J Vet Intern Med 2004;18:510–514

Tracheal Signs and Associated Vascular Anomalies in Dogs with Persistent Right Aortic Arch

James W. Buchanan

Medical records of 55 dogs with 1 or more vascular rings around the esophagus and trachea were reviewed to determine the nature and frequency of related vascular anomalies and to determine the reliability of tracheal deviation on radiographs for the diagnosis of persistent right aortic arch (PRAA). Fifty-two (95%) of the 55 dogs had PRAA. Of the 52 dogs with PRAA, 44% had coexisting compressive arterial anomalies: 17 had retroesophageal left subclavian artery and 6 had double aortic arch with atretic left arch. Characteristic tracheal deviation was consistently present in dogs with PRAA. Moderate or marked focal leftward curvature of the trachea near the cranial border of the heart in dorsoventral (DV) or ventrodorsal (VD) radiographs was found in 100% of available radiographs of 27 dogs with PRAA. Moderate or marked focal narrowing of the trachea also was noted in 74% of DV or VD radiographs and 29% of lateral radiographs of the dogs. Tracheal position in 30 of 30 dogs with megaesophagus and 62 of 63 control dogs was midline or rightward in VD or DV radiographs. Histology in a neonatal dog with PRAA revealed evidence of tracheal deviation and compression even before birth. Focal leftward deviation of the trachea near the cranial border of the heart in DV or VD radiographs is a reliable sign of PRAA in young dogs that regurgitate after eating solid food, and contrast esophagrams are not necessary to confirm the diagnosis of vascular ring compression.

Key words: Congenital heart disease; Esophageal compression; Megaesophagus; Radiographic diagnosis; Vascular rings.

Persistent right aortic arch (PRAA) is the most common vascular ring anomaly in dogs.[1] Because vascular rings cause esophageal compression, regurgitation soon after eating solid food is the principal clinical sign of PRAA in young dogs. Barium esophagrams usually are performed to detect esophageal enlargement and to differentiate generalized megaesophagus from vascular ring anomalies in which the stricture site is over the base of the heart and esophageal enlargement occurs only cranial to the heart. Barium esophagrams sometimes are not diagnostic of vascular rings because of inadequate filling or minimal dysfunction and enlargement.

Tracheal abnormalities also may be observed in dogs with PRAA because the trachea is an outgrowth of the primitive gut tube and is incorporated within vascular rings along with the esophagus.[2] The hypothesis was made that leftward displacement of the trachea by a PRAA could be identified on plain dorsoventral (DV) or ventrodorsal (VD) radiographs, which would obviate the time and expense of a contrast radiographic study for the diagnosis of PRAA. To test this hypothesis, available survey radiographs of 27 dogs with PRAA, 30 dogs with megaesophagus, and 63 control dogs were reexamined to determine the presence of tracheal abnormality sufficient to make a reliable diagnosis of PRAA and to differentiate the condition from generalized megaesophagus. Associated vascular anomalies in dogs with PRAA also have been observed, but the frequency of such anomalies has not been reported.[1] The types and frequency of other vascular anomalies associated with PRAA in the 52 dogs are presented along with evidence of severe tracheal changes in utero in a neonatal dog.

From the Section of Cardiology, School of Veterinary Medicine, University of Pennsylvania, Philadelphia PA. To be presented at the ACVIM Forum, June 9–12, 2004, Minneapolis, MN.

Reprint requests: James W. Buchanan, DVM, M Med Sci, Professor of Cardiology, ACVIM Dip Cardiology, School of Veterinary Medicine, University of Pennsylvania, 3900 Delancey, Philadelphia, PA 19104; e-mail: jwb@vet.upenn.edu.

Submitted October 29, 2003; Revised January 2, 2004; Accepted February 4, 2004.

