Persistent Right Aortic Arch in Foals

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ABSTRACT: This article describes the normal embryology of the aortic arches as well as the development of the right aortic arch anomaly. An overview of the diagnosis and causes of persistent right aortic arch (PRAA) in foals is presented. Treatment of PRAA depends on early diagnosis and involves surgery and supportive care.

Persistent right aortic arch (PRAA) is a congenital anomaly in which a vestigial structure persists when it should have disappeared.¹ In PRAA, the esophagus is constricted, primarily by a combined arterial channel that later fibroses to form the ligamentum arteriosum, resulting in esophageal dilatation and dysphagia. PRAA has been reported in small animals² as well as in a calf,³ a bull,⁴ a llama,⁵ a bison,⁶ and foals.⁷ The reports of PRAA in large animals fail to show a favorable outcome (Table 1). Various breeds can be affected. Male animals are most frequently affected. In dogs, PRAA may be attributable to single or multiple recessive gene defects.⁸

NORMAL EMBRYOLOGIC DEVELOPMENT

In equine embryos with a crown–rump length (CRL) of 5 mm (around 21 days’ gestational age), the dorsal aortic roots are reduced cranially to the small regressing first and second aortic arches (Figure 1). A ventrally located aortic sac extends cephalad from the truncus arteriosus. Five pairs of aortic arches (first, second, third, fourth, sixth) develop, extending from the aortic sac to the right or left aortic roots. Fifth aortic arches usually fail to develop in mammals; however, the last pair is still designated as sixth. By the time an embryo reaches 12 mm CRL (around 32 days’ gestational age), the third, fourth, and sixth pairs of aortic arches are completely developed.⁹

The third aortic arches contribute to the formation of the internal carotid arteries. The left fourth aortic arch forms the definitive aortic arch. Pulmonary arteries arise from the proximal parts of the sixth aortic arches. The distal part of the left sixth aortic arch remains as the ductus arteriosus, connecting the left dorsal aortic root with the pulmonary trunk (Figure 2). On the right side, this connection degenerates.¹⁰ The left and right dorsal aortic roots extend caudal to the level of the thoracic appendage buds to form a single aorta.⁹ In embryos of 11.5 to 12 mm CRL, around 32 days, spiral septation of the conus cordis and truncus arteriosus into the aorta and pulmonary trunk has occurred. The left fourth aortic arch has grown larger than the contralateral arch, which stops developing.⁹ As development continues, the right and left subclavian arteries arise from the left and right caudal dorsal cervical intersegmental arteries, respectively, and
move cephalad, following the dorsal aortic roots to join the brachiocephalic trunk (Figure 2). The right dorsal aortic root degenerates. The left dorsal aortic root continues caudad from the left dorsal aortic arch, forming the first part of the descending aorta.

**PATHOPHYSIOLOGY**

PRAA develops when, at an early gestational age (around 32 days), the disparity in growth between the opposing fourth aortic arch is reversed so that the right aortic arch forms the definitive aortic arch. If the left sixth arch completely degenerates so that it does not form a ductus arteriosus, a constricting left-to-right vessel will not be present and there will be no clinical problem.\(^\text{10}\) If the cause of the PRAA is situs inversus, in which all the viscera are reversed, the esophagus is not constricted and, obviously, no clinical signs appear.\(^\text{11}\) However, if the distal portion of the left sixth aortic arch persists as it normally does, it will develop into the ductus arteriosus, connecting the truncus arteriosus and, later, the pulmonary trunk with the left dorsal aortic root. Cranial to this junction, the left dorsal aortic root regresses between the ductus arteriosus and the origin of the left subclavian artery as the latter moves to join the brachiocephalic trunk.\(^\text{10}\) The hemodynamic flow is then directed from the ductus arteriosus caudad via the left dorsal aortic root to its junction with the right aortic root to form the aorta (Figure 2).\(^\text{10}\) Figure 3 represents the progressive shortening of the combined left-to-right arterial channel carrying blood to the aorta.

