Persistent Right Aortic Arch in a Canine Patient

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Introduction:

Vascular ring anomalies result from developmental abnormalities of the embryonic aortic arches and persist at birth.\textsuperscript{1,11} Of all animals diagnosed with vascular ring anomalies, the persistent right aortic arch (PRAA) accounts for 95% of those cases.\textsuperscript{2,3,4} PRAA occurs when the fourth aortic arch develops from the right side rather than the left side. This defect causes compression of the esophagus and trachea. Compression of these structures results in secondary esophageal dilation cranial to the base of the heart. Food is unable to progress beyond the stricture causing regurgitation. These episodes are most commonly seen when the animal is being weaned from the dam’s milk and transitioned to a dry food diet.\textsuperscript{2} The typical age at diagnosis is between 2-6 months of age but can be later in life. Young, large breed dogs are most commonly affected, with German Shepherds being the most common breed, followed by Irish Setters, then Greyhounds. A genetic predisposition has been identified in German Shepherds and is being investigated in Greyhounds.\textsuperscript{3,11,12} In cats, this anomaly is unusual, but is most commonly recognized in Siamese and Persian breeds. There is no sex predilection, with males and females being equally affected.\textsuperscript{11}

A presumptive diagnosis can be made from the patient’s signalment, history, and physical examination.\textsuperscript{1} Physical examination findings in an affected patient may include a thin body condition score, unthrifty hair coat, failure to thrive, enlarged esophagus upon palpation of the neck, and increased bronchovesicular sounds.\textsuperscript{11} If concurrent aspiration pneumonia is present, crackles may be present upon auscultation of the lungs, as well as pyrexia, coughing, or dyspnea.\textsuperscript{8,11} Complete blood count, serum chemistry, and urinalysis are usually unremarkable unless the patient is debilitated or has concurrent aspiration pneumonia.\textsuperscript{11} There are several diagnostic imaging modalities that aid in the diagnosis of a vascular anomaly. Thoracic
radiographs may reveal a dilated esophagus cranial to the base of the heart and ventral and leftward deviation of the trachea. Other diagnostics such as barium esophagrams, angiography, esophagostomy, echocardiograms, contrast tomography, and magnetic resonance imaging are more specific for making a diagnosis and help rule out foreign bodies or masses. Definitive diagnosis of persistent right aortic arch can be obtained by surgical exploration of the thoracic cavity.\textsuperscript{4,11}

Medical and surgical management are used in the treatment of affected patients, though medical management alone is not recommended.\textsuperscript{8,11} Surgery is the treatment of choice and provides a better long-term prognosis.\textsuperscript{3,7,9,12} Surgical approach for this particular anomaly is a left lateral thoracotomy incision at the fourth intercostal space.\textsuperscript{11} Long-term prognosis for patients is good to excellent with surgical correction, though all clinical signs may not fully resolve. Patients that continue to have chronic regurgitation may require medical management for life. Owner compliance, dedication, and attention to detail are extremely important in the long-term management of these patients.\textsuperscript{3,11} This report describes the surgical treatment and management of a complicated case of persistent right aortic arch in a 2-month-old Great Dane puppy.

**Case Report:**

A two-month-old, male intact, merle Great Dane who had recently transitioned from dam’s milk to solid food three weeks prior, was presented to MSU-AHC. The breeder noticed acute regurgitation after eating. After examination by his primary veterinarian, he was diagnosed with megaesophagus and advised to feed a diet of gruel consistency only. This diet was maintained until he accidentally consumed 2.5 cups of dry dog food after being acquired by his new owners on January 28, 2018. Following this incident, he began retching white foam, and his
new owners immediately elected to take him to the Animal Emergency & Referral Center (AERC) in Flowood, Mississippi.

Upon presentation to AERC, thoracic radiographs were performed and revealed an extremely dilated esophagus cranial to the heart base and a ventrally displaced trachea. He was quickly referred to the Mississippi State University Animal Health Center (MSU-AHC) for further diagnostics and surgery for megaesophagus secondary to a suspected PRAA.

