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Double Aortic Arch Presenting as Neonatal Stridor

YT ***Chan,*** DKK ***Ng,*** ASF ***Chong,*** JCS ***Ho***

Abstract Stridor is a common complaint in neonates. Association with respiratory distress mandates early flexible bronchoscopic examination of the airway. We report a case of double aortic arch who presented with stridor and respiratory distress. Management of double aortic arch was discussed. Persistence of stridor after surgery and its management was highlighted.

Key words CPAP; Double aortic arch; Newborn; Non-invasive ventilation; Stridor

Introduction

Amongst respiratory symptoms, stridor is a common complaint in a full-term neonate. Congenital cardio-vascular anomalies are rare but treatable causes of stridor in the pediatric population. The reported incidence of aortic vascular rings was 0.3-0.6%.[1] Double aortic arch was the most common type of congenital vascular ring anomalies.[2,3] We report here the management of a neonate with double aortic arch that presented as neonatal stridor.

Case Report

A newborn was referred to our unit for respiratory distress at 1 hour of life. He was a full term baby with birth weight of 3.09 Kg. He was born by vaginal delivery. Physical examination revealed a normal looking neonate with signs of respiratory distress, i.e. marked subcostal insucking. Examination of other systems were unremarkable. He was found to have cyanosis during feeding. SpO_2 during rest was 100% and during feeding was 90%. Chest X-radiograph showed bilateral hyper-inflated chest with no other abnormalities. Complete blood picture was normal. The provisional diagnosis was sepsis. The baby was put on nasal continuous positive airway pressure (CPAP) support on day 2. His respiratory distress was improved but he still had subcostal insucking. Inspiratory stridor was noticed on day three of life. Bronchoscopy (Figure 1) showed a short segment tracheomalacia at mid-trachea caused by an extra-mural pulsatile mass on day 5. Subsequent CT thorax (Figure 2) on the same day showed the pulsatile mass to be double aortic arch.

The infant underwent elective operation on day 8 of life. Division of right arch was done, which was complicated by right chylothorax. Chest drain was inserted over right chest for one week and the baby was put on Pregestimil® (Mead Johnson, Holland). Right chylothorax resolved after one week. The respiratory distress decreased after the operation and he could tolerate slow-rate oral feeding. However, he still had difficult feeding and inspiratory stridor. MRI thorax (Figure 3) and bronchoscopy (Figure 4) were repeated. Persistent tracheomalacia was identified in bronchoscopy. Video fluoroscopic swallowing study was normal and the feeding difficulty was likely due to respiratory distress. In view of the persistent respiratory distress, he was put on

Department of Paediatrics, Kwong Wah Hospital, 25 Waterloo Road, Kowloon, Hong Kong, China

YT **Chan** （陳日東） *MBBS*
DKK **Ng** （吳國強） *M Med Sc, FRCP, FHKAM(Paed)*
JCS **Ho** （何誌信） *MBBS, FRCP, FRCR*

Department of Radiology, Kwong Wah Hospital, 25 Waterloo Road, Kowloon, Hong Kong, China

ASF **Chong** （莊瑞芬）*MBBS, FRCR, FHKAM(Radiol)*

Correspondence to: Dr DKK **Ng**

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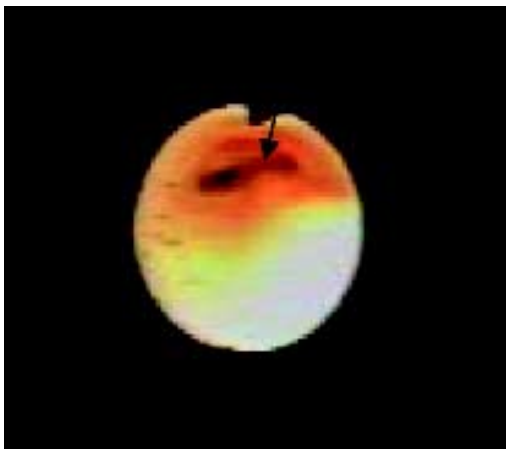


Figure 1 Bronchoscopy showed marked compression of trachea from 10 o'clock to 4 o'clock (arrow) by an extra-mural pulsating mass.