Postpartum closure and fibrosis of the ductus arteriosus and left dorsal aortic root result in the formation of the constricting ligamentum arteriosum. Necropsy of a foal at 2 months postpartum (Figure 4) showed dorsal compression of the esophagus by the ligamentum arteriosum. The vascular ring exerting pressure on the esophagus and trachea was completed by the left pulmonary artery, the PRAA, and the base of the heart.\(^\text{12}\) In another foal aged 5 months,\(^\text{13}\) the ligamentum arteriosum was described as a long, thick (2 cm in width), fibrous band located dorsally below the vertebral bodies rather than laterally as described in dogs and cats.\(^\text{14}\) Other references to the left-to-right arterial channel label it simply ductus arteriosus.\(^\text{14,15}\) In one account, left-to-right “crossing over” of the ductus arteriosus is noted,

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**Table 1. Summary of Reported Cases of Persistent Right Aortic Arch in Large Animals**

<table>
<thead>
<tr>
<th>Breed/Species</th>
<th>Sex</th>
<th>Age at Diagnosis</th>
<th>Treatment</th>
<th>Outcome</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lipizzaner</td>
<td>Male</td>
<td>6 months</td>
<td>Surgery</td>
<td>Died at surgery</td>
<td>Petrick et al</td>
</tr>
<tr>
<td>Tennessee Walking horse</td>
<td>Female</td>
<td>2 days</td>
<td>None</td>
<td>Euthanized</td>
<td>Bartels and Vaughan</td>
</tr>
<tr>
<td>Arabian</td>
<td>Male</td>
<td>5 months</td>
<td>Surgery</td>
<td>Survived</td>
<td>Mackey</td>
</tr>
<tr>
<td>Arabian</td>
<td>Female</td>
<td>14 months</td>
<td>None</td>
<td>Euthanized</td>
<td>Buchanan</td>
</tr>
<tr>
<td>Arabian</td>
<td>Male</td>
<td>2 days</td>
<td>Surgery</td>
<td>Died 3 months postoperatively</td>
<td>Bauer et al</td>
</tr>
<tr>
<td>Llama</td>
<td>Male</td>
<td>9 years</td>
<td>None</td>
<td>Euthanized</td>
<td>Butt et al</td>
</tr>
<tr>
<td>Holstein</td>
<td>Male</td>
<td>3 days</td>
<td>None</td>
<td>Euthanized</td>
<td>Rooney and Watson</td>
</tr>
<tr>
<td>Guernsey</td>
<td>Male</td>
<td>5 years</td>
<td>None</td>
<td>Euthanized</td>
<td>Roberts et al</td>
</tr>
<tr>
<td>Bison</td>
<td>Male</td>
<td>16 months</td>
<td>None</td>
<td>Died</td>
<td>Peters et al</td>
</tr>
</tbody>
</table>

**Persistent right aortic arch is a congenital anomaly that constricts the esophagus, resulting in esophageal dilatation and dysphagia.**
and a remnant of a left-to-right dorsal aortic connection is described. 14
Prepartum, PRAA is not detrimental to the fetus because nourishment through the umbilical cord is consistent. Birth defects develop before completion of the affected body system and, therefore, are present before birth. 16 However, clinical signs are usually recognized soon after birth. When affected foals nurse, the proper flow of milk is compromised by the constricting vascular ring around the esophagus, and dysphagia and regurgitation of milk occur. This can result in aspiration of the milk and subsequent pulmonary pathology. Consistent dilatation of the esophagus impairs intrinsic esophageal musculature and results in loss of parasympathetic innervation, which may become irreversible. 17
Because the trachea is also constricted, respiratory signs may also be evident. Dogs with severe tracheal compression probably do not survive the first few days of life. 18 Constriction of the trachea may not be pronounced in some animals because the constricting ligament is longer.

**CLINICAL SIGNS**

The first noted sign is usually nasal regurgitation of milk. However, foals with PRAA may not show clinical signs until weaning or until they begin to consume solid food. 19 A 14-month-old filly showed respiratory dysfunction before dysphagia. 20 Over time, foals develop a bulging swelling at the left midcervical area (Figure 5). The size can fluctuate during the day, depending on nursing habits. The distended esophagus is soft and can be fluid filled. Affected foals have a poor body condition score, stunted growth, and a coarse haircoat. They may cough after nursing. The most common respiratory complication of equine esophageal disorders is aspiration pneumonia. 21 If aspiration pneumonia develops, crackles and wheezes can be heard on auscultation. Results of auscultation of the heart are normal.