On January 29, 2018, the patient presented to MSU-AHC Emergency Services. He was bright, alert, and responsive on admission. He weighed 13.8 kilograms with a body condition score of 4/9. He was normothermic with a temperature of 102.4 degrees Fahrenheit, tachycardic with a pulse of 180 beats per minute, and slightly tachypneic with a respiratory rate of 44 breaths per minute. Cardiac auscultation revealed no murmurs or arrhythmias and femoral pulses were strong and synchronous. Pulmonary auscultation revealed harsh bronchovesicular sounds and crackles in all lung fields, with mildly increased effort, and an SpO2 of 97%. The dog was mildly hypotensive [119/51 (74), 143/42 (76)]. His mucous membranes were pink, slightly tacky, with a capillary refill time (CRT) of less than 2 seconds and he was determined to be approximately 5% dehydrated. There was obvious esophageal distension on the left side of the neck. Abdominal palpation revealed no pain or organomegaly. Thoracic FAST scan revealed lung rockets bilaterally, and there was obvious distortion of the thoracic anatomy in the cranial thorax most likely due to his extremely dilated esophagus. Abdominal FAST scan revealed a scant amount of fluid cranial to the liver. The remainder of the patient’s physical examination was unremarkable. A complete blood count and serum chemistry were performed and revealed normal values for a young puppy. He was administered maropitant citrate (1 mg/kg SC) for nausea and vomiting. He was then transferred to the Small Animal Surgery Service to undergo a left lateral thoracotomy.
On Monday, January 29, 2018, the patient was anesthetized and taken to endoscopy to try and decompress his esophagus, as well as perform a wash for cytology and culture. He was then taken to surgery and placed in right lateral recumbency. A left lateral thoracotomy incision was made at the fourth intercostal space. After opening the thoracic cavity, the left cranial lung lobe was retracted caudally and packed off using moist laparotomy sponges. Exposure of the mediastinum dorsal to the heart was obtained. The extremely dilated esophagus was identified along with the aorta, pulmonary artery, and vagus nerve. A window was carefully dissected into the mediastinum to expose the ligamentum arteriosum. The ligamentum arteriosum was double ligated using 2-0 silk and transected then released in a controlled manner. The impacted esophageal contents were carefully milked down into the stomach. The thoracic cavity was lavaged with sterile saline solution and the lungs lobes were repositioned. A 14-gauge Mila thoracostomy tube was then placed, and the fourth intercostal space was closed in a 4-layer fashion. Negative pressure within the thoracic cavity was reestablished. During the procedure, the patient was intermittently hypoxic and hypotensive.

Upon extubation, the patient regurgitated a large volume of food material and subsequently went into respiratory distress. An arterial blood gas and SpO2 were obtained and indicated poor oxygenation. The patient was then placed in an oxygen cage and nasal cannulas were placed in both nostrils. He was maintained throughout the evening on a Fentanyl CRI at 1.5 ml/hr (5 mcg/kg/hr), norepinephrine CRI at 1.5 ml/hr (0.36 mcg/kg/hr), Plasmalyte with 2 meq/ml added potassium chloride at maintenance rate (50 ml/hr), Unasyn (30 mg/kg) IV q8h, pantoprazole (1 mg/kg) IV q24h, maropitant citrate (1 mg/kg) IV q24h, albuterol 1 puff IN q6h, and he was nebulized and coupaged q4h. His chest tube was aspirated every 12 hours.
Throughout the night, arterial blood gases were obtained periodically to monitor oxygenation with continuous invasive blood pressure and ECG.

Early the next morning, his respiratory rate and effort increased, and he was not oxygenating appropriately. Due to his declining state, it was decided to anesthetize him and place him on the ventilator. He was maintained on a propofol CRI at 2ml/hr (0.06 mg/ml/min), dexmedetomidine CRI at 0.10 ml/hr (0.5 mg/ml), Plasmalyte with 20 meq/L added potassium chloride at maintenance rate (50 ml/hr), Fentanyl CRI at 1.5 ml/hr (5 mcg/kg/hr), norepinephrine CRI at 1.5 ml.hr (0.36 mcg/kg/hr), pantoprazole (1 mg/kg) IV q24h, maropitant citrate (1 mg/kg) IV q24h, albuterol 1 puff IN q6h, and Unasyn (30 mg/kg) IV q8h.