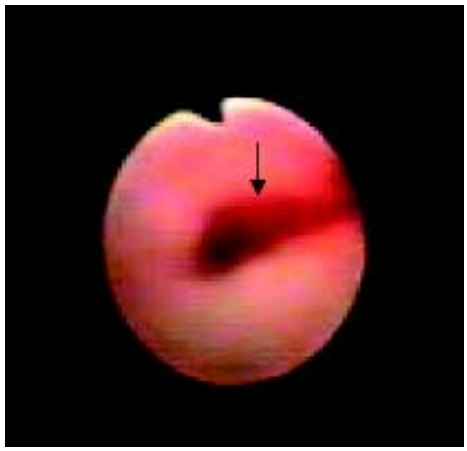
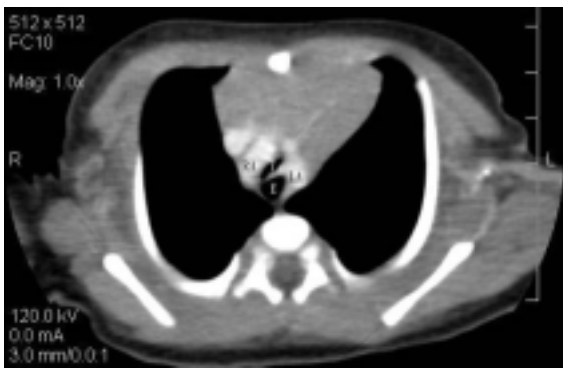
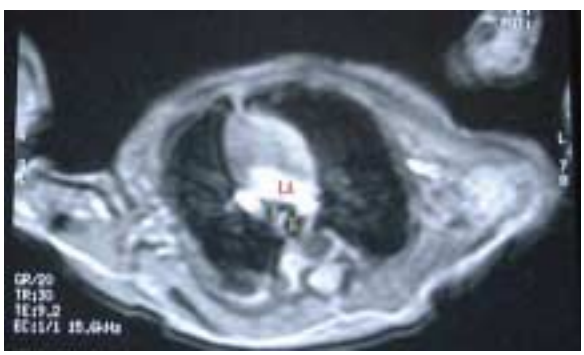


Figure 4 Bronchoscopy showed tracheomalacia (arrow) during inspiration.



RA: Right arch; LA: Left arch; T: Trachea; E: Esophagus

Figure 2 CT thorax showed double aortic arch causing focal tracheal narrowing.



LA: Left arch; T: Trachea; E: Esophagus

Figure 3 MRI showed mild narrowing of middle and distal trachea.

intermitent CPAP. Inspiratory stridor and feeding difficulty improved after the baby was put on intermittent CPAP, two hours during daytime and during sleep. CPAP pressure was 6 cmH_2O. He was thriving afterwards. Nocturnal CPAP was tailed off at nine months of age and he was free of symptoms at nine-month of age. During last follow-up at nine-month-old, his body weight was at 80th percentile, compared with body weight at 3rd centile at three-month-age. His feeding was satisfactory and no stridor was detected.

Discussion

Double aortic arch is due to failure of regression of the right aortic arch. Normally, absorption of the right (posterior) arch takes place between the right subclavian artery and its junction with the descending aorta. The remnant of the right arch becomes the right innominate artery and leaves a left (anterior) arch in normal development, freeing the trachea and esophagus. Failure of this process of absorption in the right arch produces a vascular ring, the esophagus and trachea are completely encircled and compressed, leading to severe respiratory and feeding difficulties.

