CASE REPORT

Vascular Ring Anomalies: Case Report and Brief Review

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Summary

During early fetal life six pair of aortic arches surround the esophagus and trachea. Normal maturation and selective regression of these structures form the adult vasculature. Abnormal location or development of the aortic arches may result in pressure on adjacent organs.

Vascular ring anomalies must be considered with any patient with a history of regurgitating food shortly after eating. Physical examination, test feedings, survey and contrast radiographs may give an accurate impression of the problem but a final diagnosis can only be made following surgical exploration. In the case presented, the dog had all the clinical and diagnostic signs suggestive of a vascular ring anomaly. Thoracotomy and elimination of the vascular constriction around the esophagus was both diagnostic and therapeutic for the condition. It is important that owners be made aware that surgical correction of the stenosis does not guarantee a successful conclusion to the case. If the dilation of the esophagus cranial to the stenosis is severe, accumulation of food with subsequent regurgitation may persist. A dilation of the esophagus caudal to the stenosis is present in a large percentage of cases and this also may result in an unrelenting problem. Unfortunately, the probability of these complications cannot be accurately evaluated prior to treatment.

The hereditary potential for this defect must also be considered. Congenital vascular anomalies such as patent ductus arteriosus would seem to have a hereditary basis. Therefore, it is probably correct to advise against breeding affected animals. Further, the inbreeding of nonaffected animals which come from litters containing affected animals should be avoided.

Résumé

Anomalies vasculaires en forme d'anneau: Description d'un cas et brève revue

Au début de la vie foetale, six paires d'arches aortiques entourent l'oesophage et la trachée. La maturation normale et la régression sélective de ces structures donnent la vascularisation définitive. Une localisation ou un développement anormal de ces arches peut provoquer une pression sur les organes adjacents.

Il faut penser aux anomalies vasculaires en forme d'anneau, chaque fois que l'anamnèse d'un patient révèle de la régurgitation qui se produit tôt après un repas. Un examen physique, un repas de vérification et des radiographies de contrôle ou de contraste peuvent fournir une opinion assez exacte du problème; on ne peut toutefois poser un diagnostic final qu'à la suite d'une chirurgie exploratrice. Dans ce cas-ci, le chien présentait tous les signes cliniques suggestifs d'une anomalie vasculaire en forme d'anneau. Une thoracotomie et l'élimination de la constriction vasculaire encerclant l'oesophage, corrigèrent la condition. Il importe de prévenir les propriétaires que la correction chirurgicale de la sténose n'en garantit pas automatiquement la guérison. Lors d'une dilatation grave de l'oesophage, en avant de la sténose, l'accumulation de nourriture et sa régurgitation subséquente peuvent persister. Dans beaucoup de cas, la dilatation oesophagienne se situe en arrière de la sténose et peut résulter en un problème irréductible. On ne peut malheureusement pas percevoir l'éventualité de ces complications, avant le traitement.

Il faut aussi penser à la transmission héréditaire possible de ces conditions. Des anomalies vasculaires congénitales telles qu'un *ductus arteriosus* béant, semblent en effet attribuables à l'hérédité. Il faudrait par conséquent probablement déconseiller l'accouplement d'animaux atteints de cette anomalie. Il faudrait de plus éviter d'accoupler des sujets normaux, mais issus de portées dont certains membres souffrent de cette anomalie.

Introduction

Congenital vascular ring anomalies of the intrathoracic vessels are not necessarily a complete ring as the name implies. A major vessel in an abnormal location so that it mechanically interferes with the esophagus, trachea or other related structures could be termed a vascular ring. It has been suggested that 0.5 to one percent of the general canine population have some form of congenital cardiac defect (4). Approximately ten percent of these anomalies are vascular ring formations with the persistent right aortic arch being the most common type (1). This is a case report of an unusual vascular ring which was corrected surgically and a summary of the abnormal embryologic development which could lead to this anomaly.