The patient was maintained on the ventilator throughout the day, and minor adjustments to drug rates and the ventilator settings were made when needed. Thoracic radiographs were performed and revealed a severe unstructured interstitial pattern coalescing into an alveolar pulmonary pattern. These findings were consistent with aspiration pneumonia and prolonged recumbency. Following radiographs, he regurgitated once more, and his esophagus was suctioned to clear debris. A urinary catheter and a nasal esophageal feeding tube were placed. He was started on a CliniCare CRI at 20 ml/hr to maintain nutritional needs but was later discontinued due to him regurgitating. Arterial blood gases were taken every 4 hours to monitor oxygenation and lung function. Additional antibiotics, Clindamycin (11 mg/kg) and Cefotaxime (60 mg/kg), were started once the bacterial culture and sensitivity from the wash were returned and revealed light growth of *Streptococcus sp.* and gram-negative bacilli.

On February 1, 2018, the chest tube was removed, and a PEG tube was placed endoscopically to use for feeding once he was weaned from the ventilator. He was slowly weaned off of the ventilator and was extubated late that evening. He was placed in an oxygen
cage and closely monitored overnight. The following day, nasal cannulas were placed in both nostrils so that the patient had sufficient oxygen outside of the oxygen cage. He was later moved to a regular kennel for better monitoring and handling.

Throughout the weekend, he was weaned from all pain and sedative medications, and his supplemental oxygen was decreased as blood gases and SpO2 determined that he was oxygenating appropriately. Feedings through his PEG tube were started as well with gruel consistency Purina CN.

Throughout the following week, repeat radiographs were performed and showed improvement in all lung fields, and his nasal cannulas were removed due to him adequately oxygenating. He became brighter and continued to ambulate better. All intravenous medications were switched to oral medications and were administered through his PEG tube. Oral medications included Cefpodoxime (7 mg/kg) 1 tablet PO q24h, maropitant citrate (2 mg/kg) 1 tablet PO q24h, and omeprazole (1 mg/kg) PO q24h. His urinary catheter was removed. The patient had intermittent, small volume, regurgitation that was clear. Because of his improved clinical status, it was decided to discharge him on February 9, 2018.

At discharge, the owners were given strict instructions for incision care, feeding, monitoring, and managing him at home. He was sent home with Cefpodoxime (7 mg/kg) 1 tablet PO q24h, maropitant citrate (2 mg/kg) 1 tablet PO q24h, and omeprazole (1 mg/kg) PO q24h. They were advised to bring him back in 2 weeks to check his incision and repeat thoracic radiographs. Later that evening while in his owner’s care, he regurgitated several times, and his owners were concerned that they would not be able to manage his treatments at home. They ultimately decided to return to the MSU-AHC that night with him for medical boarding and PEG tube management.
Upon presentation, he was bright, alert, and responsive. He immediately regurgitated several times a large amount of clear liquid. He weighed 12.3 kg with a body condition score of 4/9. He was normothermic at 102.5 degrees Fahrenheit, normocardic with a pulse of 100 beats per minute, and eupneic with a respiratory rate of 24 breaths per minute. His mucous membranes were pink, moist, and had a capillary refill time of less than 2 seconds. Cardiac auscultation revealed no murmurs or arrhythmias and femoral pulses were strong and synchronous. Pulmonary auscultation revealed increased lung sounds throughout all lung fields and was likely due to referred upper airway noise. No crackles or wheezes were appreciated, and his SpO2 measurement was 100%. The remainder of the patient’s physical examination was unremarkable.

He was admitted into the ICU and PEG tube management, nebulization and coupage q4-6hr, albuterol 1 puff q6hr, Cefpodoxime (7 mg/kg) 1 tablet PO q24h, maropitant citrate (2 mg/kg) 1 tablet PO q24h, and omeprazole (1 mg/kg) PO q24h were continued throughout the weekend. The frequency and amount of regurgitation decreased, and he was urinating and defecating normally. On Monday February 12, 2018, he was transferred to the Surgery Service to continue medical management. Bailey chair training was initiated, and the patient was fed small meatballs of Purina CN. After being fed, he was held upright for an additional 30 minutes to allow food to pass through his esophagus. Despite being held upright, he would still regurgitate following his oral feedings. On February 16, 2018, it was elected to have an esophagram with positive contrast performed to evaluate his esophageal function. The esophagram revealed little to no function and there was significant dilation of the entire esophagus. Due to these findings, oral feedings were discontinued, and he was only fed through his PEG tube.