The first successful surgical treatment of a double aortic arch was reported by Dr. Gross in 1945.[4] Double aortic arch usually exists as an isolated problem, although the presence of associated congenital heart disease such as tetralogy of Fallot, ventricular septal defect, and transposition of great arteries were reported.[5]

Respiratory symptoms at birth or soon after should alert the pediatrician to the possibility of vascular ring compression. Presenting symptoms included stridor (100%), persistent cough (75%), chronic dyspnoea (75%), reflex apnoea (60%), recurring respiratory tract infections (56%) and dysphagia (25%).[6] The onset of symptoms from double aortic arch tended to be earlier than other type of vascular rings.[3] Majority of patients with double aortic arch have surgical correction before one-year-old, whereas the average age for congenital vascular ring anomalies (non-double aortic arch) patients was four-years-old. It was suggested that compression of the trachea and esophagus from double aortic arch was more severe than that with other types of vascular rings.[7]

Tracheal compression may cause significant morbidity and mortality.[8] CPAP represent[5] an important non-invasive method to relieve airway obstruction before surgery.

Flexible bronchoscopy would be the first line of investigation for neonatal stridor. Typical compression of the esophageus and the trachea was visualized by esophagoscopy and bronchoscopy which were indicated for respiratory or feeding difficulties. Nature of extra-mural obstruction would be best shown by spiral CT scan, magnetic resonance imaging (MRI) or color-coded 3D echocardiogram reconstruction or angiography. Angiography confirms the diagnosis by demonstrating the details of the anomaly. Spiral CT scan and color-coded 3D echocardiogram reconstruction represent important non-invasive techniques used in evaluation of vascular anomalies of the thoracic aorta and pulmonary arteries in infants and children.[9] One case series showed that angiography, magnetic resonance imaging, and computed tomography were reliable tests and magnetic resonance imaging was found to be the investigation of choice.[10] However, magnetic resonance imaging requires a long acquisition time and is prone to motion artifacts. A case of double aortic arch was reported to be missed on magnetic resonance imaging.[11] Hence, spiral computed tomography would be a better choice of non-invasive tool for delineation of nature of obstruction demonstrated by the flexible bronchoscopy.

Surgical division of the vascular ring was indicated in patients with symptoms of airway or esophageal compression. Opening the ring was accomplished by dividing the arch and patent ductus or ligamentum arteriosus. Further dissection along the trachea and esophagus was carried out to release any constriction or fibrous band.[12] A complete release of the constriction by operation may not result in the immediate and total resolution of symptoms because the prolonged compression of trachea often results in tracheomalacia and lung infection.[13] Other operative complications include chylothorax, diaphragmatic paresis and vocal cords paresis. Complication rate of chylothorax had been reported to be 2 out of 42 cases.[12] In another case series, 84% of post-operative chylothorax cases responded to conservative management. The average duration of lymph leak was 13.7 days.[14]

Surgical division of double aortic arch has been performed without mortality in a case series of 42 patients.[12] In another case series of 39 patients with congenital aortic arch anomalies, two deaths occurred: 1 infant had undergone emergency operation for control of hemorrhage from an aorto-tracheal fistula due to tracheostomy tube erosion, and the other had multiple associated congenital heart defects. Median length of follow-up was 12.5 months, with at least 97% of survivors completely or nearly completely free of symptoms from the vascular ring.[15] Another case series also reported symptom-free evolution in most patients over a three to 90 months follow-up period.[16]

CPAP would be a good non-invasive method to relieve airway obstruction secondary to the short segment tracheomalacia associated with double aortic arch. This was well shown by our patient whose clinical response was satisfactory with catch-up growth.

In conclusion, tracheobronchial compression by vascular structures in childhood is uncommon and may be masked by non-specific respiratory symptoms. Diagnosis required a high index of suspicion. Early bronchoscopy is essential to confirm the vascular compression of trachea. Imaging techniques like spiral CT scan or MRI can clearly delineate the causative pathologic vascular structures. Surgical treatment is effective and safe. Tracheobronchial malacia may persist for several months after the operation and CPAP is an important tool to relief airway obstruction during the pre-operation and post-operation period.

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