**DIAGNOSIS**

**Differential Diagnosis**

Because the most common clinical sign in foals with PRAA is nasal regurgitation, the diagnostic differentials include cleft palate, hypoplasia of the palatal shelf, esophageal stricture, and esophageal dilatation/mega-esophagus. 22 Usually, the suspicion of esophageal obstruction is raised during passage of a nasogastric tube. During endoscopy, a vascular ring constriction is indicated by a narrowing of the esophagus at the heart base. 23 This should not be confused with normal dilation of the lumen in the midthoracic region. 24

A differential diagnosis for a cranial esophageal lesion with or without pulmonary lesions should include all causes of esophageal stenosis. A stricture at this location could be congenital or developmental and not associated with a vascular ring anomaly. A more common etiology of esophageal stenosis is foreign body obstruction of the esophagus. Clinical signs of foreign body obstruction are more acute, with less time for the development of secondary pulmonary lesions.

**Plain Radiography**

Radiographs will probably not distinguish a PRAA from other vascular ring anomalies. However, they should be taken when PRAA is suspected and should include both the cervical and cranial thoracic regions. Radiographs of equine patients with suspected PRAA have been published. 13, 15 Depending on its size, the patient may be imaged in a recumbent or standing pos-
tion. Smaller patients can be placed in a recumbent position to obtain lateral and ventrodorsal views; if the patient remains standing, only lateral views can be obtained. Larger patients must be imaged standing, and again, only lateral views can be taken. Because of the importance of evaluating the esophagus, radiographs of standing patients may be more diagnostic, even though the number of views is limited. This is because the esophagus is in a normal anatomic position in standing patients; in recumbent foals, it can shift to a more dependent position.

A prominent radiographic change suggestive of PRAA is esophageal dilatation in the caudal cervical region extending into the thoracic inlet. A change of this type is more accurately evaluated in standing studies. The dilatated esophagus can also be seen within the cranial thorax to the level of the second or third rib. The opacity of the contents of the cervical esophagus as seen on the noncontrast study depend on the foal's diet (i.e., fluid [milk] or fluid mixed with grain). The volume of the esophageal contents depends on the completeness of the stricture and the amount of prior regurgitation. The chronicity of the lesion determines the degree of sacculation of the prestenotic esophagus. The sacculation can reach into the cranioventral mediastinum.

Prepartum, persistent right aortic arch is not detrimental to the fetus; however, clinical signs (e.g., nasal regurgitation) are usually recognized soon after birth.

The radiographic study should include evaluation of the thorax to rule out aspiration pneumonia, which is diagnosed by an increase in fluid density within the lungs. An airway pattern with prominent bronchial walls as well as pneumonic segments in the more ventral parts of the lungs may be seen. With time, the pneumonic region may assume a more uniform density, suggesting
pulmonary abscess formation. Detecting pulmonary lesions is important in determining a prognosis.

**Contrast Radiography**

In standing foals, after identification of the fluid-filled, prestenotic esophageal dilatation, approximately 50 ml of barium sulfate suspension can be administered using a dose syringe or an esophageal tube. A dose syringe can be used successfully if care is taken to avoid spilling the contrast agent on the patient, where it creates confusing radiopaque shadows. Barium sulfate should also be administered carefully. During administration, the foal’s head should be elevated to encourage most of the fluid to pass into the esophagus. After the foal attempts to swallow, its head can be lowered and a lateral radiograph taken with the x-ray beam centered on the thoracic inlet (Figure 6). The radiographic technique can be increased because of the muscle thickness at this site. Passage of the barium suspension identifies the site of the stricture. If the esophageal saculation is large and the contrast agent fills only the distended esophagus, preventing it from reaching the lesion, additional contrast agent can be added; however, the possibility of regurgitation before radiography can be completed increases.

Continued administration of contrast agent may result in some of the contrast meal passing through the stricture site. The amount of contrast agent that passes through adds little information relative to either diagnosis or determination of the prognosis. Subsequent swallowing attempts may introduce the barium suspension into the upper airways, which may be seen on radiographs. The contrast agent is rather innocuous and probably does not severely affect existing pulmonary lesions.

Smaller foals that are examined in a recumbent position may inhale more of the contrast meal, resulting in less contrast outlining the dilated esophagus and stricture site. In these foals, it may be more useful to administer the contrast agent through an esophageal tube passed until the tip is at the stricture site.

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**Figure 3.** Positional migration and shortening of the left dorsal aortic root bring the left ductus arteriosus to the right side (dorsal view).