For the next several weeks, the patient received appropriate caloric intake for his weight and age through a CN slurry administered through his PEG tube. He was maintained on albuterol
1 puff q6hr, Cefpodoxime (7 mg/kg) 1 tablet PO q24h, maropitant citrate (2 mg/kg) 1 tablet PO q24h, and omeprazole (1 mg/kg) PO q24h. Sildenafil (1 mg/kg) PO q12h and Cisapride (0.2 mg/kg) PO q8h were started to try and improve tone of the lower esophageal sphincter and function. During that time, he experienced chronic regurgitation that varied in consistency and color and would regurgitate on average 8-10 times daily. Starting February 17, 2018 for the next 2 weeks, he intermittently became lethargic, would tremble, and would become pyrexic (103-104 degrees Fahrenheit). His respiratory rate and effort would increase during these episodes but would resolve. On March 2, 2018, he became lethargic and uncomfortable once again. He was hyperthermic at a temperature of 105 degrees Fahrenheit, tachycardic, and tachypneic.

Bloodwork was performed and revealed severely inadequate oxygen saturation of his blood (SO2 of 70%). The owner was contacted, and oxygen cage therapy was recommended. Due to the patient’s declining condition, the owners elected to humanely euthanize. He was administered propofol (6mg/kg) IV followed by euthanasia solution (1 ml/10lbs) IV.

On March 3, 2018, necropsy was performed. The lungs were diffusely mottled pink to red. The right cranioventral lung lobes and parts of the left cranioventral lung lobes were firm, consolidated, and sank when placed in formalin. On cut surface, caseous material could be expressed. The esophagus was diffusely dilated and filled with food and measured approximately 6 centimeters in diameter. The aorta was located on the right side of the esophagus, and the ligamentum arteriosum was not present. Within the stomach in the area of the pyloric antrum, a firm, round, red rubber toy was observed.

Discussion:

Vascular ring anomalies are congenital malformations that result from abnormal embryonic development of the great vessels and associated branches in utero and persists at
birth. These anomalies occur during weeks two and three of gestation.\textsuperscript{1,11} Persistent right aortic arch is the most common vascular anomaly reported and accounts for approximately 95\% of the total number in dogs and cats.\textsuperscript{5,7,11} In 44\% of cases diagnosed with persistent right aortic arch, other vascular anomalies are present.\textsuperscript{11} It is thought that German Shepherds may be genetically predisposed, and the heritability in Greyhounds is being investigated.\textsuperscript{3,11,12} One study revealed there was a monogenic autosomal recessive inheritance associated with this congenital defect in two unrelated German Pinscher puppies.\textsuperscript{5} In cats, this anomaly is rare, but is most commonly recognized in Siamese and Persian breeds. Both males and females are equally affected. If patients survive, owners should be advised against breeding these animals.\textsuperscript{11}

In the physiologically normal patient, there are 6 corresponding pairs of aortic arches that come together during embryonic development. As these vessels join, they form a ring around the embryonic gut. This later develops into the esophagus and trachea.\textsuperscript{4,11} As the fetus continues to evolve, the previously joined aortic arches begin to regress forming normal adult vasculature. The first and second aortic arches disappear. The third aortic arches join the dorsal aortic arch to form the right and left internal carotid arteries cranially, as well as, the brachiocephalic trunk.\textsuperscript{11} The left fourth aortic arch and the dorsal aortic root persist and forms the permanent aortic arch, while the right arch regresses from its corresponding arch to become the right subclavian artery.\textsuperscript{1,4,11} The fifth arches are incomplete. The left sixth arch forms into the ductus arteriosus and the right contributes to the right subclavian artery. This regression, in turn, allows the esophagus and trachea to be released. Vascular anomalies result from defects in arches three, four, and six.\textsuperscript{5,8,11}

