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# Vascular Rings – A Rare Phenomenon

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**Abstract**

Diverticulum-related aortic arch anomalies are quite uncommon. We discuss an instance of a right-sided aortic arch with Kommerell's diverticulum that was accidentally identified.

Dysphagia and persistent and recurrent respiratory issues are two conditions that can occur from vascular ring, which is caused by abnormal aortic arch regression during foetal life.

This case study details a 6-month-old infant with a vascular ring who was treated for recurrent respiratory symptoms and admitted to the paediatric critical care unit with bronchilitis.

**Keywords**: NIL

## Introduction

The clinician faces a hurdle while trying to differentiate between asthma and other causes of persistent wheeze in an infant and determine the cause of airway blockage.

It is possible to hear wheezing with or without a stethoscope. Wheezing is a protracted (200 msec) or continuous melodic adventitial lung sound of different intensity. The degree of airway blockage and the speed at which air is forced through the restricted airway affect the wheezing's pitch. Wheezing typically occurs during expiration, however it can also occur during inspiration or both.

Thymus enlargement, abnormally big arteries in the superior mediastinum, or congenital goitres can all put pressure on the trachea.

Because their symptoms are similar to those of more common paediatric disorders, vascular rings are challenging to detect and may result in a delay in treatment.

Vascular rings account for about 1% of all congenital heart defects [1] and may go undiagnosed until symptoms are severe enough to demand testing. According to postmortem studies, congenital arch malformations affect 3% of the population, and two-thirds of these cases go unnoticed. [1]

Compression of the trachea and oesophagus is the cause of the symptoms, which manifest as severe disease in infants as stridor that gets worse while they eat and becomes better when they are calm or asleep.

Lung infections are most likely to arise when coughing is frequent.

Most infants with congenital vascular rings pass away before turning one year old, generally from hypoxia or chronic lung infections.

## Case

In this case, a 6-month-old child with bronchitis and a 4-month history of persistent coughing was admitted to the paediatric intensive care unit.

A full-term male infant who was six months old and experiencing respiratory distress was brought in. Since birth, his history had been marked by noisy breathing, a symptom that changed with feeding and sleeping. He received a routine otolaryngologist evaluation when he was a month old, and the results were ordinary. Before her current appearance, he had

been suffering from significant tachypnea for a number of days.

He was afebrile, had a heart rate of 167 beats per minute, a blood pressure of 102/74 millimetres of mercury, a respiratory rate of 82 beats per minute, and a nasal continuous positive airway pressure oxygen saturation of 97% at the time of her initial admission.

He had apparent congestion, loud breath sounds bilaterally, mild inspiratory stridor, and respiratory distress.

He didn't have murmur, and his rate and rhythm were well felt. Bilaterally, both the proximal and distal pulses were clearly audible.

The abdomen had no organomegaly and was soft. No obvious skeletal abnormalities were present. He was alert aside from it. He moved his extremities symmetrically and normally.

The initial lab tests revealed a striking diagnosis of leukocytosis.

On a chest X-ray, the lungs were well-expanded and hyperinflated without any localised consolidation, pneumothorax, or pleural effusion.

The heart's size was remarkably normal. It was unclear how many sides the aortic arch had. The air column in the trachea was challenging to see.

By using CT angiography, the right dominant aortic arch was identified, and a 3D reconstruction revealed that the distal trachea had dramatically as a result of vascular compression. Further examination did not reveal the presence of any aortic or branch anomalies.

The most accurate way to diagnose a vascular ring is through CT imaging, which may clearly show the vascular ring's architecture and any concomitant tracheal disease. For our patient, a Kommerell diverticulum—a structure—was identified in the right aortic arch.

**Discussion**

Early infancy is a time when symptoms including stridor, feed vomiting, and cough are typical. Although the aetiology is mostly benign, in just a few per cent of children, these symptoms are brought on by the existence of a complete or partial vascular ring compressing the trachea or oesophagus.

It is challenging to differentiate these patients from the much larger group of children with non-serious illnesses, and doing so calls both a high index of suspicion as well as familiarity with the various investigative methods.

The phrase "vascular ring" describes congenital vascular abnormalities of the aortic arch system that compress oesophagus and trachea, resulting in symptoms involving those two structures.

Our case he had a 4-month history of persistent coughing, wheezing, and dyspnea. His respiratory issues were initially diagnosed as bronchiolitis, but tracheitis and pneumonia worsened his clinical status.

Due to difficulties with respiration, our patient liked to eat slowly while reclining with his heads retracted. Dysphagia frequently appeared as a symptom when solid or semi-solid foods were added to the diet.

In 1787, Bayford(2) documented an autopsy case of "obstructed deglutition" brought on by a right aberrant subclavian artery (ASCA) flowing posterior to the oesophagus. Arkin came up with the term "dysphagia lusoria" and called the abnormal ASCA "arteria lusoria."(3)But it wasn't until 1936 that Dr. Burkhard F. Kommerell established the first clinical diagnostic of dysphagia lusoria brought on by an ASCA and an aortic diverticulum compressing the oesophagus in a living subject.(4)

Due to failed regression, the Kommerell diverticulum (KD) is a persisting remnant of the fourth primitive dorsal arch.The right dorsal aorta involutes close to the right subclavian artery in the case of a left aortic arch with KD, leaving the subclavian artery connected to the left descending aorta via the distal section of the right dorsal aorta. The left dorsal arch between the carotid and subclavian arteries shrinks in patients with the right aortic arch in KD, allowing the right subclavian to originate from the residual right dorsal arch.(6)

According to the aetiology, Salomonowitz et al. (7) divided the KD into three categories: (1) KD in the left aortic arch with right ASCA; (2) KD in the right aortic arch with left ASCA; and (3) aortic diverticulum without ASCA.

In relation to the prevalence of KD and ASCA, a higher proportion of female patients (65%) were



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found in our group.(10,11), but in this instance, we report a unique case of a male child.

Due to the prevalence of concomitant congenital heart disease, patients with vascular rings should all get an echocardiography. This exemplifies how variable echocardiography is among operators. However, if the diagnosis of vascular ring has been established, it does play a significant impact given the high prevalence of congenital heart disease in this group.

Using CT or MRI is one of the accessible imaging techniques that is possibly the most alluring. Surgery planning may benefit from accurate imaging provided by MRI and CT [12].

In order to investigate the possibility of an anatomical aberration, a three-dimensional chest computed tomography scan of our patient showed a Kommerell's diverticulum and an aortic arch vascular ring pressing against the trachea on the right side.

This is important in order to prepare for the proper counselling and follow-up after a diagnosis has been established, it is crucial to be aware of the results and long-term challenges of surgical treatment.

## Conclusion

A high index of suspicion for a vascular ring should be held when a clinician is faced with a newborn or toddler who has a chronic cough, noisy breathing, or dysphagia. A chest radiograph is performed as part of the initial diagnostic assessment. On the basis of pictures from an echocardiography or bronchoscopy, the diagnosis may be suspected in other kids. Currently, we favour using CT imaging to make the diagnosis. This clearly illustrates the vascular ring's architecture and any related tracheal disease.

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