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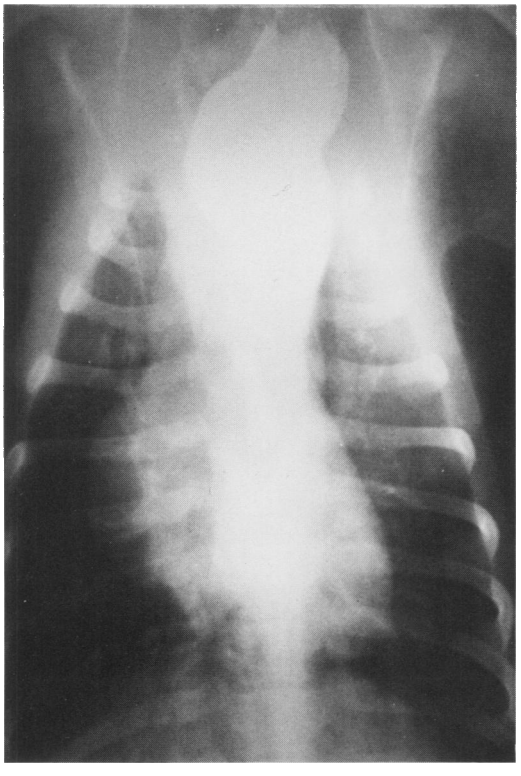

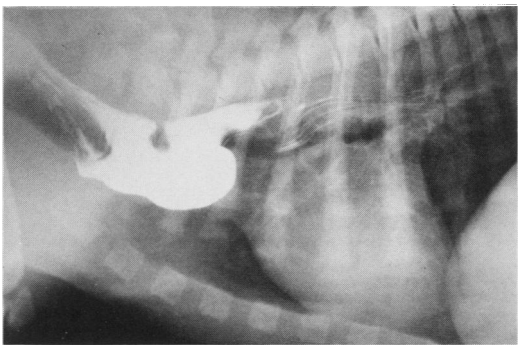

FIGURE 1A and B. Dorsoventral and lateral thoracic radiographs after swallow of a thick barium suspension, showing sacculation of the esophagus cranial to the base of the heart.

Case Report

A three month old, male, German Shepherd dog was presented to the Western College of Veterinary Medicine with a two month history of regurgitation which occurred within minutes after eating solid food. The puppy was alert and in reasonably good physical condition. A slight bilateral, serous nasal discharge was noted. There was a gas-filled structure, thought to be the esophagus, at the left side of the thoracic inlet. All other physical examination findings were normal. A test meal of dog food was given and the dog regurgitated masticated, nondigested food a few minutes after eating.

Clinical pathological findings were within normal limits. Survey radiographs of the thorax were taken. A dilated esophagus and a narrowed trachea were apparent just cranial to the base of the heart. A barium swallow followed by thoracic radiographs substantiated these radiographic findings (Figure 1A and B).

A tentative diagnosis of a vascular ring anomaly causing constriction of the esophagus and trachea was made and surgical exploration undertaken. The thorax was entered on the left side through the fourth intercostal space. A distended esophagus was readily apparent (Figure 2). The fascia over the esophagus was carefully dissected beginning at the dilated portion and moving caudally. An aberrant branch of the aorta was found three centimeters caudal to the left subclavian artery passing anteromedially over the dorsal aspect of the esophagus. After further dissection it was found that this vessel was an anastomosis between the aorta and the brachiocephalic artery ventral to the esophagus. A palpable pulse was present in this constricting vessel. Two transfixing ligatures were placed around it (Figure 3) and the vessel transected between the ligatures. The esophagus dilated at that point without major adhesions remaining. The thorax was closed and a chest drain kept in place for 24 hours.

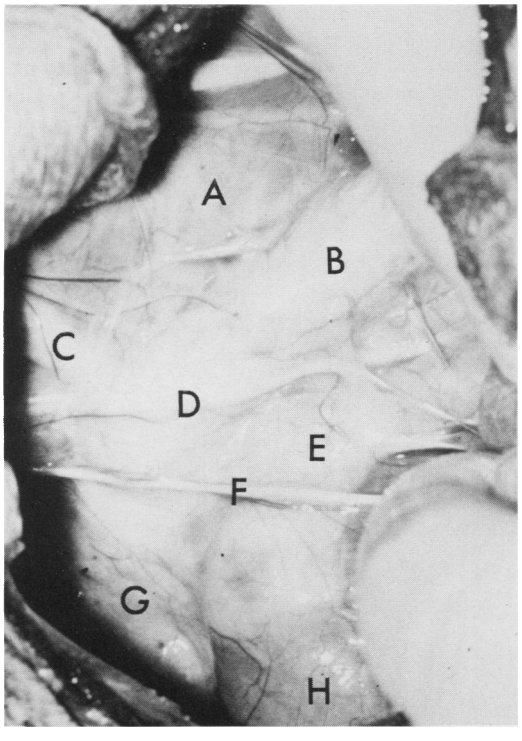

FIGURE 2. Incision through the fourth left intercostal space allowing inspection of: A — dilated esophagus, B — aorta, C — left subclavian artery, D — left vagus nerve, E — pulmonary artery, F — phrenic nerve, G — thymus and H — right ventricle.