Persistent right aortic arch is caused by abnormal development of the fourth aortic arch. This anomaly is created when the right arch is retained and forms the aorta, while the left arch
forms the left subclavian artery.\textsuperscript{10,11} This defect in combination with a normal ligamentum arteriosum (remnant of the ductus arteriosus), formulates a ring around the esophagus and trachea. Encirclement of the esophagus and trachea within the vascular ring can cause the esophagus to stricture.\textsuperscript{1,10,11} When the animal is weaned and starts consuming a solid diet, food cannot pass through the stricture causing dilation of the esophagus cranial to the base of the heart.\textsuperscript{1,11} Acute onset of regurgitation of undigested food shortly after a meal is a hallmark clinical sign. Affected individuals may look thinner and malnourished compared to littermates. If tracheal stenosis coincides, respiratory distress can occur as well.\textsuperscript{11}

Signalment, history, and a thorough physical examination are the basis for a presumptive diagnosis.\textsuperscript{1} Affected animals may have a history of chronic regurgitation following meals, especially shortly after being weaned from the dam’s milk.\textsuperscript{1,2,12} They may be small in size compared to their littermates, unable to gain weight despite a ravenous appetite, and have an unthrifty hair coat.\textsuperscript{8} An enlarged esophagus may be noticed upon palpation of the neck and thoracic inlet. Though rare, few animals may have a continuous heart murmur present if multiple vascular anomalies are present. If concurrent aspiration pneumonia is present, the patient may have pyrexia, crackles upon bronchovesicular auscultation, coughing, or dyspnea.\textsuperscript{8,11}

A complete blood count, serum chemistry, and urinalysis are expected to be normal for patients of this age unless they are extremely debilitated or have severe aspiration pneumonia. Animals with severe aspiration pneumonia may have a neutrophilia on complete blood count, while debilitated patients may be hypoproteinemic on serum chemistry.\textsuperscript{11}

Several means of diagnostic imaging can be advantageous in determining a diagnosis. Thoracic radiographs can be helpful in ruling out primary megaesophagus versus a vascular ring anomaly. Primary megaesophagus will show generalized esophageal dilation throughout.
Radiographs indicative of a vascular ring anomaly may reveal a dilated esophagus cranial to the heart with an obvious stricture located at the base of the heart. The esophagus may contain air, food, or water.\textsuperscript{4,11} One study stated that on dorsoventral and ventrodorsal radiographic views, leftward deviation of the trachea near the cranial border of the heart is strongly suggestive of a persistent right aortic arch and may alleviate the need for barium esophagrams.\textsuperscript{4} Thoracic radiographs will also show if aspiration pneumonia is present. Typical presentation includes a bronchoalveolar pattern characterized by air bronchograms within cranioventral lung fields.\textsuperscript{6} Barium esophagrams can be useful for detecting esophageal enlargement and degree of dilation, though it may be non-diagnostic in determining if a vascular ring anomaly is present if there is inadequate filling. Fluoroscopy is valuable in evaluating esophageal function and motility. An extremely dilated esophagus may have impaired peristaltic contractions. Angiography, though not commonly performed, is useful in identifying the type of vascular ring anomaly that may be present, as well as, reveal any other concurrent cardiac abnormalities. Computed tomography or magnetic resonance imaging are also valuable tools in identifying abnormal vasculature. An echocardiogram may be recommended to rule out the possibility of other congenital heart defects. Esophagoscopy can confirm esophageal stricture, and identify esophagitis or ulcers present throughout the esophagus. It may rule out an obstruction from a possible foreign body or mass.\textsuperscript{4,11} The previously described methods are advantageous in presumptively diagnosing a vascular anomaly, but the only way to confirm the diagnosis is through surgical exploration of the thoracic cavity.\textsuperscript{8}

