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Case Report

# The Hidden Arch: An Incidental Vascular Surprise in a Toddler Undergoing Cleft Palate Repair

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**ABSTRACT**

Double aortic arch (DAA) is a rare congenital vascular anomaly that forms a complete vascular ring encircling the trachea and esophagus, potentially leading to respiratory and feeding difficulties. We present a case of a 15-month-old male with an incomplete cleft palate who had an incidental finding of codominant double arch of aorta on preoperative imaging. The child underwent successful cleft palate surgery under general anesthesia with appropriate airway precautions and multidisciplinary planning. This report highlights the anesthetic implications of vascular rings and the need for thorough preoperative assessment in children presenting with non-specific symptoms like recurrent upper respiratory tract infections (URTI) and feeding intolerance.

## Introduction

Double aortic arch (DAA) is the most common type of complete vascular ring, accounting for approximately 50% of all vascular ring anomalies. It results from the persistence of both the right and left fourth embryonic aortic arches, leading to the formation of a ring that can encircle and compress the trachea and esophagus [1].

Symptoms often begin in infancy and may include stridor, wheezing, recurrent respiratory infections, and feeding difficulties. The diagnosis can be elusive, especially when the child has coexisting structural anomalies such as a cleft palate, which can independently contribute to similar symptoms. Timely diagnosis plays an important role in preventing complications and helps in planning appropriate perioperative management [2].
This case highlights the need to pay attention to subtle clinical findings that could indicate an underlying vascular anomaly, as well as the anesthetic challenges encountered during surgery.

## Case Report

A 15-month-old male child weighing 8.2 kg was scheduled for surgical correction of an incomplete cleft palate (Figure 1).

**Figure 1- Incomplete cleft palate**

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A detailed pre-anesthetic check-up (PAC) was conducted. The child had a significant history of feeding intolerance and recurrent upper respiratory tract infections since infancy. The child was delivered at term with a birth weight of 3.1 kg and had a smooth perinatal period. He did not experience neonatal jaundice, cyanosis, seizures, respiratory infections, or require NICU care.

All routine preoperative tests, such as complete blood count, renal and liver function tests, ECG, and chest X-ray, were normal. During the standard echocardiographic evaluation for cleft palate, a mirror-image branching of the great vessels was noted, raising the possibility of an underlying vascular anomaly. CT aortography was advised for further evaluation, which revealed a double aortic arch causing mild narrowing of the adjoining thoracic trachea and compression on the thoracic oesophagus (Figure 2).

**Figure 2- CT Aortograph**

A cardiology consultation recommended proceeding with palatoplasty under high-risk consent, with ICU and postoperative ventilator support available due to the presence of a vascular ring.

On the day of surgery, after confirming nil per os (NPO) status and establishing a patent and secure intravenous (IV) line, the patient was shifted inside the operating room after premedicating with Inj. Glycopyrrolate 0.004 mg/kg, Inj. Midazolam 0.02 mg/kg, and Inj. Ketamine 1 mg/kg.

All essential equipment was prepared in the operating room, including the emergency drug trolley and the difficult airway trolley equipped with a CMAC pediatric video laryngoscope, to manage any unexpected airway challenges.

Following attachment of routine ASA monitors, preoxygenation with 100% oxygen was performed before anesthesia induction. Inj. Fentanyl 1 mcg/kg was given. Inj. Propofol 3 mg/kg was used for induction, followed by laryngoscopy with a McIntosh blade no. 2, which revealed an elongated uvula, but the vocal cords were visualized. A 5 mm uncuffed South Pole endotracheal tube was secured in place (Figure 3). Inj. Atracurium 0.5 mg/kg was used as a muscle relaxant. Inj. Hydrocortisone 16 mg IV and Inj. Dexamethasone 0.8 mg IV were given. The intubation procedure proceeded without any complications

**Figure 3- South pole endotracheal tube insitu**

Intraoperatively, anesthesia was maintained with oxygen, air, and sevoflurane. The airway pressures were normal even during insertion of the mouth gag and were maintained below 20 mm of Hg with no immediate signs of airway resistance or obstruction. To facilitate smoother intraoperative hemodynamics along with analgesia, a low-dose dexmedetomidine infusion was initiated after induction at 0.3 mcg/kg/hr. Bilateral greater palatine nerve blocks were administered by the surgeons for analgesia. The surgery proceeded without any complications. The child remained hemodynamically stable throughout the procedure. Blood loss was minimal and was substituted with intravenous fluids in the form of Ringer's lactate.

**Figure 4- Repaired Cleft Palate**

Following the completion of surgery (Figure 4), extubation was performed using Inj. Neostigmine 0.4 mg and Inj. Glycopyrolate 0.06 mg. The extubation process was uneventful. Postoperatively, the child was monitored in the recovery area for airway obstruction, stridor, or dysphagia. Supportive measures included humidified oxygen, head-end elevation, and IV dexamethasone. Pain was managed with IV paracetamol, and hydration was maintained with appropriate IV fluids.

## Discussion

The child was initially evaluated for a cleft palate, and a detailed cardiovascular assessment to rule out other congenital anomalies led to the diagnosis of a double aortic arch. This case underlines the importance of maintaining a high index of suspicion for vascular rings in infants with non-specific symptoms, especially when coexisting anomalies such as cleft palate are present. In such children, echocardiography, while useful, may not be sufficient to define the arch anatomy, and a CT aortography is the gold standard for confirming the diagnosis [1-2].

Double aortic arch (DAA) presents specific anesthetic challenges because of the risk of dynamic airway compression, particularly during induction or emergence from anesthesia [3]. Clinical signs such as stridor, wheezing, feeding difficulties, and recurrent respiratory infections often resemble common pediatric airway problems, which can lead to delayed diagnosis [4]. The severity and onset of symptoms usually reflect the extent of tracheoesophageal compression and may change with growth or body positioning [5].

Perioperative care in these patients requires careful coordination among anesthesiologists, surgeons, and cardiologists. Airway management should anticipate potential difficulties with mask ventilation or intubation. Having advanced airway equipment, such as pediatric videolaryngoscopes, and being prepared for postoperative ventilation are important considerations [6]. In our case, the patient had minimal airway compression, allowing safe induction and extubation, but this is not guaranteed in patients with a dominant arch or a tighter vascular ring.

Early recognition of vascular rings can improve outcomes by preventing recurrent respiratory problems and growth delays related to feeding difficulties [7]. Incidental findings on echocardiography or chest imaging should prompt thorough evaluation, especially in children with syndromic features or other midline anomalies [8]. While some children with DAA remain asymptomatic for years, surgical correction is generally recommended in symptomatic cases to relieve the compressive effects [9].

Interestingly, in this child, cleft palate-associated symptoms masked the underlying vascular ring, reinforcing the value of comprehensive preoperative imaging and multidisciplinary care.

## Conclusion

This case emphasizes the importance of careful preoperative evaluation and airway planning in children with vascular rings, even when diagnosed incidentally. Early recognition and coordinated multidisciplinary management can help ensure safe anesthetic care and favorable surgical outcomes in such patients.

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