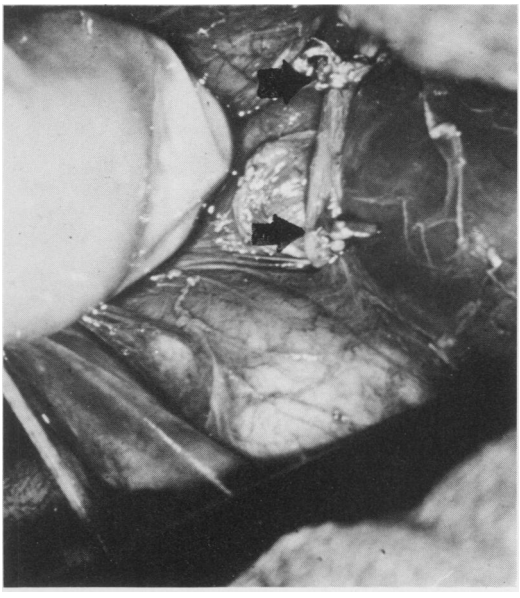

FIGURE 3. Patient heart to the left. The aorta is seen as a light colored structure with the aberrant vessel passing over the dorsum of the esophagus. Two ligatures have been placed prior to sectioning of the constricting band (arrows).

The dog was fed canned dog food 24 hours after the surgery and did not regurgitate. Three months after the surgery barium swallow radiographs were made. There was easy passage of material into the stomach and a decrease in size of the dilated cranial esophagus was seen. For ten months postoperatively, the dog has had no recurrence of regurgitation after eating.

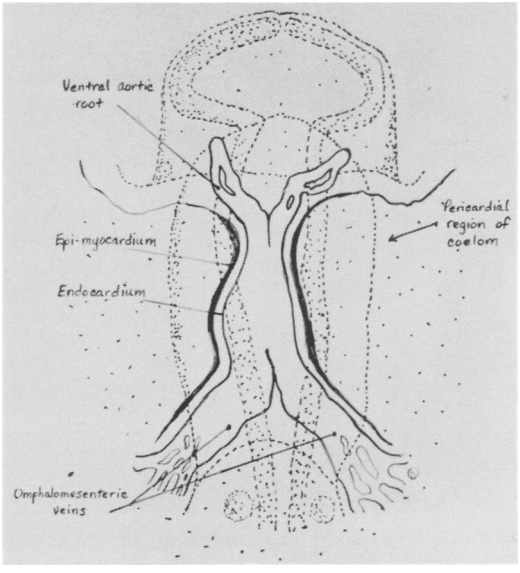

FIGURE 4. Primordial heart with paired ventral aortas forming cranially and omphalomesenteric (Yolk) veins caudally.

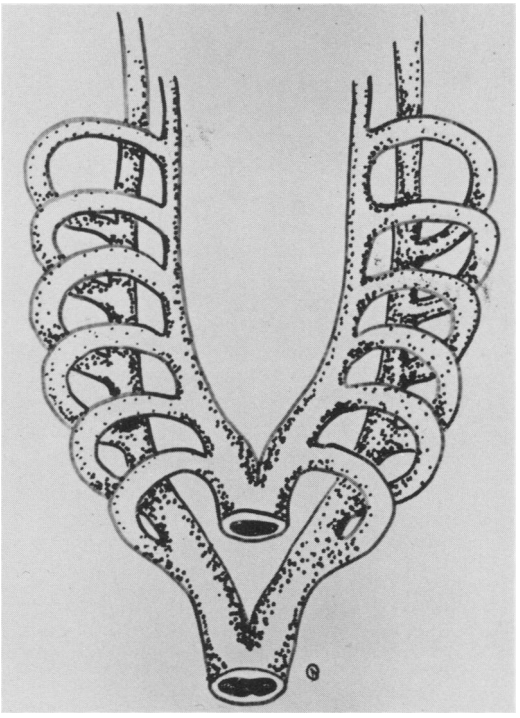

FIGURE 5. Artist's concept of the ventral and dorsal aortas with all six aortic arches. At no time will this configuration occur as the cranial arches mature and regress as the caudal arches develop.