Patients diagnosed with vascular anomalies are handled medically and surgically.\textsuperscript{11} Medical management alone is not recommended and considered only palliative therapy. Medical management includes treatment of concurrent aspiration pneumonia and insuring that the animal
maintains proper nutritional requirements. Owners should be informed that this congenital defect holds a poor prognosis without surgical intervention.\textsuperscript{8,11} Strict feeding instructions and dedicated owner compliance are essential. Animals must be fed a diet of gruel consistency while standing in an upright position with an elevated food dish. The bailey chair is a product that was created to allow patients to eat in an upright positioned with an elevated food bowl. It is important to maintain patients in this position for twenty to thirty minutes following meals to allow gravity to aid in the movement of food into the stomach without regurgitation. In animals with no esophageal function or extremely debilitated, a gastrostomy tube may be placed to assist with feedings.\textsuperscript{11}

Surgical intervention is the treatment of choice and provides a better long-term prognosis.\textsuperscript{3,7,9,12} Surgical approach for a persistent right aortic arch is a left lateral thoracotomy incision at the fourth intercostal space. After careful dissection into the thoracic cavity, the left cranial lung lobe has to be retracted caudally to visualize the mediastinum. Once exposure of the mediastinum is obtained, it important to identify the aorta, pulmonary artery, ligamentum arteriosum, and vagal and phrenic nerves. A window is carefully dissected into the mediastinum to expose the ligamentum arteriosum. Once identified, a double ligature is made using silk and then transected with controlled release. A balloon catheter or tube can be passed into the esophagus to aid in the relief of fibrous bands. The thoracic cavity should be steriley lavaged and lungs lobes repositioned into their appropriate anatomical locations. A thoracostomy tube is placed and the thorax is closed routinely. Once evacuated, it is important to reestablish negative pressure within the thoracic cavity by aspirating the thoracostomy tube.\textsuperscript{11}

Post-operatively, proper analgesia should be provided, and the patient should be closely monitored for signs of respiratory distress. Supplemental oxygen via oxygen cage or nasal
cannulas may be warranted if the patient is not oxygenating efficiently. In patients who are severely debilitated, as in this patient’s case, ventilator therapy may be required though this is extremely rare. Thoracostomy tubes should be aspirated regularly and fluid and/or air totals should be documented. These tubes can generally be removed one to three days following surgery if fluid and/or air totals are physiologically normal. Young patients must be monitored for hypoglycemia and treated accordingly. Appropriate antibiotics should be administered if aspiration pneumonia is present or if contamination during surgery occurred.\textsuperscript{3,7,8,11}

A gruel consistency diet should be continued following surgery. This should be fed in an upright position and maintained in that position for twenty to thirty minutes after each meal as previously described earlier. Over time, owners may gradually try to transition these patients to a more solid diet if regurgitation is kept at bay. If solid food is handled well, the animal may be fed from a bowl from the floor. If chronic regurgitation continues, the patient’s esophagus should be reevaluated to assess esophageal dilation and function.\textsuperscript{3,7,8,11}

One retrospective study containing fifty-two dogs diagnosed with persistent right aortic arch that underwent surgical treatment, revealed that 30\% of them had an excellent long-term outcome, while 57\% were good, and 13\% were a poor outcome. The overall survival rate in this study was 72\%. Some owners revealed that their dogs never regurgitated following surgery even on their original diet, while some revealed intermittent regurgitation occurred. Few continued to regurgitate chronically, even after surgical intervention.\textsuperscript{3} Other studies have yielded higher overall survival rates. The overall long-term prognosis for patients with persistent right aortic arch is good to excellent with surgical correction, though clinical signs may not fully resolve. In cases with continued regurgitation post-operatively, medical management may have to be
instituted for the duration of its life. Dedication and owner compliance are key in the management of these patients long-term.\textsuperscript{11}

Conclusion:

A 2-month-old Great Dane puppy presented to his primary veterinarian with acute onset of regurgitation following eating. After accidentally consuming dry dog food, the patient was referred to the Mississippi State University Animal Health Center (MSU-AHC) for further diagnostics and surgical correction of a persistent right aortic arch with secondary megaesophagus. A left lateral thoracotomy with ligamentum arteriosum ligation and division was performed. Over the next several weeks, his condition fluctuated but ultimately deteriorated due to complications of aspiration pneumonia from complete loss of esophageal function. The patient was ultimately humanely euthanized. This case report describes the surgical treatment and management of an extremely complicated case of persistent right aortic arch with secondary megaesophagus and aspiration pneumonia.
References:


