorsal aorta and surround the embryonic pharynx.[2] Vascular ring anomalies result from abnormal development of arches 3, 4, and 5.[2] Vascular ring anomalies can be classified as type 1 through 7 and are classified by which vessels entrap the intrathoracic esophagus. Double aortic arch is considered a type 4 vascular ring anomaly.[3] Double aortic arches have been reported in humans,[4] dogs,[5-8] a monkey,[4] and a cat.[5] Of the reported cases of double aortic arches in small animals, the diagnosis was determined at necropsy in 1 dog and 1 cat, 1 dog was euthanatized before recovery from surgery at the owners request, 1 dog died 3 hours after surgery, and 1 dog died 3 days after surgery.[5-8] To the authors' knowledge, there have been no previous reports of dogs that have survived long enough to be discharged from the hospital after surgical correction of double aortic arches. Therefore, no long-term complications after surgical correction of double aortic arches have been reported.

Clinical signs of vascular ring anomalies are caused by esophageal constriction with dilation of the esophagus cranial to the constriction. Clinical signs are usually seen at the time of weaning. The most common clinical sign is regurgitation associated with eating. As the condition persists, the esophagus dilates cranial to the heart base. Dilation is usually pouch-like, and continued medical management may be required for months or for the entire life on the animal after surgical correction because dilation and hypomotility of the esophagus are often irreversible.[2] The prognosis for

surgical correction of persistent right aortic arch in dogs varies.[9] Some dogs continue to regurgitate, and clinical signs in others resolve after surgery. However, the prognosis is generally regarded as poor for dogs after surgical correction of double aortic arches because no dogs have reportedly survived after surgical correction of double aortic arches. The severity of esophageal dilation before surgery is also an important prognostic factor, and it is believed that esophageal dilatation rarely resolves completely.[10,11] However, in the dog reported here, clinical or radiographic evidence of esophageal dilation was not seen after surgical correction of the vascular ring. This indicates that megaesophagus caused by vascular ring anomalies can resolve with early surgical correction. Severe hypertension after ligation of 1 arch has also been reported.[7] When there is no appreciable difference in size between the 2 arches, it is believed that ligation of 1 arch results in hypertension, with increased afterload and work on the left ventricle eventually leading to left heart failure.[7] In the dog in this report, ligation of the smaller right aortic arch was performed and complications were not seen.

Radiography of the thorax and an esophagram are typically performed to confirm a vascular ring anomaly. However, they do not provide information as to what type of vascular ring anomaly is present before surgery. Although 95% of the vascular ring anomalies in dogs are type 1 persistent right aortic arch with ligamentum arteriosum, choosing the appropriate surgical approach is important for surgical correction of the less common vascular ring anomalies.[7] Angiography may provide an accurate evaluation of the aortic arch anatomy and thus a diagnosis of the rare types of aortic arch anomalies.[12] Angiography is also helpful in determining the diameters of both arches and thus the surgical approach to the smaller arch.[12] Two-dimensional echocardiography has also been useful and fairly specific in humans with double aortic arches but is less frequently used in dogs and cats.[13]

Vascular ring anomalies are uncommon in humans and represent < 1% of congenital cardiovascular defects.[14] In humans, many imaging modalities are useful for diagnosing double aortic arches. Imaging modalities used in humans include esophagrams, angiography, **computed tomography (CT)**, and **magnetic resonance imaging (MRI)**.[12] However, the preferred method for diagnosing congenital heart disease in humans is MRI. Magnetic resonance imaging defines anatomic structures better than echocardiography and is less invasive than angiography.[15] Magnetic resonance imaging is considered advantageous over CT because CT requires contrast enhancement to depict the vessel lumen, whereas MRI can be used with or without contrast.[16] Although MRI is not as readily available in veterinary medicine, use of MRI for the diagnosis of aortic arch anomalies would provide information about vascular anatomy and permit determination of the appropriate surgical approach. Without this information, it is preferable to perform a left thoracotomy because most vascular ring anomalies are persistent right aortic arches.

In humans, various risk factors that influence surgical outcome have been identified.[17] Results of a retrospective study[17] of patients with interrupted aortic arch indicated that low cardiac output required inotropic support and low blood pressure often led to renal insufficiency or failure. Of those patients, 55% died postoperatively. Sepsis and weight < 2.4 kg (5.3 lb) also negatively influenced surgical outcome. However, early surgical intervention is the preferred treatment for dogs and cats because this will help normalize esophageal function.

[a]Foley catheter, CR Bard Inc, Covington, Ga.
[b]Trocar thoracic catheter, Sherwood Medical, St Louis, Mo.

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