0891-6640/04/1804-0010/$3.00/0

Materials and Methods

Fifty-five dogs with vascular ring anomalies form the basis of this report. Fifty-two dogs with PRAA were identified from medical records, surgery logs, teaching files, and surgery or postmortem examination photographs. Two additional dogs had vascular rings composed of left-sided aortas and right-sided patent ductus arteriosus.[3] Another dog had a left-sided aorta, retroesophageal right subclavian artery, and a right-sided ligamentum arteriosum. Age, breed, sex, and associated vascular anomalies were tabulated from the various sources. Available thoracic radiographs of 27 of the dogs were reexamined in a nonblinded fashion and scored for tracheal deviation and narrowing by using a standardized scale of 1–3 for mild, moderate, or marked tracheal deviation or narrowing. Focal leftward curvature of the trachea near the cranial border of the heart in DV or VD radiographs was scored 1 if leftward curvature was less than 25% of the diameter of the trachea (mild), 2 if leftward curvature was 25–100% of the diameter of the trachea (moderate), and 3 if leftward curvature was more than 100% of the diameter of the trachea (marked) (Fig 1). Narrowing of the trachea in the area of leftward curvature was scored 1 if the tracheal diameter in the narrowed segment was reduced less than 10% of the tracheal diameter cranial and caudal to the narrowed segment, 2 if the diameter was reduced 10–25%, and 3 if the diameter was reduced more than 25%. On lateral radiographs, ventral curvature of the trachea cranial to the heart was scored 1 if tracheal displacement was less than 100% of tracheal diameter, 2 if tracheal displacement was 100–200% of tracheal diameter, and 3 if tracheal displacement was more than 200% of its diameter (Fig 2). Focal narrowing of the trachea on lateral radiographs was scored the same way as on DV and VD radiographs. As a control group, thoracic radiographs of 63 dogs younger than 1 year, excluding dogs with a history of trauma, diaphragmatic hernia, peritoneopericardial diaphragmatic hernia, congenital heart disease, pneumothorax, or megaesophagus were examined and scored for tracheal deviation and narrowing in the same manner. All dogs with a diagnostic code in their medical records indicating megaesophagus were identified and thoracic radiographs of the most recent 30 dogs were evaluated in the same manner. Serial section histology of the trachea and adjacent vessels of a 1-day-old dog with PRAA and a normal stillborn dog were compared to assess the effects of aortic arch compression on tracheal morphology at the tissue level.

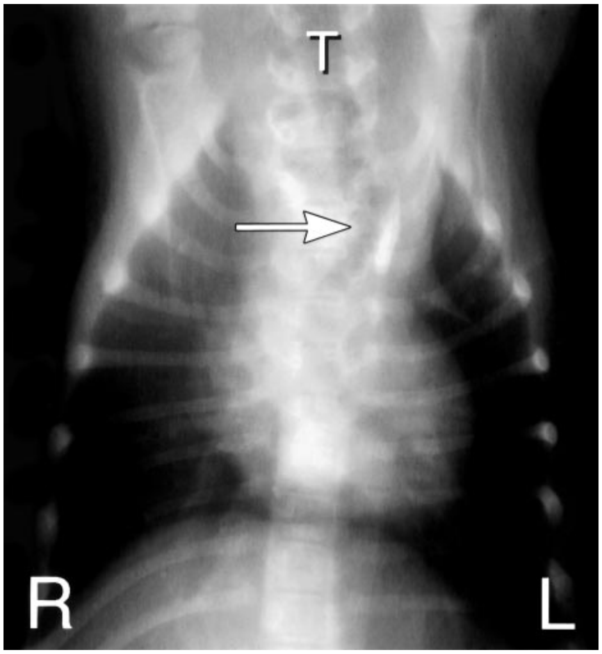

Fig 1. Ventrodorsal radiograph of a 10-week-old Golden Retriever with persistent right aortic arch confirmed at surgery. The trachea (T) is curved leftward more than 100% of the tracheal diameter near the cranial border of the heart (arrow) and is narrowed more than 25% in the deviated segment.

Results

Age, Breed, and Sex

The average age of 44 dogs with PRAA excluding the 1-day-old neonate and a 7-year-old dog was 4.2 months. Sixteen were German Shepherd Dogs, 5 were Labrador Retrievers, and 5 were Irish Setters. Other breeds were represented by only 1 or 2 dogs. The 41 dogs for which sex information was available included 23 females and 18 males.

Associated Vascular Abnormalities

Seventeen (33%) of 52 dogs with PRAA had a retroesophageal left subclavian artery in addition to PRAA (Fig 3; Table 1). Six dogs had patent ductus arteriosus and 6 dogs had 2 aortic arches with an atretic left arch. Six dogs had persistent left cranial vena cava and 3 dogs had a left hemiazygos vein. Two dogs had generalized megaesophagus in addition to PRAA.