Embryology

The heart and great vessels arise from cords of mesodermal cells which form paired tubes located ventrolateral to the pharynx. These tubes become fused as the foregut closes and give rise to the heart (Figure 4). Six pair of aortic arches appear in the course of fetal maturation (3). These arches arise from the ventral aortic roots and pass dorsolaterally on both sides of the pharynx to discharge blood into the developing pair of dorsal aortic roots. Although a total of six pairs of arches are formed, they are not present simultaneously since the cephalic arches undergo regression into their mature structure as the final arches are being formed (Figure 5). All arch formation occurs cranial to the forelimbs with the dorsal aortas brought into apposition and fused into a single structure caudal to this level.

The fate of the aortic arches in the mature animal may be summarized as follows (Figure 6):

The FIRST and SECOND arches regress and the internal and external carotids are formed by the dorsal and ventral aortas respectively. The dorsal aortas between the third and fourth arches regress leaving the THIRD arch as part of the internal carotid artery. The left FOURTH arch enlarges to form the main aortic arch while the right FOURTH arch becomes the right subclavian artery. The FIFTH arch is vestigial and of no apparent clinical significance. The

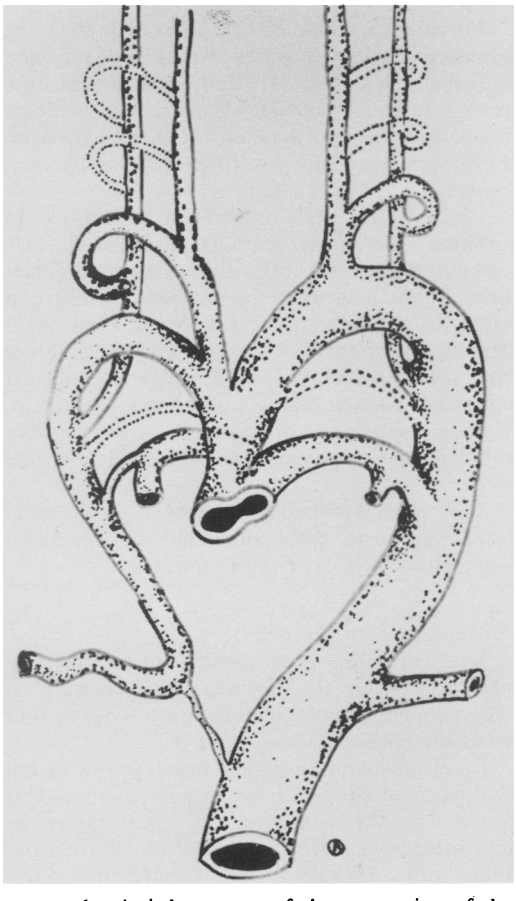

FIGURE 6. Artist's concept of the maturation of the mammalian aortic arches.

SIXTH arches become the pulmonary arteries. The left SIXTH arch retains its connection with the left dorsal aorta as the ductus arteriosus.

Many variations of the vascular ring anomaly based on degrees of patency and association with other cardiac lesions have been described (2, 5). However, five main forms are of concern:

1. Double aortic arch: both fourth arches maintain their attachment to the adult descending aorta.
2. Right aortic arch with left ductus arteriosus: ring formed by the normal ductus arteriosus from the left pulmonary arch joining to the abnormal right aortic arch.
3. Left aortic arch with right ductus arteriosus: ring formed by the abnormal communication between the right pulmonary arch and the normal aortic arch.
4. Aberrant right subclavian artery: origin of the artery such that the vessel must pass retro-esophageal and thus put pressure on the esophagus, trachea, etc.
5. Aberrant left subclavian artery: as for the aberrant right subclavian artery.

Discussion

The aberrant vessel in this case appeared to be the right dorsal aorta. This anastomotic vessel arose from the aorta 3 cm caudal to the left subclavian artery, passed dorsally over the esophagus and joined the brachiocephalic artery ventromedially. Figure 6 shows the normal vestigial tag still attached to the aorta and to what will become the brachiocephalic trunk. The right and left dorsal aortas failed to resolve their attachment completely as the left became the aorta and the right the subclavian. The aberrant vessel, which in this case completed the vascular ring around the esophagus, is postulated to be the right dorsal aorta.

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