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Persistent right Aortic Arch in Dogs

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Key Points

- Embryological Defect & Clinical Presentation: Persistent Right Aortic Arch (PRAA) is a congenital vascular ring anomaly—most prevalent in breeds like German Shepherds—where embryonic persistence of the right fourth aortic arch forms a constricting ring around the oesophagus, resulting in post-weaning regurgitation and cranial megaesophagus.
- Surgical Intervention & Management: Early diagnosis via thoracic radiography/contrast esophagography followed by prompt surgical division of the ligamentum arteriosum offers an 80% favorable prognosis, which must be supported by lifelong elevated feeding protocols using specialized devices like Bailey chairs.

Abstract

Persistent Right Aortic Arch (PRAA) is a congenital vascular ring anomaly resulting from abnormal persistence of the right fourth aortic arch during embryonic development. The anomalous vessel, together with the ligamentum arteriosum, forms a vascular ring that constricts the esophagus at the level of the heart base. This results in partial esophageal obstruction, progressive dilation of the cranial thoracic esophagus, and secondary megaesophagus. Clinical signs are most commonly observed in young dogs following the transition from milk to solid food and include regurgitation, poor growth, weight loss and, in severe cases, aspiration pneumonia. Diagnosis is primarily based on thoracic radiography and contrast oesophagography, with advanced imaging and endoscopy providing additional information when required. Surgical correction, principally by division of the ligamentum arteriosum and release of the vascular constriction, is the treatment of choice. Early intervention improves the likelihood of recovery, although persistent esophageal dysfunction and megaesophagus may occur in chronic cases. A thorough understanding of the embryology, anatomical relationships, clinical presentation, diagnosis and surgical management of PRAA is therefore essential for achieving a favorable outcome.

Introduction

- Persistent Right Aortic Arch is a congenital vascular anomaly resulting in compression of the oesophagus.
- This type of ring anomaly is most commonly found in dogs which leads to regurgitation and oesophageal dilation.

Embryology

Normal Embryology:

The aortic arches initially develop symmetrically within the early embryo.The left fourth aortic arch develops into the normal, definitive systemic aortic arch. The right aortic arch regresses and degenerates.As a result, the esophagus and trachea develop freely without external vascular compression.

Persistent Right Aortic Arch (PRAA):

The aortic arches initially develop symmetrically within the early embryo. The right fourth aortic arch abnormally persists and becomes the functional aortic arch. The left aortic arch regresses instead. As a result, a vascular ring forms (typically with the ligamentum arteriosum and heart base), encircling and compressing the esophagus, which often leads to clinical signs like regurgitation and megaesophagus.

Etiology and Predisposition

Genetics: This condition is a congenital defect with a strongly suspected genetic predisposition.

Breeds: Certain dog breeds are overrepresented in these cases, most notably German Shepherds (accounting for 23% of cases), Irish Setters and Boston Terriers.

Timing: Signs typically begin to manifest between 2 and 6 months of age, particularly when the puppy transitions from milk to solid food.

Pathophysiology

The sequence of the vascular reaction begins when a vascular ring creates a focal structure at the base of the heart, acting as a mechanical block that initiates a series of secondary changes.

1. Esophageal Compression: The mechanical obstruction causes compression of the esophagus.
2. Food Accumulation: This blockage prevents normal passage, leading to the accumulation of food.
3. Cranial Esophageal Dilation: As food continues to back up, it causes cranial esophageal dilation.
4. Regurgitation: Ultimately, the progressive blockage and dilation result in regurgitation.

Clinical Signs

- Regurgitation: This involves the passive expulsion of undigested food shortly after eating.
- Failure to Thrive: Affected animals show a severely thin body appearance despite having a voracious appetite.
- Stunted Growth: There is a marked difference in size and overall development compared to healthy littermates.

Physical Examination

PARAMETER	COMMON FINDINGS	CLINICAL SIGNIFICANCE
Body condition	Thin, stunted or underweight for age	Malnutrition due to oesophageal block
Cervical area	Palpable dilated oesophagus (intermittent)	Evidence of cranial mega oesophagus
Auscultation	Crackles or wheezes in lung fields	Secondary Aspiration Pneumonia
Appetite	Voracious (ravenously hungry)	Nutritional deprivation

Diagnosis

- Radiography visualises a gas or fluid filled oesophagus located cranial to the heart.
- Barium contrast is the gold standard for identifying focal narrowing.
- Endoscopy allows direct visualisation of luminal narrowing.

Differential Diagnosis is used in conditions such as:

- Congenital Megaesophagus is a generalised esophageal motility disorder that occurs without a mechanical obstruction.
- Esophagitis is an inflammation that causes painful swallowing (odynophagia) and regurgitation.
- Esophageal Foreign Body presents as an acute onset of regurgitation, typically with a history of ingestion.

Surgical Treatment

Procedure: A left lateral thoracotomy is performed, typically at the 4th intercostal space (ICS).

Action: The surgeon ligates and transects the ligamentum arteriosum.

Goal: The objective is to achieve complete release of the mechanical esophageal constriction.

Post-operative management

- Elevated Feeding: Elevated feeding is essential to allow gravity to move food past the dilation.
- Bailey Chair: A Bailey chair is a specialised device used for upright eating.
- Diet: The diet should consist of soft or semi-solid gruel or meatballs to ensure easy digestion.

Prognosis

Overall, 80% of cases have a good to excellent prognosis outcome.

Early Diagnosis: The best results are achieved if the condition is treated before 4 months of age.

Pre-Op Status: A favorable outcome depends on the absence of aspiration pneumonia prior to surgery.

Chronic Cases: The prognosis is guarded if megaesophagus becomes irreversible.

Complications

Aspiration Pneumonia: This represents the most serious risk and can be fatal if not identified and treated aggressively with antibiotics.

Persistent Signs: A failure of esophageal motility may lead to lifelong regurgitation requiring ongoing management.

Malnutrition: Patients may fail to thrive post-operatively if dietary management is inadequate.

Conclusion

PRAA is a congenital vascular ring anomaly caused by abnormal persistence of the right fourth aortic arch. German Shepherds and Irish Setters are among the commonly reported predisposed breeds. It causes esophageal compression followed by regurgitation and secondary megaesophagus. Clinical signs commonly appear after weaning with introduction of solid food. Early diagnosis and surgical correction are essential for a favorable outcome.

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