Tracheal Deviation and Narrowing

In 62 of 63 control dogs, the trachea was midline or curved rightward on VD or DV radiographs (Table 2). Focal tracheal narrowing was not evident on any of the radiographs. On lateral radiographs, mild ventral depression of the trachea was noted in 1 dog. In 30 of 30 dogs with megaesophagus, the trachea also was midline or curved rightward (Table 2). In 27 of 27 dogs with PRAA, the trachea was curved leftward near the cranial border of the heart (Fig 1; Table 2). This finding differed dramatically from control dogs and dogs with megaesophagus. On lateral radiographs, ventral curvature of the trachea cranial to the heart was mild in 9 dogs, moderate in 6 dogs, and marked in 11 dogs (Fig 2). The trachea was straight in 1 dog. Focal narrowing of the trachea cranial to the heart was mild in 13 dogs, moderate in 6 dogs, and marked in 3 dogs. The

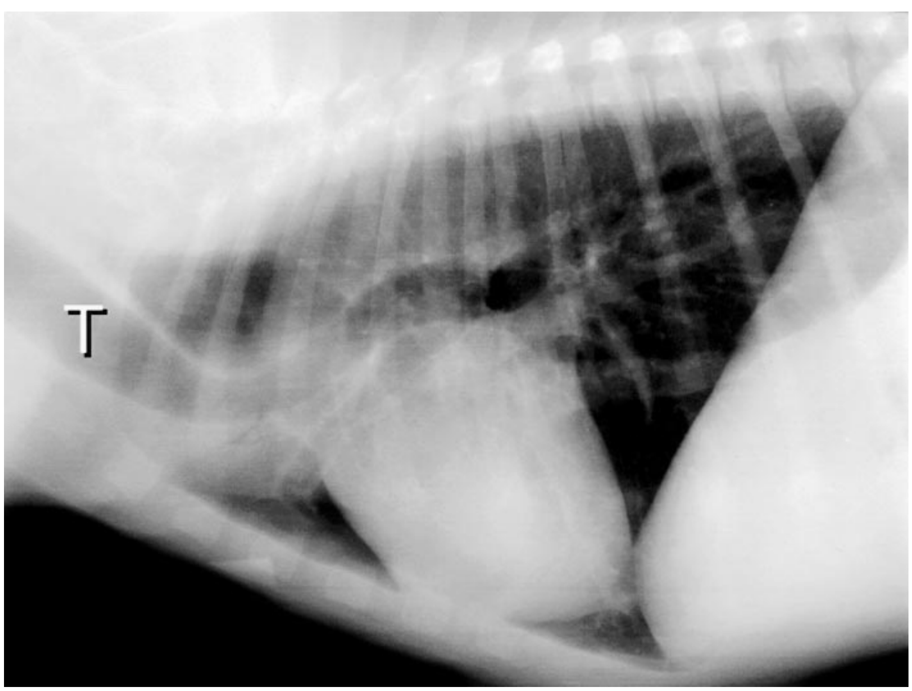

Fig 2. Lateral radiograph of a 3-month-old German Shepherd Dog with persistent right aortic arch and retroesophageal left subclavian artery. The trachea (T) is deviated ventrally more than 200% of its diameter and narrowed more than 25% in the deviated segment in comparison to the tracheal diameter just cranial to the carina and in the cervical area.

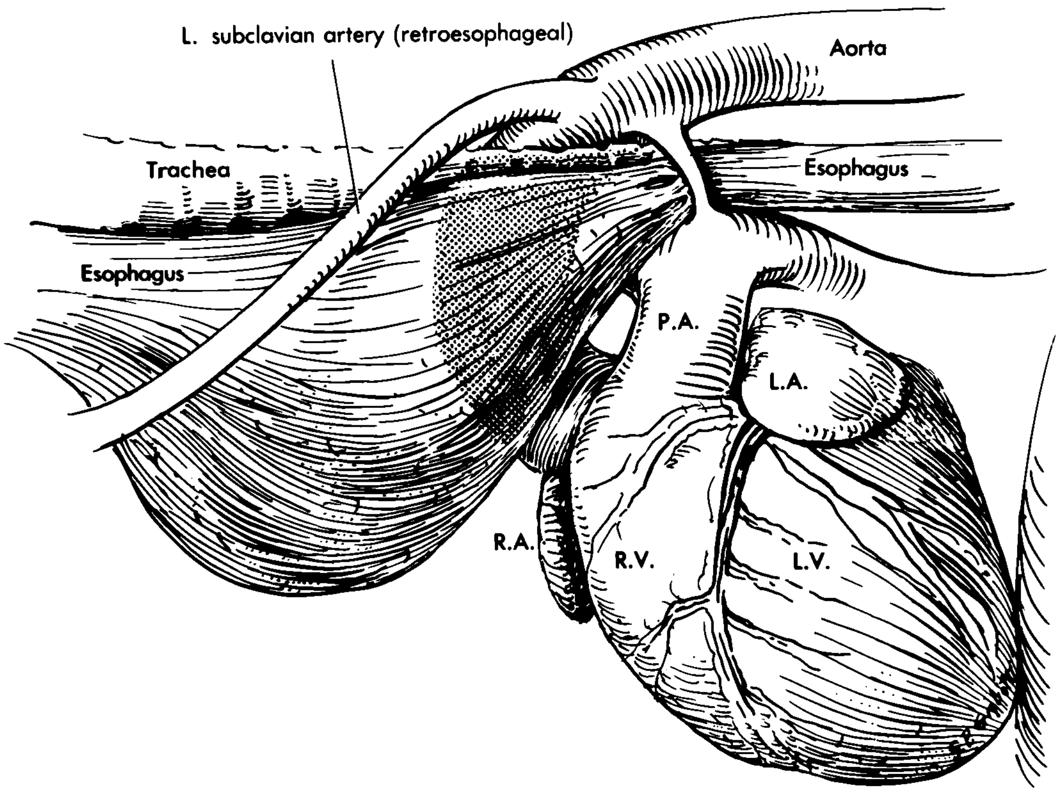

Fig 3. Diagram of vascular pathology in persistent right aortic arch viewed from the left side. The esophagus and trachea are surrounded by the pulmonary artery (P.A.) on the left and ventrally, the aorta on the right, and the ligamentum arteriosum dorsally. A retroesophageal left subclavian artery also may be present and compressing the esophagus. L.A., left atrium; L.V., left ventricle; R.A., right atrium; R.V., right ventricle.

trachea was not narrowed in 4 dogs and it was obscured by barium in 1 dog.

Serial Section Histology

In the normal stillborn dog (Fig 4), the aorta, ductus arteriosus, and pulmonary artery were situated on the left side of the trachea and separated from it. The trachea was straight and tracheal rings were symmetrical, equal in size, and contained uniform amounts of cartilage. The mucosa was thin and without folds. In the neonate with PRAA (Fig 5), the trachea was deviated leftward and severely compressed between the aortic arch on the right side and the constricted ductus arteriosus on the left side. The tracheal rings were partially overlapped in the compressed segment and asymmetrical in size, shape, and thickness and longitudinal folds were present in the tracheal mucosa. The esophagus also was severely compressed and had a thin wall in the compressed segment.

Table 1. Vascular anomalies in 55 dogs with vascular rings.

	Number	%
Persistent right aortic arch	52	95
Number with coexisting compressive anomalies:		
Retroesophageal left subclavian artery	17	33
Double aortic arch	6	12
Patent ductus arteriosus	6	12
Persistent left cranial vena cava	6	12
Left hemiazygous vein	3	6
Retroesophageal right patent ductus arteriosus	2	4
Retroesophageal right subclavian artery and right ligamentum arteriosum	1	2

Discussion

The trachea in dogs usually is straight and on the midline or curved to the right of the midline.[4] These positions were confirmed on radiographs of 62 of 63 control dogs and in 30 of 30 dogs with megaesophagus. In contrast, all 27 dogs with PRAA had moderate or marked leftward curvature of the trachea near the cranial border of the cardiac silhouette.

Table 2. Tracheal position in 120 dogs.

	Controls	Megaesophagus	PRAA
Marked rightward	3	4	
Moderate rightward	11	9	
Mild rightward	24	14	
Midline	24	3	
Mild leftward	1		
Moderate leftward			7
Marked leftward			20
Total	63	30	27

PRAA, persistent right aortic arch.

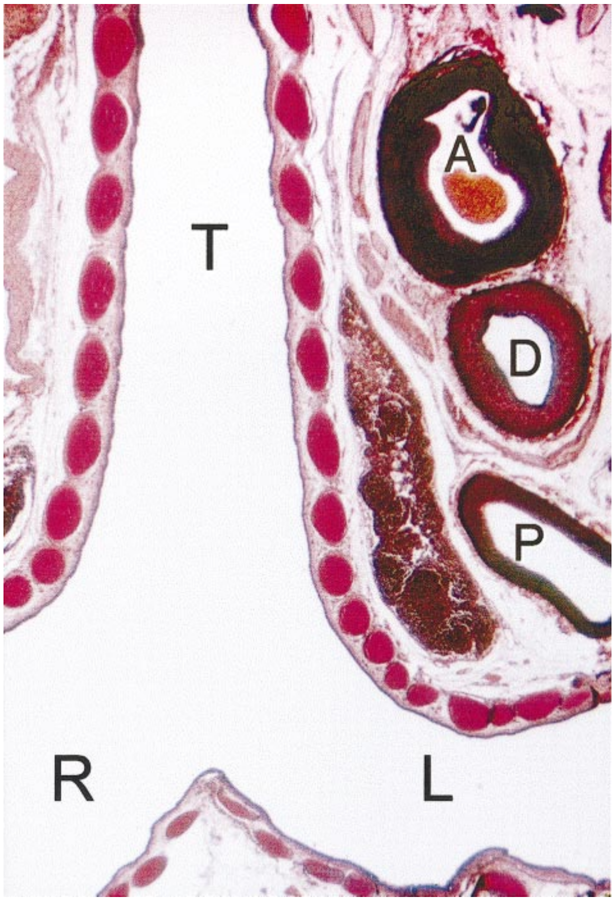

Fig 4. Histologic section of the trachea and adjacent blood vessels in a normal stillborn dog. The trachea (T) is straight and tracheal rings are uniform in size and shape. The aorta (A), ductus arteriosus (D), and pulmonary artery (P) are all situated on the left side of the trachea. Verhoeff elastic stain, 4×.

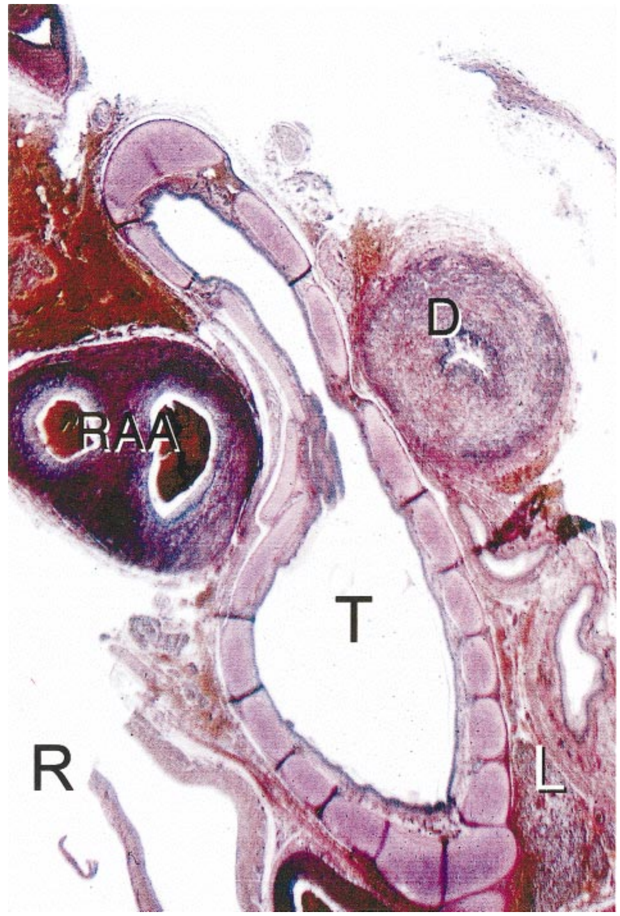

Fig 5. Histologic section of the trachea and adjacent blood vessels in a 1-day-old dog with persistent right aortic arch. The trachea (T) is deviated leftward and is severely compressed between the right aortic arch (RAA) and the contracted ductus arteriosus (D). Tracheal rings next to the aorta in the compressed segment are thin, lack dense cartilage, and have overriding edges. The mucosa in the compressed segment is thick and folded. A retroesophageal left subclavian artery and persistent left cranial vena cava also were present in this dog but are not seen in this section. Verhoeff elastic stain, 4×.

In 1 dog with double aortic arch, the site of leftward curvature was more cranial than in dogs with isolated PRAA. Tracheal narrowing in the deviated segment also was common and usually was more apparent in DV or VD views than on lateral views. Overexposed DV or VD radiographs in some of the dogs proved helpful because overexposure reduced opacity of the superimposed vertebral column and sternum and improved visibility of the air-filled trachea.

Primary generalized megaesophagus often is apparent on plain radiographs of some dogs and contrast esophagrams to confirm its diagnosis are unnecessary. However, contrast esophagrams have been recommended in the past to differentiate PRAA from generalized megaesophagus when the diagnosis is uncertain. The results of this study indicate that leftward curvature of the trachea reliably distinguishes PRAA from generalized megaesophagus and obviates the need for contrast esophagrams.

Vascular rings surround both the esophagus and trachea because the trachea is an outgrowth of the embryonic gut tube surrounded by evolving aortic arches.[2] The embryology of normal arches and the embryogenesis of PRAA and related vascular anomalies are illustrated and discussed in an accompanying cardiac surgery Web site.[a] All dogs in this report had a functioning right aortic arch, including the 6 dogs with 2 aortic arches. The left arch in each of these 6 dogs was atretic and caused esophageal compression separate from and cranial to the ligamentum arteriosum. In such cases, division of the ligamentum arteriosum without recognizing and dividing the atretic left arch will not relieve esophageal compression and probably accounts for the unsuccessful outcome of surgery in some dogs with PRAA.

The coexistence of a retroesophageal left subclavian artery (Fig 3) in 33% of the dogs with PRAA often is not recognized by surgeons. Failure to divide the anomalous subclavian artery as well as the ligamentum arteriosum allows continued dorsal compression of the esophagus and incomplete relief of the esophageal obstruction in some dogs. Five of 6 dogs with a retroesophageal left subclavian artery and 1 of 3 dogs with double aortic arch had marked ventral curvature of the trachea on lateral radiographs (Fig 2). Therefore, marked ventral curvature of the trachea should prompt a thorough search for these additional abnormalities when dividing the ligamentum arteriosum in dogs with PRAA.

The frequency of associated arterial and venous abnormalities in dogs with PRAA probably is greater than those tabulated in this report because several dogs were operated on by residents and junior staff surgeons who have less experience in recognizing coexistent anomalies. Retroesophageal left subclavian arteries are not obvious in some dogs unless specific dissection dorsal to the esophagus is carried out to look for them. Similarly, atretic left aortic arches can be overlooked in dogs with double aortic arch. A persistent complete or incomplete left cranial vena cava also is commonly present in dogs with PRAA but may not be recognized or recorded.[5]

The apparently nonrandom breed distribution in this case series corresponds to previously reported breed predispositions in which the relative risk odds ratio in German Shepherd Dogs was 4.5 times that found in other breeds.[1] Irish Setters were predisposed to PRAA 20–40 years ago[6] and the Irish Setters tabulated in this report were operated on during that period. A more recent tabulation of 60 dogs with PRAA did not include any Irish Setters.[7] The fact that 52 (95%) of 55 dogs with vascular ring anomalies in this report had PRAA confirms an estimate published 35 years ago suggesting that PRAA accounts for 95% of vascular ring anomalies in dogs.[8]

The histologic findings in the 1-day-old neonate with PRAA explain the puzzling observation that stridorous respiration usually is the presenting sign of vascular ring anomalies in human infants, especially those with double aortic arch,[9,10] whereas esophageal dysfunction is the usual presenting sign in dogs. The neonatal puppy with PRAA (Fig 5) had protracted, wheezing expirations producing a sound like a ‘‘whistle on the end of a balloon’’ during the 24 hours it lived, according to the owner. The vascular ring probably became tighter as the ductus arteriosus constricted, but tracheal compression must have been present even before ductus closure based on the abnormal tracheal ring histology. Other dogs with a short ductus arteriosus and severe tracheal compression probably do not survive to an age when veterinary examinations are performed. Postmortem examinations seldom are performed in neonatal dogs and may account for the lack of reports of stridorous respiration caused by vascular rings in the veterinary literature. Dogs with PRAA that survive to weaning and the intake of solid food must have a relatively longer ligamentum arteriosum and less severe compression of the trachea and esophagus than that observed in the 1-day-old neonate. A 2nd factor contributing to the species difference in clinical signs is that human infants ingest only liquids for many months and evidence of esophageal dysfunction may not be manifested.

A relatively long ligamentum arteriosum has been found at surgery in dogs with PRAA that have minimal esophageal enlargement and minimal tracheal deviation (unpublished observation). An example was a 2-year-old dog with a distinctly long ligamentum arteriosum that had minimal esophageal dysfunction and only regurgitated once a week. A barium esophagram was inconclusive, but the DV radiograph showed leftward curvature of the trachea suggestive of PRAA. Surgery was performed and PRAA with a long ligamentum arteriosum was confirmed.

Some authors have recommended presurgical angiography to determine the nature of vascular rings when their presence is suspected. This technique has not been helpful in the author's experience because vascular rings involve 3-dimensional relationships that are difficult to characterize even with biplane angiography.[11] Because the large majority of vascular rings involve PRAA and related vessels, a surgeon with a good understanding of aortic arch embryology can determine the nature of the vascular abnormality by thoracotomy through the left 4th intercostal space and correct whatever vascular abnormality is found.[a,3,8,12]

Footnote

[a] http://cal.vet.upenn.edu/cardiosf

References

1. Buchanan JW. Prevalence of cardiovascular disorders. In: Fox P, Sisson D, Moise NS, eds. Canine and Feline Cardiology, 2nd ed. Philadelphia, PA: WB Saunders; 1999:457–470.
2. Carlson BM. Patten's Foundations of Embryology, 6th ed. New York, NY: McGraw-Hill; 1996:548.
3. Holt D, Heldmann E, Michel K, Buchanan JW. Esophageal obstruction caused by a left aortic arch and an anomalous right patent ductus arteriosus in two German Shepherd littermates. Vet Surg 2000; 29:264–270.
4. Kneller SK. The larynx, pharynx and trachea. In: Thrall DE, ed. Textbook of Veterinary Diagnostic Radiology, 2nd ed. Philadelphia, PA: WB Saunders; 1994:229.
5. Buchanan JW. Persistent left cranial vena cava in dogs: Angiocardiography, significance, and co-existing anomalies. J Am Vet Radiol Soc 1963;4:1–8.
6. Patterson DF. Epidemiologic and genetic studies of congenital heart disease in the dog. Circ Res 1968;23:171–202.
7. Buchanan JW. Causes and prevalence of heart disease. In: Kirk RW, Bonagura JD, eds. Current Veterinary Therapy XI. Philadelphia, PA: WB Saunders; 1992:647–655.
8. Buchanan JW. Patent ductus arteriosus and persistent right aortic arch surgery in dogs. J Small Anim Pract 1968;9:409–428.
9. Nugent EW, Plauth WH Jr, Edwards JE, et al. Congenital heart disease. In: Hurst JW, ed. The Heart Arteries and Veins, 6th ed. New York, NY: McGraw-Hill; 1986:624.
10. Fleenor JD, Weinberg PM, Kramer SK, Fogel M. Vascular rings and their effect on tracheal geometry. Pediatr Cardiol 2003;24:430–435.
11. Buchanan JW, Patterson DF. Selective angiography and angiocardiography in dogs with congenital heart disease. J Am Vet Radiol Soc 1965;6:21–39.
12. Buchanan JW, Lawson DD. Cardiovascular system. In: Archibald J, ed. Canine Surgery, 2nd Archibald ed. Santa Barbara, CA: American Veterinary Publications; 1974:429–480.