**Figure 4.** In a 2-month-old Norwegian colt with PRAA, the combined channel of the ductus arteriosus and the remnant of the left dorsal aortic root fibrosed to form a ligamentum arteriosum (arrow) that compressed the esophagus (arrowhead) dorsally (left lateral view).
Compromise the cardiovascular system. Foals may regurgitate milk after induction of anesthesia. Muzzling the foal 1 hour before induction reduces the risk for regurgitation. Anesthesia may be induced with a face mask while the foal is still standing. Immediately after induction of anesthesia, the foal should remain in sternal position with the head elevated. This prevents esophageal contents from flowing into the trachea. Placement of an endotracheal tube and inflation of the cuff further prevents aspiration of milk. Once under general anesthesia, the foal is placed in right lateral recumbency. Positive-pressure ventilation should be available or used from the induction of anesthesia until recovery. Additional analgesia is accomplished with intercostal nerve blocks and intrapleural injection of local anesthetics. Surgery in foals is facilitated because of their size.

Computed Tomography
The use of computed tomography to diagnose PRAA in a neonatal foal was recently reported. For precise diagnosis, contrast medium injected into the ascending aorta can determine the exact location of the anomaly. The foal should be tranquilized and a catheter placed in the right carotid artery. The foal is then placed over an automatic cassette changer, and approximately 25 ml of contrast medium is injected during the aortogram. The tip of the catheter should be visualized in the left ventricle.

TREATMENT
Medical management may be effective when pathologic changes in esophageal dilatation and motility are minimal. One affected llama was successfully fed small meals from an elevated feeder for 2½ years. Surgery allows more comprehensive treatment. Any concurrent aspiration pneumonia should be treated accordingly.

Vascular ring anomalies are treated by surgical ligation and division of the constricting vascular ring. Any fluid or electrolyte imbalances should be corrected before induction of anesthesia. PRAA is not associated with hemodynamic problems and, therefore, usually does not compromise the cardiovascular system. Foals may regurgitate milk after induction of anesthesia. Muzzling the foal 1 hour before induction reduces the risk for regurgitation. Anesthesia may be induced with a face mask while the foal is still standing. Immediately after induction of anesthesia, the foal should remain in sternal position with the head elevated. This prevents esophageal contents from flowing into the trachea. Placement of an endotracheal tube and inflation of the cuff further prevents aspiration of milk. Once under general anesthesia, the foal is placed in right lateral recumbency. Positive-pressure ventilation should be available or used from the induction of anesthesia until recovery. Additional analgesia is accomplished with intercostal nerve blocks and intrapleural injection of local anesthetics. Surgery in foals is facilitated because of their size.

Several surgical techniques have been described for thoracotomy in the horse. Use of thoracoscopy to free the constricted esophagus has been described in dogs with PRAA. The skin incision for thoracotomy is created caudal to the triceps brachii muscles over the

**Surgery allows more comprehensive treatment than medical management, but affected foals have a poor prognosis.**
sixth rib, beginning adjacent to the caudal border of the scapular cartilage and extending ventrad toward the olecranon. The latissimus dorsi and serratus ventralis muscles are incised. The periosteum of the sixth rib is cut and separated from the bone using a periosteal elevator. The proximal end of the bone is cut with a Gigli wire or an oscillating saw. Rib resection is completed by disarticulation at the costochondral junction. Finocchietto self-retaining retractors are used to spread the fifth and seventh ribs. A nasogastric tube or an endoscope is advanced within the esophagus toward the constricted area. The ligamentum arteriosum is identified by sharp and blunt dissection. Two ligatures are then placed around the ligamentum arteriosum, and the ligament is incised. Any remaining periesophageal fibrous tissue is divided. The dilated portion of the esophagus is usually not resected because of possible leakage. Plication can be attempted, taking care not to penetrate the lumen during suture placement. Resection or plication reduces redundant tissue but does not restore peristalsis.

The incision is closed beginning with the periosteum, using a simple interrupted suture pattern and size 2 absorbable suture material. Before complete closure, the lungs are inflated and a chest tube is placed in the thoracic cavity through a separate stab incision. Muscle layers and the subcutaneous layer are closed with a continuous suture pattern. After skin closure, any air remaining in the thoracic cavity is suctioned out and the chest tube removed.

**POSTOPERATIVE CARE**

Limited information exists regarding postoperative care of foals recovering from PRAA. There are more reports of postoperative management in small animals. The goal is to provide proper nutrition and prevent regurgitation. A liquid diet should be given frequently and in small amounts from an elevated position. A nasogastric tube enables feeding a gruel diet or milk. To avoid ingestion of bedding material by the patient, the stall should not be bedded. Antibiotics for management of aspiration pneumonia are administered based on results of culture and sensitivity tests. Postoperative analgesia should eliminate the pain resulting from thoracic surgery. Additionally, intercostal nerves adjacent to the incision site can be infiltrated with local anesthetic.

If surgical treatment of PRAA is successful, subsequent contrast studies of the esophagus made with the patient in a standing position can be used to evaluate any persistent postoperative esophageal stenosis or dilatation. For these studies, the barium sulfate should be mixed with grain or gruel to create a bolus that will permit determination of the character of the esophageal lumen and extent of esophageal distention at the site of the earlier stricture. The passage of barium liquid alone does not determine the ability of the esophagus to distend enough to permit passage of a solid bolus. These radiographic studies can also permit reevaluation of bronchial or pulmonary lesions.

**PROGNOSIS**

Because PRAA is a rare condition that may go unnoticed for some time, the onset of clinical signs may be insidious. The extent of aspiration pneumonia greatly
influences the timing of treatment. Severe aspiration pneumonia compromises treatment efforts; therefore, surgical treatment is seldom pursued. In dogs, the prognosis after surgery is good to excellent.³⁸ Foals with PRAA have a poor prognosis for long-term survival.³⁹ Often, large animals are presented for diagnosis or treatment when they are months or years old. This usually results in the animals being euthanized because of the severity of clinical signs.

**SUMMARY**

PRAA is the result of abnormal embryonic development in which the disparity in growth of the aortic arches is reversed. An arterial channel formed by the ductus arteriosus and left dorsal root conveys blood from the pulmonary trunk to the aorta. This arterial channel and the arterial ligament that develops from it constrict the esophagus, causing esophageal dilatation, dysphagia, and regurgitation. Aspiration pneumonia is a common complication. Successful treatment depends on early diagnosis, surgery, and supportive care. Affected foals are considered to have a poor prognosis.

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**REFERENCES**


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1. The descending aorta is formed by the convergence of the
   a. left sixth aortic arch and right dorsal aortic root.
   b. right and left dorsal aortic roots.
   c. left dorsal aortic root and left ductus arteriosus.
   d. right dorsal aortic root and right ductus arteriosus.

2. In the development of a PRAA in which the esophagus is compressed, the ductus arteriosus derived from the left sixth aortic arch (pulmonary arch) is moved to the descending aorta on the right side of the embryo. This is accomplished via the connection of the ductus arteriosus with the
   a. migrating right subclavian artery.
   b. developing right fourth aortic arch.
   c. left dorsal aortic root.
   d. right sixth aortic arch.

3. Which of the following occurs in the early development of PRAA?
   a. incomplete septation of the truncus arteriosus and conus cordis
   b. invariable persistence of the right sixth aortic arch, forming the ductus arteriosus
   c. failure of the right subclavian artery to join the brachiocephalic trunk
   d. degeneration of the left dorsal aortic root between the ductus arteriosus and the left subclavian artery

4. The special imaging technique that would most easily and accurately assist in diagnosing PRAA in a foal is a(n)
   a. standing lateral radiographic study.
   b. barium sulfate esophagogram.
   c. angiogram made following a jugular vein injection of contrast agent.
   d. computed tomographic study.

5. _____ is a common feature of PRAA in a foal seen on lateral radiographs.
   a. Ventral displacement of a narrowed trachea
   b. Dorsocaudal pneumonia
   c. Pleural fluid
   d. Pneumomediastinum

6. All of the following are possible clinical signs of PRAA in foals except
   a. dysphagia.  c. coughing.
   b. cardiac arrhythmia.  d. poor body condition score.

7. _____ is not a diagnostic differential for PRAA in foals.
   a. Cleft palate  c. Congestive heart failure
   b. Esophageal stricture  d. Megaesophagus

8. The most frequently noticed complication of PRAA in foals is
   a. situs inversus.  c. tracheal compression.
   b. aspiration pneumonia.  d. colic.

9. The surgical approach for treatment of PRAA is
   a. over the left sixth rib.
   b. over the right fifth rib.
   c. between the fourth and fifth ribs.
   d. through a parasternal incision.

10. What is the prognosis of PRAA in foals?
    a. excellent  c. fair
    b. good  d. poor